As filed with the Securities and Exchange Commission on December 23, 2013.

Registration No. 333-192244

UNITED STATES
SECURITIES AND EXCHANGE COMMISSION

Washington, D.C. 20549

Amendment No. 1
to
FORM S-1

REGISTRATION STATEMENT UNDER THE SECURITIES ACT OF 1933

ULTRAGENYX PHARMACEUTICAL INC.

(Exact name of Registrant as specified in its charter)

Delaware	**2834**	**27-2546083**
(State or other jurisdiction of incorporation or organization)	*(Primary Standard Industrial Classification Code Number)*	*(I.R.S. Employer Identification Number)*

60 Leveroni Court
Novato, CA 94949
(415) 483-8800
(Address, including zip code, and telephone number, including area code, of Registrant's principal executive offices)

Emil D. Kakkis, M.D., Ph.D.
President and Chief Executive Officer
Ultragenyx Pharmaceutical Inc.
60 Leveroni Court
Novato, CA 94949
(415) 483-8800
(Name, address, including zip code, and telephone number, including area code, of agent for service)

Copies to:

Ryan A. Murr, Esq. **Lisa M. Kahle, Esq.** **Ropes & Gray LLP** **Three Embarcadero Center, Third Floor** **San Francisco, CA 94111** **Telephone: (415) 315-6300** **Facsimile: (415) 315-6350**	**Shalini Sharp** **Chief Financial Officer** **Ultragenyx Pharmaceutical Inc.** **60 Leveroni Court** **Novato, CA 94949** **Telephone: (415) 483-8800** **Facsimile: (415) 483-8810**	**B. Shayne Kennedy, Esq.** **Brian J. Cuneo, Esq.** **Latham & Watkins LLP** **650 Town Center Drive, 20th Floor** **Costa Mesa, CA 92626** **Telephone: (714) 540-1235** **Facsimile: (714) 755-8290**

Approximate date of commencement of proposed sale to the public:
As soon as practicable after the effective date of this Registration Statement.

If any of the securities being registered on this Form are to be offered on a delayed or continuous basis pursuant to Rule 415 under the Securities Act of 1933, check the following box. ☐

If this Form is filed to register additional securities for an offering pursuant to Rule 462(b) under the Securities Act, please check the following box and list the Securities Act registration statement number of the earlier effective registration statement for the same offering. ☐

If this Form is a post-effective amendment filed pursuant to Rule 462(c) under the Securities Act, check the following box and list the Securities Act registration statement number of the earlier effective registration statement for the same offering. ☐

If this Form is a post-effective amendment filed pursuant to Rule 462(d) under the Securities Act, check the following box and list the Securities Act registration statement number of the earlier effective registration statement for the same offering. ☐

Indicate by check mark whether the registrant is a large accelerated filer, an accelerated filer, a non-accelerated filer, or a smaller reporting company. See the definitions of "large accelerated filer," "accelerated filer" and "smaller reporting company" in Rule 12b-2 of the Exchange Act.

Large accelerated filer ☐	Accelerated filer ☐	Non-accelerated filer ☒ (Do not check if a smaller reporting company)	Smaller reporting company ☐

The Registrant hereby amends this Registration Statement on such date or dates as may be necessary to delay its effective date until the Registrant shall file a further amendment which specifically states that this Registration Statement shall thereafter become effective in accordance with Section 8(a) of the Securities Act of 1933 or until the Registration Statement shall become effective on such date as the Commission, acting pursuant to said Section 8(a), may determine.

Explanatory Note

This Amendment No. 1 is being filed for the purpose of filing Exhibits 10.1, 10.2, 10.3, 10.6, 10.7, 10.8 and 10.9. No changes or additions are being made hereby to the Prospectus constituting Part I of the Registration Statement (not included herein) or to Part II of the Registration Statement other than with respect to Item 15 of Part II.

PART II
Information Not Required in Prospectus

Item 13. Other Expenses of Issuance and Distribution.

The following table sets forth the costs and expenses, other than the underwriting discounts and commissions, payable by the registrant in connection with the sale of common stock being registered. All amounts are estimates except for the Securities and Exchange Commission, or SEC, registration fee, the FINRA filing fee and The NASDAQ Global Market listing fee.

Item	Amount to be paid
SEC registration fee	$11,109
FINRA filing fee	13,438
The NASDAQ Global Market Listing fee	*
Printing and engraving expenses	*
Legal fees and expenses	*
Accounting fees and expenses	*
Blue Sky, qualification fees and expenses	*
Transfer Agent fees and expenses	*
Miscellaneous expenses	*
Total	$ *

* To be provided by amendment

Item 14. Indemnification of Directors and Officers.

Section 145(a) of the Delaware General Corporation Law provides, in general, that a corporation may indemnify any person who was or is a party or is threatened to be made a party to any threatened, pending or completed action, suit or proceeding, whether civil, criminal, administrative or investigative (other than an action by or in the right of the corporation), because he or she is or was a director, officer, employee or agent of the corporation, or is or was serving at the request of the corporation as a director, officer, employee or agent of another corporation, partnership, joint venture, trust or other enterprise, against expenses (including attorneys' fees), judgments, fines and amounts paid in settlement actually and reasonably incurred by the person in connection with such action, suit or proceeding, if he or she acted in good faith and in a manner he or she reasonably believed to be in or not opposed to the best interests of the corporation and, with respect to any criminal action or proceeding, had no reasonable cause to believe his or her conduct was unlawful.

Section 145(b) of the Delaware General Corporation Law provides, in general, that a corporation may indemnify any person who was or is a party or is threatened to be made a party to any threatened, pending or completed action or suit by or in the right of the corporation to procure a judgment in its favor because the person is or was a director, officer, employee or agent of the corporation, or is or was serving at the request of the corporation as a director, officer, employee or agent of another corporation, partnership, joint venture, trust or other enterprise, against expenses (including attorneys' fees) actually and reasonably incurred by the person in connection with the defense or settlement of such action or suit if he or she acted in good faith and in a manner he or she reasonably believed to be in or not opposed to the best interests of the corporation, except that no indemnification shall be made with respect to any claim, issue or matter as to which he or she shall have been adjudged to be liable to the corporation unless and only to the extent that the Court of Chancery or other adjudicating court determines that, despite the adjudication of liability but in view of all of the circumstances of the case, he or she is fairly and reasonably entitled to indemnity for such expenses which the Court of Chancery or other adjudicating court shall deem proper.

Section 145(g) of the Delaware General Corporation Law provides, in general, that a corporation may purchase and maintain insurance on behalf of any person who is or was a director, officer, employee or agent of

the corporation, or is or was serving at the request of the corporation as a director, officer, employee or agent of another corporation, partnership, joint venture, trust or other enterprise against any liability asserted against such person and incurred by such person in any such capacity, or arising out of his or her status as such, whether or not the corporation would have the power to indemnify the person against such liability under Section 145 of the Delaware General Corporation Law.

Article VI of our amended and restated certificate of incorporation (the "Charter"), provides that no director of our company shall be personally liable to us or our stockholders for monetary damages for any breach of fiduciary duty as a director, except for liability (1) for any breach of the director's duty of loyalty to us or our stockholders, (2) for acts or omissions not in good faith or which involve intentional misconduct or a knowing violation of law, (3) in respect of unlawful dividend payments or stock redemptions or repurchases, or (4) for any transaction from which the director derived an improper personal benefit. In addition, our Charter provides that if the Delaware General Corporation Law is amended to authorize the further elimination or limitation of the liability of directors, then the liability of a director of our company shall be eliminated or limited to the fullest extent permitted by the Delaware General Corporation Law, as so amended.

Article VI of the Charter further provides that any repeal or modification of such article by our stockholders or amendment to the Delaware General Corporation Law will not adversely affect any right or protection existing at the time of such repeal or modification with respect to any acts or omissions occurring before such repeal or modification of a director serving at the time of such repeal or modification.

In connection with the sale of common stock being registered hereby, we have entered into indemnification agreements with each of our directors and our executive officers. These agreements will provide that we will indemnify each of our directors and such officers to the fullest extent permitted by law and the Charter.

We also maintain an insurance policy that covers certain liabilities of directors and officers of our company arising out of claims based on acts or omissions in their capacities as directors or officers.

In any underwriting agreement we enter into in connection with the sale of common stock being registered hereby, attached hereto as exhibit 1.1, the underwriters will agree to indemnify, under certain conditions, us, our directors, our officers and persons who control us within the meaning of the Securities Act against certain liabilities.

Item 15. Recent Sales of Unregistered Securities.

In the three years preceding the filing of this registration statement, we or our California corporation predecessor have issued the following securities that were not registered under the Securities Act. Issuances made prior to June 2011 were made by Ultragenyx Pharmaceutical, Inc., a California corporation. In connection with our reincorporation into Delaware in June 2011, each outstanding share of common stock was converted into 80 shares of the new Delaware corporation.

Common Stock Purchase Agreements

On April 27, 2010, we issued 100,000 shares of common stock for consideration of $0.01 per share, for an aggregate purchase price of $1,000.00 to Emil D. Kakkis. Emil D. Kakkis is our current President and Chief Executive Officer and one of our directors.

On November 19, 2010, we issued 3,000 shares of common stock for consideration of $0.01 per share, for an aggregate purchase price of $30.00 to Nobelpharma Co. Ltd.

On February 11, 2011, we issued 1,030 shares of common stock for consideration of $0.01 per share, for an aggregate purchase price of $10.30 to William E. Aliski. Mr. Aliski is one of our directors.

On February 28, 2011, we issued 1,030 shares of common stock for consideration of $0.01 per share, for an aggregate purchase price of $10.30 to Jonathan K. Wright.

On March 11, 2011, we issued 1,667 shares of common stock for consideration of $0.01 per share, for an aggregate purchase price of $16.67 to William E. Aliski. Mr. Aliski is one of our directors.

On March 28, 2011, we issued 1,030 shares of common stock for consideration of $0.10 per share, for an aggregate purchase price of $103.00 to Steven Jungles. Mr. Jungles is our Senior Vice President, Technical Operations.

On April 6, 2011, we issued 11,300 shares of common stock for consideration of $0.01 per share, for an aggregate purchase price of $113.00 to John Klock.

We claimed exemption from registration under the Securities Act for the sale and issuance of these shares of common stock by virtue of Section 4(a)(2) and/or Regulation D promulgated thereunder as transactions not involving any public offering. All of the purchasers of the shares of common stock for which we relied on Section 4(a)(2) and/or Regulation D represented that they were accredited investors as defined under the Securities Act. We claimed such exemption on the basis that (a) the purchasers in each case represented that they intended to acquire the securities for investment only and not with a view to the distribution thereof and that they either received adequate information about the registrant or had access, through employment or other relationships, to such information and (b) appropriate legends were affixed to the stock certificates issued in such transactions.

Convertible Notes and Series A Convertible Preferred Stock Warrants

On June 30, 2010, we entered into a Note and Warrant Purchase Agreement with Emil D. Kakkis, our President and Chief Executive Officer and one of our directors. Pursuant to the Note and Warrant Purchase Agreement, we issued a convertible promissory note in the amount of $1.0 million to Dr. Kakkis and also issued him a warrant to purchase up to 241,803 shares of our Series A convertible preferred stock.

On February 15, 2011, we entered into a Note and Warrant Purchase Agreement with the John and Cynthia Klock Trust. Pursuant to the Note and Warrant Purchase Agreement, we issued a convertible promissory note in the amount of $1.5 million to the John and Cynthia Klock Trust and also issued to the trust a warrant to purchase up to 507,786 shares of our Series A convertible preferred stock.

On February 23, 2011, we entered into a Note and Warrant Purchase Agreement with William Aliski, one of our directors. Pursuant to the Note and Warrant Purchase Agreement, we issued a convertible promissory note in the amount of $250,000 to Mr. Aliski and also issued him a warrant to purchase up to 84,631 shares of our Series A convertible preferred stock.

On June 14, 2011, we entered into a Note and Warrant Purchase Agreement with Emil D. Kakkis, our President and Chief Executive Officer and one of our directors. Pursuant to the Note and Warrant Purchase Agreement, we issued a convertible promissory note in the amount of $300,000 to Dr. Kakkis and also issued him a warrant to purchase up to 72,541 shares of our Series A convertible preferred stock.

On June 14, 2011, we entered into a second Note and Warrant Purchase Agreement with Emil D. Kakkis, our President and Chief Executive Officer and one of our directors. Pursuant to the Note and Warrant Purchase Agreement, we issued a convertible promissory note in the amount of $500,000 to Dr. Kakkis and also issued him a warrant to purchase up to 120,901 shares of our Series A convertible preferred stock.

We claimed exemption from registration under the Securities Act for the sale and issuance of these securities by virtue of Section 4(a)(2) and/or Regulation D promulgated thereunder as transactions not involving any public offering. All of the purchasers of unregistered securities for which we relied on Section 4(a)(2) and/or Regulation D represented that they were accredited investors as defined under the Securities Act. We claimed such exemption on the basis that (a) the purchasers in each case represented that they intended to acquire the securities for investment only and not with a view to the distribution thereof and that they either received adequate information about the registrant or had access, through employment or other relationships, to such information and (b) appropriate legends were affixed to the stock certificates issued in such transactions.

Series A Convertible Preferred Stock Financing

On June 16, 2011, we sold an aggregate of 18,052,464 shares of our Series A convertible preferred stock to eight investors at a purchase price of $1.034 per share, for an aggregate purchase price of approximately $15.0 million in cash and $3.7 million in converted bridge notes. On July 16, 2012, we sold, pursuant to a second tranche closing, an aggregate of 14,604,895 shares of our Series A convertible preferred stock to six investors at a purchase price of $1.034 per share, for an aggregate purchase price of $15.1 million in cash. We claimed exemption from registration under the Securities Act for the sale and issuance of these securities by virtue of Section 4(a)(2) and/or Regulation D promulgated thereunder as transactions not involving any public offering. All of the purchasers of unregistered securities for which we relied on Section 4(a)(2) and/or Regulation D represented that they were accredited investors as defined under the Securities Act. We claimed such exemption on the basis that (a) the purchasers in each case represented that they intended to acquire the securities for investment only and not with a view to the distribution thereof and that they either received adequate information about the registrant or had access, through employment or other relationships, to such information and (b) appropriate legends were affixed to the stock certificates issued in such transactions.

Series A Paid-in-Kind Dividends

On August 16, 2012, we issued an aggregate of 1,193,088 shares of our Series A convertible preferred stock to holders of our Series A convertible preferred stock in accordance with Article IV.B.1.(a) of our Certificate of Incorporation that was filed with the Secretary of State of the State of Delaware on June 13, 2011. We claimed exemption from registration under the Securities Act for the sale and issuance of these securities by virtue of Section 4(a)(2) and/or Regulation D promulgated thereunder as transactions not involving any public offering.

On December 14, 2012, we issued an aggregate of 499,447 shares of our Series A convertible preferred stock to holders of our Series A convertible preferred stock in accordance with Article IV.B.1.(a) of our Certificate of Incorporation that was filed with the Secretary of State of the State of Delaware on June 13, 2011. We claimed exemption from registration under the Securities Act for the sale and issuance of these securities by virtue of Section 4(a)(2) and/or Regulation D promulgated thereunder as transactions not involving any public offering.

Series B Convertible Preferred Stock Financing

On December 18, 2012, we sold an aggregate of 27,081,680 shares of our Series B convertible preferred stock to 34 investors at a purchase price of $2.7694 per share, for an aggregate purchase price of approximately $75 million in cash. We claimed exemption from registration under the Securities Act for the sale and issuance of these securities by virtue of Section 4(a)(2) and/or Regulation D promulgated thereunder as transactions not involving any public offering. All of the purchasers of unregistered securities for which we relied on Section 4(a)(2) and/or Regulation D represented that they were accredited investors as defined under the Securities Act. We claimed such exemption on the basis that (a) the purchasers in each case represented that they intended to acquire the securities for investment only and not with a view to the distribution thereof and that they either received adequate information about the registrant or had access, through employment or other relationships, to such information and (b) appropriate legends were affixed to the stock certificates issued in such transactions.

Stock Options

From November 17, 2011 through December 19, 2013, we granted stock options to employees under our 2011 Equity Incentive Plan, as amended, covering an aggregate of 10,089,000 shares of common stock, at a weighted-average average exercise price of $0.81 per share. Of these, options covering an aggregate of 821,667 shares were cancelled without being exercised and we sold an aggregate of 2,280,945 shares of common stock to employees for cash consideration in the aggregate amount of $0.3 million upon the exercise of stock options.

We claimed exemption from registration under the Securities Act for these sales and issuances under Section 4(a)(2) of the Securities Act in that such sales and issuances did not involve a public offering or under

Rule 701 promulgated under the Securities Act, in that they were offered and sold either pursuant to written compensatory plans or pursuant to a written contract relating to compensation, as provided by Rule 701.

Item 16. Exhibits and Financial Statement Schedules.

(a) Exhibits.

See the Exhibit Index attached to this Registration Statement, which is incorporated by reference herein.

(b) Financial Statement Schedules.

Schedules not listed above have been omitted because the information required to be set forth therein is not applicable or is shown in the financial statements or notes thereto.

Item 17. Undertakings.

The undersigned Registrant hereby undertakes to provide to the underwriters at the closing specified in the underwriting agreement certificates in such denominations and registered in such names as required by the underwriters to permit prompt delivery to each purchaser.

Insofar as indemnification for liabilities arising under the Securities Act may be permitted to directors, officers and controlling persons of the Registrant pursuant to the foregoing provisions, or otherwise, the Registrant has been advised that in the opinion of the SEC such indemnification is against public policy as expressed in the Securities Act, and is, therefore, unenforceable. In the event that a claim for indemnification against such liabilities (other than the payment by the Registrant of expenses incurred or paid by a director, officer, or controlling person of the Registrant in the successful defense of any action, suit or proceeding) is asserted by such director, officer or controlling person in connection with the securities being registered, the Registrant will, unless in the opinion of its counsel the matter has been settled by controlling precedent, submit to a court of appropriate jurisdiction the question of whether such indemnification by it is against public policy as expressed in the Securities Act and will be governed by the final adjudication of such issue.

The undersigned Registrant hereby undertakes that:

1. For purposes of determining any liability under the Securities Act, the information omitted from the form of prospectus filed as part of this Registration Statement in reliance upon Rule 430A and contained in a form of prospectus filed by the Registrant pursuant to Rule 424(b)(1) or (4) or 497(h) under the Securities Act shall be deemed to be part of this Registration Statement as of the time it was declared effective.
2. For the purpose of determining any liability under the Securities Act, each post-effective amendment that contains a form of prospectus shall be deemed to be a new registration statement relating to the securities offered therein, and the offering of such securities at that time shall be deemed to be the initial bona fide offering thereof.

The undersigned Registrant hereby undertakes to provide to the underwriters at the closing specified in the underwriting agreement certificates in such denominations and registered in such names as required by the underwriters to permit prompt delivery to each purchaser.

Signatures

Pursuant to the requirements of the Securities Act of 1933, as amended, the Registrant has duly caused this Amendment No. 1 to Registration Statement on Form S-1 to be signed on its behalf by the undersigned, thereunto duly authorized, in Novato, California, on December 23, 2013.

ULTRAGENYX PHARMACEUTICAL INC.

By: /s/ EMIL D. KAKKIS
Emil D. Kakkis, M.D., Ph.D.
President and Chief Executive Officer

Pursuant to the requirements of the Securities Act, this Amendment No. 1 to Registration Statement has been signed by the following persons in the capacities and on the dates indicated.

Signature	Title	Date
/S/ EMIL D. KAKKIS Emil D. Kakkis, M.D., Ph.D.	Director, President and Chief Executive Officer *(Principal Executive Officer)*	December 23, 2013
/S/ SHALINI SHARP Shalini Sharp	Senior Vice President, Chief Financial Officer *(Principal Financial and Accounting Officer)*	December 23, 2013
* Eran Nadav, Ph.D.	Chairman of the Board	December 23, 2013
* Benjamin Auspitz	Director	December 23, 2013
* Mårten Steen, M.D., Ph.D.	Director	December 23, 2013
* William Aliski	Director	December 23, 2013

*By: /s/ SHALINI SHARP
Shalini Sharp
Attorney-in-fact

Exhibit Index

Exhibit Number	Exhibit Description	Incorporated by Reference			Filed Herewith
		Form	Date	Number	
1.1*	Form of Underwriting Agreement				
3.1	Amended and Restated Certificate of Incorporation, as currently in effect	S-1	11/8/2013	3.1	
3.2	Bylaws, as currently in effect	S-1	11/8/2013	3.2	
3.3*	Amended and Restated Certificate of Incorporation, to be in effect immediately prior to the completion of this offering				
3.4*	Amended and Restated Bylaws, to be in effect immediately prior to the completion of this offering				
4.1	Reference is made to Exhibits 3.1 through 3.4				
4.2	Form of Common Stock Certificate	S-1	11/8/2013	4.2	
4.3	Warrant to purchase Series A Preferred Stock of the Registrant, dated as of June 30, 2010, issued to Emil D. Kakkis, M.D., Ph.D.	S-1	11/8/2013	4.3	
4.4	Warrant to purchase Series A Preferred Stock of the Registrant, dated as of February 15, 2011, issued to the John and Cynthia Klock Charitable Trust	S-1	11/8/2013	4.4	
4.5	Warrant to purchase Series A Preferred Stock of the Registrant, dated as of February 23, 2011, issued to William E. Aliski	S-1	11/8/2013	4.5	
4.6	Warrant to purchase Series A Preferred Stock of the Registrant, dated as of June 14, 2011, issued to Emil D. Kakkis, M.D., Ph.D.	S-1	11/8/2013	4.6	
4.7	Warrant to purchase Series A Preferred Stock of the Registrant, dated as of June 14, 2011, issued to Emil D. Kakkis, M.D., Ph.D.	S-1	11/8/2013	4.7	
4.8	Amended and Restated Investors' Rights Agreement, dated December 18, 2012, among the Registrant and the investors named therein	S-1	11/8/2013	4.8	
5.1*	Opinion of Ropes & Gray LLP				
10.1†	Collaboration and License Agreement, dated as of August 29, 2013, between the Registrant and Kyowa Hakko Kirin Co., Ltd.				X
10.2†	License Agreement, dated as of March 1, 2011, between the Registrant and AAIPharma Services Corp.				X
10.3†	License Agreement, dated as of September 20, 2012, between the Registrant and Baylor Research Institute				X
10.4†	Amendment to the License Agreement, dated as of March 22, 2013, between the Registrant and Baylor Research Institute	S-1	11/8/2013	10.4	
10.5†	Exclusive License Agreement, dated as of April 23, 2012, between the Registrant and HIBM Research Group	S-1	11/8/2013	10.5	
10.6†	Collaboration and License Agreement, dated as of September 30, 2010, between the Registrant and Nobelpharma Co., Ltd.				X
10.7†	License Agreement, dated as of September 1, 2012, between the Registrant and St. Jude Children's Research Hospital				X

Exhibit Number	Exhibit Description	Incorporated by Reference			Filed Herewith
		Form	Date	Number	
10.8†	Exclusive License Agreement, dated as of November 22, 2010, between the Registrant and Saint Louis University				X
10.9†	Supply Agreement, dated as of November 19, 2012, between the Registrant and CREMER OLEO GmbH & Co KG				X
10.10†	Development and Clinical Supply Agreement, dated as of August 31, 2012, between the Registrant and Rentschler Biotechnologie GmbH	S-1	11/8/2013	10.10	
10.11#	2011 Equity Incentive Plan (including forms of Stock Option Grant Notice and Stock Option Agreement thereunder)	S-1	11/8/2013	10.11	
10.12#	Amendment to the 2011 Equity Incentive Plan	S-1	11/8/2013	10.12	
10.13#*	2014 Incentive Plan				
10.14#*	Form of Incentive Stock Option Award Agreement				
10.15#*	Form of Non-Qualified Stock Option Award Agreement				
10.16#*	Form of Restricted Stock Unit Award Agreement				
10.17#*	2014 Employee Stock Purchase Plan				
10.18#	Executive Employment Agreement, dated as of June 15, 2011, between the Registrant and Emil D. Kakkis, M.D., Ph.D.	S-1	11/8/2013	10.18	
10.19#	Offer Letter, dated as of October 31, 2011, between the Registrant and Thomas Kassberg	S-1	11/8/2013	10.19	
10.20#	Offer Letter, dated March 12, 2012, between the Registrant and Shalini Sharp	S-1	11/8/2013	10.20	
10.21#*	Form of Indemnification Agreement				
10.22	Standard Lease, dated as of July 5, 2011, between the Registrant and Condiotti Enterprises, Inc.	S-1	11/8/2013	10.22	
10.23	License and Services Agreement, dated as of September 24, 2010, between the Registrant and The Buck Institute for Age Research	S-1	11/8/2013	10.23	
10.24	Amendment No. 1 to License and Services Agreement, dated as of September 4, 2012, between the Registrant and The Buck Institute for Research on Aging	S-1	11/8/2013	10.24	
10.25#*	Corporate Bonus Plan				
23.1	Consent of independent registered public accounting firm	S-1	11/8/2013	23.1	
23.2*	Consent of Ropes & Gray LLP (included in Exhibit 5.1)				
24.1	Power of Attorney	S-1	11/8/2013	24.1	
99.1	Consent of Matthew Fust	S-1	11/8/2013	99.1	

* To be filed by amendment.

† Portions of this exhibit (indicated by asterisks) have been omitted pursuant to a request for confidential treatment and this exhibit has been filed separately with the SEC.

# Indicates management contract or compensatory plan.

Exhibit 10.1

COLLABORATION AND LICENSE AGREEMENT

by and between

KYOWA HAKKO KIRIN CO., LTD.

and

ULTRAGENYX PHARMACEUTICAL INC.

Dated August 29, 2013

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

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COLLABORATION AND LICENSE AGREEMENT

This Collaboration and License Agreement (this "**Agreement**") is made effective as of August 29, 2013 (the "**Effective Date**"), by and between Kyowa Hakko Kirin Co., Ltd., a company organized and existing under the laws of Japan, with an address at 1-6-1 Ohtemachi, Chiyoda-ku, Tokyo, 100-8185, Japan ("**KHK**"), and Ultragenyx Pharmaceutical Inc., a company organized and existing under the laws of California, U.S.A., with an address at 60 Leveroni Ct. Novato, CA 94949, U.S.A. ("**UGNX**"). KHK and UGNX are sometimes hereinafter referred to each as a "**Party**" and collectively as the "**Parties**."

WITNESSETH:

WHEREAS, KHK desires to grant UGNX, and UGNX desires to accept, certain licenses and other rights subject to the terms of this Agreement regarding the Development and Commercialization of the Licensed Products in the Territory and the European Territory (each as defined below); and

WHEREAS, the Parties desire to set forth the terms and the conditions of such licenses and other rights;

NOW, THEREFORE, in consideration of the foregoing premises and the mutual covenants herein contained, the Parties hereby agree as follows:

ARTICLE 1.
DEFINITIONS

1.1 **Definitions**. When used in this Agreement, capitalized terms will have the meanings as defined below and throughout the Agreement.

1.1.1 "**Affiliate**" with respect to a Party means an individual, trust, business trust, joint venture, partnership, corporation, association or other legal entity that, directly or indirectly, through one or more intermediaries, controls, is controlled by, or is under common control with that Party. For purposes of this definition only, "control" and, with correlative meanings, the terms "controlled by" and "under common control with" means (a) the possession, directly or indirectly, of the power to direct the management or policies of a legal entity, whether through the ownership of voting securities or by contract relating to voting rights or corporate governance, or (b) the ownership, directly or indirectly, of fifty percent (50%) or more of the voting securities or other ownership interest of a legal entity.

1.1.2 "**Applicable Laws**" means any federal, state, local, national and supra-national laws, statutes, rules and/or regulations, including any rules, regulations, guidance, guidelines or requirements of Regulatory Authorities, national securities exchanges or securities listing organizations, that may be in effect from time to time during the Term and apply to a particular activity hereunder and including laws, regulations and guidelines governing the import, export, development, manufacture, marketing, distribution and/or sale of Licensed Products.

1.1.3 "**Business Day**" means a day that is not a Saturday, Sunday or a day on which banking institutions in either Tokyo, Japan, or San Francisco, U.S.A., are required by law to remain closed.

1.1.4 "**Calendar Quarter**" means the period beginning on the Effective Date and ending on the last day of the Calendar Quarter in which the Effective Date falls, and thereafter each successive period of three (3) consecutive calendar months ending on March 31, June 30, September 30 or December 31.

1.1.5 "**Calendar Year**" means the period beginning on the Effective Date and ending on the last day of the Calendar Year in which the Effective Date falls, and thereafter each successive period of twelve (12) months commencing on January 1 and ending on December 31.

1.1.6 "**CKD**" means chronic kidney disease.

1.1.7 "**Clinical Trial**" means a Phase 1 Clinical Trial, a Phase 2 Clinical Trial, a Phase 3 Clinical Trial, a Phase 4 Clinical Trial, a Phase 5 Clinical Trial or a combination of any of the foregoing.

1.1.8 "**Commercialization Costs**" means all costs and expenses, including [***], incurred by or on behalf of a Party in accordance with this Agreement and attributable to, or reasonably allocable to, the Commercialization of the Licensed Products in the Field, including [***] as further set forth in the Financial Exhibit attached as Exhibit A to this Agreement.

1.1.9 "**Commercialization**" means any and all activities relating to marketing, selling, promoting, distributing, importing, detailing, offering to sell, having sold, and/or selling the Licensed Products, whether before or after the Marketing Approvals and Pricing and/or Reimbursement Approvals for such Licensed Product have been obtained. When used as a verb, "**Commercialize**," means to engage in Commercialization.

1.1.10 "**Commercially Reasonable Efforts**" means, with respect to a Party in the performance of its obligations hereunder, the application by or on behalf of such Party of a level of efforts that a similarly-situated pharmaceutical or biotechnology company, as the case may be, would apply to such activities in relation to a similar pharmaceutical product owned by it or to which it has rights, which product is at a similar stage in its development or product life and is of similar market potential, profit potential and strategic value (in each case as compared to the Licensed Products) taking into account efficacy, safety, expected labeling, the competitiveness of alternative products in the marketplace, the patent and other proprietary position of the product, the likelihood of regulatory approval given the regulatory structure involved, the profitability of the product including the royalties payable to licensors, or patent or other intellectual property rights, alternative products and other relevant factors, based on conditions then prevailing.

1.1.11 "**Confidential Information**" means information of a confidential or proprietary nature disclosed by a Party to the other Party hereunder, whether disclosed in oral, written, graphic or electronic form provided that Confidential Information disclosed in written, graphic or electronic form expressly indicates the confidential nature of such information and that Confidential Information disclosed orally is reduced to writing in summary form indicating the confidential nature of such information within [***] Business Days of disclosure, in connection with this Agreement or the performance of its obligations hereunder, including any such information related to any scientific, clinical, engineering, manufacturing, marketing, financial or personnel matters relating to a Party, or related to a Party's present or future products, sales, suppliers, customers, employees, investors, business plans, Know-How, regulatory filings, data, compounds, research projects, work in progress, future developments or business, in all such cases whether disclosed in oral, written, graphic or electronic form; provided, however, that in any event, Confidential Information excludes any information that: (a) is known by recipient at the time of its receipt, and not through a prior disclosure by or on behalf of the disclosing Party, as documented by contemporaneous business records; (b) is or becomes properly in the public domain through no fault

of the recipient; (c) is subsequently disclosed to the recipient by a Third Party who may lawfully do so and is not directly or indirectly under an obligation of confidentiality to the disclosing Party, as documented by written business records in existence prior to the receipt of such information from the disclosing Party; or (d) is developed by the recipient independently of, and without reference to or use of, Confidential Information received from the disclosing Party.

1.1.12 "**Control**" means with respect to any Patent Rights, Know-How or other intellectual property rights, possession of the right, whether directly or indirectly, and whether by ownership, license or otherwise, to grant a license, sublicense or other right to or under, such Patent Rights, Know-How or other intellectual property rights as provided for herein without violating the terms of any agreement or other arrangements with any Third Party at the time when such license, sublicense or other rights are first granted hereunder.

1.1.13 "**Development**" means, with respect to the [***] and the [***], including the [***]. When used as a verb, "**Develop**" means to engage in Development.

1.1.14 "**Development Costs**" means all costs and expenses, including [***], incurred by or on behalf of a Party in accordance with this Agreement and attributable to, or reasonably allocable to, the [***] of the [***] in the [***] and that are [***], including costs and expenses [***].

1.1.15 "**Drug Substance**" means the recombinant human IgG1 monoclonal antibody targeting FGF23 identified as KRN23 with the amino acid sequence set forth on Schedule 1.1.15.

1.1.16 "**European Centralized Approval**" means a [***].

1.1.17 "**European Core Territory**" means [***].

1.1.18 "**European Territory**" means (a) the European Core Territory, if any; (b) any other member states of the European Union (if applicable); and (c) Switzerland and Turkey.

1.1.19 "**European Union**" means the European Union consisting of all member states thereof as of the Effective Date.

1.1.20 "**European Transition Date**" means the date on which Marketing Approval for a Licensed Product for the First Indication is obtained in the European Territory on a country-by-country basis (or, in the case of the European Core Territory, the date on which the European Centralized Approval is obtained for a Licensed Product for the First Indication). By way of illustration, if UGNX [***].

1.1.21 "**FDA**" means the United States Food and Drug Administration or any successor agency thereto.

1.1.22 "**Field**" means the treatment and/or prevention of: (a) any Orphan Disease (whether or not listed in the following clauses); (b) XLH; (c) [***]; (d) [***]; (e) [***]; and (f) [***]; provided, however, that the Field will not include either (i) [***] or (ii) [***].

1.1.23 "**Financial Exhibit**" means the exhibit attached hereto as Exhibit A.

1.1.24 "**First Commercial Sale**" means, with respect to a country in the [***] or [***], the first commercial sale of any Licensed Product to a Third Party for use, consumption or resale in that country after obtaining Marketing Approval and Pricing and/or Reimbursement Approval in that country. For the avoidance of doubt, [***].

1.1.25 "**First Indication**" means [***].

1.1.26 "**First Pediatric Study**" means a first Clinical Trial in pediatric patients in the First Indication.

1.1.27 "**First Pediatric Study Deadline**" is defined in Section 15.3.1(a).

1.1.28 "**Force Majeure Event**" is defined in Section 17.2.

1.1.29 "**FTE**" means a full time equivalent person year (consisting of a total of [***] per year) for personnel supporting [***] and/or [***] and/or [***] of [***] in [***] in accordance with this Agreement. For the avoidance of doubt, [***].

1.1.30 "**FTE Costs**" means all costs for FTEs calculated by multiplying (a) [***] by (b) the [***], provided that, to the extent either Party is unable to fully track the number of FTEs utilized, the Parties shall agree on a mechanism for estimating such number. For clarity, FTE Costs shall in no case include [***], provided that costs [***], may be included.

1.1.31 "**FTE Rate**" means, except as specified in the Financial Exhibit with respect to the Sales Force FTE Rate therein defined, [***] for each of KHK and UGNX (such amount premised on the accuracy of information disclosed and used to determine the amount), with respect to the [***]. With respect to [***]. At the time of any [***] proposed by either Party, the other Party ("**Reviewing Party**") shall have the right to review, upon written request, the

information (including actual costs and applicable assumptions) used by the proposing Party to determine its FTE Rate. The proposing Party shall provide any such information (in an accurate manner) reasonably requested by the Reviewing Party, and in the event the Reviewing Party reasonably believes that additional discussions are required to confirm the basis for the determination of such FTE rate, the Parties shall engage in such discussions in good faith. For clarity, the FTE Rate shall in no case include [***], provided that costs [***], may be included.

1.1.32 "**GAAP**" means generally accepted accounting principles applicable to a Party in a particular country (e.g., Japanese Accounting Standards or U.S. Generally Accepted Accounting Principles) as consistently applied throughout the applicable periods indicated herein by or on behalf of the relevant Party.

1.1.33 "**GMP**" means the then-current good manufacturing practices required by the FDA and as set forth in the laws and regulations in the United States with respect thereto, for the manufacture and testing of pharmaceutical materials, and comparable Applicable Laws and requirements of Regulatory Authorities applicable to the manufacture and testing of pharmaceutical materials in jurisdictions within the Territory, as they may be updated from time to time, including applicable rules and guidelines promulgated under the International Conference on Harmonization.

1.1.34 "**IND**" means an Investigational New Drug Application filed with the FDA pursuant to 21 CFR 312.20, or the corresponding filing required for the clinical testing in humans of a pharmaceutical product in any country or regulatory jurisdiction other than the United States.

1.1.35 "**Initiation**" means, with respect to [***], the date on which [***]. "**Initiate**" shall have a correlative meaning.

1.1.36 "**In-Licenses**" means: (a) [***]; (b) [***]; and (c) [***].

1.1.37 "**Joint Invention**" means any invention made by at least [***].

1.1.38 "**Key Latin American Country**" means any of the following countries: [***].

1.1.39 "**KHK Invention**" means any invention or discovery that relates to a [***] (a) a [***] or (b) [***].

1.1.40 "**KHK Non-Core Development Plan**" means KHK's plan for the KHK Non-Core Development Activities.

1.1.41 "**KHK Out-of-Field Products**" means [***] for use outside of the Field.

1.1.42 "**KHK Regulatory Data**" means all regulatory filings made by or on behalf of KHK (including by its Affiliates and licensees other than UGNX) with respect to the Licensed Products and all data generated by or on behalf of KHK (including by its Affiliates and licensees) in the performance of non-clinical and clinical development activities or regulatory activities, including any information contained in any regulatory filings with respect to the Licensed Products and the results of, and all information generated in connection with, any non-clinical or clinical studies or regulatory activities performed by or on behalf of KHK with respect to the Licensed Products.

1.1.43 "**KHK Trademark**" means any trademark or trade name, including registrations and applications therefor, owned or Controlled by KHK covering KHK's corporate name and/or company logo.

1.1.44 "**Know-How**" means any non-public knowledge, experience, know-how, technology, information and data (including pharmacological, toxicological and clinical data and analytical and quality control data), trade secrets, formulas and formulations, processes, techniques, unpatented inventions, methods, discoveries, specifications, formulations, compositions, materials, ideas, developments, test procedures and results, together with all documents and files embodying the foregoing, but excluding, in any event, any patent rights in any of the foregoing.

1.1.45 "**Latin America**" means Central America and South America, where "**Central America**" means Belize, Costa Rica, El Salvador, Guatemala, Honduras, Mexico, Nicaragua and Panama; and "**South America**" means Argentina, Bolivia, Brazil, Chile, Columbia, Ecuador, Guyana, Paraguay, Peru, Surinam, Uruguay and Venezuela.

1.1.46 "**Licensed Know-How**" means Know-How Controlled by KHK or its Affiliates at any time during the Term which (a) relates to the Drug Substance and/or the Licensed Products, and (b) is necessary or useful for the Development, use or Commercialization of the Drug Substance and/or Licensed Products in the Field. Without limiting the foregoing, "Licensed Know-How" shall include the KHK Clinical Data and KHK Regulatory Data.

1.1.47 "**Licensed Patent Rights**" means all patents and patent applications (and patents issued from the foregoing) Controlled by KHK or its Affiliates at any time during the Term which (a) cover the Drug Substance and/or the Licensed Products or their use and (b) are necessary or useful for the Development, use or Commercialization of the Drug Substance and/or the Licensed Products in the Field. A list of Licensed Patent Rights as of the Effective Date is attached as Schedule 1.1.47.

1.1.48 "**Licensed Products**" means any formulation or product that is comprised of, in whole or in part, or containing the Drug Substance as an active ingredient (regardless of dosage, strength or size).

1.1.49 "**Licensed Technology**" means the Licensed Patent Rights and the Licensed Know-How.

1.1.50 "**Marketing Approval**" means, with respect to a Licensed Product in a country or region, all registrations or authorizations (other than Pricing and/or Reimbursement Approvals) required from Regulatory Authority(ies) to market and sell such Licensed Product in such country or region. For the avoidance of doubt, with respect to the European Core Territory, "Marketing Approval" may consist of a European Centralized Approval.

1.1.51 "**Marketing Materials**" is defined in Section 6.4.

1.1.52 "**Medical Geneticist**" means a medical professional who specializes in the diagnosis, treatment, counseling and/or management of patients with hereditary disorders.

1.1.53 "**Named Patient Sale(s)**" means the sale of a Licensed Product in a country on a "named-patient" basis to meet the needs of particular patients under the order of a physician or other health care professional.

1.1.54 "**Net Sales**" means, with respect to any Licensed Product, the gross amounts invoiced by a Party or its Affiliates **(**"**Selling Party**"**)** to any Third Party for sales of such Licensed Product in any indication in the Field in the Territory or in the European Territory, as applicable, including Named Patient Sales, less the following items, provided that they are bona fide:

(a) actual credits, refunds or allowances to Third Party customers for spoiled, damaged, rejected, recalled, outdated and reasonably returned Licensed Products;

(b) discounts, including cash, volume, quantity and other trade discounts, charge-back payments, and rebates and allowances actually granted, incurred or allowed in the ordinary course of business, as well as government-required discounts and allowances (including government rebates and other price reductions), and other reductions, concessions and allowances that effectively reduce the selling price to the Selling Party;

(c) transportation charges, freight, postage and insurance (but only insurance related to protecting the particular shipment against physical loss or damage); and

(d) sales, use or excise Taxes and import/export duties or tariffs and similar governmental charges actually due or incurred in connection with the sales of such Licensed Products.

Components of Net Sales shall be determined in the ordinary course of business in accordance with GAAP (as applicable in the country of sale), consistently applied. Net Sales shall include, for any Licensed Product, [***]. Notwithstanding the above, to the extent KHK and/or its Affiliates enter into an arrangement that calls for or permits the manufacturing and/or sale of Licensed Products by a Third Party (whether or not a Sublicensee) in consideration for a running royalty or other consideration that operates as a substitute thereof (other than milestone payments or upfront payments) then such Third Party shall be the "Selling Party" and its sales or resales shall be included in definition of "Net Sales". Furthermore, the Parties acknowledge and agree that, with respect to the [***], Net Sales shall be determined based on [***], provided that, in the [***]. For purposes of determining when a sale of any Licensed Products occurs for purposes of calculating Net Sales, the sale will be deemed to occur on date the Licensed Products are shipped. No deductions shall be made for commissions paid to individuals or agents, nor shall any deductions be permitted for the cost of collections. For purposes of determining Net Sales, a "sale" shall not include transfers or dispositions, at no cost or below cost, of Licensed Products for charitable, non-clinical, clinical or regulatory purposes or for promotional samples or free goods. Amounts invoiced by the Selling Party for the sale of Licensed Products to another Affiliate for resale to a Third Party shall not be included in the computation of Net Sales hereunder. In the event that the Selling Party sells the Licensed Products: (a) to a Third Party in a bona fide arm's length transaction, for material consideration, in whole or in part, other than cash (but excluding, for the avoidance of doubt, consideration in the form of non-financial legal terms and conditions incident to sale including, for clarity, the supply of Licensed Products for non-commercial purposes substantially at cost); (b) to a Third Party in other than a bona fide arm's length transaction; or (c) with discounts of Licensed Products that are disproportional to the discounts of other products sold by the Selling Party in conjunction with such Licensed Products, the Net Sales price for such Licensed Product shall be deemed to be the standard invoice price then being invoiced by the Selling Party in an arm's length transaction with similar customers in the same country within the Territory and/or the European Territory. In the event that the Selling Party includes one or more Licensed Product as part of a bundle of products, the price for such Licensed Products shall be deemed to be the standard invoice price for such Licensed Products when sold separately and not as part of a bundle of products. In the event that no separate prices are charged in the applicable transaction, then Net Sales for such bundle shall be determined based on the list price for the Licensed Product and the other products or services in the relevant country during the accounting period in which the sale was made. If no list price exists in such country for the Licensed Product or the other products or services that are part of the bundle, then Net Sales for such bundle shall be equitably determined based on the fair market value of the Product relative to that of the other products or services. Any dispute between the Parties with respect to the determination of such market value shall be finally resolved pursuant to Section 16.4.

1.1.55 "**Non-Core Development Plan**" means, as applicable, the UGNX Non-Core Development Plan and/or the KHK Non-Core Development Plan.

1.1.56 "**Orphan Disease**" means a disease or condition for which in any given country a pharmaceutical treatment meets, as of the Effective Date, the definition of an "orphan product" or "orphan drug" for treatment of a "rare disease" under the U.S. Orphan Drug Act or Regulation No. 141/2000 of the European Parliament and of the Council of 16 December 1999 on orphan medicinal products, each as amended, or any successor laws or regulations thereto.

1.1.57 "**Option Negotiation Right Field**" means all [***] and the Licensed Products outside of the Field, [***]. Option Negotiation Right Field shall not include [***].

1.1.58 “**Patent Rights**” means all issued patents and patent applications, certificates of invention, or applications for certificates of invention, together with any extensions, registrations, confirmations, provisionals, divisionals, continuations, continuations in part (to the extent the claims in such continuation-in-part application are directed to subject matter specifically described in such prior patent application), and patents issuing therefrom, reissues, divisions, continuations, or continuations-in-part, reexaminations, substitutions, renewals, restorations, additions, registrations, and foreign counterparts thereof, as well as extensions and supplementary protection certificates based thereon

1.1.59 “**Person**” means any individual, corporation, association, partnership (general or limited), joint venture, trust, estate, limited liability company, limited liability partnership, unincorporated organization, government (or any agency or political subdivision thereof) or other legal entity or organization.

1.1.60 “**Phase 1 Clinical Trial**” means a human clinical trial performed in accordance with Applicable Laws that provides for the first introduction of a Licensed Product into humans for the purpose of determining human toxicity, metabolism, biomarker, absorption, elimination and other pharmacological action.

1.1.61 “**Phase 2 Clinical Trial**” means a human clinical trial performed in accordance with Applicable Laws in patients with a particular disease or condition which is designed to establish the safety, appropriate dosage, efficacy, and tolerability of a Licensed Product given its intended use and to initially explore its efficacy for such disease or condition and will include such a clinical trial intended to be a pivotal trial.

1.1.62 “**Phase 3 Clinical Trial**” means a registration or pivotal clinical trial performed in accordance with Applicable Laws and conducted in subjects with a particular disease or condition which is designed to establish the efficacy and safety of a Licensed Product given its intended use and to define warnings, precautions and adverse events that are associated with such Licensed Product in the dosage range intended to be prescribed.

1.1.63 “**Phase 4 Clinical Trial**” means a post-registration clinical trial or post-marketing surveillance study performed in accordance with Applicable Laws and required as a condition to, or for the maintenance of, any Marketing Approval or Pricing and/or Reimbursement Approval for a Licensed Product.

1.1.64 “**Phase 5 Clinical Trial**” means a post-registration clinical trial that is not required as a condition to, or for the maintenance of, any Marketing Approval or Pricing and/or Reimbursement Approval for a Licensed Product. Phase 5 Clinical Trials are commonly referred to as “post-marketing clinical trials”.

1.1.65 “**Pricing and/or Reimbursement Approvals**” means, with respect to a Licensed Product in any country or region where Regulatory Authorities may approve or determine pricing or pricing reimbursement for such Licensed Product, such approval or determination.

1.1.66 “**Product Trademarks**” means any trademarks or trade name, and registrations and applications therefor, for a Licensed Product selected in accordance with Section 2.4 for use in connection with the Commercialization of such Licensed Product in the Territory or the European Territory in the Field (excluding, in any event, KHK Trademarks and UGNX Trademarks).

1.1.67 “**Profit Share Territory**” means the U.S. and Canada.

1.1.68 “**Profit Share Territory Transition Date**” means the fifth (5th) anniversary of the First Commercial Sale in the U.S. in the First Indication.

1.1.69 “**Regulatory Authority**” means any applicable governmental regulatory authority involved in granting approvals for the manufacture, Commercialization, reimbursement and/or pricing a Licensed Product. Regulatory Authority includes the FDA in the U.S. or the applicable governmental regulatory authority in any country or region, or any successor agency of the foregoing having regulatory jurisdiction over the manufacture, distribution and sale of drugs in any country or region.

1.1.70 “**Regulatory Filings**” means any filings that may be required for any Marketing Approval, Pricing and/or Reimbursement Approval or otherwise filed or submitted to a Regulatory Authority in an effort to comply with Applicable Laws with respect to the Development or Commercialization of a Licensed Product. Regulatory Filings include any IND (or similar filing outside the U.S.), Biologic Licensing Application (or similar filing outside the U.S.), clinical study protocols and Regulatory Authority briefing books.

1.1.71 “**Rest of the World**” means all countries and territories outside the [***] and the [***].

1.1.72 “**Royalty Term**” means: (a) with respect to each country in the [***] and [***], the period of time commencing on the date of the First Commercial Sale in that country and continuing for as long as any Licensed Product is sold by UGNX or its Affiliates, or KHK or its Affiliates or other Selling Party, as applicable, in or to such country, and (b) with respect to each country in the [***], the period of time commencing on the [***] and continuing for as long as any Licensed Product is sold by KHK or its Affiliates or other Selling Party, as applicable, in any country of the [***].

1.1.73 “**Specifications**” means the specifications of the Licensed Products and the active pharmaceutical ingredient form of the Drug Substance (“**API**”), as determined and updated based on KHK’s actual specifications thereof, that include the minimum shelf life therefor and the tests, references to analytical procedures, and appropriate acceptance criteria that (a) are numerical limits, ranges, or other criteria for the tests, analytical procedures and other criteria described, and (b) establish the set of criteria to which the Licensed Product or the API should conform when tested by the tests, analytical procedures and acceptance criteria listed in the Specifications.

1.1.74 “**Sublicensee**” means a Third Party to whom KHK or any of its Affiliates has granted a license or a sublicense under the Licensed Technology to Develop and/or Commercialize Licensed Products in the Field, but excluding, for clarity, any Third Party distributor that has no rights other than to resell the Licensed Products, and for which resale KHK, its Affiliates or Sublicensees, as applicable, receive no further consideration (such as royalties) beyond the price for the initial sale to the distributor.

1.1.75 “**Tax**” or “**Taxes**” means any taxes, assessments or the like on income, profits, gross receipts, net proceeds, sales, value-added, ad valorem, withholding, or other such taxes or governmental charges.

1.1.76 “**Territory**” means the Profit Share Territory and Latin America, except for any country removed from the Territory in accordance with the express terms and conditions of Section 15.3.

1.1.77 "**Third Party**" means a Person other than KIM, UGNX or their respective Affiliates.

1.1.78 "**Type-C Meeting**" means a meeting (other than a Type A or Type B meeting) between the FDA and a sponsor or applicant regarding the development and review of a product in a human drug.

1.1.79 "**UGNX Non-Core Development Plan**" means UGNX's plan for the UGNX Non-Core Development Activities.

1.1.80 "**UGNX Invention**" means any invention or discovery that relates to a Licensed Product and that is conceived, made or generated during the Term in the performance of activities undertaken pursuant to this Agreement solely by employees, agents, or independent contractors of UGNX or its Affiliates (including any enhancement or modification of a Licensed Product's use, dosage form or formulation).

1.1.81 "**UGNX Regulatory Data**" means all Regulatory Filings made by or on behalf of UGNX with respect to the Licensed Products and all data generated by or on behalf of UGNX in the performance of UGNX Core Development Activities or Regulatory Activities, including any information contained in any Regulatory Filings with respect to Licensed Products and the results of, and all information generated in connection with, any UGNX Core Development Activities or Regulatory Activities performed by or on behalf of UGNX pursuant to this Agreement. UGNX Regulatory Data shall be jointly owned by UGNX and KIM and used in accordance with Section 4.8.1.

1.1.82 "**UGNX Trademark**" means any trademark or trade name, and registrations and applications therefor, owned or Controlled by UGNX in the Territory and covering UGNX's (or its Affiliate's) corporate name or company logo.

1.1.83 **"United States**" or "**U.S.**" means the United States of America and its territories and possessions.

1.1.84 "**XLH**" means x-linked hypophosphatemic rickets/osteomalaica.

1.1.85 "**001 and 002 Studies**" means the Phase 1/2 study (KRN23-INT-001) and the Phase 1/2 extension study (KRN23-INT-002) in adult XLH patients undertaken by KHK as of the Effective Date.

1.2 **Additional Definitions**. In addition, each of the following definitions will have the respective meanings set forth in the Section of this Agreement indicated below:

Definitions	Section
Additional Indication	4.5
Agreement	Preamble
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1.3 **Financial Terms**. All financial and accounting terms not otherwise defined in this Agreement, whether capitalized or not, will have the meanings assigned to them in accordance with GAAP.

ARTICLE 2.
LICENSES

2.1 **Licenses Regarding Licensed Technology**.

2.1.1 **Licenses for Profit Share Territory**. Subject to the terms and conditions of this Agreement, KHK hereby grants to UGNX, without right to sublicense: (a) an exclusive (except for KHK and its subcontractors, solely to the extent necessary to exercise KHK's rights and to perform KHK's obligations under this Agreement), royalty-free license under the Licensed Technology to Develop the Licensed Products in the Field in the Profit Share Territory until the later of (i) the day immediately preceding the Profit Share Territory Transition Date and (ii) if applicable, the completion of the UGNX Core Development activities in the Profit Share Territory under this Agreement; and (b) subject to the restrictions in Section 6.1.2, a non-exclusive license, royalty-free under the Licensed Technology to promote the Licensed Products in the Field in the Profit Share Territory until the end of the Term.

2.1.2 **Licenses for Latin America**. Subject to the terms and conditions of this Agreement, KHK hereby grants to UGNX and its Affiliates, without right to sublicense, an exclusive, royalty-bearing (as described in Section 7.2.3) license under the Licensed Technology to Develop, Commercialize, import and export the Licensed Products in the Field in Latin America until the end of the Term.

2.1.3 **Licenses for European Territory**. Subject to the terms and conditions of this Agreement, KHK hereby grants to UGNX a non-exclusive, royalty-free license under the Licensed Technology to Develop the Licensed Products in the Field in the European Territory, on a country-by-country basis, until the later of (a) the day immediately preceding the applicable European Transition Date and (b) if applicable, the completion of the UGNX Core Development activities in the European Territory under the Agreement.

2.1.4 **In-Licenses**. KHK shall not terminate or breach the In-Licenses (or otherwise cause them to be terminated), or amend the In-Licenses in any way that could reasonably be expected to materially conflict or adversely affect the performance of the activities contemplated by or rights granted under this Agreement. For clarity, this Section 2.1.4 shall be subject to Section 17.2 (Force Majeure).

2.2 **No Sublicense Rights**. Subject to Section 13.6, it is agreed between the Parties that UGNX shall exercise the licensed rights granted hereunder and that UGNX shall perform the Development and Commercialization authorized under this grant of license. Accordingly, no sublicense rights are granted hereunder to UGNX, provided that for the avoidance of doubt, UGNX may (a) subject to the obligation to inform the JDC about the engagement of CROs (Contract Research Organizations), use subcontractors for specific functions in connection with Development

and/or Commercialization of Licensed Products in accordance with the terms of the Agreement (including Section 17.10) and/or (b) Commercialize Licensed Products in accordance with the terms of the Agreement through commercially reasonable distributors and/or resellers.

2.3 **KHK Trademarks.**

2.3.1 **License**. Subject to the terms and conditions of this Agreement, KHK hereby grants to UGNX a non-exclusive (except in Latin America, where such license shall be exclusive in the Field) license to use the KHK Trademarks solely in connection with UGNX's exercise of the licenses granted to it pursuant to Section 2.1 above. UGNX shall use the KHK Trademarks: (a) solely in the manner specified in this Agreement in connection with Licensed Products and not for any other goods or services; and (b) in each case, with KHK's Trademark above UGNX's Trademark if aligned vertically or to the left of UGNX's Trademark if aligned horizontally and otherwise only in the form and manner as reasonably prescribed in writing to UGNX in advance from time to time by KHK (provided, however, that UGNX shall have a reasonable period of time to modify any of its promotional, marketing, regulatory or other practices, including in light of Applicable Laws, and/or cease use of the KHK Trademarks, as may be reasonably necessary to comply with any such form and manner prescriptions and/or any changes thereto). Without limiting the foregoing, any use by UGNX of a KHK Trademark for Licensed Products should be accompanied by a trademark notice that states that such KHK Trademark is a trademark (or a registered trademark, if applicable) of Kyowa Hakko Kirin Co., Ltd. Any use by UGNX of the KHK Trademarks, and KHK's maintenance of the KHK Trademarks, shall be in compliance with all Applicable Laws, including those relating to the licensing of trademarks, in the Territory and the European Territory. UGNX agrees to promptly correct any failure to comply with this Section 2.3 (subject to the cure period provided for in Section 15.2.1).

2.3.2 **No Ownership of KHK Trademarks**. UGNX acknowledges KHK's ownership of all right, title and interest in and to the KHK Trademarks, and agrees that it will do nothing inconsistent with such ownership, that all use of the KHK Trademarks by UGNX will inure to the benefit of and be on behalf of KHK, and that any goodwill associated with the use of any KHK Trademark by UGNX will inure to the benefit of KHK. UGNX agrees that nothing in this Agreement will give UGNX any right, title or interest in the KHK Trademarks other than the right to use the KHK Trademarks in accordance with this Agreement. Anything in this Agreement to the contrary notwithstanding, if by virtue of UGNX's use of the KHK Trademarks, UGNX acquires any equity, title or other rights in or to the KHK Trademarks, UGNX hereby agrees all such equity, title or other rights in or to the KHK Trademarks belong to KHK upon creation of the value, and UGNX agrees to and hereby does assign and transfer any such KHK Trademark rights to KHK. UGNX agrees not to use or file any application to register any trademark or trade name that is confusingly similar to any KHK Trademark.

2.4 **UGNX Trademarks.**

2.4.1 **License**. Subject to the terms and conditions of this Agreement, UGNX hereby grants to KHK a non-exclusive, royalty-free license to use the UGNX Trademarks solely to promote Licensed Products in the Field in accordance with this Agreement.

2.4.2 **No Ownership of UGNX Trademarks**. KHK acknowledges UGNX's ownership of all right, title and interest in and to the UGNX Trademarks, and agrees that it will do nothing inconsistent with such ownership, that all use of the UGNX Trademarks by KHK will inure to the benefit of and be on behalf of UGNX, and that any goodwill associated with the use of any UGNX Trademark by KHK will inure to the benefit of UGNX. KHK agrees that nothing in this

Agreement will give KHK any right, title or interest in the UGNX Trademarks other than the right to use the UGNX Trademarks in accordance with this Agreement. Anything in this Agreement to the contrary notwithstanding, if by virtue of KHK's use of the UGNX Trademarks, KHK acquires any equity, title or other rights in or to the UGNX Trademarks, KHK hereby agrees all such equity, title or other rights in or to the UGNX Trademarks belong to UGNX upon creation of the value, and KHK agrees to and hereby does assign and transfer any such UGNX Trademark rights to UGNX. KHK agrees not to use or file any application to register any trademark or trade name that is confusingly similar to any UGNX Trademark.

2.5 **Use of KHK Trademarks and UGNX Trademarks.**

2.5.1 **Product Packaging**.

(a) Subject to Section 2.3 and the requirements of Applicable Laws, for so long as UGNX promotes Licensed Products to Medical Geneticists, UGNX may request KHK to include one or more UGNX Trademarks on the product packaging for all Licensed Products in the Field to be Commercialized in the Profit Share Territory, and KHK shall use Commercially Reasonable Efforts to include such UGNX Trademarks in an appropriate manner.

(b) Subject to Section 2.3 and the requirements of Applicable Laws, UGNX may include one or more UGNX Trademarks on the product packaging for all Licensed Products in the Field to be Commercialized in Latin America.

2.5.2 **Marketing Materials**.

(a) Subject to Section 2.3 and the requirements of Applicable Laws, UGNX shall have the right to use one or more UGNX Trademarks on Marketing Materials to be used (i) in the Profit Share Territory through the day immediately preceding the Profit Share Territory Transition Date and (ii) in Latin America, and UGNX shall reasonably consider any request by KHK to include one or more KHK Trademarks on such Marketing Materials.

(b) Subject to Section 2.4 and the requirements of Applicable Laws, KHK shall have the right to use one or more KHK Trademarks on Marketing Materials to be used in the Profit Share Territory beginning on the Profit Share Territory Transition Date and in the European Territory, and shall reasonably consider any request by UGNX to include one or more UGNX Trademarks on such Marketing Materials.

2.6 **Product Trademarks**. The Parties will jointly select the Product Trademarks for use in connection with the Commercialization of the Licensed Products in the Territory and the European Territory in the Field (which Product Trademarks may or may not be the same as the names, marks and logos used for the Licensed Products in the Rest of the World) provided, however, that in the event of a dispute with respect to such selection, KHK will have the authority to make any final decision. Furthermore, KHK will be responsible for (and will control) the filing, prosecution, maintenance and defense of all registrations of the Product Trademarks in the Territory and the European Territory, and will be solely responsible for the payment of any costs incurred by KHK relating to filing, prosecution, maintenance, defense and enforcement of the Product Trademarks in the Territory and the European Territory. KHK hereby grants to UGNX a non-exclusive license to use the Product Trademarks in the Territory solely in connection with UGNX's exercise of the licenses granted to it pursuant to this Article 2 or otherwise in accordance with this Agreement. The use of the Product Trademarks by UGNX in the Profit Share Territory will be

royalty-free and the royalties for use of the Product Trademarks by UGNX in Latin America will be deemed to be included within the royalty amounts set forth in Section 7.2.3. In the event that UGNX wishes to use any trademark(s) owned by UGNX as Product Trademarks for any Licensed Products, following the Parties' mutual agreement to use such trademark(s), UGNX shall transfer the rights to such trademark(s) free-of-charge to KHK and KHK shall license such trademark(s) to UGNX for use in accordance with this Agreement.

2.7 **Reservation of Rights.**

2.7.1 **Development and Commercialization**. Except as expressly set forth in this Agreement, all rights to Develop and Commercialize the Licensed Products will be retained by KHK. For the avoidance of doubt, subject to the terms of this Agreement, KHK retains the exclusive right to Commercialize the Licensed Products in the European Territory (including through commercially reasonable distributors and/or resellers) and the exclusive right to Develop and Commercialize the Licensed Products in Japan. In the event KHK decides to license or sublicense its right to Develop and/or Commercialize the Licensed Products in the European Territory, (a) each license or sublicense granted by KHK shall be subject to the terms of this Agreement, and KHK shall remain responsible for compliance with the relevant terms of this Agreement by each Sublicensee, and (b) KHK shall provide to UGNX a copy of each license and sublicense agreement within thirty (30) days of its execution, which copy may be reasonably redacted to exclude confidential information of the applicable Sublicensee.

2.7.2 **Licensed Technology, Confidential Information and Regulatory Data**. Except for the rights and licenses specifically granted in this Agreement, KHK reserves all rights to the Licensed Technology, Confidential Information of KHK, KHK Regulatory Data and any regulatory filings owned or held by KHK, and, except for such specifically granted rights and licenses, this Agreement does not include the grant of any right or license, express or implied, to any other intellectual property or other rights owned or Controlled by KHK. Notwithstanding any provision to the contrary, KHK retains all rights under the Licensed Technology, Confidential Information of KHK, KHK Regulatory Data and any regulatory filings owned or held by KHK, as may be required for KHK to perform its express obligations under this Agreement.

ARTICLE 3.
JOINT STEERING COMMITTEE

3.1 **Joint Steering Committee.**

3.1.1 **Establishment and Membership**. Within thirty (30) days of the Effective Date, the Parties shall establish a joint steering committee (the "**JSC**"**)** to coordinate and oversee the Development and Commercialization of the Licensed Products as contemplated by this Agreement.

3.1.2 **Purpose of JSC**. The purposes of the JSC will be: (a) to keep each Party informed about the manufacturing, Development and Commercialization of the Licensed Products; (b) to (i) review any updates or amendments to the Core Development Plan and (ii) approve any material updates or amendments to the Core Development Plan, including any updates or amendments to budgets; (c) to review (but not approve) the Marketing Plans (including monthly sales forecasts) in the Profit Share Territory and the European Territory, and any proposed material updates or amendments proposed thereto; (d) to review and approve the Marketing Budgets in the Profit Share Territory in accordance with Section 6.1.7; (e) to review (but not approve) the

Marketing Plans of each of KHK and UGNX outside the Profit Share Territory and the European Territory, and the Non-Core Development Plans, as well as any proposed material updates or amendments thereto, including the overall strategy for development activities (including proposed pricing, and forecasts); (f) to review the overall strategy for clinical and commercial manufacturing of the Licensed Products, including plans for packaging, labeling, supply chain and trade and distribution activities for the Profit Share Territory and the European Territory, and risk mitigation strategies related thereto. UGNX and KHK agree to carry out all Development of Licensed Products in the Field in the Territory and the European Territory in consultation with the JSC and the JDC, and (g) to (i) provide a forum for the Parties to discuss the working relationship between the Parties' respective personnel, and (ii) provide recommendations to address any issues arising therefrom.

3.1.3 **Membership**. Each Party shall designate an equal number of representatives, to be three (3) each unless the Parties agree otherwise, with appropriate expertise to serve as members of the JSC, and each of such representatives will be an employee of such Party and will have sufficient seniority within the applicable Party to make decisions arising within the scope of the JSC's responsibilities. Subject to the foregoing, each Party may replace its representatives on the JSC at any time upon at least ten (10) days advance Notice to the other Party. The JSC may change its size from time to time by mutual consent of its members.

3.1.4 **Administrative Chair**. One member of the JSC will serve as the administrative chair of the JSC. The administrative chair will be responsible for organizing meetings and preparing and circulating an agenda in advance of each meeting of the JSC and preparing minutes of each meeting, to be approved by both Parties. For the first year, the Party at whose offices the meetings are held will appoint the administrative chair of the JSC. Thereafter, the Parties will alternate in appointing the administrative chair for one (1) year terms.

3.2 **Subcommittees.**

3.2.1 **Establishment**. The JSC may establish and disband the JDC, the JCC and such other subcommittees as deemed necessary by the JSC, provided that neither the JDC nor the JCC may be so disbanded before the later of (a) the end of the Profit-Sharing Period and (b) the end of the last to occur European Transition Date. Each such subcommittee will consist of the same number of representatives designated by each Party, which number will initially be three (3) each, and thereafter shall be as is mutually agreed by the Parties. Each Party will be free to change its representatives on Notice to the other or to send a substitute representative to any subcommittee meeting; provided, however, that each Party shall ensure that at all times during the existence of any subcommittee, its representatives on such subcommittee are appropriate in terms of expertise and seniority for the then-current stage of Development and/or Commercialization of the Licensed Products in the Field in the Territory and the European Territory and have the authority to bind such Party with respect to matters within the purview of the relevant subcommittee. Each Party's representatives and any substitute for a representative will be bound by the obligations of confidentiality set forth in Article 11. Except as expressly provided in this Agreement, no subcommittee will have the authority to bind the Parties hereunder, and each subcommittee will

report to, and any decisions will be made by, the JSC. The initial subcommittees of the JSC will be the Joint Development Committee ("**JDC**") and the Joint Commercialization Committee ("**JCC**"). The JSC, JDC and JCC shall also be referred to as, each, a "**Committee**".

3.2.2 **Joint Development Committee**.

(a) The JDC will oversee Development of Licensed Products in the Field in the Territory and the European Territory. As soon as practicable following the Effective Date (but in no event more than thirty (30) days following the Effective Date), each Party shall designate its initial three (3) representatives on the JDC, and each of such representatives will be an employee of such Party and will have sufficient seniority within the applicable Party to make decisions arising within the scope of the JDC's responsibilities.

(b) Each Party shall appoint a person from among its representatives on the JDC to serve as a co-chairperson of the JDC. The co-chairpersons will not have any greater authority than any other representative on the JDC and shall conduct the following activities of the JDC: (i) calling meetings of the JDC; (ii) preparing and issuing minutes of each such meeting within thirty (30) days thereafter; (iii) preparing and circulating an agenda for the upcoming meeting; and (iv) ensuring that any decision-making delegated to the JDC is carried out in accordance with Section 3.5.

(c) The JDC will have responsibility for: (i) overseeing, reviewing and coordinating the Development of the Licensed Products (including all Clinical Trials and reviewing the protocols, statistical analysis plans and clinical study reports for such Clinical Trials developed by the applicable Lead Development Party) in the Field; (ii) developing and submitting the Core Development Plan, including the budget and proposed amendments or updates thereto, to the JSC; and (iii) as applicable, overseeing, reviewing and coordinating the work being done under the Core Development Plan and any Non-Core Development Plan(s).

(d) UGNX shall periodically, and at least semi-annually, submit comprehensive and complete reports (i) to the JDC, regarding activities previously approved by the JSC and undertaken by or on behalf of UGNX with respect to the Development of Licensed Products in the Field, including their progress, status and outcome as well as major findings and major decision points, as applicable, so as to keep the JDC fully advised of UGNX's Development activities with respect to the Licensed Products in the Field and (ii) to the JCC with respect to the Commercialization of Licensed Products in the Field, including Sales, Promotion and Marketing Activities, and the date of the First Commercial Sale in a country (if applicable) in the relevant half year.

(e) KHK shall periodically, and at least semi-annually, submit reasonably detailed reports (i) to the JDC, regarding activities undertaken by KHK with respect to KHK's Development activities and (ii) to the JCC with respect to KHK's Commercialization activities.

3.2.3 **Joint Commercialization Committee**.

(a) The JCC will oversee the Commercialization of the Licensed Products in the Field in the Territory and the European Territory. Prior to the commencement of the Commercialization of the Licensed Products in the Territory or the European Territory, at a time agreed to by the Parties, each Party shall designate its initial three (3) representatives on the JCC, and each of such representatives will be an employee of such Party and will have sufficient seniority within the applicable Party to make decisions arising within the scope of the JCC's responsibilities.

(b) Each Party shall appoint a person from among its representatives on the JCC to serve as a co-chairperson of the JCC. The co-chairpersons will not have any greater authority than any other representative on the JCC and shall conduct the following activities of the JCC: (i) calling meetings of the JCC; (ii) preparing and issuing minutes of each such meeting within thirty (30) days thereafter; (iii) preparing and circulating an agenda for the upcoming meeting; and (iv) ensuring that any decision-making delegated to the JCC is carried out in accordance with Section 3.5.

(c) The JCC will have responsibility for: (i) overseeing the Commercialization of the Licensed Products; (ii) setting overall strategic objectives and plans (including pricing and reimbursements) related to Commercialization of the Licensed Products in the Field in the Territory and the European Territory; (iii) developing and submitting to the JSC for review the annual Marketing Plan in the Territory at least ninety (90) days prior to the start of each Calendar Year; (iv) developing and submitting to the JSC for review and approval the annual Marketing Budget in the Profit Share Territory in accordance with Section 6.1.7; (v) reviewing marketing plans for the European Territory and proposed material amendments or updates thereto submitted to the JCC by KHK in accordance with Section 6.1.8; (vi) reviewing Commercialization issues for the Licensed Products in the Field in the Territory and the European Territory; (vii) providing a forum for the Parties to discuss the Commercialization of the Licensed Products in the Field in the Territory and the European Territory; and (viii) such other responsibilities as may be assigned to the JCC pursuant to this Agreement or as may be mutually agreed upon by the Parties from time to time.

3.3 **Committee Meetings**. Each Committee shall meet at least once each Calendar Quarter, or more or less often as agreed to by the Parties; provided that either Party may also call a special meeting of any Committee on an *ad hoc* basis upon at least seven (7) days' prior Notice in order to address urgent matters which cannot be reasonably postponed until the next meeting of such Committee. Meetings may be held in person or by means of electronic communication (including telephone, video or web conferences). Each Committee may invite non-members (including consultants and advisors of a Party who are under an obligation of confidentiality consistent with this Agreement) to participate in the discussions and meetings of such Committee, as non-voting observers; provided, however, that all such Third Party attendees at such meetings must be subject to obligations of confidentiality and non-use applicable to the Confidential Information of each Party that are at least as stringent as those set forth in Article 11. Following any Committee meeting, the administrative chair, in the case of the JSC, and a co-chairperson, on an alternating basis, in the case of the JDC or JCC, will be responsible for preparing and issuing minutes of such meeting within thirty (30) days thereafter. Such minutes will not be finalized until a representative of the other Party has reviewed and confirmed the accuracy of such minutes in writing. If a disagreement regarding the accuracy of such minutes cannot be resolved, the minutes will reflect such disagreement.

3.4 **Limitations of Powers**. The JSC and the subcommittees will have only such powers as are specifically delegated to them hereunder and will not be a substitute for the rights of the Parties. Without limiting the generality of the foregoing, neither the JSC nor any subcommittee will have any power to amend, modify or waive compliance with this Agreement (without limiting the right of the JSC to approve amendments to the Core Development Plan) and the JSC and subcommittees are otherwise subject to the express terms and conditions of this Agreement. Any amendment to the terms and conditions of this Agreement may only be implemented pursuant to Section 17.7 below.

3.5 **Decision-Making.**

3.5.1 **Actions by Unanimous Vote**. Subject to the terms of this Section 3.5, the JSC and the subcommittees will make decisions by unanimous vote with each Party having a single vote, irrespective of the number of representatives actually in attendance at a meeting, or by a written resolution signed by the designated representatives to the JSC or a subcommittee, as applicable, of each of the Parties. For the avoidance of doubt, the approval of all Development budgets and Marketing Budgets and any updates or amendments thereto by the JSC will require the unanimous vote with each Party having a single vote. The JSC members shall use good faith efforts to reach agreement on any and all matters submitted to the JSC. If the JSC fails to reach unanimous consent on a particular matter within thirty (30) days of a Party having requested a formal vote on such matter (or, if such matter is urgent, within seven (7) days of such request) or within thirty (30) days of referral of a matter from a subcommittee, then such dispute will be subject to the resolution procedures described in Sections 3.5.2 and 3.5.3.

3.5.2 **Referral of Disputes for Resolution**. If any subcommittee fails to reach unanimous agreement on a matter after it is presented for decision for a period in excess of thirty (30) days (or, if such matter is urgent, within seven (7) days of such request), the matter will be referred to the JSC. Unless the JSC decides to continue discussing the issue, the JSC will have meetings within thirty (30) days of receiving the dispute submission to reach unanimous consensus on a resolution. If the JSC is unable to reach a resolution of any matter referred up to the JSC (either after the first meeting or any mutually agreed continued discussions), then the matter shall be promptly discussed by the senior executives of the Parties.

3.5.3 **Final Resolution of Disputes**. In the event that any dispute within the JSC or a subcommittee is not resolved pursuant to the terms of Sections 3.5.1 and 3.5.2 then final decision-making authority will be as follows: (a) if the matter pertains to a dispute with respect to [***], then the dispute shall be resolved by [***]; provided, however, that [***] will have the authority to make the final decision (except that increases to the unanimously approved budget shall not exceed [***] of the [***] budget without KHK's prior written consent) until the earlier of (i) [***] and (ii) [***] (subject to extensions, if any, applicable pursuant to Sections 15.3.1(b) or 15.3.1(c)(i)) and that [***] will have the authority to make the final decision in the [***] on and after the [***] and in the [***] on and after the [***] except with respect to the [***]; (b) if the matter pertains to a dispute with respect to [***] in the [***], then [***] will have the authority to make the final decision in the [***] through the day immediately preceding the [***], and [***] will have the authority to make the final decision in the [***] beginning on the [***]; (c) if the matter pertains to a dispute with respect to [***] in the [***], then the dispute shall be resolved by [***] through the day immediately preceding the [***] (subject to Section 6.1.7(b)), and, beginning on the [***], [***] will have the authority to make the final decision with respect to such dispute in the [***]; (d) if the matter pertains the [***], then [***] will have the authority to make the final decision; provided that in each case such decision will be consistent with Applicable Laws; and (e) if the matter pertains to the [***], then [***] will have the authority to make the final decision, provided that in each case such decision will be consistent with Applicable Laws.

3.5.4 **Disputes regarding this Agreement**. Notwithstanding this Section 3.5, any dispute regarding the interpretation of this Agreement, the performance or alleged nonperformance of a Party's obligations under this Agreement, or any alleged breach of this Agreement will be resolved in accordance with the terms of Article 16 and will not be subject to resolution by the JSC.

3.6 **Expenses**. Each Party will be responsible for all of its own travel and other costs and expenses for its respective Committee members, designees and non-JSC or subcommittee invitees to attend meetings of, and otherwise participate on, the JSC and any subcommittees or working groups.

3.7 **Relationship of Parties**. Nothing contained in this Agreement shall be deemed to make any member of the JSC or any subcommittee (or any other committees or sub-teams of thereof) a partner, agent or legal representative of the other Party, or to create any fiduciary relationship for any purpose whatsoever. Except as may be explicitly provided in this Agreement, no member of the JSC, any subcommittee (or any other committee or sub-team of thereof) will have any authority to act for, or to assume any obligation or responsibility on behalf of, any other member of the JSC, any subcommittee (or any other committee of sub-team of thereof) of the other Party. For the avoidance of doubt, this Agreement is not intended to create and may not be construed to create a partnership, joint venture, or entity of any kind between the Parties for any legal purpose and/or for any federal, state or local income tax purposes in any jurisdiction in which either of the Parties is resident or may be subject to taxation. The Parties agree not to treat the Agreement as giving rise to a partnership for any U.S., Japanese or other tax purposes, unless otherwise required by a determination of a taxing authority.

ARTICLE 4.
DEVELOPMENT OF PRODUCTS

4.1 **General**. Pursuant to and subject to the terms of this Agreement, the Parties agree to collaborate with respect to the Development of Licensed Products for the First Indication as well as for other indications in the Field, in accordance with the terms set forth in this Article 4.

4.2 **Core Development Plan.**

4.2.1 The Parties shall prepare in writing an overall Development plan and budget (as such plan and budget may be amended from time to time in accordance with this Agreement, the "**Core Development Plan**") covering the entire Development period and the Development activities and costs required in order to obtain and maintain the Marketing Approvals and (if applicable) the Pricing and/or Reimbursement Approvals for the Licensed Products (including Phase 4 Clinical Trials, if applicable) for the First Indication in the Profit Share Territory and the European Territory (such activities, collectively, "**Core Development Activities**"). The Parties acknowledge and agree that it is their intent to seek Marketing Approval in the First Indication for a label that is as broad as reasonably possible (including, for clarity, broad use by age), taking into account, among other things, the requirements of Applicable Laws and the interest in making Licensed Products in the Field commercially available in a timely manner. In addition to Clinical Trial(s) designed to obtain Marketing Approval for pediatric patients from the age of five (5) through the age of eighteen (18), unless otherwise agreed upon in writing by the Parties, the Core Development Plan shall include a Clinical Trial for pediatric patients below the age of 5. For clarity, the Core Development Plan and Core Development Activities shall cover the 001 and 002 Studies. The initial Core Development Plan has been mutually agreed in writing by the Parties as of the date of signing this Agreement, and shall be the operative Core Development Plan until amended with the approval of the JSC.

4.2.2 With the exception of the 2013 and 2014 Budgets (each as defined below), each year by [***], the Parties shall prepare and approve a detailed annual plan and budget covering the portion of the Core Development Plan and Core Development Activities that

will be performed during the [***] months of the following Calendar Year. The detailed annual plan and budget covering the portion of the Core Development Plan and Core Development Activities that will be performed for the period from the Effective Date until December 31, 2013 and that has been agreed upon by the Parties in writing as of the Effective Date (the "**2013 Budget**"). The detailed annual plan and budget covering the portion of the Core Development Plan and Core Development Activities that will be performed during Calendar Year 2014 (the "**2014 Budget**") shall be prepared and updated by the Parties by [***], 2013. The annual plan and budget will also include an allocation of Development activities between the Parties including the number of allocated full time equivalent personnel and the applicable FTE Rate and other expenses to be incurred by each Party during such period.

4.3 **Core Development Activities.**

4.3.1 Subject to the terms of this Agreement and the requirements of Applicable Laws, and except as otherwise mutually agreed upon in writing by the Parties, the Parties Development responsibilities shall be allocated as follows:

(a) UGNX shall be the lead Party for the Development of Licensed Products in the Field in the Profit Share Territory and the European Territory ("**Lead Development Party**") for the following activities beginning on the Effective Date (collectively, "**UGNX Core Development Activities**"): (i) [***] conducted in the Profit Share Territory [***]; and (ii) [***] conducted in the European Territory [***]; provided, however, that UGNX shall in each case under (i) and (ii) continue to be the Lead Development Party for any Phase 4 Clinical Trials and/or Clinical Trials for additional indications in the Field (but excluding for clarity Phase 5 Clinical Trials), if any, commenced (Initiation) by [***] in the [***] or the [***] prior to the [***] or the applicable [***], as applicable, until completion of such respective Clinical Trial ("**On-Going Clinical Trials**").

(b) [***] shall be the Lead Development Party for the following activities (collectively, "[*****] Core Development Activities**"): (i) [***]; (ii) [***]; and (iii) [***], all of the Core Development Activities conducted in the [***], until completion of such studies, excluding, for clarity, in the case of (ii) and (iii), any On-Going Clinical Trials.

4.3.2 The Lead Development Party shall use Commercially Reasonable Efforts to conduct (or have conducted) the Core Development Activities allocated to it under this Section 4.3 (i.e., the UGNX Core Development Activities in the case of UGNX and the KHK Core Development Activities in the case of KHK) in accordance with the Core Development Plan and Applicable Laws. The non-Lead Development Party shall provide the Lead Development Party such timely assistance as reasonably requested by the Lead Development Party to enable such Party to perform its obligations and accomplish the activities allocated to such Party under the Core Development Plan. Subject to the terms of this Agreement and the requirements of Applicable Laws, the Lead Development Party shall make all decisions relating to the Day-to-Day Core Development Activities allocated to it under this Section 4.3, including all decisions related to the matters set forth on Schedule 4.3.2, provided such decisions are consistent with the then-current Core Development Plan. In addition, UGNX will update the JDC or KHK as reasonably requested with respect to the UGNX Core Development Activities. The Lead Development Party shall inform the JDC in writing as soon as reasonably practicable about any unforeseen and/or material results, problems, difficulties or issues in connection with its respective Core Development Activities.

4.4 **INTENTIONALLY DELETED.**

4.5 **Additional Indications**. At any time prior to [***] months before the [***], either Party may propose to Develop in the Profit Share Territory Licensed Products for one or more additional indications in the Field (i.e., in addition to the First Indication) by submitting to the JDC a written proposal for the Development thereof, including a proposed work plan, budget and timeline. Upon mutual written agreement by the Parties to Develop such indication(s), the JDC shall update the Core Development Plan to include such indication consistent with Section 4.2. Until the Profit Share Territory Transition Date, UGNX shall use Commercially Reasonable Efforts to Develop at least one additional indication mutually agreed-upon by the Parties pursuant to the foregoing sentence (each such indication, an "**Additional Indication**") and shall complete any On-Going Clinical Trials. For the avoidance of doubt, subject to the terms of this Agreement, on and after the [***].

4.6 **KHK Manufacturing-Related Development Activities**. Subject to the terms of this Agreement and the requirements of Applicable Laws, KHK will retain all rights and responsibility to conduct at its cost (i.e., not subject to profit/loss sharing under Section 7.1.1) all necessary Development activities related to the manufacture and supply of the Licensed Products, including process development, manufacturing scale-up, development-stage and commercial-stage manufacturing, quality assurance/quality control procedure development, and compilation and reporting of CMC information ("**Manufacturing-Related Development Activities**").

4.7 **Initial Information and Regulatory Filings Transfer**. Within a reasonable period of time after the Effective Date and in accordance with the Core Development Plan, KHK shall provide to UGNX (a) the data (tables, listings and figures) and final report on completed Clinical Trial, (b) the interim data (tables, listings and figures) on Phase 1/2 extension study (KRN23-INT-002), and (c) the final reports on completed non-clinical studies related to the Licensed Products. In addition, within a reasonable period of time following the Effective Date, KHK shall transfer to UGNX the registration as IND holder for the Licensed Products in the U.S. and all Regulatory Filings (for the Profit Share Territory, the European Territory and Latin America), including any INDs, CTAs or their equivalents, including all such filings related to the 001 and 002 Studies, in each case solely for the purpose of the Development and Commercialization of the Licensed Products by UGNX as contemplated by this Agreement. For the avoidance of doubt, KHK will have no obligation to translate any such results and other information into English (or any other language).

4.8 **Development Data.**

4.8.1 **Obligations of UGNX**. The results of all Core Development Activities, including all data collected or analyzed with respect thereto, and all study reports analyzing such data (collectively, "**UGNX Clinical Data**") will be jointly owned by UGNX and KHK. Subject to Applicable Laws, UGNX shall provide KHK free-of-charge with copies of UGNX Clinical Data (in electronic form if requested by KHK and in or reasonably convertible to such electronic form). KHK may use UGNX Clinical Data free-of-charge, and KHK and its Affiliates, licensees and commercialization and development partners will have a right of access, a right of reference and a right to use and incorporate all UGNX Clinical Data in any Regulatory Filings for the Licensed Products in accordance with Applicable Laws in the Profit Share Territory, the European Territory and the Rest of the World. For the avoidance of doubt, KHK may provide the foregoing information (and extend the foregoing rights) to its licensees or commercialization and development partners for the purposes specified in this Section 4.8.1 and otherwise in accordance with this Agreement.

4.8.2 **Obligations of KHK**. During the Term, and subject to Applicable Laws, KHK shall provide UGNX free-of-charge with copies of information or data with respect to the Licensed Products (including non-clinical and clinical study data and results and including all data collected or analyzed with respect thereto, and all study reports analyzing such data) that is (or was prior to the Effective Date) generated by KHK or any Affiliates or licensees or commercialization or development partners of KHK, or by any Third Parties acting on their behalf, which is necessary or useful for UGNX to conduct the UGNX Core Development Activities and UGNX Non-Core Development Activities, or to Commercialize Licensed Products in the Field in the Territory (collectively, "**KHK Clinical Data**"). KHK Clinical Data includes the clinical study reports and Regulatory Filings. However, KHK will have no obligation to provide any translation of KHK Clinical Data to UGNX. UGNX and its Affiliates will have a right of access, a right of reference and a right to use and incorporate all KHK Clinical Data in any Regulatory Filings for the Licensed Products in accordance with Applicable Laws solely for the purposes specified in this Section 4.8.2 and otherwise in accordance with the Agreement.

4.9 **Development Costs**. The obligations of the Parties with respect to Development Costs of the Licensed Products are set forth in this Section 4.9.

4.9.1 **Funding of Core Development Activities**. Development Costs for Core Development Activities shall be allocated as follows:

(a) Pre-Transition Funding. UGNX and KHK shall share equally (50/50) the Development Costs for all of the Core Development Activities incurred after the Effective Date through the day immediately preceding the Profit Share Transition Date (in the Profit Share Territory) or the applicable European Transition Date (in the European Territory), including, for clarity, the 001 and 002 Studies and any On-Going Clinical Trials, until completion of such studies.

(b) Post-Transition Funding. Beginning on the Profit Share Transition Date (in the Profit Share Territory) or the applicable European Transition Date (in the European Territory), KHK will be responsible for [***] ([***]%) of the Development Costs for the Core Development Activities conducted in the Profit Share Territory and the European Territory, respectively, excluding, for clarity, any On-Going Clinical Trials.

(c) Manufacturing-Related Development Costs. Beginning on the Effective Date, KHK shall be solely responsible for one hundred percent (100%) of the Development Costs for any and all Manufacturing-Related Development Activities.

4.9.2 **Funding of Development in Latin America**. UGNX shall be solely responsible for one hundred percent (100%) of the Development Costs for Development activities (other than Core Development Activities) with respect to Licensed Products in the Field (including Development Costs for Phase 4 Clinical Trials) conducted in Latin America.

4.9.3 **Funding of Development for Injection Devices**. The Parties shall separately discuss and agree upon development and cost-sharing for the development of injection devices to be used in connection with the Licensed Products.

4.9.4 **Development Cost Reports**. Each Party shall prepare and deliver to the other Party a [***] report detailing its Development Costs incurred during such period with respect to activities covered by the Core Development Plan. Each Party shall submit any supporting information reasonably requested by the other Party related to such Development Costs included in its report within [***] days after its receipt of such request. The Parties shall conduct a reconciliation of such Development Costs aiming toward agreement within [***] days after receipt

of all such supporting information, and an invoice shall be issued for any unpaid share of the Development Costs identified in such reconciliation. Each Party shall pay all amounts due and payable by such Party under any such invoice within [***] days after its receipt of such invoice and neither Party will have a set-off right with respect to amounts payable pursuant hereto. Each Party shall have the right to audit the records of the other Party with respect to any purported Development Costs included in the reports prepared by such Party in accordance with Section 9.2.

4.10 **No Debarred Personnel**. In performing Development activities under this Agreement, neither Party will use the services of any employee or consultant, who has been debarred by the FDA or any Regulatory Authority or is the subject of debarment proceedings by the FDA or any other Regulatory Authority.

4.11 **Non-Core Development Activities.**

4.11.1 As between UGNX and KHK, subject to the terms of this Agreement and the requirements of Applicable Laws:

(a) KHK will be solely responsible and will have sole discretion and control for all non-clinical, clinical and other Development or Commercialization activities (including regulatory activities) with respect to (i) [***] and (ii) [***] (collectively, the "**KHK Non-Core Development Activities**").

(b) UGNX will be solely responsible and will have sole discretion and control for all non-clinical, clinical and other Development or Commercialization activities (including regulatory activities) with respect to Licensed Products in the Field in Latin America (collectively, the "**UGNX Non-Core Development Activities**").

4.11.2 The responsible Party (i.e., KHK for the KHK Non-Core Development Activities, and UGNX for the UGNX Non-Core Development Activities) ("**Responsible Party**"), through the JDC, shall keep the other Party reasonably informed of all material events and developments, occurring in the course of the non-clinical, clinical and other development activities for the Licensed Products. The Responsible Party shall consider in good faith any recommendations that the other Party may make to the Responsible Party regarding such activities; provided, however, that subject to the terms of this Agreement and the requirements of Applicable Laws, the Responsible Party will have the final decision-making authority with respect thereto.

4.11.3 Notwithstanding the foregoing: (a) KHK shall use Commercially Reasonable Efforts not to cause a material adverse effect on the Development and/or Commercialization of Licensed Products in the Field in the Territory or the European Territory; and (b) UGNX shall use Commercially Reasonable Efforts not to cause a material adverse effect on the Development and/or Commercialization of Licensed Products in the Field in the Territory or the European Territory.

4.12 **Regular Development Updates**. The Parties agree to discuss, through appropriate employees in person or by video or teleconference, the current status of the Development of the Licensed Products in the Field in the Territory and the European Territory as reasonably required or requested by either Party. Updates shall include discussions and consultations between the Parties on topics, in each case as applicable, including: (a) protocol drafting and/or finalization; (b) establishment of, or changes to, the Statistical Analysis Plan (SAP); (c) initial investigator selection after the Effective Date; (d) major investigator meetings; (e) Clinical Study Report (CSR) drafting and/or finalization; (f) regulatory strategy with respect to agency interactions; (g) regulatory meeting requests; (h) agency submissions; and (i) agency responses.

ARTICLE 5.
REGULATORY MATTERS

5.1 **Regulatory Activities**. The obligations of the Parties with respect to Regulatory Activities related to the Licensed Products are set forth in this Article 5.

5.1.1 **Regulatory Activities**. Beginning on the Effective Date and to the extent UGNX remains the Lead Development Party with respect to a particular territory, subject to and in accordance with the terms and conditions of this Agreement and the requirements of Applicable Laws, UGNX, shall: (a) use Commercially Reasonable Efforts to file (or have filed) all Regulatory Filings with respect to the Licensed Products in the Field in order to obtain Marketing Approvals in each country in the Territory and the European Territory (or to obtain the European Centralized Approval in the European Core Territory) and in order to obtain Pricing and/or Reimbursement Approvals in the Profit Share Territory; (b) respond in a timely fashion to requests for data and information from Regulatory Authorities with respect to the Licensed Products in the Field in the Territory and the European Territory; and (c) meet with officials of the Regulatory Authorities at such times as may be requested by such Regulatory Authorities with respect to the Core Development Activities ("**Regulatory Activities**"), provided that KHK will have primary responsibility for obtaining, and UGNX shall provide all assistance reasonably requested by KHK, in relation to Pricing and/or Reimbursement Approvals for the Licensed Products in the Field in the European Territory. For the avoidance of doubt, UGNX will be responsible for obtaining, and KHK will provide all assistance reasonably requested by UGNX, in relation to Pricing and/or Reimbursement Approvals, if any, for the Licensed Products in the Field in the Profit Share Territory as part of the UGNX Core Development Activities, it being understood that the costs incurred by UGNX in connection with such activities will be shared equally (50/50). All such Regulatory Activities will be conducted in a manner consistent with the Core Development Plan and coordinated by the JSC in accordance with Article 3. Without limiting the applicability of the foregoing and the remainder of this Article 5, UGNX shall interface with the applicable Regulatory Authority(ies) and, through the JDC, shall keep KHK reasonably informed of all material events and developments occurring in the course of the Regulatory Activities, including scheduled UGNX regulatory strategy discussions and meetings with Regulatory Authorities in the Territory and the European Territory relating to the Licensed Products in the Field.

5.1.2 **Transfer Regulatory Approvals and Data**. Subject to, and in accordance with, the terms and conditions of this Agreement and the requirements of Applicable Laws, UGNX shall take all steps reasonably necessary to transfer and assign the Marketing Approvals for the Licensed Products in the Field in the Profit Share Territory and the European Territory, the Pricing and/or Reimbursement Approvals for the Licensed Products in the Field in the Profit Share Territory and the UGNX Regulatory Data for the Licensed Products in the Field in the Profit Share Territory and the European Territory to KHK within [***] days after UGNX has obtained Pricing and/or Reimbursement Approval (for the Profit Share Territory) and [***] days after UGNX has obtained Marketing Approval (for the European Territory), in each applicable territory or country, or after such other period required by the regulatory process of such territory or country.

(a) UGNX shall transfer to KHK or KHK's designee all Regulatory Filings (including any INDs, CTAs or their equivalents) unless KHK has provided its prior written consent to the retention of any of the foregoing by UGNX until the day immediately preceding the Profit Share Territory Transition Date or for such longer period as necessary for UGNX to perform Development activities under this Agreement in accordance with Applicable Laws and Marketing Approvals and Pricing and/or Reimbursement Approvals owned by UGNX for the Commercialization of Licensed Products in the Field if and to the extent such transfer is possible, or, if such transfer is not possible under Applicable Laws, then UGNX shall, at KHK's discretion: (i) [***]; or (ii) [***].

(b) UGNX shall provide KHK with copies of all material correspondence between UGNX and any Regulatory Authorities with respect to such Regulatory Filings, Marketing Approvals and Pricing and/or Reimbursement Approvals for the Licensed Products in the Field and all other clinical and non-clinical data, records and tabulations, in all such cases with respect to the Licensed Products in the Field.

(c) Promptly after the Profit Share Territory Transition Date in each country in the Profit Share Territory and the applicable European Transition Date in the European Territory, except to the extent UGNX retains responsibility for the conduct of On-Going Clinical Trials for Licensed Products in the Field, UGNX shall assign to KHK all agreements specific to the conduct of such Clinical Trials in the applicable territory or country (to the extent assignable and excluding any such agreements that also involve clinical trials for other UGNX products that are not Licensed Products), including agreements or contracts with contract research organizations, clinical sites and investigators, between UGNX and any Third Party, subject to any consent required by such Third Party, which consent UGNX will use Commercially Reasonable Efforts to obtain on behalf of KHK.

(d) UGNX shall provide KHK with copies of all reports and data obtained by UGNX or its Affiliates pursuant to this Agreement regarding the Development and the Commercialization of the Licensed Products in the Field under this Agreement, including any UGNX Clinical Data.

5.1.3 **KHK Assistance**. Upon request of UGNX, KHK shall use Commercially Reasonable Efforts to assist UGNX in connection with any meetings with, or requests from, Regulatory Authorities in the Territory and the European Territory related to Licensed Products in the Field; provided, however, that the costs in connection with the Core Development Activities shall be shared equally (50/50), the costs in connection with CMC activities shall be [***] and the costs in connection with the UGNX Non-Core Development Activities shall be [***].

5.1.4 **UGNX Assistance**. Upon request of KHK, following the conclusion of the Profit Sharing Period, UGNX shall use Commercially Reasonable Efforts to assist KHK at KHK's cost in connection with any meetings with, or requests from, Regulatory Authorities with respect to Licensed Products in the Field in any country of the world where KHK is the Lead Development Party.

5.2 **Regulatory Data and Approvals.**

5.2.1 **Regulatory Filings**.

(a) Review. The JDC will coordinate communication and the exchange of information between the Parties with respect to Regulatory Filings to be prepared and submitted by or for UGNX for Licensed Products in the Field in the Territory and the European Territory; and without limiting the foregoing, UGNX shall provide KHK with copies of all such Regulatory Filings (including any protocols to be included in such filings) (in English) in the Territory and the European Territory to review prior to filing thereof. It is acknowledged by the Parties that given time constraints, the Regulatory Filing actually submitted by UGNX to a Regulatory Authority may vary from the matters discussed by the Parties in the JDC because of changes resulting from interactions with Regulatory Authorities and from continued work on such filings by UGNX's regulatory personnel.

(b) Copies. Subject to Applicable Laws, UGNX shall provide to KHK: (i) [***]; (ii) [***]; and (iii) [***]. KHK will have a right of access, a right of reference and the right to use and incorporate all UGNX Regulatory Data in connection with the Licensed Products and in a manner consistent with the relevant terms of this Agreement (e.g., outside the Territory except to the extent required to perform any of its obligations hereunder). For the avoidance of doubt, subject to Article 11 and the relevant terms of the Agreement, KHK may provide copies of such Regulatory Filings (and extend its right of access, right of reference and the right to use and incorporate all UGNX Regulatory Data in connection with Licensed Products into regulatory submissions outside the Territory) to its licensees or commercialization or development partners solely for the Commercialization or the Development of Licensed Products. KHK will have no obligation to translate Regulatory Filings outside the Territory into English (or any other language).

5.2.2 **Regulatory Meetings**. UGNX shall provide KHK with advance notice of any formal, scheduled meetings with any Regulatory Authority in the Territory or the European Territory with respect to Licensed Products in the Field (including any meetings related to the final positioning of labeling and safety claims within the original and subsequent regulatory submissions), and UGNX shall provide a brief description of the topics to be presented or discussed at that meeting. Subject to Applicable Laws, UGNX shall allow KHK and/or its Affiliates to attend any such meeting (without any obligation on KHK to do so), and to participate in the meeting preparation process.

5.3 **Provision of Regulatory Information to UGNX**. KHK shall use Commercially Reasonable Efforts to support UGNX in connection with its conduct of all Regulatory Activities by providing relevant documents and information in KHK's possession. Furthermore, KHK shall provide to UGNX manufacturing and CMC information solely for fulfilling regulatory requirements (e.g., for Regulatory Filings) in the Profit Share Territory, the European Territory and Latin America.

5.4 **Safety; Safety Agreement**. Within [***] months of the Effective Date, the JSC will develop a mutually acceptable safety agreement (to be agreed upon and executed by both Parties) setting forth the Parties' respective obligations in detail regarding pharmacovigilance and the exchange of drug safety data for Licensed Products (the "**Safety Agreement**"). The Safety Agreement will include applicable timelines and scope for reporting (including adverse event data collection and analysis) between UGNX and KHK (or their respective Affiliates) that will: (a) enable each Party to comply with its respective reporting requirements to Regulatory Authorities in the Territory and the European Territory and to satisfy its duty of care with respect to the Drug Substance and the Licensed Products as required by Regulatory Authorities in the Territory and the European Territory (it being understood that, following the transfer of the Marketing Approvals and the Pricing and/or Reimbursement Approvals to KHK, KHK shall be responsible for complying with all associated regulatory requirements, including safety reporting requirements); (b) enable KHK to comply with its reporting requirements to Regulatory Authorities outside the Territory, and (c) ensure worldwide safety surveillance.

5.5 **Recalls and Voluntary Withdrawals.**

5.5.1 For each country in the Territory, UGNX and KHK shall, through the JSC, confer and coordinate regarding their respective internal standard operating procedures (and any changes thereto) regarding product recalls and the treatment of and response to product complaints and inquiries as to safety, quality or efficacy that may be relevant to the Licensed Products.

5.5.2 If either Party becomes aware of information about a Licensed Product indicating that it may not conform to the applicable Specifications or that there are potential adulterations, misbranding and/or other material adverse issues regarding safety of a Licensed Product (or otherwise that a recall or withdrawal of a Licensed Product is potentially at issue), such Party shall notify the other Party as soon as practical (but in any event within such period as the Parties may mutually establish in order to ensure their respective compliance with Applicable Laws). With respect to in the Territory, the Parties shall promptly meet to discuss such circumstances and to consider appropriate courses of action, including Licensed Product recalls.

5.6 **Inspection Rights**. Not more than [***] per year, if either Party (the "**Inspecting Party**") has any reasonable concerns regarding the other Party's or its Affiliates' storage, handling or manufacturing of any Licensed Products, the Inspecting Party will have the right, at the Inspecting Party's expense and on not less than [***] days' prior notice, to inspect the facilities where the other Party or its Affiliates store, handle or manufacture, or have stored, handled or manufactured, any Licensed Products and to audit the procedures of such other Party or its Affiliates for the storage, handling and/or manufacturing of Licensed Products for purposes of quality control. The Safety Agreement will contain additional customary mutual provisions addressing inspection rights for cause.

5.7 **Governmental Inspections and Inquiries**. Each Party shall advise the other Party promptly, but in no event later than [***] days after such Party's receipt of notice thereof, of any planned Regulatory Authority visit to the portion of the facilities of the receiving Party or its Affiliates where Licensed Products are Developed, stored or handled or any material written inquiries by a Regulatory Authority concerning such facilities, the procedures of the receiving Party or its Affiliates for the Development, storage or handling of Licensed Products, or the Commercialization of Licensed Products in the Territory or the European Territory, as applicable. If the Regulatory Authority makes an unannounced or unplanned visit, or if the receiving Party does not have at least [***] days notice of the visit, the receiving Party shall inform the other Party of the visit within one (1) Business Day after the receiving Party obtains actual knowledge of the visit. The receiving Party shall inform the other Party as soon as practicable regarding the purpose and result of such visit or inquiry, and shall provide to the other Party copies of any minutes of the inspection generated by the receiving Party promptly following such inspection and any report or correspondence provided by the receiving Party, or any Affiliate, as the case may be, to the Regulatory Authority or issued by or provided by the Regulatory Authority to the receiving Party, or any Affiliate, as the case may be, in connection with such visit or inquiry. The receiving Party shall advise the other Party of the material aspects of such minutes and correspondence at the next JSC meeting.

5.8 **Regulatory Matters in the Rest of the World**. KHK, through the JSC, shall keep UGNX reasonably informed of significant events occurring in the course of the regulatory activities with respect to the Licensed Products in the Rest of the World or any KHK Out-of-Field Products, to the extent relevant to issues pertaining to the Licensed Products in the Territory and the European Territory. UGNX shall provide all reasonable cooperation to KHK with respect to regulatory activities related to the Licensed Products in the Rest of the World including providing access to UGNX's relevant facilities in the event of an inspection by applicable Regulatory Authorities.

ARTICLE 6.
SALES, PROMOTION AND MARKETING; DILIGENCE OBLIGATIONS

6.1 **Sales, Promotion and Marketing Activities**. The obligations of the Parties with respect to Sales, Promotion and Marketing Activities related to the Licensed Products are set forth in this Section 6.1.

6.1.1 **General**. Subject to and in accordance with the terms and conditions of this Agreement and the requirements of Applicable Laws, each Party shall promote, market and sell, as applicable, the Licensed Products in its respective territory (“**Sales, Promotion and Marketing Activities**”) as set forth in this Section 6.1, and shall keep the other informed through the JCC as to plans and activities regarding Commercialization of the Licensed Products. As appropriate, the Parties shall discuss shared global commercialization activities that benefit the Licensed Products both in the Territory and outside the Territory.

6.1.2 **Sales, Promotion and Marketing in the Profit Share Territory**. KHK shall book sales of the Licensed Products in the Field in the Profit Share Territory. During the [***] following the First Commercial Sale in the U.S., UGNX will have the exclusive right and responsibility, and shall use Commercially Reasonable Efforts, to promote the Licensed Products in the Field in the Profit Share Territory in order to maximize sales of the Licensed Products in the Profit Share Territory. During the period of time commencing on [***], KHK will have the right to increasingly participate in the promotion of the Licensed Products in the Profit Share Territory [***], and UGNX shall continue to promote the Licensed Products in the Profit Share Territory and use Commercially Reasonable Efforts in assisting KHK in the transition of the promotion activities for the Licensed Products in the Profit Share Territory. UGNX shall provide all information, documents (including originals or copies, as applicable) and other assistance reasonably requested by KHK in order to allow KHK to market and promote the Licensed Products in the Profit Share Territory as of the Profit Share Territory Transition Date. Beginning on or after the Profit Share Territory Transition Date, KHK will have the exclusive right, and shall use Commercially Reasonable Efforts, to market and promote the Licensed Products in the Field in the Profit Share Territory [***], provided that UGNX will have the right to continue to promote the Licensed Products in the Profit Share Territory [***] using its own sales force with respect to activity targeting Medical Geneticists and UGNX shall use Commercially Reasonable Efforts to promote the Licensed Products to such key subscribers for Medical Geneticists. During the Profit Sharing Period, if KHK fails to use Commercially Reasonable Efforts to sell the Licensed Products in the Field in the Profit Share Territory, KHK will follow reasonable suggestions from UGNX regarding how KHK should satisfy this obligation.

6.1.3 **Sales, Promotion and Marketing in Latin America**. UGNX shall book sales of the Licensed Products in the Field in Latin America. During the Term, UGNX will have the exclusive right and responsibility to sell, promote and market the Licensed Products in the Field in Latin America. Such right shall include the right to sell and otherwise provide Licensed Products as part of Named Patient Sales in each country in Latin America in accordance with Applicable Laws, and shall be responsible for all associated costs.

6.1.4 **Sales, Promotion and Marketing in the European Territory**. KHK shall book sales, sell, market and promote the Licensed Products in the Field in the European Territory.

6.1.5 **Named Patient Sales in the European Territory**. KHK retains the exclusive right and responsibility to sell and otherwise provide the Licensed Products in the European Territory as part of Named Patient Sales, and shall be responsible for all associated costs (i.e., not subject to cost-sharing). For clarity, UGNX shall not have the right to sell or otherwise provide Licensed Products in the European Territory as part of Named Patient Sales.

6.1.6 **Marketing Guidelines**. Without limiting the generality of the other provisions of this Article 6, the Parties shall agree upon guidelines for the marketing and sale of Products in the Territory and the European Territory at least [***] prior to the expected date of the First Commercial Sale of the Licensed Products in the Territory and the European Territory. Such guidelines shall be included in the relevant Marketing Plans and UGNX and KHK shall abide by the guidelines.

6.1.7 **Marketing Plans and Marketing Budgets**.

(a) Marketing Plans. Without limiting the generality of the other provisions of this Article 6, UGNX shall prepare and submit to the JCC for review (but not approval) a plan containing the strategy and proposed activities (described generally) for marketing and selling Licensed Products in each country in the Territory (as updated pursuant to this Section 6.1.7, the "**Marketing Plan**"). UGNX shall submit a proposed draft of the Marketing Plan, including Phase 5 Clinical Trials, samples, supplies provided to patients as part of "compassionate use" programs in the Profit Share Territory to the JCC for review (but not approval) no later than [***] months prior to the anticipated date of the First Commercial Sale of any Licensed Product in the applicable country. The initial Marketing Plan shall cover the period through the day before the Profit Share Territory Transition Date. UGNX shall annually by [***] update the Marketing plan covering the rest of the period through the day before the Profit Share Territory Transition Date and prepare each year a detailed annual Marketing Plan for each country in the Territory covering the following Calendar Year. This will be subject to review (but not approval) by the JSC. All decisions regarding the Sales, Promotion and Marketing Activities within Latin America consistent with the Marketing Plan will be determined solely by UGNX. Sales forecasts with respect to the Profit Share Territory and Latin America shall be made on a [***] updated [***].

(b) Marketing Budgets. Without limiting the generality of the other provisions of this Article 6, UGNX shall prepare and submit to the ICC for review and approval with each Marketing Plan a marketing budget (the "**Marketing Budget**"), which shall set forth the budgeted amounts for Commercialization Costs for Licensed Products in the Profit Share Territory, it being understood that personnel costs will be allocated based on the percentage of time dedicated by such personnel to the Commercialization of such Licensed Products. Such Marketing Budget shall contain an annual breakdown for the period from the date when pre-commercial Commercialization Costs are posted for the first time through the Profit Share Territory Transition Date. At the end of [***] of each year thereafter, UGNX shall update such Marketing Budget and prepare a detailed annual Marketing Budget with a detailed [***] breakdown covering the following Calendar Year in the Profit Share Territory. Notwithstanding foregoing, in the event actual costs and expenses are expected to exceed the annual Marketing Budget, the solution and/or amendment of the Marketing Budget, shall be promptly agreed at the JSC. The initial Marketing Budget and each annual Marketing Budget in the Profit Share Territory will be subject to the unanimous approval of the JSC. In addition, except as otherwise mutually agreed upon by the Parties, headcount for all sales representatives (including medical liaisons) shall not exceed [***]. UGNX shall submit a proposed draft of the Marketing Budget for a country, or countries, in the Profit Share

Territory to the JCC for review and no later than [***] months prior to the anticipated date when pre-commercial Commercialization Costs are posted for the first time for any Licensed Product in such country. If the initial submission timing falls in [***], the proposed draft shall contain [***] breakdown of the following Calendar Year's budget. In the event that the JSC does not unanimously approve the initial Marketing Budget thereafter, pursuant to Section 3.5.1, either Party may initiate procedures to finally resolve the issue pursuant to Section 16.4.

6.1.8 **Marketing Plans for the European Territory**. KHK shall prepare and submit to the JCC for review (but not approval) a plan containing the strategy and proposed activities (described generally) for marketing and selling Licensed Products in each country in the European Territory no later than [***] months prior to the anticipated date of the First Commercial Sale of any Licensed Product in such country. All decisions regarding the Sales, Promotion and Marketing Activities including budget within the European Territory will be determined solely by KHK.

6.1.9 **Product Complaints**. UGNX shall report to KHK in a manner reasonably designated by KHK from time to time all complaints that are received from any direct or indirect purchasers of the Licensed Products from UGNX involving labeling or packaging of the Licensed Products or other problems not related to the medical effectiveness of the Licensed Products and that could reasonably negatively affect the high-quality image of the Licensed Products, KHK or UGNX. Such reports shall include the following information and/or other information as reasonably specified by KHK: (a) the person or entity that initiated the complaint; (b) the nature of the complaint; (c) the geographic area in which the complaint originated; and (d) whether any return samples of the Licensed Product are available.

6.2 **Sales Forecasts**. UGNX shall provide forecasts with respect to its commercial requirements for sale of the Licensed Products in the Territory in accordance with the terms and conditions to be set forth in the Commercial Supply Agreement.

6.3 **Labeling and Patent Rights Marking**. Subject to Applicable Laws, UGNX shall identify KHK as the licensor or manufacturer of the Licensed Products using the KHK Trademarks as designated by KHK for such use in certain mutually agreed promotional materials for Licensed Products in the Territory where such identification is appropriate, in a manner approved in advance in writing by both Parties and in accordance with (and subject to) the trademark license set forth in Section 2.3. To the extent reasonably and customary in the industry for such products, UGNX shall mark all Licensed Product sold by UGNX with appropriate Product Trademarks and patent numbers, to the extent permitted by Applicable Laws in the country within the Territory where such Licensed Products are sold.

6.4 **Marketing Materials.** All marketing and promotional literature related to the Licensed Products for use in the Field in the Territory and the European Territory by the Parties ("**Marketing Materials**") will be prepared in consultation with the JCC in a manner consistent with Applicable Laws, provided that UGNX will have the authority to make any final decision (a) in Latin America and (b) in the Profit Share Territory through the day immediately preceding the Profit Share Territory Transition Date, and KHK shall have the authority to make any final decision (a) in the European Territory and (b) in the Profit Share Territory beginning on the Profit Share Territory Transition Date. In particular, UGNX shall use Commercially Reasonable Efforts in facilitating a smooth transition in preparation for KHK's Sales, Promotion and Marketing Activities in respect of the Profit Share Territory.

ARTICLE 7.
FINANCIAL TERMS

7.1 **Profit-Sharing.**

7.1.1 Profit Sharing Calculation.

(a) UGNX and KHK shall share equally (50/50) in the net income or net loss derived from sales of Licensed Products in the Profit-Sharing Period in the Field in the Profit Share Territory with net income or loss calculated as follows:

(i) From the Effective Date through [***]:

Net Sales – [***]% of [***]

(ii) From [***] through [***]:

Net Sales – [***]% of [***]

7.1.2 **Commercialization Cost Reports**. Within [***] days following the end of each [***], each Party shall submit to the other Party a [***] report detailing its Commercialization Costs incurred during such period with respect to activities covered by the applicable Marketing Plan for the Licensed Products in the Field in the Profit Share Territory on a country-by-country basis. Each Party shall submit any supporting information reasonably requested by the other Party related to such Commercialization Costs included in its report within [***] days after its receipt of such request. The Parties shall conduct a reconciliation of such Commercialization Costs aiming to reach agreement within [***] days after receipt of all such supporting information, and an invoice will be issued for any unpaid share of the Commercialization Costs identified in such reconciliation. Each Party will have the right to audit the records of the other Party with respect to any purported Commercialization Costs included in the reports prepared by the other Party in accordance with Section 9.2.

7.1.3 **Profit Sharing Period**. The profit-sharing period for the Licensed Products in the Field in the Profit Share Territory means the period commencing on the First Commercial Sale in the U.S. and ending on the day immediately preceding the [***] anniversary of the First Commercial Sale in the U.S. **(**"**Profit-Sharing Period**").

7.1.4 **Pre-Commercial Activity Cost Sharing**. Development Costs will be funded and shared as set forth in Section 4.9. The Parties shall share equally (50/50) all reasonable, mutually agreed upon in writing, pre-commercial Commercialization Costs (other than Development Costs) incurred by UGNX or KHK, if any, in the Field in the Profit Share Territory.

7.2 **Sales Royalties and Revenue Share.**

7.2.1 **Royalties Payable to UGNX in the European Territory.** Subject to the terms and conditions of this Agreement, KHK shall pay royalties to UGNX for each Calendar Year during the applicable Royalty Term in an amount equal to [***] percent ([***]%) of Net Sales of any Licensed Products in the Field in a country in the European Territory ("**UGNX Sales**

Royalties in the European Territory"), provided that, if KHK or any of its Affiliates or Sublicensees obtains Marketing Approval for an additional indication in the Field, the payment of the UGNX Sales Royalties in such country for such additional indication shall be conditioned on UGNX having obtained the Marketing Approval in such country in the First Indication.

7.2.2 **Revenue Share Payable to UGNX in the Profit Share Territory.** Subject to the terms and conditions of this Agreement, KHK shall pay a revenue share to UGNX for each Calendar Year on Net Sales of any Licensed Products in the Field at the rates set forth in this Section 7.2.2 during the applicable Royalty Term in the Profit Share Territory ("**UGNX Revenue Share in the Profit Share Territory**"), provided that, if KHK or any of its Affiliates or Sublicensees obtains Marketing Approval for an Additional Indication, the payment of the UGNX Revenue Share in a country in the Profit Share Territory for such Additional Indication shall be conditioned on UGNX having obtained the Marketing Approval in such country in the First Indication. For the avoidance of doubt, Section 7.1 will not apply in the Profit Share Territory during such Royalty Term.

Net Sales (US$)	Revenue Share (% of Net Sales)
Portion of Net Sales less than or equal to [***]	[***]%
Portion of Net Sales more than [***] but less than or equal to [***]	[***]%
Portion of Net Sales in excess of [***]	[***]%

7.2.3 **Royalties Payable to KHK**. Subject to the terms and conditions of this Agreement, UGNX shall pay royalties to KHK for each Calendar Year during the applicable Royalty Term in an amount equal to [***] percent ([***]%) of the Net Sales of the Licensed Products in the Field in Latin America on a country-by-country basis ("**KHK Sales Royalties**").

7.2.4 **Payments and Reports**. Each royalty or revenue share payment will be non-refundable and non-creditable against any other payments due hereunder. Each Party will make royalty or revenue share payments contemplated by this Section 7.2 in arrears, within [***] days from the end of each Calendar Quarter in which the underlying Net Sales occur. Each royalty or revenue share payment will be accompanied by a report for each country in the Territory and the European Territory in which sales of any Licensed Products occurred in the Calendar Quarter, specifying: (a) the gross sales (if available) and the Net Sales (including a statement of the aggregate deductions taken from gross sales in the calculation of Net Sales) on a Licensed Product-by-Licensed Product and a country-by-country basis, in each country's currency; (b) the applicable royalty or revenue share rate under this Agreement; (c) the royalties or revenue share payable in the country's currency where the Net Sales occurred; (d) the applicable exchange rate to convert from each country's currency to U.S. dollars pursuant to Section 7.5; and (e) the royalties or revenue share payable in U.S. Dollars. For the avoidance of doubt, no royalties or revenue share shall be due or payable by either Party to the other Party with respect to Net Sales of Licensed Products in a given country before or after the Royalty Term for such country.

7.3 **Method of Payment**. Unless otherwise expressly provided, each Party shall make payments owed to the other Party under this Article 7 in arrears, within [***] days from the end of each Calendar Quarter in which such payment accrues. All payments due hereunder will be made by wire transfer of immediately available funds in U.S. Dollars to a bank account or bank accounts designated by the Party to whom the payment is owed.

7.4 **Interest on Overdue Payments**. Any undisputed amounts not paid by either Party when due under this Agreement will be subject to interest from and after the date payment is due through and including the date upon which such Party makes such payment in immediately available funds at an annual rate equal to the sum of 300 basis points over the prime rate of interest quoted in the Money Rates section of the *Wall Street Journal* (New York Edition), calculated daily on the basis of a three hundred sixty (360) day year, or a similar reputable data source, or if lower, the maximum rate permitted by Applicable Laws.

7.5 **Foreign Currency Exchange**. For any currency conversion from the currency of one country in which the Licensed Products are sold into U.S. Dollars (or another currency if applicable) required in determining the amount of Net Sales or any royalties or revenue share due hereunder, such conversion shall be calculated at the conversion rate as reported in the *Wall Street Journal* (New York Edition) (or if that is no longer published, at the exchange rate reported by The Bank of Tokyo-Mitsubishi UFJ, Ltd.) on the last Business Day of the applicable quarterly period in which the Net Sales are determined, or such other method agreed to by the Parties in writing. Likewise in any other situation in which a currency conversion calculation is required, the conversion rate published on an appropriate date by the *Wall Street Journal* (New York Edition) (or if that is no longer published, at the exchange rate reported by The Bank of Tokyo-Mitsubishi UFJ, Ltd.) shall be used.

7.6 **Taxes.**

7.6.1 **No Withholdings.** All payments required to be made by one Party to the other Party under this Agreement shall be made free and clear of, and without reduction for, withholding Tax or similar Taxes; provided, however, that if a Party or any of its Affiliates is required by Applicable Laws to deduct or withhold any such Taxes from payments made under this Agreement, then such Party or its Affiliates, as applicable, shall make such deduction or withholding from such payment and pay the full amount deducted or withheld to the relevant governmental authority in accordance with Applicable Laws. If, under Applicable Laws, such Tax is required to be deducted or withheld, the paying Party or its Affiliates shall promptly furnish the other Party with reasonable evidence of such deduction or withholding and payment thereof to the relevant governmental authority, in electronic or written form. The Parties shall reasonably cooperate in completing and filing documents required under the provisions of any Applicable Laws in connection with the making of any required Tax payment or withholding payment, or in connection with any claim to an exemption from, reduction of, or a refund of or credit for any such payment to the extent available under Applicable Laws.

7.6.2 **Other Tax Liability**. Except as provided to the contrary in this Agreement, each Party shall be solely responsible for all federal, state and local Tax liability arising from this Agreement imposed on such Party by the taxing authority of a jurisdiction in which such Party is resident or is otherwise subject to such Tax liability. In the case of value added or similar Taxes incurred by a Party with respect to payments made to a Party hereunder or the activities underlying such payments ("**VAT**"), each Party and their Affiliates shall use Commercially Reasonable Efforts to secure available exemption(s) from VAT and/or to cooperate with the other Party's efforts to obtain maximum recovery of VAT paid or incurred by such Party or any Affiliate, to the extent permitted by Applicable Laws.

7.6.3 **Treatment of Royalties and Revenue Shares for Tax Purposes**. To the extent permitted by Applicable Laws, the Parties intend and agree to treat for income Tax purposes (i) the revenue shares payable pursuant to Sections 7.2.2 and sub-Sections (d) and (e) of Section 15.5 as business profits within the meaning of Article 7 of the Income Tax Convention for

the Avoidance of Double Taxation between Japan and the United States **(**"**U.S.-Japan Treaty**") and the corresponding (Business Profits) article of any equivalent income tax treaties that may apply to such payments and (ii) the royalties payable pursuant to Sections 7.2.1, 7.2.3 and sub-Sections (a), (b), (c), (f) and (g) of Section 15.5 as consideration for the use of, or the right to use, a patent or patents, a secret process, or information concerning industrial, commercial or scientific experience within the meaning of U.S.-Japan Treaty and any equivalent income tax treaties that may apply to such payments.

7.7 **Prohibited Payments**. Notwithstanding anything to the contrary in this Agreement, if either Party is prohibited from making any payments by virtue of the statutes, laws, codes or governmental regulations of the country from which the payment is to be made, then such payment may be paid by depositing funds in the currency in which it accrued to the other Party's account in a bank reasonably acceptable to the other Party in the country whose currency is involved.

ARTICLE 8.
KHK SUPPLY OF LICENSED PRODUCT; SPECIFICATIONS

8.1 **KHK Obligation to Supply Licensed Product**. UGNX shall obtain one hundred percent (100%) of its requirements of the Licensed Products for Development and Commercialization in the Field in the Territory and the European Territory from KHK, and KHK agrees to manufacture or have manufactured and to supply to UGNX all of UGNX's requirements of the Licensed Products for Development and Commercialization in the Field in the Territory and the European Territory, in all such cases except to the extent otherwise provided in, and in any event subject to and in accordance with, the terms of this Article 8.

8.2 **Supply of Licensed Products for Development**. Subject to the terms and conditions of this Agreement, KHK shall use Commercially Reasonable Efforts to supply UGNX with quantities of the Licensed Products (for purposes of this Section 8.2, "Licensed Products" shall be deemed to refer to "unlabeled, GMP vialed Licensed Products." KHK currently has Licensed Product in the following concentrations and vial sizes: [***]. KHK shall use Commercially Reasonable Efforts to provide Licensed Product, Drug Substance and any placebo required for UGNX's Development activities with respect to the Licensed Products in the Territory and the European Territory in accordance with UGNX's forecasts and orders therefor, as provided below. UGNX shall use the Licensed Products supplied by KHK pursuant to this Section 8.2 solely in order to conduct Development activities in accordance with the terms and conditions of this Agreement, and shall not use such Licensed Products for any other purpose.

8.2.1 **Forecasts and Orders.** UGNX shall keep KHK reasonably informed of its anticipated requirements of the Licensed Products for Core Development Activities and Non-Core Development Activities through the JDC and the Core Development Plan and Non-Core Development Plan by providing a good faith estimate of such requirements on a [***] basis, the first [***] months of which shall be binding, to be updated [***], (collectively, the "**UGNX Forecast**"). UGNX shall order Licensed Products for use in Development from KHK by providing KHK with a binding purchase order (consistent with the terms and conditions of this Agreement) indicating the quantities of the Licensed Products ordered for Development purposes, the requested delivery date and the destination delivery location. Upon receipt of any such binding purchase order, KHK shall use Commercially Reasonable Efforts to manufacture and supply the Licensed Products in accordance therewith. Within [***] Business Days of receiving a binding purchase order, KHK shall notify UGNX with confirmation of such purchase order, it being understood that KHK may not reject a purchase order if such order is materially consistent with the UGNX Forecast and the required delivery date is at least [***] after the date of such purchase order.

8.2.2 **Cost of Supply of Products for Development**. The price for the Licensed Products, the Drug Substance and any placebos required in order for UGNX to conduct non-clinical and clinical studies required for Development will be at (a) [***] and (b) [***].

8.2.3 **Delivery**. KHK shall: (a) deliver the Licensed Products FCA KHK's or the Third Party manufacturer's facility selected in accordance with Section 8.5.1 (Incoterms 2000); (b) if requested by UGNX, obtain any necessary export permits for such delivery, at UGNX's sole expense; (c) deliver all Licensed Products by the delivery dates established under Section 8.2.1 and otherwise in conformance with UGNX's order (except that KHK may elect to split the order into multiple shipments so long as the delivery dates for the entire order are not materially delayed thereby). Title to and risk of loss in the Licensed Products will pass to UGNX upon delivery to the common carrier for delivery to UGNX or its designee (each, a "**Delivery**"), and UGNX shall be responsible for paying any freight, delivery and insurance charges incurred in delivering the Licensed Products to UGNX's designated delivery destination(s) (subject to cost-sharing except in Latin America), provided that KHK shall provide reasonable assistance to UGNX in identifying appropriate carriers. KHK shall provide appropriate documentation (including certificates of analysis, GMP declaration statements and other documentation required by Regulatory Authorities or otherwise to comply with Applicable Laws of the Profit Share Territory and European Territory) with all shipments of Licensed Products hereunder. KHK shall not be responsible for complying with any additional or differing legal or regulatory requirements specific to Licensed Products for Latin America.

8.2.4 **Responsibilities of UGNX**. With respect to the importation of the Licensed Products to Latin America, UGNX will be responsible for the following on a case-by-case basis: (a) obtaining all necessary permits for the importation of the Licensed Products; (b) all customs, duties and other governmental charges relating to the importation of the Licensed Products from the manufacturing site to UGNX; (c) storing and clearing the Licensed Products through all customs and importation requirements; (d) having the Licensed Products delivered to UGNX's labeling and packaging facility; and (e) conducting quality control testing, retention of samples and lot release, labeling and packaging of the Licensed Products for distribution, and conducting any and all release testing required in Latin America, all in full compliance with all Applicable Laws. Notwithstanding the foregoing, based on good faith discussions between the Parties, KHK shall, at [***], either (i) transfer to UGNX any testing procedures and assays solely to the extent reasonably necessary for UGNX to Develop and Commercialize Licensed Products in Latin America, or (ii) conduct any release testing required for any country in Latin America on behalf of UGNX, solely to the extent required by relevant Regulatory Authorities. KHK shall not be responsible for complying with any requirements that are specific to Latin America and are in addition to, or differ from, legal or regulatory requirements applicable to the Profit Share Territory and the European Territory.

8.2.5 **Manufacturing Compliance and Quality Assurance by KHK**. KHK shall manufacture or have manufactured Licensed Products in accordance with GMP and Applicable Laws of the Profit Share Territory and the European Territory. For all Licensed Products delivered to UGNX under this Section 8.2, KHK shall conduct quality control, or will cause its Third Party Manufacturing contractor to conduct such testing, for compliance with Specifications and testing required for compliance with GMP and Applicable Laws of the Profit Share Territory and the European Territory. KHK shall conduct a quality assurance review of all applicable documents and activities for compliance with Applicable Laws of the Profit Share Territory and European Territory, including GMP, with respect to the Licensed Products prior to shipment thereof. KHK shall not be responsible for compliance with Applicable Laws of Latin America but the Licensed Products must comply with Applicable Laws that would have applied had the Licensed Products been made for the Profit Share Territory or European Territory. Further requirements with respect to Licensed Product to be Developed and Commercialized in Latin America shall be set forth in the Commercial Supply Agreement.

8.2.6 **Limited Warranties**. KHK represents and warrants to UGNX that the Licensed Products supplied pursuant to this Section 8.2 will (a) be manufactured and tested in accordance with GMP and Applicable Laws of the Profit Share Territory or European Territory (but not of Latin America), (b) conform to the Specifications therefore in all material respects, and (c) have a shelf life of at least [***] from shipment. Notwithstanding the above, UGNX agrees that initial shipment(s) to UGNX which are tentatively planned to be made prior to the Type C Meeting will only contain vials with shelf life until [***], provided that, for clarity, UGNX may order after the initial shipment(s) additional Licensed Products with a shelf life of at least [***] from shipment as required for Development pursuant to Section 8.2. KHK further agrees to maintain in inventory sufficient bulk Drug Substance and/or vialed, Licensed Product, in KHK's discretion, the aggregate amount of which shall be equivalent to a [***] month supply of vialed Licensed Product, based on UGNX's most recent forecast for the Territory and the European Territory.

8.2.7 **Nonconforming Licensed Product**.

(a) **Acceptance and Rejection by UGNX**. Each shipment of Licensed Product will contain such quality control documentation (e.g., certificates of analysis and compliance, batch release records) as are necessary to show that Licensed Product conforms to the Specifications in all material respects at the time of Delivery and were manufactured in compliance with GMP and Applicable Laws of the Profit Share Territory or European Territory. In the event that UGNX determines within [***] days of UGNX's receipt of each shipment of Licensed Products that any such Licensed Products did not materially conform with the Specifications at the time of Delivery or otherwise comply with the product warranty in Section 8.2.6 or the requirements of the order (such as matching the quantities ordered), UGNX shall provide Notice to KHK thereof, and, if requested by KHK, ship a sample portion of the affected Licensed Products to KHK or its designated Third Party manufacturing site, freight prepaid and properly insured, along with a reasonably detailed statement of the claimed non-conformity and copy of KHK's invoice therefor. UGNX shall retain the balance of the Licensed Products that are subject to review subject to resolution of the rejection and further disposition in accordance with this Section 8.2.7.

(b) **Replacement by KHK**. In the event that KHK agrees that the returned Licensed Products were non-conforming (or such non-conformance is confirmed under Section 8.2.7(c) below), KHK shall replace all of such non-conforming units of Licensed Product, at no cost to UGNX, and KHK shall as soon as practicable deliver to UGNX, freight prepaid, all replacement units of the Licensed Products, along with reimbursement of the shipment and insurance charges for return of the non-conforming Licensed Product. UGNX shall dispose of all non-conforming Licensed Product at KHK's expense. In the event that the quantities of the Licensed Products delivered to UGNX (in one or more shipments) do not match the quantity ordered in any material respect, KHK shall promptly ship (such shipping at KHK's sole expense) the additional Licensed Products required to make up such shortfall; or if the amount shipped exceeds the amount ordered by a material amount, accept only the amount ordered, in which case upon KHK's request and at KHK's sole expense, such additional quantities shall be returned to KHK.

(c) **Disputes Over Non-Conforming Licensed Product**. In the event that KHK disagrees with UGNX's rejection because the Licensed Products are in fact conforming, the Parties shall cooperate to have both UGNX's returned samples and KHK's retained samples from the same production batch of the Licensed Products in dispute analyzed by a mutually acceptable independent testing laboratory of recognized reputation in the pharmaceutical industry,

using the analytical methods, tests and criteria for conformance set forth in the Specifications. The out-of-pocket external costs of such arrangement shall be shared [***] by the Parties, unless and until an alternative determination is made as provided below. The results of such laboratory testing shall be conclusive and binding on the Parties on the issue of compliance of such units of Licensed Product with the Specifications at the time of Delivery. If such independent testing laboratory determines that UGNX's returned samples of such Licensed Product conform to the Specifications, then (i) the applicable Licensed Product shall be deemed to have been improperly rejected by UGNX, and (ii) UGNX shall bear the cost of the independent laboratory testing and all out-of-pocket external costs and expenses of returning the improperly rejected Licensed Product to UGNX. If such independent testing laboratory determines that UGNX's returned samples of such Licensed Product did not conform to the Specifications and that such returned samples conform to the samples for such batch retained by KHK, then KHK shall bear the cost of the laboratory testing, as well as the costs associated with properly-rejected Licensed Product described in Section 8.2.7(b), and KHK shall promptly supply to UGNX conforming Licensed Products in accordance with Section 8.2.7(b).

(d) **Sole Remedy.** Except for Third Party Claims otherwise indemnified under this Agreement, the remedies expressly provided in this Section 8.2.7 will be UGNX's sole and exclusive remedy for the breach of Section 8.2.6 as a result of the delivery by KHK of non-conforming Licensed Products.

(e) **No Liability for Subsequent Events.** In no event will KHK be liable under this Section 8.2.7 for Licensed Products that conformed to the Specifications at the time of Delivery but that ceased to so conform as a result of any event or occurrence, or any action or omission by UGNX, its Affiliate or a Third Party, following Delivery of such License Products.

8.2.8 **Invoice and Payment.** KHK shall invoice UGNX for each shipment of the Licensed Products upon shipment to UGNX at the supply price (if applicable) determined as set forth herein.

8.3 **Commercial Supply of Licensed Product**. Subject to the terms and conditions of this Agreement, UGNX and its Affiliates shall purchase, and KHK shall supply UGNX and its Affiliates, with all quantities of Licensed Products (for purposes of this Section 8.3 "Licensed Products" shall be deemed to refer to "unlabeled, GMP vialed Licensed Products" in the concentrations and vial sizes to be agreed upon by the Parties as required for UGNX's and its Affiliates' Commercialization of Licensed Products in Latin America). UGNX shall use the Licensed Products supplied by KHK under this provision solely to conduct its Commercialization activities, in accordance with the terms and conditions of this Agreement, and shall not use such Licensed Products for any other purpose.

8.3.1 **Commercial Supply Agreement**. The Parties shall start negotiations as soon as practical after the Effective Date and shall execute a definitive commercial supply agreement ("**Commercial Supply Agreement**") for the supply by KHK of the Licensed Products to UGNX and its Affiliates for marketing and sale of such Licensed Products in Latin America. Such Commercial Supply Agreement shall contain the terms and conditions set forth in this Section 8.3, consistent with those set forth in the remainder of this Article 8 (except as provided otherwise in this Section 8.3) and other reasonable and customary terms and conditions. In the event the Parties fail to enter into such a Commercial Supply Agreement for Latin America, and without diminishing the Parties' obligation to enter into such agreement, UGNX will be obligated to purchase from KHK, and KHK will be obligated to sell to UGNX, all of the requirements of UGNX and its Affiliates for the Licensed Products pursuant to the terms of this Article 8, and either Party may refer the matter for resolution as a Dispute under Section 16.2 (Regular Arbitration).

8.3.2 **Cost of Supply of Licensed Products for Commercial Sales in Latin America, and Non Commercial Uses**.

(a) During the Term, KHK shall supply the Licensed Products to UGNX and its Affiliates for commercial sales (including Named Patient Sales) in Latin America at a cost (the "**Commercial Supply Cost**") based on a percentage of Net Sales where such percentage is [***] percent ([***]%) of Net Sales for Licensed Products sold prior to [***] and [***] percent ([***]%) of Net Sales for Licensed Products sold thereafter.

(b) During the Term, KHK shall supply Licensed Products to UGNX and its Affiliates for non-commercial uses in each country in the Territory following receipt of Marketing Approval (i.e., Phase 5 Clinical Trials, samples, supplies provided to patients as part of "compassionate use" programs in the U.S.) at a cost to be determined by the Parties.

8.3.3 **Term**. The term of Commercial Supply Agreement will be co-terminus with the Term (plus any extensions as described below), unless the Parties mutually agree to extend the term of the Commercial Supply Agreement to cover periods after this Agreement is terminated or expires. The Commercial Supply Agreement will provide that the term of that agreement will be automatically extended on an annual basis after the end of the Term, unless KHK elects to terminate such Commercial Supply Agreement with at least [***] advance Notice (e.g., [***]); provided that KHK's obligation to supply Licensed Product thereunder shall expire upon the earlier of the end of such [***] notice period or the date on which UGNX has transitioned the manufacture of Licensed Products to a new facility or manufacturer.

8.3.4 **Priority Status**. To the extent the available supply of, or capacity to manufacture, the Licensed Products is less than the requirements of UGNX and its Affiliates hereunder together with the requirements of KHK and its Affiliates and their licensees, KHK shall allocate the available Licensed Product in a fair and reasonable manner as between all such interested entities, which shall not be deemed a breach of KHK's supply obligations under the Commercial Supply Agreement. In the event of any shortage in availability of supply of, or capacity to manufacture, Licensed Products as contemplated by this Section 8.3.4, the Parties will discuss appropriate actions in good faith and will cooperate with each other in order to resolve such situation.

8.4 **Handling of Licensed Products**. UGNX shall use Commercially Reasonable Efforts to maintain all registrations necessary in the Territory and the European Territory for the lawful handling of the Licensed Products other than such registrations as have been transferred to KHK pursuant to Section 5.1.2 and shall immediately notify KHK of any denial, revocation or suspension of any such registration. Standard instructions for safe handling of the Licensed Products will be set forth in instructions provided by KHK to UGNX from time to time.

8.5 **General Manufacturing and Supply Provisions.**

8.5.1 Third Party Manufacturer. UGNX hereby acknowledges and agrees that KHK will be entitled, in its sole discretion, to perform any or all of its obligations under this Article 8 by subcontracting any or all of such obligations to Third Party manufacturers (each, a "**Third Party Manufacturer**") in any country. In the event that KHK elects to subcontract any or all of its

obligations under this Agreement to a Third Party Manufacturer, KHK shall use Commercially Reasonable Efforts to: (a) coordinate the transfer of such obligations with UGNX, including provision to UGNX of the right to audit any facility of the Third Party Manufacturer used or to be used for the manufacture of the applicable Licensed Products and (b) involve UGNX in decisions regarding process changes that could affect the comparability of the Licensed Products. KHK will be solely responsible for Development costs resulting from any change to a Third Party Manufacturer.

8.5.2 **Quality Agreement**. The Parties shall enter into a reasonable and customary GMP quality agreement with respect to the Licensed Products to be manufactured by or for KHK and supplied to UGNX hereunder for use in Development of the Licensed Products in the Territory and in the European Territory within [***] months of the Effective Date. A separate GMP quality agreement with respect to Licensed Products supplied to UGNX for Commercial use in Latin America shall be executed between the Parties within [***] months of execution of a Commercial Supply Agreement between the Parties for Latin America.

ARTICLE 9.
RECORDS AND REPORTING

9.1 **Records**. Each Party shall keep and maintain complete and accurate books and records necessary to permit calculation and verification of amounts due under Sections 4.9, 7.1, 7.2 or 15.5. Each Party shall maintain such books and records for five (5) years after the applicable book or record was created, or such longer period as may be required by Applicable Laws.

9.2 **Audits.**

9.2.1 Each Party may, at the electing Party's expense and upon not less than [***] days' prior written Notice to the other Party, elect to have an independent certified public accountant selected by the electing Party and reasonably acceptable to other Party examine, during regular business hours and under a customary non-disclosure agreement, the books and records of the other Party required to be maintained pursuant to Section 9.1 at the electing Party's expense, not more often than [***] each Calendar Year, for the sole purpose of verifying the accuracy of the payments made under Sections 4.9, 7.1, 7.2 or 15.5, and the associated reports furnished by the other Party with respect thereto solely for prior periods covering no more than the last [***] full Calendar Years. Any amounts shown to be owed but unpaid as a result of such audit shall be paid within [***] days from the accountant's report (plus interest on such amounts pursuant to Section 7.4), unless challenged as provided below. Any amounts shown to have been overpaid shall be refunded to the Party that overpaid within [***] days from the accountant's report. The electing Party shall bear the full cost of such audit unless such audit discloses an underpayment of the amount actually owed during the applicable Calendar Year of more than [***] percent [***]%), in which case the other Party shall bear the full out-of-pocket, external cost of such audit.

9.2.2 If the Party that is the object of an audit (the "**Audited Party**") challenges the results of the audit in good faith, such Party shall be entitled at its own cost and expense to obtain a second independent certified public accountant to confirm the accuracy of the first audit. If the results of the confirmatory audit are substantially similar to the results of the first audit, any amounts owed or overpaid by the Audited Party shall be paid or refunded in accordance with the procedures above. If the results of the confirmatory audit are not substantially similar to the results of the first audit, each Party shall

cause its respective auditors to identify the discrepancy and to agree on a final amount owed or overpaid (as the case may be) by the Audited Party that shall be final and binding on the Parties. If the auditors cannot resolve the discrepancy, the Parties shall mutually agree on a third independent certified public accountant to audit the discrepancy and provide a final amount owed or overpaid (as the case may be) by the Audited Party, which shall be binding on the Parties. The costs of such third audit shall be shared [***] by the Parties. Amounts owed or overpaid as determined by such final audit shall be paid or refunded in accordance with the procedures above.

ARTICLE 10.
INTELLECTUAL PROPERTY PROVISIONS

10.1 **Patent Prosecution and Maintenance.**

10.1.1 KHK shall diligently prepare, file, prosecute and maintain the Licensed Patent Rights at KHK's sole cost and expense. Patent counsel selected by KHK will handle all patent filings and prosecutions. KHK shall provide to UGNX a reasonable opportunity to review and comment on any material filings and correspondence with applicable patent offices with respect thereto, and KHK shall use good faith efforts to consider any such comments. KHK shall notify UGNX in the event KHK desires to abandon its efforts to prosecute and maintain Licensed Patent Rights, or decides not to file patent applications for Licensed Patent Rights (other than [***]), which notification will be given within a reasonable period (i.e., with sufficient time for UGNX to take whatever reasonable action may be necessary) prior to the date on which such Licensed Patent Rights will lapse, go abandoned (other than to file a continuation application for the same subject matter) or otherwise diminish UGNX will then have the right, exercisable upon written notification to KHK, to assume full responsibility, at its discretion and its sole cost and expense, to file, prosecute and maintain the Licensed Patent Rights (other than the [***]) in such country or countries. Should UGNX exercise such right, KHK shall execute such documents and perform such acts as may be reasonably necessary for UGNX to so file, prosecute and maintain such Licensed Patent Rights. With respect to the [***], in the event that KHK desires to abandon its efforts to prosecute and maintained such Licensed Patent Rights, KHK shall work with UGNX in good faith and use Commercially Reasonable Efforts to provide UGNX with the opportunity to discuss directly with [***].

10.1.2 KHK will be responsible for any royalty or any other payments required with regard to patents owned by the licensors under the In-Licenses relevant to the Licensed Products in the Field in the Territory and the European Territory.

10.1.3 If it is determined that a license under the [***] patent owned by [***] is required, KHK shall enter into a license for the applicable patent with [***] as necessary to allow the Parties to exercise the rights and perform the obligations set forth in this Agreement, and KHK shall pay the necessary royalties or any other payments under such license. If it is determined that a license under any Third Party patent is required, Section 10.6 shall apply.

10.2 **Ownership of Inventions.**

10.2.1 KHK will retain ownership of all KHK Inventions and UGNX will retain ownership of all UGNX Inventions and the Parties will jointly own Joint Inventions. The Parties shall reasonably cooperate with respect to, and [***], the preparation, filing, prosecution and maintenance of any patents and/or patent applications on any such Joint Inventions. In connection with the foregoing, the Parties shall agree upon a lead Party to administer such filing, prosecution and maintenance of any such patent applications and/or patents on Joint Inventions and the lead Party shall provide the non-lead Party a reasonable opportunity to review, comment on and approve

(such approval not to be unreasonably withheld) in advance any material filings and correspondence with applicable patent offices with respect thereto. Each Party will have the right to abandon any such patents or patent applications on a patent-by-patent (or application-by-application) or country-by country basis, provided that, if the abandoning Party is the lead Party, the lead Party shall provide reasonable advance notification to the non-lead Party (i.e., with sufficient time for the non-lead Party to take whatever reasonable action may be necessary) prior to the date on which such Licensed Patent Rights will lapse, go abandoned (other than to file a continuation application for the same subject matter) or otherwise diminish. Subject to the licenses granted to each Party hereunder in their respective territories, each Party will have full rights to exploit and license such Joint Inventions (and joint any patent rights therein), without any obligation or requirement of an accounting to the other Party, and each Party hereby consents to such exploitation and licensing of the other Party for Joint Inventions. For the avoidance of doubt, all KHK Inventions (including KHK's rights to any Joint Inventions, but on a royalty-free basis) will be included within the Licensed Technology hereunder and licensed to UGNX pursuant to Article 2.

10.2.2 Subject to the terms and conditions of this Agreement, UGNX hereby grants to KHK and its Affiliates a non-exclusive, royalty-free, sublicenseable (except as provided below) license under the UGNX Inventions (including UGNX's rights to any Joint Inventions) to develop, use, sell, offer for sale, make, import and export (and to have such actions taken on its behalf by agents, contractors and other Third Party service providers) the Drug Substance and the Licensed Products for all indications and all fields in the Rest of the World, and in the Profit Share Territory and the European Territory.

10.2.3 Each Party shall cause all Persons who perform development activities or regulatory activities for such Party relating in whole or in part to the Licensed Products or otherwise to under this Agreement to assign to such Party all their rights and title in any inventions conceived, made or generated or to be conceived, made or generated by them and resulting in whole or in part from such activities, except to the extent Applicable Laws prohibit such a requirement, and further except in the case of governmental, not-for-profit and public institutions which have standard policies against such an assignment (in which case a suitable license, or right to obtain such a license, shall be obtained and maintained).

10.3 **Disclosure**. Each Party ("**Inventing Party**") shall promptly disclose to the other Party, in writing, and shall cause its Affiliates, agents, and independent contractors to disclose to the Inventing Party, all inventions related the Licensed Products conceived, made or generated by the Inventing Party which are, in the Inventing Party's reasonable judgment, potentially patentable.

10.4 **Cooperation**. Each Party agrees to reasonably cooperate in the preparation, filing, prosecution and maintenance of the Licensed Patent Rights in the Territory and the European Territory under this Agreement and in the obtaining and maintenance of any patent extensions, supplementary protection certificates and the like with respect to the Licensed Patent Rights in the Territory and the European Territory. Such cooperation includes: (a) executing all papers and instruments, or requiring its employees or contractors to execute such papers and instruments, in order to effectuate the ownership of inventions set forth in Section 10.2 and of Patents claiming or disclosing such inventions, and to enable the other Party to apply for and to prosecute patent applications in any country, to the extent provided for in this Agreement; (b) consistent with this Agreement, assisting in any license registration processes with applicable governmental authorities that may be available in the Territory or the European Territory for the protection of a Party's interests in this Agreement; and (c) promptly informing the other Party of any matters coming to such Party's attention that may materially affect the preparation, filing, prosecution or maintenance of any such Licensed Patent Rights in the Territory or the European Territory.

10.5 **Enforcement of Licensed Patent Rights against Infringement.**

10.5.1 Initiation. UGNX shall promptly notify KHK of any alleged or threatened infringement of the Licensed Patent Rights by a Third Party, or any alleged or threatened assertion of invalidity of any of the Licensed Patent Rights by a Third Party, in all such cases of which UGNX becomes aware in the Field and in the Territory or the European Territory ("**Competing Product Infringement**"). KHK will have the sole discretionary right, but not the obligation, to prosecute any such infringement at its expense. UGNX will have the right to join as a party to any suit initiated by KHK to recover its damages and participate with its own counsel; provided that KHK will retain control of the prosecution of such suit. KHK agrees to keep UGNX informed with respect to such enforcement action.

10.5.2 **Cooperation**. In the event that KHK initiates an infringement action pursuant to this Section 10.5, UGNX shall cooperate fully with respect to such action, including, if required to bring such action, the furnishing of a power of attorney solely for such purpose or to join or be named as a party such action as a necessary party.

10.5.3 **Recoveries.** Any recoveries resulting from such an action relating to a claim of Competing Product Infringement of the Licensed Patent Rights shall be first applied against repayment of each Party's actual out-of-pocket costs and expenses, or proportionate percentages thereof, in connection therewith. Any remainder will be shared as follows: [***].

10.6 **Defense of Infringement Claims.**

10.6.1 **Infringement in Profit Share Territory.** If the manufacture, sale or use of a Licensed Product in the Field in the Profit Share Territory pursuant to this Agreement results in, or may result in, any claim, suit, or proceeding by a Third Party alleging patent infringement by UGNX (or its Affiliates), the Party that learns of this claim, suit or proceeding shall promptly notify the other Party in writing. KHK will be responsible for the defense of such claim, suit or proceeding using counsel reasonably acceptable to both Parties and both Parties shall share [***] the cost and expense (including attorneys' fees) of such defense and damages or royalties (if any) payable to such Third Party in connection with such claim, suit or proceeding, provided that UGNX shall have the opportunity to provide input with respect to the strategy for such defense as well as the material pleadings, and KHK shall use Commercially Reasonable Efforts to take such input into consideration. The Party controlling the defense shall keep the other Party reasonably informed of all material developments in connection with any such claim, suit or proceeding. Each Party agrees to provide the other Party with copies of all pleadings filed in such action and to allow the other Party reasonable opportunity to participate in the defense of the claims. Any recoveries of attorneys' fees or costs in defense of a claim under this Section 10.6.1, and any sanctions awarded to the Party defending the claim and against a party asserting a claim being defended under this Section 10.6.1.

10.6.2 **Infringement in Latin America.** UGNX acknowledges and agrees that KHK has made no representations or warranties to UGNX in this Agreement or otherwise with respect to infringement (or the lack thereof) of Third Party patents in Latin America. If the manufacture, sale or use of a Licensed Product in the Field in Latin America pursuant to this Agreement results in, or may result in, any claim, suit or proceeding by a Third Party alleging patent infringement by UGNX (or its Affiliates), UGNX shall promptly notify KHK in writing. UGNX will be responsible for the defense of any such claim, suit or proceeding at its own cost and expense

(including attorneys' fees and damages or royalties (if any) payable to such Third Party in connection with such claim, suit or proceeding), using counsel of its own choice. KHK may participate in any such claim, suit or proceeding with counsel of its choice at its own expense. UGNX shall keep KHK reasonably informed of all material developments in connection with any such claim, suit, or proceeding. UGNX agrees to provide KHK with copies of all pleadings filed in such action and to allow KHK reasonable opportunity to participate in the defense of the claims. Any recoveries by UGNX of attorneys' fees or costs in defense of a claim under this Section 10.6.2, and any sanctions awarded to UGNX and against a party asserting a claim being defended under this Section 10.6.2, shall be retained by UGNX.

10.6.3 **Infringement in the European Territory**. If the manufacture, sale or use of a Licensed Product in the Field in the European Territory pursuant to this Agreement results in, or may result in, any claim, suit or proceeding by a Third Party alleging patent infringement by KHK (or its Affiliates), KHK shall promptly notify UGNX in writing. KHK will be responsible for the defense of any such claim, suit or proceeding at its own cost and expense (including attorneys' fees and damages or royalties (if any) payable to such Third Party in connection with such claim, suit or proceeding), using counsel of its own choice. UGNX may participate in any such claim, suit or proceeding with counsel of its choice at its own expense. KHK shall keep UGNX reasonably informed of all material developments in connection with any such claim, suit, or proceeding. KHK agrees to provide UGNX with copies of all pleadings filed in such action and to allow UGNX reasonable opportunity to participate in the defense of the claims. Any recoveries by KHK of attorneys' fees or costs in defense of a claim under this Section 10.6.3, and any sanctions awarded to KHK and against a party asserting a claim being defended under this Section 10.6.3, shall be retained by KHK.

ARTICLE 11.
CONFIDENTIALITY, PUBLICATION AND PUBLICITY

11.1 **Confidentiality**. All Confidential Information disclosed by or on behalf of one Party to the other Party hereunder will be maintained in confidence by the receiving Party and will not be disclosed to a Third Party or used for any purpose other than for purposes of exercising a Party's rights or performing a Party's obligations hereunder pursuant to the terms of this Agreement, except as follows:

11.1.1 If the Confidential Information is required to be disclosed by Applicable Laws or court order, including by governmental or other regulatory agencies in order to obtain patents, to obtain approval to conduct Clinical Trials or to market the Licensed Products (or to otherwise perform a Party's obligations hereunder) or to comply with applicable securities exchange or U.S. Securities and Exchange Commission regulations (or the regulations of counterpart agencies within the Territory or the European Territory), then notice of such disclosure shall be promptly delivered to the disclosing Party to the extent reasonable practicable in order to provide such disclosing Party with an opportunity to challenge or limit the disclosure obligations, provided that the receiving Party works in good faith with the disclosing Party to seek confidential treatment of such disclosure and to disclose only to the extent reasonably necessary to comply with the Applicable Law or court order, such Confidential Information may be disclosed to the extent legally required; and

11.1.2 If it is necessary or useful to disclose the Confidential Information to employees, agents, consultants, Affiliates and/or other Third Parties for the purpose of conducting activities permitted or required in accordance with this Agreement, Confidential Information may be

disclosed to such employees, agents, consultants, Affiliates and/or other Third Parties only to the extent necessary, and only if such Persons agree to be bound by confidentiality obligations at least as protective of such Confidential Information as the terms herein provided that the disclosing Party will be responsible for any disclosure of Confidential Information by any such Person inconsistent with the confidentiality obligations owed by the disclosing Party hereunder.

11.2 **Disclosure of Agreement**. Neither Party will release to any Third Party or publish in any way any non-public information regarding the terms and conditions of this Agreement without the prior written consent of the other Party, which consent will not be unreasonably withheld, conditioned or delayed, except: (a) for KHK 's disclosure to its relevant licensors who are subject to a signed confidentiality agreement; (b) pursuant to Sections 11.3 through 11.5; (c) to the extent required to comply with Applicable Laws (including securities laws, regulations and guidances) or by the requirements of any nationally recognized securities exchange, quotation system or over-the-counter market on which such Party has its securities listed or traded, provided that the disclosing Party shall make Commercially Reasonable Efforts to provide the other Party with Notice beforehand in order to permit the other Party to challenge the necessity of such disclosure and/or to coordinate with the other Party with respect to the wording and timing of any such disclosure; or (d) to the disclosing Party's legal and financial advisors, and to any actual or prospective acquirers, investors, collaborators and lenders (as well as and to their respective legal and financial advisors) who are obligated to keep such information confidential provided that the disclosing Party will be responsible for any disclosure of Confidential Information by any such Person inconsistent with the confidentiality obligations owed by the disclosing Party hereunder.

11.3 **Publications.**

11.3.1 **Publication**. Each Party shall submit any proposed publication or presentation concerning the Drug Substance or the Licensed Products (a "**Publication**") to the other Party at least [***] prior to submitting it to any Third Party (including any editing Person) for publication or presentation. In the event that KHK submits a Publication to UGNX in the Japanese language, KHK shall also submit an abstract of such Publication to UGNX in the English language. For the avoidance of doubt, Publications exclude marketing materials.

11.3.1.1 The other Party will have [***] days after receipt of the draft Publication to review and comment on such draft.

11.3.1.2 Upon Notice within such [***] day period by the other Party that the other Party reasonably believes the Publication would amount to the public disclosure of the other Party's Confidential Information and/or negatively impact the other Party's intellectual property position, submission of the concerned Publication to Third Parties will be delayed for a [***] day period from the date of said Notice for appropriately deleting Confidential Information from the proposed Publication or drafting and filing a patent application with respect to any subject matter to be made public in such Publication. Notwithstanding the foregoing, neither Party will be restricted hereunder from making any publication or disclosure to extent required to comply with Applicable Laws.

11.3.2 For all proposed Publications, each Party shall cooperate in good faith to achieve the business objectives of the proposed Publication and the publishing Party will in good faith take into account reasonable comments from the other Party.

11.4 **Press Releases**. UGNX and KHK shall issue a press release within thirty (30) days of the execution of this Agreement in a form reasonably agreeable to both Parties. Thereafter, UGNX and KHK shall consult in advance and reasonably cooperate in the method and manner of any press release regarding this Agreement and any activity related thereto. For clarity, following any press release issued pursuant to this Section 11.4, UGNX and KHK may each disclose to Third Parties the information set forth in such press release without the need for further approval by the other.

11.5 **Employees and Consultants**. Each Party hereby agrees and covenants that all of its employees and consultants and all of the employees and consultants of its Affiliates who participate in any activities under this Agreement or have access to any Confidential Information are or will, prior to their participation or access, be bound by written obligations to maintain such Confidential Information in confidence and not to use or transfer such information or materials except as expressly permitted hereunder. Each Party agrees to enforce, and to cause its Affiliates to enforce, such obligations.

ARTICLE 12.
REPRESENTATIONS AND WARRANTIES

12.1 **Mutual Representations and Warranties**. Each Party hereby represents and warrants to the other Party that as of the Effective Date:

12.1.1 **Corporate Existence and Power**. It is a corporation duly organized, validly existing and in good standing under the laws of the jurisdiction in which it is incorporated, and has full corporate power and authority and the legal right to own and operate its property and assets and to carry on its business as it is now being conducted and as contemplated in this Agreement, including the right to grant the licenses granted hereunder.

12.1.2 **Authority and Binding Agreement**. (a) It has the corporate power and authority and the legal right to enter into this Agreement and to perform its obligations hereunder; (b) It has taken all necessary corporate action on its part required to authorize the execution and delivery of this Agreement and the performance of its obligations hereunder; and (c) This Agreement has been duly executed and delivered on behalf of such Party, and constitutes a legal, valid and binding obligation of such Party that is enforceable against it in accordance with its terms.

12.1.3 **No Conflict**. It has not entered into any agreement with any Third Party that is in conflict with the rights granted to the other Party under this Agreement, and has not taken any action that would in any way prevent it from granting the rights granted to the other Party under this Agreement, or that would otherwise materially conflict with or adversely affect the rights granted to the other Party under this Agreement. Its performance and execution of this Agreement will not result in a breach of any other contract to which it is a Party.

12.1.4 **No Litigation**. It is aware of no action, suit, inquiry or investigation instituted by any Third Party which questions or threatens the validity or enforceability of this Agreement.

12.1.5 **Consents**. All necessary consents, approvals and authorizations of all governmental authorities and other persons or entities required to be obtained by such Party in connection with the execution and delivery of this Agreement have been obtained.

12.2 **UGNX's Representations and Warranties**. UGNX hereby represents and warrants to KHK that as of the Effective Date there is no pending filing, complaint, matter or action against or involving either UGNX or its Affiliates with any Regulatory Authority that could be reasonably anticipated to have a material adverse effect on its ability to obtain Marketing Approvals and Pricing and/or Reimbursement Approvals for the Licensed Products in any country or region of the Territory or the European Territory.

12.3 **KHK's Representations and Warranties**. KHK hereby represents and warrants to UGNX as of the Effective Date:

12.3.1 **Licensed Patent Rights; Licensed Technology**. KHK owns or Controls the Licensed Patent Rights listed on Schedule 1.1.47 and Schedule 1.1.47 is a complete list of all patents and patent applications owned or Controlled by KHK as of the Effective Date which claim or cover the Drug Substance or the manufacture or use thereof or any Licensed Product in the Territory or the European Territory.

12.3.2 **Title; Encumbrances; Third Party Claims**.

12.3.2.1 KHK has sufficient legal and/or beneficial title, ownership or license, free and clear from any encumbrances of the Licensed Technology, including under the In-Licenses, to grant the licenses to UGNX as purported to be granted pursuant to this Agreement.

12.3.2.2 To KHK's knowledge, the Development and Commercialization of Licensed Products do not infringe any intellectual property rights owned or possessed by any Third Party in the Profit Share Territory or the European Territory and do not and will not breach or infringe any obligation of confidentiality or non-use owed by KHK or its Affiliates to a Third Party in the Profit Share Territory or the European Territory, provided, however, that KHK does not make any representation or warranty under this Section 12.3.2.2 with respect to those Third Party patents disclosed by KHK to UGNX in writing prior to the Effective Date;

12.3.2.3 To KHK's knowledge, there are no pending claims, judgments or settlements against or owed by KHK or its Affiliates, or, to KHK's knowledge, threatened claims or litigation; in each case relating to the Licensed Patent Rights or Licensed Know-How; and

12.3.2.4 To KHK's knowledge, none of the issued Licensed Patent Rights are invalid or unenforceable.

12.3.3 **In-Licenses**. Neither KHK nor its Affiliates have in-licensed any Licensed Patent Rights or Licensed Know-How other than under the In-Licenses.

12.4 **Limitation on Warranties; No Implied Warranties**. EXCEPT AS EXPRESSLY PROVIDED IN THIS AGREEMENT, EACH PARTY MAKES NO AND EXPRESSLY DISCLAIMS ALL OTHER REPRESENTATIONS AND WARRANTIES WITH RESPECT TO THE LICENSED PRODUCTS, THE LICENSED TECHNOLOGY, THE LICENSED PATENT RIGHTS OR ANY OTHER SUBJECT MATTER OF THIS AGREEMENT, WHETHER EXPRESS, IMPLIED OR STATUTORY, INCLUDING WARRANTIES OF MERCHANTABILITY, FITNESS FOR A PARTICULAR PURPOSE, NON-INFRINGEMENT OR NON-

MISAPPROPRIATION OF THIRD PARTY INTELLECTUAL PROPERTY RIGHTS. EXCEPT TO THE EXTENT EXPRESSLY PROVIDED FOR HEREIN, NOTHING IN THIS AGREEMENT WILL BE CONSTRUED AS A REPRESENTATION OR WARRANTY BY KHK THAT THE LICENSED PATENT RIGHTS OR THE LICENSED TECHNOLOGY IS NOT INFRINGED BY ANY THIRD PARTY OR THAT THE PRACTICE OF SUCH RIGHTS DOES NOT INFRINGE ANY INTELLECTUAL PROPERTY RIGHTS OF ANY THIRD PARTY. UGNX HAS CONDUCTED ITS OWN INDEPENDENT DUE DILIGENCE REGARDING RISKS OF INFRINGEMENT OF INTELLECTUAL PROPERTY RIGHTS OF THIRD PARTIES AND HAS MADE ITS OWN INDEPENDENT DETERMINATION OF POTENTIAL RISKS WITH THE ASSISTANCE OF COUNSEL.

ARTICLE 13.
OTHER COVENANTS AND AGREEMENTS

13.1 **KHK Licenses to Third Parties.**

13.1.1 **Right of First Negotiation**. In the event that KHK or any of its Affiliates decides to grant to a Third Party a license to develop, sell, offer for sale, import or export Licensed Product(s) in (a) any indication in the Option Negotiation Right Field or (b) any indication in the Field in the Rest of the World, before commencing discussions with respect to any such license, KHK shall provide Notice to UGNX of such decision. For the avoidance of doubt, except as set forth in this Section 13.1, nothing in this Agreement will be deemed to restrict the activities of KHK or its Affiliates in (i) the Option Negotiation Right Field, or (ii) within the Field in the Rest of the World.

13.1.2 **Exercise of Right**. UGNX will have [***] days following receipt of the Notice delivered pursuant to Section 13.1.1 to provide Notice to KHK about whether UGNX is interested in negotiating with KHK for the exclusive development and Commercialization rights in the Option Negotiation Right Field or in the Field in the Rest of the World. If UGNX elects to exercise its right, UGNX and KHK shall negotiate in good faith on the terms for such license for not less than [***] days from the date of UGNX's receipt of the Notice (the "**Negotiation Period**").

13.1.3 **Decision not to Exercise Right**. If UGNX provides Notice to KHK that UGNX is not interested in exercising the right contemplated by this Section 13.1, if the Parties fail to reach a non-binding term sheet at the end of the Negotiation Period, or if no Notice is provided by UGNX to KHK prior to end of the [***] day response period, KHK will be free to negotiate a license with any Third Party; provided, however, that KHK will not enter into a definitive agreement with any Third Party on more favorable terms than those last offered to UGNX for a period of [***] months from the end of the first to occur of [***], the [***], and, [***].

13.2 **Mutual Covenants**. Each Party hereby covenants and agrees during the Term that such Party: (a) shall carry out the Sales, Promotion and Marketing Activities and its other obligations or activities hereunder in accordance with (i) the terms of this Agreement, (ii) accepted pharmaceutical industry practices and (iii) all Applicable Laws; (b) shall use Commercially Reasonable Efforts to undertake its Development and regulatory responsibilities under this Agreement; (c) shall not enter into any agreement with a Third Party which, in any way, will limit such Party's ability to perform all of the obligations undertaken by it hereunder.

13.3 **Exclusivity**. During the period within the Term in which a Licensed Patent Right exists in any country in the Territory and/or the European Territory, on a country-by-country basis, neither Party nor its Affiliates will engage directly or indirectly in, or grant to any Third Party any right with respect to, the [***] (other than Licensed Products Commercialized under this Agreement) [***]. For the avoidance of doubt, the Parties' obligations under this Section 13.3 will expire on the expiration or earlier termination of this Agreement.

13.4 **Mutual Policing Obligations.**

13.4.1 During the Term, UGNX and its Affiliates shall not knowingly sell, transfer or otherwise provide any Licensed Products, directly or indirectly to any Third Party (a) outside of Latin America or, (b) in Latin America for (i) use outside the Field or (ii) for export outside of Latin America.

13.4.2 During the Term, KHK and its Affiliates shall not knowingly sell, transfer or otherwise provide, directly or indirectly (a) to any Third Party in any country of the world, any KHK Out-of-Field Products for use in the Field, or (b) to any Third Party in the Rest of the World or the European Territory, any Licensed Products for export into Latin America for use in the Field.

13.4.3 In the event of any sales or transfers by a Third Party prohibited by Sections 13.4.1 and 13.4.2, the Parties shall discuss in good faith ways to prevent such sales or transfers in the future and shall use Commercially Reasonable Efforts to prevent such future sales or transfers, in each case subject to and in a manner consistent Applicable Laws.

13.5 **Annual Meetings**. To the extent consistent with Applicable Laws, (a) the Parties, including UGNX's CEO and KHK's senior management shall meet upon KHK's request (but no more often than [***] per Calendar Year) at a mutually agreeable time at UGNX's offices (unless otherwise agreed upon by the Parties), at which time and location UGNX will, on a confidential basis, update KHK on the strategy, plans and other aspects of UGNX's business as reasonably requested by KHK, including UGNX's financing plans and budgets, and an update on UGNX's clinical and R&D programs (provided, however, that UGNX may elect not to disclose any information that it reasonably considers to be competitive or privileged) and (b) UGNX shall provide KHK with copies of financial statements on a quarterly basis. Notwithstanding the above, following the closing of a UGNX IPO, UGNX shall not be required to share with KHK under this Section 13.5 any non-publicly available information.

13.6 **Performance Through Affiliates**. Each Party may discharge any obligation and exercise any right hereunder through any of its Affiliates (without an assignment of this Agreement), provided that such Party shall be responsible for its Affiliates' compliance with the terms of this Agreement.

ARTICLE 14.
INDEMNIFICATION AND INSURANCE

14.1 **Indemnity by UGNX**. UGNX hereby agrees to defend, hold harmless and indemnify KHK and its Affiliates, agents, directors, officers and employees (the "**KHK Indemnitees**") from and against any and all Third Party suits, claims, actions and proceedings and associated expenses (including court costs, legal expenses and attorneys' fees) and damages and

recoveries awarded with respect thereto (collectively "**Losses**") incurred by a KHK Indemnitee in connection with any and all Third Party claims arising or resulting from: (a) any misrepresentation (or alleged misrepresentation) or breach (or alleged breach) of any of the representations, warranties, covenants or agreements made by UGNX under this Agreement; (b) any injury, damage or health complication suffered (or alleged to be suffered) as a result of the Development or Commercialization of the Licensed Products by or for UGNX in the Territory or the Development of the Licensed Products by or for UGNX in the European Territory, as applicable, or (c) any violation by UGNX or its Affiliates of laws and regulations applicable to Named Patient Sales in connection with any such Named Patient Sales made by UGNX or its Affiliates under this Agreement in Latin America, except in the case of either (a) or (b), to the extent such Losses arise from KHK's breach of its representations, warranties or covenants under this Agreement or from KHK's failure to supply Licensed Products hereunder in conformance with the Specifications therefor and in compliance with Applicable Laws (including GMP) in the Profit Share Territory or the European Territory (provided that KHK shall not be responsible for complying with any additional requirements specifically applicable to Licensed Products for Development or Commercialization purposes in Latin America) where such failure could not have been identified or detected by UGNX through its application of reasonable and customary quality assurance and quality control practices, or other inspections or activities required under such Applicable Laws with respect to such Licensed Products, prior to distribution of the Licensed Products in Latin America, and (only with respect to clinical supplies) in the Profit Share Territory or the European Territory.

14.2 **Indemnity by KHK**. KHK hereby agrees to defend, hold harmless and indemnify UGNX and its Affiliates agents, directors, officers and employees (the "**UGNX Indemnitees**") from and against any and all Losses incurred by a UGNX Indemnitee in connection with any and all Third Party claims arising or resulting from (a) any misrepresentation (or alleged misrepresentation) or breach (or alleged breach) of any of the representations, warranties, covenants or agreements made by KHK under this Agreement other than such Losses arising from KHK's failure to supply Licensed Products hereunder in conformance with the Specifications therefor and in compliance with Applicable Laws (including GMP) in the Profit Share Territory or the European Territory (provided that KHK shall not be responsible for complying with any additional requirements specifically applicable to Licensed Products for Development or Commercialization purposes in Latin America) where such failure could have been identified or detected by UGNX through its application of reasonable and customary quality assurance and quality control practices, or other inspections or activities required under such Applicable Laws with respect to such Licensed Products, prior to distribution of the Licensed Products in Latin America, and (only with respect to clinical supplies) in the Profit Share Territory or in the European Territory, as applicable; (b) any injury, damage or health complication suffered (or alleged to be suffered) as a result of the development, commercialization, use, promotion, marketing, distribution or sale of Licensed Product by or for KHK or its Affiliates or licensees in the Rest of the World or in the European Territory; (c) any violation by KHK or its Affiliates of laws and regulations applicable to Named Patient Sales in connection with any such Named Patient Sales made by KHK or its Affiliates under this Agreement in the European Territory, or (d) or UGNX's reasonable and appropriate use of any Product Trademarks or KHK Trademarks in accordance with KHK's instructions in connection with any Development or Commercialization of the Licensed Products in the Territory or in the European Territory, as applicable, except in the case of either (a) or (b), to the extent such Losses arise from UGNX's breach of its representations, warranties or covenants.

14.3 **Procedure for Indemnification.** If a KFIK Indemnitee or UGNX Indemnitee (as the case may be, an "**Indemnitee**") wishes to seek indemnification hereunder, such Indemnitee shall inform the Party obligated to indemnify the Indemnitee hereunder (the "**Indemnifying Party**") of the Third Party claim giving rise to the obligation to indemnify as soon as reasonably practicable

after receiving Notice of such Third Party claim. The Indemnifying Party will have the right to assume and control the defense of any such Third Party claim for which it is obligated to indemnify the Indemnitee under this Agreement. The Indemnitee will cooperate with the Indemnifying Party (and its insurer) as the Indemnifying Party may reasonably request, and at the sole cost and expense of the Indemnifying Party. The Indemnitee will have the right to retain its own counsel, at the expense of the Indemnifying Party, if representation of such Indemnitee by the counsel retained by the Indemnifying Party would be inappropriate because of actual or potential differences in the interests of such Indemnitee and any other Party represented by such counsel. In all other cases, the Indemnitee will have the right to participate in such defense, subject to the Indemnifying Party's control, using its own counsel at its own expense. The Indemnifying Party will have no obligation to indemnify any Indemnitee in connection with any settlement made without the Indemnifying Party's prior written consent; provided that the Indemnifying Party does not unreasonably withhold or delay any such written consent. The Indemnifying Party shall seek the prior written consent of the Indemnitee for any settlement of a Third Party claim subject to indemnification hereunder (such consent to not be unreasonably withheld, delayed or conditioned) if such settlement would materially diminish or materially adversely affect the scope, exclusivity or duration of any intellectual property licensed under this Agreement, would require any payment by such Indemnitee, would require an admission of legal wrongdoing in any way on the part of an Indemnitee, or would affect an amendment of this Agreement (otherwise, no such consent shall be required). If the Indemnifying Party does not assume and conduct the defense of the Third Party claim as provided above, (a) the Indemnitee may defend against, and consent to the entry of any judgment or enter into any settlement with respect to the claim in any manner the Indemnitee may deem reasonably appropriate (and the Indemnitee need not consult with, or obtain any consent from, the Indemnifying Party in connection therewith), and (b) the Indemnifying Party will remain responsible to indemnify the Indemnitee as provided in this Article 14.

14.4 **Losses for Product Liability Claims**. Losses from any Third Party claim alleging product liability, product defect, design, packaging or labeling defect, failure to warn, or any similar action relating to the use or safety of a Licensed Product in the Field in any country of the world (each a "**Product Liability Claim**") shall be allocated as follows:

(a) to the extent the Product Liability Claim is covered by a Party's indemnity obligations under Sections 14.1 or 14.2, the Indemnifying Party shall [***];

(b) to the extent a Product Liability Claim is not covered by either Party's indemnity obligations under Sections 14.1 or 14.2, then:

(i) For Product Liability Claims arising or resulting from Licensed Products sold in the Field in the Profit Share Territory, the Parties shall [***].

(ii) For Product Liability Claims arising or resulting from Licensed Product sold in the Field in the European Territory (except as set forth in Section 14.4(b)(i) above) and/or the Rest of the World, KHK shall [***]; and

(iii) For Product Liability Claims arising or resulting from Licensed Products sold in the Field in Latin America, UGNX shall [***].

14.5 **Insurance**. Each Party shall procure and maintain, at its sole cost and expense, comprehensive general liability insurance (which may be self insurance), including product liability insurance, with bodily injury, death and property limits and amounts of coverage consistent with industry standards, including contractual liability and product liability coverage. It is understood that such insurance will not be construed to create a limit of a Party's liability with respect to its indemnification obligations hereunder. Each Party shall provide the other Party with written evidence of such insurance upon request.

ARTICLE 15.
TERM AND TERMINATION

15.1 **Term**. The term of this Agreement ("**Term**") will commence on the Effective Date and, unless earlier terminated as expressly provided below in this Article 15, will continue for so long as either Party is selling Licensed Products in the Field in the Territory or in the European Territory.

15.2 **Termination by Either Party for Breach or Insolvency**. Either Party will have the right to terminate this Agreement prior to the expiration of the Term upon the occurrence of any of the following:

15.2.1 Upon the material, substantial and ongoing breach of any representation, warranty or obligation by the other Party if the breaching Party has not cured such breach within ninety (90) days after written Notice thereof (describing such breach in reasonable detail) by the non-breaching Party; provided, however, that if a breach is not reasonably capable of being cured within the 90-day cure period described above and the breaching Party is making continuing good faith efforts to cure such breach, the cure period shall be extended to one hundred eighty (180) days.

15.2.2 Immediately upon written Notice if the other Party has filed a petition in bankruptcy, or if an involuntary petition in bankruptcy has been filed against the other Party and such petition is not dismissed within sixty (60) days, or if a receiver or guardian has been appointed for the other Party, or upon or after the cessations of operations of the other Party.

15.3 **Special Termination Rights of KHK.**

15.3.1 **Failure to Initiate First Pediatric Study**.

(a) Subject to the terms of this Agreement, in the event UGNX does not Initiate a First Pediatric Study in the Profit Share Territory or the European Territory within eleven (11) months following a Type-C Meeting for such First Pediatric Study (if the meeting is conducted in the Profit Share Territory) or, if the meeting is conducted in the European Territory, by the earlier to occur of (i) the end of the eleventh (11th) months following the foreign equivalent of a Type-C Meeting and (ii) the end of the fifteenth (15th) months after the date of the Type-C Meeting in the Profit Share Territory (collectively, "**First Pediatric Study Deadline**"), KHK may terminate this Agreement by providing Notice to UGNX within thirty (30) days following the expiration of the First Pediatric Study Deadline.

(b) Notwithstanding Section 15.3.1(a) above, UGNX may extend the First Pediatric Study Deadline in order to reflect any of the following excusable delays: safety issues, KHK's failure to comply with this Agreement, including failure to timely provide information such as final 001 data and interim 002 data, and delays resulting from the application of Sections 3.5.1 and/or 3.5.2 despite each Party acting in good faith.

(c) In addition, (i) notwithstanding Section 15.3.1(a) above, UGNX may in its discretion obtain a one-time extension of the First Pediatric Study Deadline for an additional period of six (6) months upon payment by UGNX to KHK of a fee equal to One Million US Dollars ($1,000,000) and (ii) KHK may, in its discretion, grant to UGNX one or more further extensions of the First Pediatric Study Deadline. For clarity, during any extension period available under this subsection (c), Development Costs in the Profit Share Territory and European Territory shall be shared by the Parties as set forth in Section 4.9.1.

15.3.2 **Failure to obtain Marketing Approval in the European Territory**.

(a) Subject to the terms of this Agreement, in the event a first European Centralized Approval is not obtained in the European Core Territory for the First Indication by December 31, 2019 **(**"**European Marketing Approval**"), KHK may terminate this Agreement solely with respect to the European Territory by providing Notice to UGNX within thirty (30) days following the expiration of such deadline.

(b) Notwithstanding Section 15.3.2(a) above, UGNX may extend the deadline set forth in subsection (a) of this Section 15.3.2 in order to reflect the following excusable delays: safety issues, KHK's failure to comply with this Agreement, delays related to manufacturing and CMC, delays caused by KHK's failure to agree on matters subject to joint decision-making and Force Majeure Events.

15.3.3 **Failure to obtain US Marketing Approval in the U.S.**

(a) Subject to the terms of this Agreement, in the event a first Marketing Approval is not obtained in the U.S. for the First Indication by December 31, 2019 ("**US Marketing Approval**"), then, subject to subsection (b) of this Section 15.3.3, KHK may terminate this Agreement solely with respect to the Profit Share Territory by providing Notice to UGNX within thirty (30) days following the expiration of such deadline.

(b) Notwithstanding Section 15.3.3(a) above, upon receipt of KHK's Notice under the preceding sub-clause (a), UGNX may, by providing Notice to KHK within thirty (30) days of receipt of such KHK's Notice, elect to obtain a one-time extension of the deadline set forth in Section 15.3.3(a) for two (2) years in consideration for the payment to KHK of a fee equal to Ten Million US Dollars ($10,000,000) ("**US Extension Period**"). For clarity, during the US Extension Period, Development Costs in the Profit Share Territory shall be shared by the Parties as set forth in Section 4.9.1. If US Marketing Approval is not obtained by the end of the US Extension Period, KHK may terminate the Agreement solely with respect to the Profit Share Territory by providing Notice to UGNX within thirty (30) days following the expiration of the US Extension Period.

(c) In addition, UGNX may extend the deadline set forth in Section 15.3.3(a) in order to recover and reflect the following excusable delays: safety issues, KHK's failure to comply with this Agreement, delays related to manufacturing and CMC, delays caused by KHK's failure to agree on matters subject to joint decision-making and Force Majeure Events.

15.3.4 **Failure to Commercialize in Latin America**.

(a) Subject to the terms of this Agreement, in the event UGNX does not make the First Commercial Sale within two (2) years of US Marketing Approval in any Key Latin American Country, or within five (5) years of US Marketing Approval in any other country in Latin America (if applicable, pursuant to Section 15.3.4(b)), KHK may terminate this Agreement solely in the applicable country in Latin America, on a country-by-country basis, by providing Notice to UGNX within thirty (30) days following the expiration of the applicable deadline.

(b) Notwithstanding Section 15.3.4(a) above, UGNX may extend the deadline in subsection (a) above in order to reflect the following excusable delays: safety issues, regulatory requirements, KHK's failure to comply with this Agreement, delays related to manufacturing and CMC and Force Majeure Events. Notwithstanding the foregoing, as long as UGNX (i) applies for Marketing Approval, or applies for approval for Named Patient Sales within two (2) years of US Marketing Approval in a Key Latin American Country and (ii) uses Commercially Reasonable Efforts to pursue such application in such Key Latin American Country, then KHK may not terminate this Agreement in such country.

15.3.5 Requests for Extensions. In addition to other extensions available under Sections 15.3.1 through 15.3.4, if at any time UGNX believes that, notwithstanding the use of Commercially Reasonable Efforts, it will not be able meet any of the deadlines set forth in such Sections, then UGNX may request an extension of such deadline by providing Notice to KHK, including a reasonably detailed statement of the factors likely to cause such delay. In such a situation, the Parties will discuss in good faith UGNX's request, along with related alterations or supplements to the Core Development Plan where appropriate, provided that KHK may in its discretion decline UGNX's request.

15.3.6 **Excusable Delays.** The Parties shall discuss in good faith the existence or duration of any excusable delay applicable pursuant to Sections 15.3.1(b), 15.3.2(b), 15.3.3(c) and/or 15.3.4(b). Any dispute between the Parties as to such existence or duration shall be finally resolved pursuant to Sections 16.2 and 16.3. For clarity, during any extension period available under the provisions referred to in this Section 15.3.6, Development Costs in the Profit Share Territory and European Territory shall be shared by the Parties as set forth in Section 4.9.1.

15.4 **Key Man Provision.**

(a) It is the intent of the Parties that Dr. Emil Kakkis, Chief Executive Officer and President of UGNX as of the Effective Date, will remain actively involved with the Development of Licensed Products in the First Indication until the first Marketing Approval.

(b) If Dr. Emil Kakkis does not remain actively involved until at least the earlier of (i) Initiation of a pediatric pivotal study in the First Indication in the Profit Share Territory or the European Territory **(**"**Pivotal Study Initiation**") and (ii) the closing of a public offering of UGNX's securities pursuant to a registration statement under the Securities Act of 1933 (or successor thereof or foreign equivalent) ("**UGNX IPO**"), then UGNX may propose a suitable replacement during the three (3) month period following the date on which Dr. Emil Kakkis is no longer actively involved in the Development of Licensed Products in the First Indication, in which case the Parties shall discuss in good faith UGNX's proposed candidate(s). If the Parties fail to agree, then KHK may terminate this Agreement by providing Notice to UGNX within the following thirty (30) days.

15.5 **Post-Termination Commercialization by KHK**. If KHK Commercializes or permits others to Commercialize Licensed Products following the effective date of the termination of this Agreement under Sections 15.2.1, 15.3 or 15.4(b), then MAX shall make the following payments to UGNX:

(a) Upon termination under Section 15.3.1(a) and subject to UGNX's fulfillment of its obligations under Section 15.7.3 on a country-by-country basis, KHK shall (i) pay a royalty equal to [***] percent ([***]%) of Net Sales of Licensed Products by KHK or its Affiliates or other Selling Party, as applicable in the Field in the Territory and the European Territory for a period of [***] following the First Commercial Sale on a country-by-country basis and (ii) reimburse to UGNX [***] Development Costs incurred by UGNX (i.e., [***]) prior to the effective date of such termination.

(b) Upon termination under Section 15.4(b) and subject to UGNX's fulfillment of its obligations under Section 15.7.3 on a country-by-country basis, KHK shall (i) to the extent Dr. Emil Kakkis had remained actively involved with the Development of Licensed Products upon or beyond the Initiation of a First Pediatric Study in the Profit Share Territory or the European Territory, pay a royalty equal to [***] percent ([***]%) of Net Sales of Licensed Products by KHK or its Affiliates or other Selling Party, as applicable, in the Field in each country in the Territory and the European Territory, which royalty shall be payable for [***] following the First Commercial Sale in such country, on a country-by-country basis and (ii) reimburse to UGNX [***] Development Costs incurred by UGNX (i.e., [***]) prior to the effective date of such termination.

(c) Upon termination under Section 15.2.1, or 15.3.2(a), and subject to UGNX's fulfillment of its obligations under Section 15.7.3 on a country-by-country basis, if following such termination KHK or any of its Affiliates or Sublicensees obtains a European Marketing Approval based on data from a pivotal study commenced by UGNX or its Affiliates only if more than [***] percent ([***]%) of the patients to be enrolled in the Phase 3 Clinical Trial (or pivotal study) would have been dosed with study drugs (including KRN23, placebo, or comparators) at the time of the applicable deadline (a "**Qualifying Pivotal Study**"), then KHK shall pay a royalty equal to [***] percent ([***]%) of Net Sales of Licensed Products by KHK and its Affiliates or other Selling Party, as applicable, in the Field in each country of the European Territory for [***] following the First Commercial Sale in such country, on a country-by-country basis. For clarity, if no data from a Qualifying Pivotal Study is used in obtaining a European Marketing Approval for a Licensed Product in the Field, no royalties or any other amounts shall be payable pursuant hereto by KHK or its Affiliates to UGNX for any Licensed Product. For clarity, in the event of the applicability of paragraph "f" below, this paragraph "c" shall no longer apply.

(d) Upon termination under Section 15.2.1, 15.3.3(a) or 15.3.3(b) and subject to UGNX's fulfillment of its obligations under Section 15.7.3 on a country-by-country basis, if following such termination KHK or any of its Affiliates or Sublicensees obtains a US Marketing Approval based on data from a Qualifying Pivotal Study, then KHK shall pay a revenue share equal to [***] percent ([***]%) of Net Sales of Licensed Products by KHK and its Affiliates or other Selling Party, as applicable, in the Field in each country of the Profit Share Territory for [***] following the First Commercial Sale in such country, on a country-by-country basis. For clarity, if no data from a Qualifying Pivotal Study is used in obtaining a US Marketing Approval for a Licensed Product, no revenue share or any other amounts shall be payable pursuant hereto by KHK or its Affiliates to UGNX for such Licensed Product. For clarity, in the event of the applicability of paragraph "e" below, this paragraph "d" shall no longer apply.

(e) Upon termination under Section 15.2.1, 15.3.3(a) or 15.3.3(b) and subject to UGNX's fulfillment of its obligations under Section 15.7.3 on a country-by-country basis, if following such termination KHK or any of its Affiliates or Sublicensees obtains a US Marketing Approval based on a Biologic Licensing Application (or similar filing outside the U.S.) (BLA) filed by UGNX prior to the effective date of such termination, or UGNX obtains US Marketing Approval prior to the effective date of such termination, KHK shall pay a revenue share at the rates set forth in Section 7.2.2 on Net Sales of Licensed Products by KHK and its Affiliates or other Selling Party, as applicable, in the Field in each country of the Profit Share Territory for [***] following the First Commercial Sale in such country, on a country-by-country basis, or, if UGNX obtains US Marketing Approval prior to the effective date of termination, [***] following such termination, on a country by country basis. For clarity, if no data from a Qualifying Pivotal Study is used in obtaining a US Marketing Approval for a Licensed Product in the Field, no revenue share or any other amounts shall be payable pursuant hereto by KHK or its Affiliates to UGNX for any Licensed Product.

(f) Upon termination under Section 15.2.1 or 15.3.2(a), and subject to UGNX's fulfillment of its obligations under Section 15.7.3 on a country-by-country basis, if following such termination KHK or any of its Affiliates or Sublicensees obtains a European Marketing Approval based on a Marketing Authorization Application filed by UGNX prior to the effective date of such termination, or if UGNX obtains European Marketing Approval prior to the effective date of such termination, KHK shall pay a royalty equal to [***] percent ([***]%) of Net Sales of Licensed Products by KHK and its Affiliates or other Selling Party, as applicable, in the Field in each country of the European Territory for [***] following the First Commercial Sale in such country, on a country-by-country basis, or, if UGNX obtains European Marketing Approval prior to the effective date of termination, [***] following such termination, on a country by country basis. For clarity, if no data from a Qualifying Pivotal Study is used in obtaining a European Centralized Approval for a Licensed Product in the Field, no royalties or any other amounts shall be payable pursuant hereto by MHK or its Affiliates to UGNX for any Licensed Product.

(g) Upon termination under Section 15.2.1, and subject to UGNX's fulfillment of its obligations under Section 15.7.3 on a country-by-country basis, in the event UGNX has achieved a First Commercial Sale or a Named Patient Sale in any country in Latin America prior to the effective date of such termination, then KHK shall pay a royalty equal to [***] percent ([***]%) of Net Sales by KHK and its Affiliates or other Selling Party, as applicable, in such country for [***] following the effective date of termination.

(h) Except as set forth in the preceding sub-clauses of this Section 15.5, upon any termination of this Agreement, notwithstanding anything to the contrary, KIIK shall, subject to the provisions of paragraph "i" below, and subject to UGNX's fulfillment of its obligations under Section 15.7.3 on a country-by-country basis, pay a royalty equal to [***] percent ([***]%) of Net Sales of Licensed Products by KHK and its Affiliates or other Selling Party, as applicable, in the Field in each country of the Territory and/or European Territory for [***] following the First Commercial Sale in such country, on a country-by-country basis.

(i) Notwithstanding the foregoing, no payments shall be due under this Section 15.5 upon termination pursuant to Section 15.2.1 to the extent the breach results from UGNX's intentional or willful misconduct, reckless misconduct or a repeated pattern of bad faith behavior.

15.6 **Effect of Expiration or Termination of this Agreement.**

15.6.1 Upon the expiration or termination of this Agreement by either Party for any reason, the following provisions will apply:

(a) Each Party shall return the originals and any copies of the other Party's Confidential Information; provided that each Party may retain copies of any Confidential Information that is subject to a continuing license hereunder and one copy of the other Party's Confidential Information in possession of its legal counsel for the purposes of monitoring its obligations hereunder and exercising any surviving rights and complying with Applicable Laws; and

(b) Neither Party will be relieved of any liability or obligation of such Party that accrued, or which arose during or relates to any period, prior to the effective date of such termination, including any payment obligations.

15.6.2 The provisions of Sections 5.5 through 5.7, Sections 7.2 through 7.7 (to the extent applicable as a result of the application of Section 15.5), Article 9, Section 10.6 (to the extent the relevant Third-Party claim, suit, or proceeding relate to Licensed Products in the Field sold under this Agreement prior to the effective date of expiration or termination of this Agreement), Article 11, Sections 12.4 and 13.6, Sections 14.1 through 14.3, Section 14.4(a), Sections 15.5 through 15.8, Articles 16 and 17 (other than Section 17.1) will survive any expiration or termination of this Agreement and remain in full force and effect in accordance with their terms. Section 10.5 will survive any expiration or termination of this Agreement and remain in full force and effect in accordance with its terms, except that with respect to infringements covered by Section 10.5 that occur after the Term, UGNX's percentage share of any recoveries remaining after reimbursement of costs and expenses shall equal the percentage royalty or revenue share called for under Section 15.5 to be paid by KHK (if any). Section 14.4(b) will survive any expiration or termination of this Agreement and remain in full force and effect in accordance with its terms, except that KHK will bear [***] of all Losses to the extent resulting from a Product Liability Claim that arises from Licensed Product sold after the Term and that is not covered by either Party's indemnity obligations under Section 14.1 or 14.2.

15.7 **Effect of Certain Terminations**. Upon the termination of this Agreement by either Party pursuant to Section 15.2.1 for breach or Section 15.2.2 for insolvency or by KHK under Section 15.3 or 15.4, the following provisions of this Section 15.7 will apply. In the event any such early termination concerns only a specific country or countries pursuant to Section 15.3, the following shall apply solely with respect to such country(ies) and the Agreement shall otherwise remain in effect in accordance with its terms for all non-terminated countries:

15.7.1 The rights and licenses granted by KIM to UGNX hereunder and UGNX's obligations to share Development Costs pursuant to this Agreement will terminate with respect to the terminated country(ies);

15.7.2 UGNX shall immediately (a) cease conducting any Commercialization activities with respect to the Licensed Products and (b) discontinue making any representation regarding its status as a licensee or distributor of KHK in the Territory for the Licensed Products, subject, in either such case, to requirements of Applicable Laws and to a reasonable wind-down and transition period (not to exceed [***] days);

15.7.3 Subject to Applicable Laws, UGNX shall promptly :

(a) transfer to KHK or KHK's designee all Regulatory Filings (for clarity, including registrations as an IND holder or equivalent), Marketing Approvals and Pricing and/or Reimbursement Approvals owned by UGNX for the Commercialization of the Licensed Products, if such transfer is possible, or, if such transfer is not possible, then at KHK's discretion (i) withdraw any such Regulatory Filings, Marketing Approvals and Pricing and/or Reimbursement Approvals for the Commercialization of the Licensed Products in its name and take all actions necessary or useful to support KHK's or KHK's designee's submission of Regulatory Filings and the achievement of Marketing Approvals and Pricing and/or Reimbursement Approvals in the name of KHK or KHK's designee with respect to the Commercialization of the Licensed Products or (ii) provide KHK with access to, and grant KHK the right and license to use and to reference, such Regulatory Filings, Marketing Approvals and Pricing and/or Reimbursement Approvals then in its name applicable to the Commercialization of the Licensed Products;

(b) provide KHK with copies of all material correspondence between UGNX and Regulatory Authorities with respect to such Regulatory Filings, Marketing Approvals and Pricing and/or Reimbursement Approvals for the Licensed Products, and any and all other clinical and non-clinical data, records and tabulations, in all such cases with respect to the Licensed Products, that UGNX holds as of the date of termination with respect to the Licensed Products;

(c) assign to KHK all agreements specific to the conduct of Clinical Trials for the Licensed Products (to the extent assignable and excluding any such agreements that also involve Clinical Trials for other UGNX products that are not Licensed Products), including agreements or contracts with contract research organizations, clinical sites and investigators, between UGNX and any Third Party, subject to any consent required by such Third Party, which consent UGNX shall use Commercially Reasonable Efforts to obtain on behalf of KHK; and

(d) provide KHK with copies of all reports and data obtained by UGNX or its Affiliates pursuant to this Agreement regarding the Development and the Commercialization of Licensed Products, including any UGNX Clinical Data, which KHK may use for any purpose.

As promptly as possible after any such termination, UGNX shall execute any and all documents of any Regulatory Authorities so as to allow KHK to make immediate use of any data, records and Regulatory Filings transferred by UGNX to KHK pursuant to this Section 15.7.3.

15.7.4 KHK shall make the payments due under Section 15.5, if applicable under the provisions thereof, with respect to the applicable country(ies)

15.8 **Remedies Cumulative and Nonexclusive**. All of the non-breaching Party's remedies will be cumulative, and the exercise of one remedy hereunder by the non-breaching Party will not be deemed to be an election of remedies.

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

ARTICLE 16.
DISPUTE RESOLUTION

16.1 **Disputes**. The Parties recognize that disagreements as to certain matters may from time to time arise out of this Agreement. The Parties agree that such disagreements are to be governed in accordance with this Article 16. Disagreements that are claims, counterclaims, demands, causes of action, disputes or controversies both arising out of this Agreement and related to the performance, enforcement, breach or termination of this Agreement are each, a "**Dispute.**" For the avoidance of doubt, Dispute does not include any claims, counterclaims, demands, causes of action, disputes or controversies regarding a Party's use of any intellectual property rights of the other Party, where such use is not expressly granted by the other Party hereunder, including (with respect to uses by UGNX) any such use in the Rest of the World.

16.2 **Agreement to Arbitrate**. If the Parties fail to resolve any Dispute pursuant to Section 16.1, either Party may submit that Dispute for final resolution by binding arbitration administered by the International Chamber of Commerce ("**ICC**") in accordance with its Rules of Arbitration (the "**Rules**"**)** then in force, to the extent such Rules are not inconsistent with the provisions of this Agreement.

16.3 **Number and Appointment of Arbitrators**. Except as provided by this Section 16.3 or in Section 16.4, the appointment and confirmation of the arbitrators shall be made in accordance with the relevant provisions of the Rules. The arbitral tribunal shall be composed of three (3) arbitrators (the "**Tribunal**") to be appointed in accordance with the Rules, except as expressly provided for herein. Each Party shall select one (1) arbitrator from the list of available ICC arbitrators and such arbitrators shall jointly appoint the third arbitrator who shall act as the chairman of the Tribunal (the "**Chairman**"). In the event any arbitrator becomes unable to serve, that arbitrator will be replaced in the same manner in which he or she was appointed. If either Party fails to appoint an arbitrator within [***] days of the initiation of arbitration, the other Party may request the ICC to appoint such co-arbitrator (for the non-responsive Party). Such appointment shall be binding on the Parties. If the arbitrators selected by the Parties cannot agree on a Chairman within [***] days after they have been selected, then the ICC shall appoint the Chairman upon request by either Party.

16.4 **Special Rules For Certain Specific Disputes (Baseball Arbitration)**. This Section 16.4 shall only apply to the matters expressly identified in this Agreement as subject to resolution pursuant to this Section 16.4. The Tribunal for such matters shall be composed of one (1) arbitrator. In such arbitration, the arbitrator shall be an independent expert (including in the area of the dispute) in the pharmaceutical or biotechnology industry mutually acceptable to the Parties. The Parties shall use their best efforts to mutually agree upon one (1) arbitrator; provided, however, that if the Parties have not done so within [***] Business Days after initiation of arbitration hereunder, or such longer period of time as the Parties have agreed to in writing, then the ICC shall appoint such arbitrator (which arbitrator shall meet the requirements set forth in the preceding sentence) upon request by either Party. Arbitration pursuant to this Section 16.4 shall be limited to choosing one or the other from among the two proposed resolutions prepared by the two Parties with respect to all matters in dispute, and in connection therewith, each Party shall submit to the arbitrator and to each other in writing its position on and desired resolution of (including any specifics or breakdown of the amounts included within their respective figures) each such matter. Such submission shall be made within [***] Business Days of the selection or appointment of the arbitrator, and the arbitrator shall issue its resolution of all such matters within [***] days of receipt of the written submissions by both Parties. For the avoidance of doubt, the arbitrator's resolution of all such matters shall be limited to the selection of one or the other of the two competing resolution methods proposed by the Parties and other than minor modification, the arbitrator may not impose a resolution not proposed by either Party or "split the difference" by creating a compromise resolution based on the desired resolutions proposed separately by the Parties. Except as provided in the preceding sentence, such arbitration shall be conducted in accordance with the relevant provisions of the ICC Rules. The arbitrator's vote shall be final and binding upon the Parties.

16.5 **Conduct of Arbitration**. The arbitration shall be conducted and the award rendered in the English language.

16.6 **Place of Arbitration**. The arbitration shall be held in Singapore.

16.7 **Powers of the Arbitrators, Limitations on Remedies**. With respect to any arbitration conducted pursuant to Section 16.2, the Tribunal will have the power to award all remedies available under Applicable Laws, by a vote of at least two (2) of the three (3) arbitrators.

16.8 **Arbitration Award**. Except as set forth in Section 16.4 (Baseball Arbitration), the Tribunal will make all reasonable efforts to render a written, reasoned decision within [***] days following the closing of the hearing. The decision of the Tribunal will be final and binding on the Parties, and judgment upon the decision may be confirmed in, and judgment upon the award entered by, any court having jurisdiction over the Parties.

16.9 **Confidentiality**. Except to the extent necessary for proceedings relating to enforcement of the arbitration agreement, the award, or other related rights of the Parties, the fact of the arbitration, the arbitration proceeding itself, all evidence, written statements or other documents exchanged or used in the arbitration and the Tribunal's award will be maintained in confidence by the Parties to the fullest extent permitted by law. However, a violation of this covenant will not affect the enforceability of this agreement to arbitrate or of the Tribunal's award.

16.10 **Costs**. Each Party shall bear its own costs and expenses in connection with any Dispute resolution under this Article 16 and shall share equally (50/50) the fees, costs and expenses of the Tribunal and related Third Party arbitration expenses. Notwithstanding the foregoing, the prevailing Party may, as determined by the Tribunal under the circumstances, be awarded its attorneys' fees, costs and expenses of the arbitration, including the arbitrators' fees and expenses, in full. The Tribunal may also fix such costs and expenses proportionate to the extent each Party prevails in the arbitration, as the circumstances may warrant. If a Party fails to proceed with arbitration, unsuccessfully challenges the arbitration award or fails to comply with the arbitration award, the other Party will be entitled to costs, including reasonable attorneys' fees and disbursements, for having to compel arbitration or defend or enforce the award.

16.11 **Injunctive or Other Interim Relief**. Notwithstanding the provisions of this Article 16, the Parties agree that irreparable harm might accrue with respect to a Dispute absent a temporary injunctive or other interim relief and the Parties therefore shall have the right to seek such injunctive or interim relief in a court of competent jurisdiction pending the outcome of Dispute resolution hereunder. In the event interim or injunctive relief is sought by a Party as to a Dispute, and a court of competent jurisdiction grants such interim or injunctive relief, the Parties shall continue to be bound under this Article 16 to resolve by arbitration such Dispute that is the subject of interim or injunctive relief.

16.12 **Continued Performance**. Pending resolution of any Dispute covered by this Article 16, both Parties shall continue their performance under this Agreement of any obligations (including payment obligations) that are not the subject of such Dispute.

16.13 **Governing Law**. Resolution of all Disputes arising out of or related to this Agreement or the performance, enforcement, breach or termination of this Agreement and any remedies relating thereto, will be governed by and construed under the substantive laws of the State of New York, U.S.A., without reference to any choice of law principles thereof that would cause the application of the laws of a different jurisdiction.

16.14 **Separability and Survival of the Agreement to Arbitrate**. The provisions of this agreement to arbitrate are independent of the remaining provisions of this Agreement and the Parties intend that the remaining provisions shall continue in effect even though one or more of the provisions of the Agreement will be determined to be null and void. This agreement to arbitrate will also survive the termination or expiration of this Agreement.

ARTICLE 17.
OTHER PROVISIONS

17.1 **Non-Solicitation of Employees**. During the Term, neither Party nor its Affiliates shall, directly or through its representatives or agents, solicit for employment or hire any officer, director, or employee of the other Party or its Affiliates with whom it has had contact in connection with, or who otherwise is known by it to participate in, the subject matter of this Agreement or the development of the Drug Substance or a Licensed Product; provided, however, a Party will not be prohibited from soliciting and hiring through general public advertisement or other solicitation that is not directed toward the employees of the other Party, and a Party may hire any former employee of the other Party as long as the discussions with the former employee are initiated after termination of employment by the other Party.

17.2 **Force Majeure**. Both Parties will be excused from the performance of their obligations under this Agreement, other than the obligation to make monetary payments, and neither Party will be held liable or responsible to the other Party nor be deemed to have defaulted under or breached this Agreement for failure or delay in fulfilling or performing any term of this Agreement, to the extent that such performance is prevented by force majeure and the nonperforming Party promptly provides Notice thereof to the other Party. Such excuse will be continued so long as the condition constituting a force majeure event continues and the nonperforming Party uses Commercially Reasonable Efforts to remove the condition. For purposes of this Agreement, a force majeure event ("**Force Majeure Event**") will include conditions beyond the reasonable control and without the fault of a Party, such as an act of God, voluntary or involuntary compliance with any regulation, law or order of any government, war, an act of terrorism, civil commotion, labor strike or lock-out, epidemic, failure or default of public utilities or common carriers, destruction of production facilities or materials by fire, earthquake, storm or like catastrophe, inability to procure necessary raw materials in a commercially reasonable manner or default of suppliers or sub-contractors; provided, however, the payment of invoices due and owing hereunder may not be delayed by the payer because of a force majeure affecting the payer.

17.3 **Exclusions of Consequential Damages**. EXCEPT IN THE CASE OF A PARTY'S INTENTIONAL OR WILLFUL MISCONDUCT OR EGREGIOUS BAD FAITH CONDUCT OR DAMAGES AWARDED TO THIRD PARTIES COVERED BY THE INDEMNITY OBLIGATIONS SET FORTH IN SECTION 14, IN NO EVENT WILL EITHER PARTY BE LIABLE TO THE OTHER PARTY FOR LOST PROFITS OR FOR ANY INDIRECT, INCIDENTAL, CONSEQUENTIAL, SPECIAL, PUNITIVE OR EXEMPLARY DAMAGES OF THE OTHER PARTY IN CONNECTION WITH THIS AGREEMENT OR THE TRANSACTIONS CONTEMPLATED BY THIS AGREEMENT, HOWEVER CAUSED, UNDER ANY THEORY OF LIABILITY, AND WHETHER OR NOT SUCH PARTY HAD PRIOR NOTICE THEREOF.

17.4 **Assignment**. Neither Party may assign this Agreement or any of its obligations hereunder without the prior written consent of the other Party; provided, however, that either Party may assign this Agreement in its entirety without such consent to any of its Affiliates, to any purchaser of all, or substantially all, of its assets or to any successor corporation resulting from any merger, consolidation, share exchange, or other similar transaction, and provided further that either Party may assign or sell its rights to receive any amounts due hereunder. This Agreement will inure to the benefit of UGNX and IGIK and their respective successors and permitted assigns. Any assignment of this Agreement that is not made in accordance with this Section 17.4 shall be null and void and of no legal force or effect.

17.5 **Severability**. In the event any one or more of the provisions contained in this Agreement should be held invalid, illegal or unenforceable in any respect, the validity, legality and enforceability of the remaining provisions contained herein will not in any way be affected or impaired thereby, unless the absence of the invalidated provision(s) frustrates the purpose of this Agreement (in which case the Parties will attempt to replace such invalidated provision with an enforceable provision that most clearly implements such purpose). The Parties will in such an instance use their Commercially Reasonable Efforts to replace the invalid, illegal or unenforceable provision(s) with valid, legal and enforceable provision(s) that, insofar as practical, implement the purposes of this Agreement.

17.6 **Notices**. All notices, consents, approvals and other legally operative communications that are required or permitted hereunder ("**Notice**") will be in writing in the English language and sufficient if delivered personally, sent by facsimile or e-mail (and promptly confirmed by personal delivery, registered or certified mail or overnight courier), sent by nationally-recognized overnight courier or sent by registered or certified mail, postage prepaid, return receipt requested, addressed as follows:

Kyowa Hakko Kirin Co., Ltd.
1-6-1 Ohtemachi
Chiyoda-ku, Tokyo, 100-8185, Japan
Attention: Director of Business Development Department
Tel: +81-3-3282-0093
Fax: +81-3-3282-0107

Ultragenyx Pharmaceutical Inc.
60 Leveroni Court
Novato, CA 94949

Attention: Business Development Department/Tom Kassberg
Tel: 415.483.8800
Fax: 415.483.8820
E-mail: Tkassberg@ultragenyx.com

or to such other address as the Party to whom Notice is to be given may have furnished to the other Party in writing in accordance herewith. Any such communication will be deemed delivered: (a) if sent by mail, as aforesaid, on the date upon which the return receipt is signed or delivery is refused or the Notice is designated by the postal authorities as not deliverable, as the case may be; (b) if sent

by facsimile or e-mail, as aforesaid, when sent (with confirmation of receipt); or (c) if sent by courier or hand delivered, as aforesaid, when received. The cost of any translation into English of any communication, document or Notice will be borne solely by the Party providing such communication, document or Notice.

17.7 **Entire Agreement; Amendments**. This Agreement contains the entire understanding of the Parties with respect to the subject matter hereof. All express or implied agreements and understandings, either oral or written, heretofore made are expressly superseded by this Agreement. Except as expressly set forth in this Agreement, this Agreement may be amended, or any term hereof modified, only by a written instrument duly executed by both Parties.

17.8 **Headings**. The captions to the several Articles and Sections hereof are not a part of this Agreement, but are merely guides or labels to assist in locating and reading the several Articles and Sections hereof.

17.9 **Independent Contractors**. It is expressly agreed that KHK and UGNX will be independent contractors and that the relationship between the two Parties will not constitute a partnership, joint venture or agency. Neither KHK nor UGNX will have the authority to make any statements, representations or commitments of any kind, or to take any action, which will be binding on the other, without the prior consent of the other Party.

17.10 **Subcontractors**. Except as otherwise set forth in this Agreement, each Party may engage subcontractors to perform, under its direction, specific and discrete functions that are allocated to it hereunder or that it carries out in the exercise of its rights hereunder, in each case in accordance with this Section 17.10. Each Party shall be fully responsible under this Agreement for the performance hereof by its permitted subcontractors as if such Party so performed this Agreement itself.

17.11 **Waiver**. The waiver by either Party hereto of any right hereunder or the failure to perform or of a breach by the other Party will not be deemed a waiver of any other right hereunder or of any other breach or failure by said other Party whether of a similar nature or otherwise.

17.12 **Counterparts**. This Agreement may be executed in identical duplicate copies exchanged by facsimile or e-mail (PDF form) transmission. The Parties agree to execute two identical original copies of this Agreement after exchanging signed facsimile versions. Each identical counterpart will be deemed an original, but all of which together will constitute one and the same instrument.

17.13 **Waiver of Rule of Construction**. Each Party has had the opportunity to consult with counsel in connection with the review, drafting and negotiation of this Agreement. Accordingly, the rule of construction that any ambiguity in this Agreement will be construed against the drafting Party will not apply.

17.14 **Third Party Beneficiaries**. Except as otherwise expressly provided in this Agreement, nothing herein expressed or implied is intended or will be construed to confer upon or to give to any Third Party any rights or remedies by reason of this Agreement. Except as otherwise expressly provided in this Agreement, there are no intended Third Party beneficiaries under or by reason of this Agreement.

17.15 **Further Assurances**. Upon the other Party's request hereunder, each Party agrees to execute, acknowledge and deliver such further instruments, and to do all such other acts, as may be necessary or appropriate in order to carry out the purposes and intent of this Agreement.

17.16 **Construction of Agreement**. Unless context otherwise clearly requires, whenever used in this Agreement: (a) the words "include" or "including" shall be construed as incorporating, also, "but not limited to" or "without limitation;" (b) the word "day" or "year" means a calendar day or calendar year unless otherwise specified; (c) the words "hereof," "herein," "hereby" and derivative or similar words refer to this Agreement (including all Exhibits); (d) provisions that require that a Party, the Parties or any committee or team hereunder "agree," "consent" or "approve" or the like shall require that such agreement, consent or approval be specific and in writing, whether by written agreement, letter, approved minutes or otherwise; (e) words of any gender include the other gender; (f) references to any specific Applicable Law or article, section or other division thereof shall be deemed to include the then-current amendments thereto or any replacement Applicable Law thereof; and (g) references to either Party include the successors and permitted assigns of that Party. Except as otherwise stated herein, if the terms of this Agreement conflict with the terms of any Schedule or Exhibit, then the terms of this Agreement will govern.

[Remainder of this page is intentionally left blank.]

IN WITNESS WHEREOF, the Parties have executed this Collaboration and License Agreement to be effective as of the Effective Date.

KYOWA HAKKO KIRIN CO., LTD.

By: /s/ Nobuo Hannai
Name: Nobuo Hanai, Ph.D.
Title: President and CEO
Date: August 29, 2013

ULTRAGENYX PHARMACEUTICAL INC.

By: /s/ Emil D. Kakkis
Name: Emil D. Kakkis, MD, PhD
Title: CEO
Date: August 29, 2013

Schedules

Schedule 1.1.15 –Drug Substance Sequence

Schedule 1.1.47 – Licensed Patent Rights

Schedule 4.3.2 – Decision-Making for Day-to-Day Core Development Activities

Exhibits:

Exhibit A – Financial Exhibit

SCHEDULE 1.1.15

Drug Substance Sequence

Amino acid sequence of KRN23

Light chain

[***]

Heavy chain

[***]

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

SCHEDULE 1.1.47

Licensed Patent Rights

"**Licensed Patent Rights**" shall include the following patents and applications:

1. Title: [***] Applicant/Assignee: [***]

Country	Application No.	Filing Date	Patent No.	Issue Date	Status
[***]	[***]	[***]	[***]	[***]	[***]
[***]	[***]	[***]	[***]	[***]	[***]
[***]	[***]	[***]	[***]	[***]	[***]
[***]	[***]	[***]	[***]	[***]	[***]

2. Title: [***] Applicant/Assignee: [***]

Country	Application No.	Filing Date	Patent No.	Issue Date	Status
[***]	[***]	[***]	[***]	[***]	[***]
[***]	[***]	[***]	[***]	[***]	[***]
[***]	[***]	[***]	[***]	[***]	[***]
[***]	[***]	[***]	[***]	[***]	[***]

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

3. Title: [***] Applicant/Assignee: [***]

Country	Application No.	Filing Date	Patent No.	Issue Date	Status
[***]	[***]	[***]	[***]	[***]	[***]
[***]	[***]	[***]	[***]	[***]	[***]
[***]	[***]	[***]	[***]	[***]	[***]

4. Title: [***] Applicant/Assignee: [***]

Country	Application No.	Filing Date	Patent No.	Issue Date	Status
[***]	[***]	[***]	[***]	[***]	[***]
[***]	[***]	[***]	[***]	[***]	[***]
[***]	[***]	[***]	[***]	[***]	[***]
[***]	[***]	[***]	[***]	[***]	[***]
[***]	[***]	[***]	[***]	[***]	[***]
[***]	[***]	[***]	[***]	[***]	[***]

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

SCHEDULE 4.3.2

Decision-Making for Dav-to-Day Core Development Activities

The Lead Development Party shall make all decisions with respect to day-to-day Core Development Activities ("**Day-to-Day Core Development Activities**") including the following types of decisions and activities:

(a) Number of sites, site selection and site management
(b) Vendor management
(c) Site agreements
(d) Vendor agreements
(e) Site documents
(f) Safety reporting
(g) Recruitment materials
(h) Case Report Form (CRF) design
(i) Database design
(j) Tables, Listings and Graphs (TLGs) design
(k) Study unblinding procedures
(l) Data analysis and report generation
(m) Data Monitoring Committee (DMC) membership, charter and conduct
(n) Regulatory meeting preparations and conduct of meetings
(o) Patient identification
(p) Key opinion leader/Principal investigator interactions

EXHIBIT A

Financial Exhibit

Examples of Commercialization Costs

Commercialization Costs shall include, but shall not be limited to, the following, in each case attributable to, or reasonably allocable to, Commercialization of Licensed Products in the Field:

Detailing costs—costs associated with interactive face-to-face visits by a sales representative with a medical professional who has prescribing authority or is able to influence prescribing decisions, within the target audience during which approved uses, safety, effectiveness, contraindications, side effects, warnings or other relevant characteristics of a pharmaceutical product are discussed in an effort to increase prescribing preferences of a pharmaceutical product for its approved uses.

Distribution costs—costs identifiable to the distribution of a Licensed Product to a customer (and not otherwise deducted from Net Sales), including customer and specialty pharmacy services (including costs associated with establishing and running customer call centers), collection of data about sales to hospitals, clinics and other customers, order entry, billing, shipping, logistics, warehousing, product insurance, freight not paid by customers, credit and collection and other like activities the costs of which are includable in "distribution costs" in accordance with GAAP.

Compassionate Use Programs—costs associated with any program to provide patients with products free of charge. These costs include compassionate use programs, provided however that any compassionate use programs prior to Marketing Approval shall be mutually agreed upon by the Parties in advance.

Health Care Reform fees—costs for fees paid to the U.S. government as defined in the Patient Protection and Affordable Care Act and similar taxes and governmental fees.

Marketing Expenses—costs identifiable to the advertising, promotion and marketing of Licensed Products in the Field, and related professional education, including:

a. **Advertising**, which includes costs associated with media costs, direct mails, production expenses, agency fees, and medical congresses and meetings;

b. **Promotion**, which includes costs associated with professional samples, professional literature, promotional material, patient aids, detailing aids, reimbursement of patient assistance programs, public relations and communications expenses, web and social media expenses, patient advocacy support, development of information and data for national accounts, managed care organizations and group purchasing organizations;

c. **Market Research**, which includes costs associated with market information, focus groups, and market research professional staff and related out-of-pocket costs such as travel, business meals, and training;

d. **Marketing Management**, which includes the costs of product management and sales promotion management compensation and departmental expenses as well as costs associated with developing overall sales and marketing strategies and planning;

e. **Reimbursement/Access Services**, which includes costs incurred to manage reimbursement programs, marketing costs (educational material) as well as coupon or co-pay programs; and

f. **Health Policy/Advocacy**, which includes costs associated with advocacy as well as any specific policy lobbying and trade and government relations related expenses.

Medical Affairs Expenses—costs with respect to: medical affairs and other activities associated with clinical studies conducted after Marketing Approval in the Profit Share Territory (to the extent not otherwise included within Development Costs); medical and scientific information and response to external inquiries or complaints; pharmacovigilance, investigator initiated research if not covered in the Core Development Plan, Phase V Clinical Trials, costs of establishing and maintaining patient registries, medical education, Health Economics and Outcomes Research (HECOR, HEMAR), speaker programs, advisory boards, educational grants and fellowships, drug safety, government affairs (including costs associated with compliance with the Sunshine Act and other similar government regulation); and field-based medical science liaisons, medical affairs clinical trial management, MD's in field (separate from medical science liaisons), publications, medical communications and field medical education.

Recall Expenses—costs, to the extent not otherwise covered by KHK's indemnity obligations, associated with notification, retrieval and return of Licensed Products in the Field in the Profit Share Territory, destruction of such returned Licensed Products in the Field, and distribution of the replacement Licensed Products in the Field.

Sales Force FTE Costs—costs associated with the sales force, calculated by multiplying the number of sales representative FTEs dedicated to Licensed Products in the Field in the Profit Share Territory by the applicable Sales Force FTE Rate.

Sales Force FTE Rate—an agreed upon multiple of the gross salaries (including any cash bonus or other performance-based cash incentive payments, to the extent based directly on sales or promotion of Licensed Products in the Field) under and in accordance with the Marketing Plan. The sales force FTE rate includes any automobile allowance, meal expenses, travel/housing for meetings and other incidental expenses incurred by such personnel in the ordinary course of employment.

Selling Costs—all out-of-pocket costs and internal costs (other than costs for sales representatives) directly attributable to selling Licensed Product in the Field, including first line sales managers, exhibits at shows or conventions including samples, charges for space, sales aids and brochures, sales meetings, consultants, call reporting and other Third Party monitoring/tracking services, sales training, sales administration and the like.

UGNX shall reasonably consider any requests by KHK with respect to the choice of vendors or subcontractors for Commercialization activities resulting in material Distribution Costs (customer and specialty pharmacy services), Marketing Research, Reimbursement/Access Services, Medical Affairs Expenses and/or Selling Costs.

LICENSE AGREEMENT

This License Agreement is entered into as of the 1st day of March, 2011 (the "EFFECTIVE DATE") between AAIPharma Services Corp., having its principal offices at 2320 Scientific Park Drive, Wilmington, North Carolina 28405 (hereinafter "AAI"), and Ultragenyx Pharmaceutical, Inc., having principal offices at 77 Digital Drive, Suite 210, Novato, California 94949 (hereinafter "ULTRAGENYX").

WHEREAS, AAI has developed and is the owner of certain know how and proprietary intellectual property related to pharmaceutical preparations of controlled release matrix tablet drug delivery formulations; and

WHEREAS, ULTRAGENYX desires to license certain rights to use such technologies on an exclusive basis, and AAI desires to grant such license to ULTRAGENYX, subject to the terms and conditions of this AGREEMENT.

NOW, THEREFORE, in consideration of the mutual covenants and promises in this AGREEMENT and for other good and valuable consideration, the receipt and sufficiency of which are hereby acknowledged, AAI and ULTRAGENYX agree as follows:

ARTICLE I — RECITALS

1.01 Incorporation of Recitals. The foregoing recitals are hereby incorporated.

ARTICLE II — DEFINITIONS

"AAI CONFIDENTIAL INFORMATION" shall have the meaning set forth in Section 4.03 herein.

"AAI KNOW-HOW" shall mean all discoveries, methods, ideas which are conceived and reduced to practice, information, data, knowledge, processes, specification, designs, trade secrets, show-how and know-how and/or techniques whether or not patentable, which are presently owned, licensed or controlled by AAI (solely or jointly) or during the term of this AGREEMENT are invented, developed, owned, licensed or controlled by AAI (solely or jointly) and which are necessary or useful in the practice of the TECHNOLOGY or AAI PATENTS, or necessary or useful in the development, manufacture, or promotion or sale of any PRODUCT in the LICENSED FIELD, but shall not mean or include AAI PATENT RIGHTS.

"AAI PATENTS" or "AAI PATENT RIGHTS" shall mean any and all rights to all ideas and inventions embodied in the TECHNOLOGY, any and all claims in any and all patents and patent applications owned, controlled or licensed by AAI (solely or jointly) related to the TECHNOLOGY, and further including claims in any reissues, extensions, substitutions, continuations, divisions, continuations-in-part, supplementary protection certificates, registrations, revalidations, additions, renewals, substitutes in the United States and/or any foreign counterparts of any patents involving the TECHNOLOGY or other patents and patent applications encompassed under this definition. AAI PATENTS include, but are not limited to, any of the foregoing related to AAI's proprietary pharmaceutical formulations technology known as [***].

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

"AFFILIATE" shall mean: (i) a corporation or any other entity that directly, or indirectly through one or more intermediaries, owns or controls, is controlled by, or is under common control with, the designated party, but only for so long as the affiliate relationship exists, or (ii) Nobelpharma Co., Ltd. of Japan, but only for so long as the collaboration agreement between Nobelpharma Co., Ltd. and ULTRAGENYX exists. "Control" shall mean (i) the right to vote, or ownership of, shares of stock or other ownership interests or rights having at least fifty percent (50%) of the voting power entitled to vote for the election of directors or their equivalent in the case of a corporation or other entities, (ii) the right to appoint, directly or indirectly, a majority of the board of directors (or other comparable managers) or (iii) the right to control, directly or indirectly, the management or policies of the entity, whether through the ownership of voting securities, by contract or otherwise.

"AGREEMENT" shall mean this License Agreement and the exhibits hereto, as the same shall be amended from time to time.

"COMMERCIALLY REASONABLE EFFORTS" shall mean for efforts by either party hereto, those efforts that a prudent business person or company would expend in the normal course of business to accomplish an important objective (including marketing an important product, as appropriate), but shall not mean efforts that could, if carried out, have a significant negative impact on that party's relevant business unit as a whole.

"DOMINANT IP" shall mean all intellectual property and proprietary rights (whether patentable or not) that now or at any time during the term of this AGREEMENT are owned, controlled or licensed by AAI (solely or jointly) and that dominate or are otherwise required to practice any of the TECHNOLOGY, AAI KNOW-HOW or AAI PATENTS in the LICENSED FIELD.

"GOVERNMENTAL OR REGULATORY AUTHORITY" shall mean any court, tribunal, arbitrator, authority, agency, commission, official or other instrumentality of any country or any state, county, city or other political subdivision.

"LICENSED FIELD" shall mean use of Sialic Acid for treatment of distal myopathy with rimmed vacuoles or for Hereditary Inclusion Body Myopathy (HIBM), in any and all applications, markets and fields of use, whether now known or hereafter existing.

"LICENSED IP" shall mean the TECHNOLOGY, AAI KNOW-HOW, AAI PATENTS and DOMINANT IP.

"LICENSED TERRITORY" shall mean the territory set forth in Exhibit A attached hereto.

"PERSON" shall mean any natural person, corporation, general partnership, limited partnership, proprietorship, other business organization, trust, union, association or Governmental or Regulatory Authority.

"PRODUCT" shall mean any product (or any product made using a process, or any product that when used practices a method) that is covered by any of the AAI PATENT RIGHTS or DOMINANT IP, or that is based on, uses, includes, incorporates or otherwise embodies any of the LICENSED IP.

"SERVICES DATA" means all reports, analyses and data arising out of services performed by AAI for ULTRAGENYX.

"SUBLICENSE REVENUE" means revenue received by ULTRAGENYX (and its AFFILIATES) from NON-AFFILIATE SUBLICENSEES in exchange for a sublicense to practice under the LICENSED IP (including license fees, technology access fees, milestone payments and PRODUCT royalties). SUBLICENSE REVENUE does not include amounts received in exchange for equity, or amounts received for the sale, license, lease or other conveyance of know-how, patent rights or other intellectual property rights that are not LICENSED IP, or amounts received for the provision of scientific, research, development, clinical or other professional services.

"TECHNOLOGY" means AAI's controlled release matrix solid dose oral tablet consisting of certain combinations of generally recognized as safe pharmaceutically acceptable polymers in combinations that result in a controlled release of pharmaceutical active agents, which is sometimes referred to as ProCR™.

"THIRD PARTY" means an entity or person which is not party to this AGREEMENT or an AFFILIATE thereof nor a licensee nor sublicensee of such party or AFFILIATE.

"ULTRAGENYX CONFIDENTIAL INFORMATION" shall have the meaning as set forth in Section 4.02 herein. To avoid uncertainty, ULTRAGENYX CONFIDENTIAL INFORMATION includes the SERVICES DATA.

Unless the context of this AGREEMENT otherwise requires, (i) words of any gender include every other gender; (ii) the terms "hereof", "herein", "hereby" and derivative or similar words refer to this entire AGREEMENT; and (iii) the terms "Article" and "Section" refer to the specified Article and Section of this AGREEMENT. Whenever this AGREEMENT refers to a number of days, such number shall refer to calendar days unless business days are specified. The terms "including" or "includes", when used herein, shall mean "including, without limitation" and "includes, without limitation".

ARTICLE III — GRANT AND CONSIDERATION

3.01 License Grant. AAI hereby grants to ULTRAGENYX and its AFFILIATES a fully paid up, royalty-free, exclusive (even as to AAI), perpetual and irrevocable license (without right to sublicense, except as permitted in Section 3.03) to research, develop, make, have made, offer to sell, sell, have sold, use, have used and import PRODUCTS, and to otherwise practice, commercialize, exploit, use, modify, improve and make derivative works of the LICENSED IP, in each case, solely in the LICENSED FIELD and in the LICENSED TERRITORY.

3.02 Exclusivity. In granting the foregoing exclusive license under Section 3.01, AAI hereby agrees that (i) except at the request of ULTRAGENYX, it shall not research, develop, make, or have made, or (ii) offer to sell, sell, import or use, any PRODUCT (or otherwise practice, commercialize or exploit any of the LICENSED IP) in the LICENSED FIELD anywhere in the LICENSED TERRITORY, and that it shall not transfer or grant to any THIRD PARTY any right, license or option to do so, and that it shall not covenant not to sue any THIRD PARTY in respect of such prohibited activities.

3.03 Sublicense Rights. This AGREEMENT and license granted hereunder are personal to ULTRAGENYX and may not be assigned by ULTRAGENYX, except as permitted under Section 9.01. In addition, ULTRAGENYX shall not sublicense any of its rights under this AGREEMENT without AAI's written consent (such consent not be unreasonably conditioned, delayed or withheld), except that, without consent, ULTRAGENYX may sublicense any or all of its rights hereunder to: (a) any of its AFFILIATES; and/or (b) any other person or entity that is not an AFFILIATE (a "NON-AFFILIATE SUBLICENSEE"), but only pursuant to a written sublicense agreement between ULTRAGENYX and the NON-AFFILIATE SUBLICENSEE that is entered into on an arms-length basis, that includes payment by the NON-AFFILIATE SUBLICENSEE to ULTRAGENYX of a commercially reasonable fee, and that is executed by the parties thereto after the EFFECTIVE DATE. ULTRAGENYX's AFFILIATES may grant further sublicenses in accordance with the provisions of this Section 3.03, but NON-AFFILIATE SUBLICENSEES may not. ULTRAGENYX agrees not to sublicense any use or practice of the LICENSED IP outside the LICENSED FIELD.

3.04 Technology Transfer. Promptly after the EFFECTIVE DATE and on an ongoing basis during the term of this AGREEMENT (as new information becomes available), AAI shall disclose and transfer to ULTRAGENYX all tangible embodiments of the LICENSED IP in the LICENSED FIELD, including without limitation, all patent applications and patents, specifications, designs, documentation and technology implementations and ULTRAGENYX shall pay AAI for time and materials used in such technical transfer at AAI's standard rates.

3.05 Diligence. AAI acknowledges and agrees that nothing in this AGREEMENT shall be construed as ULTRAGENYX's promise or obligation to use best efforts, diligent efforts, COMMERCIALLY REASONABLE EFFORTS or any other level of diligence in commercializing the LICENSED IP or any PRODUCT in the LICENSED FIELD, and AAI hereby waives any such requirement that may be implied at law or otherwise. AAI agrees that any and all business, technical, clinical and regulatory plans, strategies, designs and decisions applicable to commercializing the LICENSED IP in the LICENSED FIELD shall be within ULTRAGENYX's sole discretion. ULTRAGENYX makes no commitment that its efforts will be continuous or successful, and it shall have the unrestricted right to redirect or abandon such activities, at any time, for any reason or no reason.

3.06 Consideration. In consideration of ULTRAGENYX conducting pre-clinical and clinical studies and making all data from such studies available to AAI for use in advancing the LICENSED IP and marketing, and in consideration of the rights and licenses granted to ULTRAGENYX hereunder and the other subject matter of this AGREEMENT, the parties agree as follows:

(a) Sublicense Revenue Share. Within [***] days after the [***], ULTRAGENYX shall pay AAI [***] of the SUBLICENSE REVENUE [***]. At the same time, ULTRAGENYX shall [***] SUBLICENSE REVENUE.

(b) Non Interference. Prior to the [***] anniversary of the EFFECTIVE DATE, AAI and its AFFILIATES shall not undertake or engage in any activity or service related to the development of modified release Sialic Acid formulations, extended release Sialic Acid formulations or other formulations of Sialic Acid, for its own benefit or the benefit of any person or entity (other than ULTRAGENYX and its AFFILIATES).

3.07 Audit. During the term of this AGREEMENT and for [***] thereafter, ULTRAGENYX agrees to keep and maintain accurate records and accounts of [***] received hereunder. Upon reasonable advance written notice (but not more than [***] in any [***] period), AAI shall have the right to have an independent certified public accountant (reasonably acceptable to ULTRAGENYX, and subject to execution of ULTRAGENYX's nondisclosure agreement) to verify ULTRAGENYX's compliance with its reporting and payment obligations under Section 3.06(a) and to report the results of its audit simultaneously to AAI and ULTRAGENYX. No period of time may be audited more than once. ULTRAGENYX shall make all directly applicable books and records available for such inspection during normal business hours at its principal place of business. Any such audit shall be at the expense of AAI, unless it discloses an error for the audited period in excess of [***] percent ([***]%), in which case ULTRAGENYX shall reimburse AAI for its reasonable out-of-pocket audit expenses.

ARTICLE IV — CONFIDENTIALITY

4.01 Disclosure. Upon execution of this AGREEMENT, and thereafter during the term hereon, at such times as the parties shall mutually agree, each party shall disclose to the other, in confidence subject to Section 4.02 and 4.03 hereof, such relevant AAI CONFIDENTIAL INFORMATION and/or ULTRAGENYX CONFIDENTIAL INFORMATION, as the case may be, as is reasonably necessary or useful to allow the other party to proceed with the activities contemplated or permitted by this AGREEMENT, subject to any existing confidentiality agreements.

4.02 Obligations of AAI. Except as specifically authorized by this AGREEMENT, AAI shall, for the term of this AGREEMENT and for [***] thereafter, keep confidential, exercise reasonable safeguards to prevent unauthorized access, use and disclosure, not disclose to others and use and copy only for the purposes provided for or permitted under this AGREEMENT, (a) any information supplied by or for ULTRAGENYX to AAI relating to ULTRAGENYX's or any AFFILIATE's business, employees, investors, finances, technologies, clinical trials or regulatory affairs, and (b) all information and data not described in clause (a) hereof but supplied by or for ULTRAGENYX to AAI under this AGREEMENT that is marked or otherwise identified as "Confidential" or proprietary at time of disclosure or that by its nature would be understood by a reasonable person to be proprietary or confidential, and (c) including all copies, analyses and derivatives thereof (collectively, "ULTRAGENYX CONFIDENTIAL INFORMATION"). Notwithstanding the above, AAI shall have no liability to ULTRAGENYX with respect to the following use, or disclosure to others not a party to this AGREEMENT or an AFFILIATE thereof, of ULTRAGENYX CONFIDENTIAL INFORMATION, as AAI can establish to,

(i) have been known by AAI on a non-confidential basis prior to communication by ULTRAGENYX to AAI;

(ii) have been a matter of public knowledge at the time of such disclosure by ULTRAGENYX to AAI;

(iii) have become a matter of public knowledge, without fault on the part of AAI or an agent thereof, subsequent to disclosure by ULTRAGENYX to AAI;

(iv) have been disclosed to AAI or an AFFILIATE thereof on a non-confidential basis from a THIRD PARTY lawfully having possession of the ULTRAGENYX CONFIDENTIAL INFORMATION without an obligation of confidentiality to ULTRAGENYX; or

(v) was independently developed by or for AAI by persons not having knowledge of such ULTRAGENYX CONFIDENTIAL INFORMATION.

4.03 Obligations of ULTRAGENYX. Except as specifically authorized by this AGREEMENT, ULTRAGENYX shall, for the term of this AGREEMENT and thereafter for so long as AAI's PATENT RIGHTS continue (but no less than [***]), keep confidential, exercise reasonable safeguards to prevent unauthorized access, use and disclosure, not disclose to others and use and copy only for the purposes provided for or permitted under this AGREEMENT, (a) any information supplied by or for AAI to ULTRAGENYX relating to LICENSED IP, (b) all information and data not described in clause (a) hereof but supplied by or for AAI to ULTRAGENYX under this AGREEMENT that is marked or otherwise identified as "Confidential" or proprietary at the time of disclosure or that by its nature would be understood by a reasonable person to be proprietary or confidential, and (c) all copies, analyses, and derivatives thereof (collectively, "AAI CONFIDENTIAL INFORMATION"). Notwithstanding the above, ULTRAGENYX shall have no liability to AAI with respect to the following use, or disclosure to others not a party to this AGREEMENT or an AFFILIATE thereof, of AAI CONFIDENTIAL INFORMATION, as ULTRAGENYX can establish to,

(i) have been known by ULTRAGENYX or any AFFILIATE on a non-confidential basis prior to communication by AAI to ULTRAGENYX;

(ii) have been a matter of public knowledge at the time of such disclosure by AAI to ULTRAGENYX;

(iii) have become a matter of public knowledge, without fault on the part of ULTRAGENYX, or agent thereof, subsequent to disclosure by AAI to ULTRAGENYX;

(iv) have been disclosed to ULTRAGENYX or any AFFILIATE on a non-confidential basis from a THIRD PARTY lawfully having possession of the AAI CONFIDENTIAL INFORMATION without an obligation of confidentiality to AAI; or

(v) was independently developed by or for ULTRAGENYX or any AFFILIATE by persons not having knowledge of such AAI CONFIDENTIAL INFORMATION.

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

4.04 Other Exceptions. In addition, notwithstanding the foregoing, AAI CONFIDENTIAL INFORMATION and/or ULTRAGENYX CONFIDENTIAL INFORMATION, may be (a) disclosed to governmental agencies and others where such AAI CONFIDENTIAL INFORMATION or ULTRAGENYX CONFIDENTIAL INFORMATION may be required to be included in regulatory filings permitted under the terms of this AGREEMENT or in patent applications filed within the United States Patent and Trademark Office or corresponding international patent offices; (b) provided to third parties under appropriate terms and conditions including confidentiality provisions substantially protective of the other party's CONFIDENTIAL INFORMATION as those in this AGREEMENT, but only if such third parties have a bona fide need to know for any purpose expressly permitted by this AGREEMENT; (c) published, if and to the extent such publication has been approved by both parties; or (d) disclosed to the extent required by applicable laws or regulations (including the regulations of the U.S. Securities and Exchange Commission, as determined by the outside SEC legal counsel for the party disclosing the other party's CONFIDENTIAL INFORMATION), pursuant to applicable legal process (including subpoenas and civil investigative demands), or pursuant to an order by a court, governmental agency or other regulatory body having competent jurisdiction, providing, however, that the party intending to disclose under Section 4.04(a) or 4.04(d) shall notify the other party prior to such disclosure and shall cooperate with such other party in the event such other party elects, by timely notice to the disclosing party prior to the disclosure, to legally contest such disclosure or request confidential treatment for or otherwise lawfully avoid disclosure of such information; and provided further, upon the request of the other party from time to time, each party agrees to confirm whether or not it has disclosed the other party's CONFIDENTIAL INFORMATION pursuant to Section 4.04(b) to any third party identified in the other party's request. In each of the foregoing cases, the recipient will use COMMERCIALLY REASONABLE EFFORTS to limit the disclosure and maintain confidentiality to the extent possible. Neither party shall disclose the existence or terms of this AGREEMENT without the other party's prior written consent, except that either party may disclose this AGREEMENT or its terms in connection with any legal or regulatory requirement, financing transaction or due diligence inquiry. In the case of any other publication pertaining specifically to or referencing any LICENSED IP or PRODUCT not covered by one of the permitted exceptions to non-publication above, each party recognizes the mutual interest in obtaining valid patent protection. Consequently, either party and its employees or consultants or any other THIRD PARTY wishing to make a publication (including any oral disclosure made without obligation of confidentiality) relating to work performed under this AGREEMENT or any DEFINITIVE AGREEMENT shall transmit to the other party (the "REVIEWING PARTY") a copy of the proposed written publication at least [***] days prior to submission for publication, or an abstract of such oral disclosure at least [***] days prior to submission of the abstract or the oral disclosure, whichever is earlier. The REVIEWING PARTY shall have the right upon timely written notice to the other party (w) to identify any of the REVIEWING PARTY's CONFIDENTIAL INFORMATION, which the publishing party shall delete and not disclose, (x) to propose modifications and redactions to the publication for patent reasons, which the publishing party shall reasonably consider, (y) to require delay in publication or presentation (for up to [***] days) in order to protect patentable information, or (z) to request that the information

be maintained as a trade secret, and, in such case, the publishing party shall consult with the REVIEWING PARTY in good faith. In the event that a party is legally required to provide a copy of this AGREEMENT or any related document to any THIRD PARTY (except in confidence as permitted by this AGREEMENT), such party shall redact CONFIDENTIAL INFORMATION from such document, except as otherwise required by law. Each party shall have the right to review and approve each redacted document prior to its submission to a THIRD PARTY, where practicable. A period of [***] days shall be allowed for review of a redacted document, with the exception of the order of a court or regulatory agency or other legal compulsion for disclosure on a shorter time basis, for which the maximum reasonable amount of time shall be afforded.

4.05 Ownership. Subject to the license rights granted hereunder, all rights, title and interests (including without limitation, intellectual property and proprietary rights) in and to the AAI CONFIDENTIAL INFORMATION and LICENSED IP shall remain with AAI. All rights, title and interests (including without limitation, intellectual property and proprietary rights) in and to all SERVICES DATA (whether currently existing or generated pursuant to future services) and other ULTRAGENYX CONFIDENTIAL INFORMATION shall remain with ULTRAGENYX. Nothing herein is intended to transfer the ownership of any party's CONFIDENTIAL INFORMATION, KNOW-HOW, or PATENT RIGHTS to the other party. In addition, each party to this AGREEMENT maintains the right to use its own CONFIDENTIAL INFORMATION, KNOW-HOW, and PATENT RIGHTS, for any purpose, except as specifically restricted herein.

ARTICLE V — WARRANTIES AND INDEMNIFICATIONS

5.01 Representations and Warranties.

(a) Representations and Warranties of AAI. AAI hereby represents and warrants, the following:

(i) AAI has and will own all right, title and interest in and to LICENSED IP and has and will maintain the right to grant to ULTRAGENYX the exclusive license set forth in Section 3.01 hereunder.

(ii) AAI has full corporate power and authority to execute and deliver this AGREEMENT and to perform its obligations hereunder and to consummate the transactions contemplated hereby.

(iii) except for ULTRAGENYX, no other PERSON presently has any license, option or other right with respect to the manufacture, use and sale of the PRODUCT or the practice of the LICENSED IP, in each case in the LICENSED FIELD and LICENSED TERRITORY.

(iv) there are no adverse actions, suits or claims pending against AAI or any of its AFFILIATES in or before any GOVERNMENTAL OR REGULATORY AUTHORITY with respect to the LICENSED IP and no such actions, suits or claims have been threatened against AAI or any of its AFFILIATES.

(v) to the best of AAI's knowledge there is no reason why any of the LICENSED IP could be rendered invalid or unenforceable upon challenge and prosecution post filing.

(b) AAI further represents and warrants that during the term of this AGREEMENT, it will not develop or commercialize another PRODUCT in the LICENSED FIELD for sale within the LICENSED TERRITORY, except on the request of ULTRAGENYX.

AAI'S WARRANTIES SET FORTH IN THIS AGREEMENT ARE ITS EXCLUSIVE WARRANTIES TO ULTRAGENYX, AND ARE GIVEN AND ACCEPTED IN LIEU OF ANY AND ALL OTHER WARRANTIES, GUARANTEES, CONDITIONS AND REPRESENTATIONS, EXPRESS OR IMPLIED, INCLUDING ANY IMPLIED WARRANTIES OF MERCHANTABILITY OR FITNESS FOR A PARTICULAR PURPOSE. AAI specifically disclaims any warranty of performance of the LICENSED IP in the LICENSED FIELD.

5.02 AAI Indemnity Obligations. AAI agrees to defend, indemnify and hold ULTRAGENYX and its AFFILIATES harmless from any and all costs, expenses, damages, judgments, and liabilities (including reasonable attorneys' fees) incurred by or rendered against ULTRAGENYX or AFFILIATE as a result of any THIRD PARTY claim or suit brought to the extent resulting from a breach by AAI of its covenants or warranties as set forth herein or AAI's negligence or willful misconduct, or arising out of any illegal or unlicensed use by AAI or its transferees or sub-licensees of ULTRAGENYX or any AFFILIATE's patent rights or other intellectual property or proprietary rights, except for claims to the extent arising from a breach of this AGREEMENT that is caused or aggravated in substantial part by ULTRAGENYX's negligence or a breach of a material representation, warranty, covenant or agreement provided herein by ULTRAGENYX. ULTRAGENYX shall give prompt written notice of any such claim or suit, and AAI shall undertake the defense thereof, at AAI's expense. ULTRAGENYX shall cooperate in such defense, to the extent reasonably requested by AAI, at AAI's expense. ULTRAGENYX shall have the right to participate in such defense, at its own expense, to the extent that in its judgment ULTRAGENYX may be prejudiced thereby. In any claim made or suit brought for which ULTRAGENYX seeks indemnification under this Section 5.02, ULTRAGENYX shall not settle, offer to settle, or admit liability or damages without the prior written consent of AAI.

5.03 Negation of Implications. Notwithstanding any provisions set forth in this AGREEMENT to the contrary, nothing in this AGREEMENT shall be construed as:

(i) a warranty or representation by AAI as to the validity or scope of any AAI PATENT RIGHTS; or

(ii) granting by implication, estoppel, or otherwise, any licenses or rights under LICENSED IP, other than as set forth in the grant provisions of Section 3.01.

Except as otherwise set forth in this AGREEMENT, AAI makes no representations or warranties, and assumes no responsibilities or liabilities whatsoever, with respect to use (including clinical efficacy), sale, or other disposition by ULTRAGENYX or its licensees, sub-licensees or vendees, or other transferees or consumers of any PRODUCT in the LICENSED FIELD.

5.04 ULTRAGENYX Warranties. ULTRAGENYX represents and warrants that:

(a) ULTRAGENYX has full corporate power and authority to execute and deliver this AGREEMENT and to perform its obligations hereunder and to consummate the transactions contemplated hereby; and

(b) ULTRAGENYX will use LICENSED IP only for PRODUCTS in the LICENSED FIELD.

ULTRAGENYX'S WARRANTIES SET FORTH IN THIS AGREEMENT ARE ITS EXCLUSIVE WARRANTIES TO AAI, AND ARE GIVEN AND ACCEPTED IN LIEU OF ANY AND ALL OTHER WARRANTIES, GUARANTEES, CONDITIONS AND REPRESENTATIONS, EXPRESS OR IMPLIED, INCLUDING ANY IMPLIED WARRANTIES OF MERCHANTABILITY OR FITNESS FOR A PARTICULAR PURPOSE.

5.05 ULTRAGENYX Indemnity Obligations. ULTRAGENYX agrees to defend, indemnify and hold AAI harmless from any and all costs, expenses, damages, judgments, and liabilities (including reasonable attorneys' fees) incurred by or rendered against AAI as a result of any THIRD PARTY claim or suit brought to the extent resulting from a breach by ULTRAGENYX of its covenants or warranties as set forth herein or ULTRAGENYX's negligence or willful misconduct, or arising out of any illegal or unlicensed use by ULTRAGENYX or its transferees or sub-licensees of LICENSED IP furnished under this AGREEMENT, and/or out of any use, sale, or other disposition in the LICENSED FIELD by ULTRAGENYX or its licensees, sub-licensees or transferees of PRODUCT(S) in the LICENSED FIELD or by any distributors and/or consumers thereof, except for claims to the extent arising from a breach of this AGREEMENT that is caused or aggravated in substantial part by AAI's negligence or a breach of a material representation, warranty, covenant or agreement provided herein by AAI. AAI shall give prompt written notice of any such claim or suit, and ULTRAGENYX shall undertake the defense thereof, at ULTRAGENYX's expense. AAI shall cooperate in such defense, to the extent reasonably requested by ULTRAGENYX, at ULTRAGENYX's expense. AAI shall have the right to participate in such defense, at its own expense, to the extent that in its judgment AAI may be prejudiced thereby. In any claim made or suit brought for which AAI seeks indemnification under this Section 5.05, AAI shall not settle, offer to settle, or admit liability or damages without the prior written consent of ULTRAGENYX.

5.06 Mitigation. In the event of any occurrence which may result in either party becoming liable under Section 5.02 or 5.05, each party shall use COMMERCIALLY REASONABLE EFFORTS to take such actions as may be reasonably necessary to mitigate the damages payable by the other party under Section 5.02 or 5.05, as the case may be.

ARTICLE VI — PATENT PROSECUTION AND INFRINGEMENT

6.01 Prosecution. Using COMMERCIALLY REASONABLE EFFORTS, AAI shall be responsible, at its sole cost and expense, except as otherwise set forth herein, for the prosecution and maintenance of all AAI PATENTS (including applying for available patent extensions) in the United States. AAI will provide to ULTRAGENYX copies of relevant patent applications and substantive communications to and from the U.S. patent offices. At least [***], AAI agrees to make its patent counsel available to meet (in person or by telephone) with ULTRAGENYX's patent counsel to share information about prosecution and maintenance of the PATENT RIGHTS (e.g., status, issues, upcoming filings) and related matters. AAI will incorporate ULTRAGENYX's reasonable suggestions in filing, prosecuting and maintaining AAI PATENTS relative to the LICENSED FIELD but AAI shall have the sole right as to the final content of any such applications and/or communications.

6.02 Infringement Actions.

(a) ULTRAGENYX and AAI each shall promptly notify each other of any infringement of the LICENSED IP, and/or unauthorized use of any LICENSED IP by one or more THIRD PARTY that may come to its respective attention. AAI shall promptly undertake COMMERCIALLY REASONABLE EFFORTS to obtain a discontinuance of the aforesaid infringement or unauthorized use and, if not successful, AAI may, but is not required to, bring suit against such infringer or unauthorized user. At ULTRAGENYX's election, ULTRAGENYX may join such suit initiated by AAI and contribute to the cost of such proceedings, and in such case ULTRAGENYX shall be entitled to share in any sums recovered pro rata in relation to the extent ULTRAGENYX has contributed following reimbursement of each party's respective costs associated with such actions under this Section 6.02(a),

(b) If AAI fails to obtain a discontinuance of said infringement or unauthorized use and elects not to bring suit against such THIRD PARTY within [***] days after notice thereof, or immediately if AAI elects not to pursue such THIRD PARTY, then in any such event AAI shall give notice in writing to ULTRAGENYX of its failure or election not to bring suit against such infringement or unauthorized use, including such evidence of infringement as AAI may possess, the numbers of the AAI PATENT(S) so infringed and the unauthorized use of LICENSED IP. ULTRAGENYX may, but is not required to, assume sole control and authority to (i) obtain a discontinuance of the infringing operation or unauthorized use or (ii) bring suit against such third party. Any suit by ULTRAGENYX may be either in the name of ULTRAGENYX, or in the name of AAI, or jointly by AAI and ULTRAGENYX, as may be required by the laws of the forum, and, in furtherance of such rights, AAI hereby agrees that ULTRAGENYX may join AAI as a party plaintiff in any such suit.

(c) It is understood and agreed that the party to this AGREEMENT that solely institutes a suit or an action hereunder, without the other party hereto joining in such action or joinder of such other party by operation of law, shall solely bear all costs and expenses associated therewith, and shall be entitled to retain and keep any and all sums received, obtained, collected or recovered whether by judgment, settlement or otherwise, as a result of such suit or action. In addition, with respect to any suit for infringement of the LICENSED IP or unauthorized use of LICENSED IP, the party that did not institute suit shall render all reasonable assistance, cooperation and information, at the instituting party's expense, including executing all documents as may be reasonably requested by the party that did institute suit and making available any relevant records, documents, information, evidence, samples and the like for the action providing such records, documents, information, evidence, samples and the like are not detrimental or adverse to the party not instituting suit.

6.03 Infringement or Unauthorized Use of THIRD PARTY Patent, or KNOW-HOW. Each party hereto shall notify the other promptly in the event of the receipt of notice of any action, suit or claim alleging infringement by the PRODUCT, or of unauthorized use, of any patent held or know-how owned, by a THIRD PARTY in the LICENSED TERRITORY arising from the manufacture, use distribution, sale, or use of the PRODUCTS. The party's whose PRODUCTS are at issue shall have the sole right and authority to defend any such action, suit or claim.

6.04 Settlements. Neither party shall settle or enter into any voluntarily final judgment regarding any action, claim or suit under Section 6.02 or 6.03 that is inconsistent with any provision of this AGREEMENT.

ARTICLE VII — FORCE MAJEURE

7.01 Event of Force Majeure. Neither party shall be responsible or liable to the other hereunder for the failure or delay in the performance of this AGREEMENT due to any civil unrest, war, governmental action, fire, earthquake, hurricane, accident or other casualty, strike or labor disturbance, act of God or the public enemy, or any other contingency beyond the party's reasonable control. In the event of the applicability of this Section 7.01, the party failing or delaying performance shall use COMMERCIALLY REASONABLE EFFORTS to eliminate, cure and overcome any of such causes and resume the performance of its obligations.

7.02 Notification. Upon the occurrence of an event of force majeure, the party failing or delaying performance shall promptly notify the other party in writing pursuant to the provisions of Section 11.04 herein, setting forth the nature of the occurrence, its expected duration, and how such party's performance is affected. The failing or delaying party shall resume performance of its obligations hereunder as soon as practicable after the force majeure event ceases.

ARTICLE VIII — TERM AND TERMINATION

8.01 Term. The Term of this AGREEMENT shall commence on the EFFECTIVE DATE and remain in effect until terminated as provided herein.

8.02 Convenience. ULTRAGENYX may terminate this AGREEMENT for any reason or no reason at any time, upon at least thirty (30) days prior written notice to AAI.

8.03 Termination for Material Breach. If either party breaches or defaults in the performance or observance of any of its material obligations under this AGREEMENT (other than pursuant to one or more events of force majeure as provided for in Article VII herein) and such material breach or default is not cured within sixty (60) days after receipt by such party of a written notice from the non-breaching party specifying the material breach or default (or such longer period as is reasonably necessary if the breach is of such nature that it cannot be reasonably cured within such sixty (60) day period), the non-breaching party shall have the right to terminate this AGREEMENT upon an additional thirty (30) days' written notice to the breaching or defaulting party.

8.04 Rights on Termination. Termination of this AGREEMENT for any reason shall terminate all rights, licenses and obligations of the parties hereunder, except that termination shall be without prejudice to:

(i) either party's rights under this AGREEMENT with respect to obligations accruing prior to termination or claims arising out of events occurring prior to termination;

(ii) any other remedies which either party may otherwise have; and

(iii) all sublicenses properly granted to NON-AFFILIATE SUBLICENSEES by ULTRAGENYX or any AFFILIATE prior to termination, which shall remain in effect in accordance with their terms, and ULTRAGENYX agrees to assign to AAI all such sublicense agreements, and AAI hereby agrees to accept assignment thereof (but not to assume ULTRAGENYX's obligations thereunder); *provided*, after any such assignment, each sublicensee may elect, at its sole discretion, to terminate its sublicense agreement upon written notice to AAI.

8.05 Return of Confidential Information. Upon termination or expiration of this AGREEMENT, and at any other time upon receipt of written notice by the party which previously, and pursuant to this AGREEMENT, disclosed its CONFIDENTIAL INFORMATION (the "DISCLOSING PARTY") to the other party hereto (the "RECEIVING PARTY"), the RECEIVING PARTY shall, as soon as reasonably practicable following receipt of such notice, return all CONFIDENTIAL INFORMATION of the DISCLOSING PARTY'S and, unless otherwise set forth herein, immediately cease and desist all use of such CONFIDENTIAL INFORMATION. Notwithstanding the foregoing, RECEIVING PARTY may continue to possess and use that portion of the other's CONFIDENTIAL INFORMATION as to which this AGREEMENT expressly provides RECEIVING PARTY a continuing right or license to use such CONFIDENTIAL INFORMATION (such as, for example, for the purposes permitted under Section 4.04(a) in connection with regulatory filings and applying for patent protection), subject always to all other restrictions in this AGREEMENT regarding the use and disclosure of CONFIDENTIAL INFORMATION.

8.06 Limitations on Liability for Proper Termination. Neither party shall incur any liability to the other by reason of the permitted termination of this AGREEMENT as provided herein, whether for loss of goodwill, anticipated profits or otherwise, and the parties shall accept all rights granted and all obligations assumed hereunder, including those in connection with such termination, in full satisfaction of any claims resulting from such permitted termination.

ARTICLE IX — ASSIGNMENT

9.01 Assignment. This AGREEMENT and its rights and obligations shall not be assigned or delegated by either party without the prior written consent of the other party, such consent not to be unreasonably withheld, except that without consent, either party may assign this AGREEMENT and its rights and duties under this AGREEMENT, in whole or in part, to any successor (including the surviving company in any consolidation, reorganization or merger), or to any assignee or transferee of all or substantially all of its assets or business, or to any AFFILIATE. This AGREEMENT will be binding upon and inure to the benefit of the successors, representatives and permitted assigns of the parties.

ARTICLE X — DISPUTE RESOLUTION

10.01 Dispute Resolution.

(a) Except as otherwise provided in Section 10.01(c), all disputes or claims which may arise under, out of or in connection with this AGREEMENT (each, a "DISPUTE") will be referred in writing by the party raising the DISPUTE to the person designated in Section 11.04 herein for attempted resolution by good faith negotiations. If the DISPUTE remains unresolved for more than [***] business days after the notice of such DISPUTE, the parties will submit the DISPUTE to the next step in the dispute resolution process set forth in subsection (b) of this Section 10.01.

(b) If any DISPUTE is not resolved in accordance with subsection (a), the DISPUTE will be referred in writing (by the party that originally raised the DISPUTE) to ULTRAGENYX's Chief Executive Officer and AAI's Chief Executive Officer for attempted resolution by good faith negotiations. If they are unable to resolve any DISPUTE within [***] business days after the referral of such DISPUTE to them, the parties will submit the DISPUTE to the next step in the dispute resolution process set forth in subsection (c) of this Section 10.01.

(c) If either party desires to pursue any DISPUTE which is no longer subject to the dispute resolution process as provided in Section 10.01 (a) or (b), such party may submit the DISPUTE to any United States District Court or state court of competent jurisdiction.

(d) No DISPUTE under this AGREEMENT will be the subject of formal judicial proceedings between ULTRAGENYX and AAI before following the dispute resolution procedures set forth in Sections 10.01(a) and (b), except for an action to seek specific performance or injunctive relief to protect CONFIDENTIAL INFORMATION or intellectual property rights pursuant to this AGREEMENT. Accordingly, each party agrees that, in the event of any breach or threatened breach of Section 3.01 or Article IV or any other infringement, misappropriation or violation of its intellectual property rights, the non-breaching party will suffer

irreparable damage for which it will have no adequate remedy at law. Accordingly, the non-breaching party shall, in addition to any other legal or equitable remedies, be entitled to seek an order for specific performance, or an injunction or similar equitable relief against such breach or threatened breach, without the necessity of posting any bond.

(e) Notwithstanding anything to the contrary contained in this AGREEMENT, in the event of a DISPUTE arising out of, relating to or in connection with termination of this AGREEMENT pursuant to Section 8.03 herein, the dispute resolution process set forth in this Section 10.01 is intended to and will toll any cure periods and any notice periods set forth in such Section 8.03.

ARTICLE XI — MISCELLANEOUS

11.01 Waiver and Amendment. Any waiver by any party hereto of a breach of any provisions of this AGREEMENT shall not be implied and no consent or waiver shall be valid unless it is recited in writing and signed by such party. Failure of any party to require, in one or more instances, performance by the other party in strict accordance with the terms and conditions of this AGREEMENT shall not be deemed a waiver or relinquishment of the future performance of any such terms or conditions or of any other terms and conditions of this AGREEMENT. A waiver by either party of any term or condition of this AGREEMENT shall not be deemed or construed to be a waiver of such term or condition for any other term. All rights, remedies, undertakings, obligations and agreements contained in this AGREEMENT shall be cumulative and no one of them shall be a limitation of any other remedy, right, undertaking, obligation or agreement of either party. This AGREEMENT may not be amended except in a writing signed by both parties.

11.02 Relationship of the Parties. For all purposes of this AGREEMENT, AAI and ULTRAGENYX shall be deemed to be independent entities and anything in this AGREEMENT to the contrary notwithstanding, nothing herein shall be deemed to constitute AAI and ULTRAGENYX as partners, joint ventures, co-owners, or an association, nor shall this AGREEMENT constitute any party hereto an employee or agent, legal or otherwise, of the other party for any purposes whatsoever. Neither party hereto is authorized to make any statements or representations on behalf of the other party or in any way obligate the other party, except as expressly authorized in writing by the other party. Anything in this AGREEMENT to the contrary notwithstanding, no party hereto shall assume nor shall be liable for any liabilities or obligations of the other party, whether past, present or future.

11.03 Headings. The headings set forth at the beginning of the various Articles and Sections of this AGREEMENT are for reference and convenience and shall not affect the meanings of the provisions of this AGREEMENT.

11.04 Notices. Notices required under this AGREEMENT shall be in writing and delivered in person or sent by registered or certified mail, postage prepaid and return receipt requested, or by facsimile and confirmed by registered or certified mail, postage prepaid and return receipt requested, or by Federal Express or UPS or other recognized express courier (prepaid with confirmation of delivery), and addressed as follows:

If to ULTRAGENYX: Ultragenyx Pharmaceutical, Inc.
77 Digital Drive, Suite 210
Novato, CA 94949
Attention: Chief Executive Officer

If to AAI: AAIPharma Services Corp.
2320 Scientific Park Drive
Wilmington, NC 28405
Attention: Legal Department

All notices shall be deemed to be effective upon receipt. Either party may change the address at which written notice is to be received pursuant to this Section 11.04.

11.05 Severability. If any provision of this AGREEMENT is held by a court of competent jurisdiction to be invalid or unenforceable, it shall be modified, if reasonably possible, to the minimum extent necessary to make it valid and enforceable or, if such modification is not reasonably possible, it shall be stricken to the minimum extent necessary such that the remaining provisions shall remain in full force and effect.

11.06 Survival. The provisions of Article II (as such definitions pertain to surviving Sections and Articles), Articles IV, V, VIII, IX, X and XI and Sections 3.06 and 3.07 shall survive any termination of this AGREEMENT.

11.07 No Conflict. Each party represents that neither this AGREEMENT nor any of its obligations hereunder will conflict or result in a breach of any arrangement or agreement between such party and any THIRD PARTY.

11.08 Entire Agreement. This AGREEMENT, including the attachments and appendices hereto, sets forth the entire understanding between the parties hereto as to the subject matter hereof and supersedes all other documents, agreements, verbal consents, arrangements and understandings by or between the parties with respect to the subject matter hereof. Prior to the execution of this AGREEMENT, the parties have had numerous discussions, conversations and negotiations, and have generated correspondence, writings and other memoranda with respect to the subject matter hereof. Notwithstanding all of such activities, this AGREEMENT (including the attachments and appendices hereto) is intended to define the full extent of the parties respective agreements, arrangements and obligations with respect to the subject matter hereof, and each party represents that it is not relying on any such other discussions, conversations, negotiations, correspondence, writings and memoranda in executing and delivering this AGREEMENT or performing its respective obligations hereunder. This AGREEMENT may be executed in one or more counterparts, each of which shall be an original, but taken together constituting one and the same instrument. Execution of a facsimile or email (.pdf) copy shall have the same force and effect as execution of an original, and a facsimile or email (.pdf) signature shall be deemed an original and valid signature.

11.09 Limitation of Grant. Except for the limited rights and licenses expressly granted hereunder, no other license is granted (by implication, estoppel, or otherwise) and no other use is permitted.

11.10 Governing Law; Courts. This AGREEMENT shall be governed by, and construed and enforced in accordance with the substantive laws of the State of Delaware, without giving effect to such state's rules concerning conflicts of law.

11.11 No Publicity. Except as may be required by applicable laws or regulations, neither party may use any name, trade name, trademark, or other designation of the other party (or its affiliates or employees) in any press release, advertising, marketing, publicity or other promotional activity without the other party's prior written consent.

IN WITNESS WHEREOF, the parties hereto have caused this AGREEMENT to be executed as of the date first written above by their duly authorized representatives.

AAIPharma Services Corp.

By: /s/ Philippe Maitre
Name: Philippe Maitre
Title: Chief Financial Officer

Ultragenyx Pharmaceutical, Inc.

By: /s/ Emil D. Kakkis
Name: Emil D. Kakkis, MD, PhD
Title: Chief Executive Officer

Exhibit A

Territory

The following countries abbreviated by their two-letter codes for the representation of states, other entities and intergovernmental organizations used by the World Intellectual Property Organization (WIPO):

[***]

The following countries abbreviated by their two-letter codes for the representation of states, other entities and intergovernmental organizations used by the African Regional Intellectual Property Org. (ARIPO):

[***]

The following countries abbreviated by their two-letter codes for the representation of states, other entities and intergovernmental organizations used by Eurasian Patent Organization (EAPO):

[***]

The following countries abbreviated by their two-letter codes for the representation of states, other entities and intergovernmental organizations used by European Patent Office (EPO):

[***]

The following countries abbreviated by their two-letter codes for the representation of states, other entities and intergovernmental organizations used by African Intellectual Property Organization (OAPI):

[***]

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

License Agreement

This License Agreement (this "**Agreement**") is made as of September 20, 2012 (the "**Effective Date**"), by and between Ultragenyx Pharmaceutical Inc., a corporation organized and existing under the laws of the State of Delaware, having its principal place of business at 60 Leveroni Court, Novato, CA 94949 ("**Ultragenyx**"), and Baylor Research Institute, a non-profit corporation organized and existing under the laws of the State of Texas, having its principal place of business at 3310 Live Oak Street, Suite 501, Dallas, Texas 75204 ("**BRI**"). Ultragenyx and BRI are referred to in this Agreement individually as a "**Party**" and collectively as the "**Parties**."

Recitals

WHEREAS, Ultragenyx is a biotechnology company focused on the discovery and development of innovative therapeutics for patients with rare and ultra-rare genetic diseases;

WHEREAS, BRI is a research center focused on finding prevention therapies and treatments for diseases and illnesses;

WHEREAS, BRI owns or controls certain intellectual property related to the Compound (as defined below); and

WHEREAS, the Parties desire for Ultragenyx to obtain certain rights and licenses to such intellectual property pertaining to the Compound in order to develop, manufacture and commercialize prophylactic, therapeutic and diagnostic products pursuant to the terms and conditions set forth herein.

NOW, THEREFORE, in consideration of the foregoing premises and the mutual covenants herein contained, the receipt and sufficiency of which are hereby acknowledged, Ultragenyx and BRI hereby agree as follows:

Article 1

Definitions

The terms in this Agreement with initial letters capitalized, shall have the meanings set forth below, or the meaning as designated in the indicated places throughout this Agreement.

1.1 "Active Ingredient" means a therapeutically active material that provides pharmacological activity in a nutritional or pharmaceutical product (excluding formulation components such as coatings, stabilizers, excipients or solvents, adjuvants or controlled release technologies).

1.2 "Affiliate" means, with respect to a Party, any Entity that controls, is controlled by, or is under common control with that Party. For the purpose of this definition, "control" means, direct or indirect, ownership of more than fifty percent (50%) of the shares of stock entitled to vote for the election of directors, in the case of a corporation, or more than fifty percent (50%) of the equity interest in the case of any other type of legal entity, status as a

general partner in any partnership, or any other arrangement whereby the Entity controls or has the right to control the board of directors or equivalent governing body of a corporation or other entity, or the ability to cause the direction of the management or policies of a corporation or other entity.

1.3 "[***]" means [***].

1.4 "[***] Option" means that certain option pursuant to the option and license agreement between BRI and [***], dated [***] and as amended by the amendment agreement dated [***], which grants [***] the option to obtain a license to develop, manufacture and commercialize Compound and Product in the [***].

1.5 "[***] Option Termination" is defined in Section 2.2(a).

1.6 "BRI Indemnitee" is defined in Section 9.2.

1.7 "BRI Know-How" means, subject to Section 10.2(b), any and all Know-How Controlled by BRI or any of its Affiliates as of the Effective Date or thereafter during the Term that relates to, or is otherwise reasonably necessary or reasonably useful for, the use, development, manufacture or commercialization of any Compound or Product.

1.8 "BRI Patents" means any and all Patents Controlled by BRI or its Affiliate(s) as of the Effective Date or thereafter during the Term that: (i) claim the composition of matter of, or the method of manufacturing or using, any Compound or Product; or (ii) that otherwise relate to, or are reasonably necessary for, the use, development, manufacture or commercialization of any Compound or Product, including the Patents set forth in Exhibit A.

1.9 "BRI Technology" means BRI Know-How and BRI Patents.

1.10 "Business Day" means a day other than (a) a Saturday or Sunday, or (b) a day on which commercial banks located in San Francisco, California are authorized or required by law to be closed.

1.11 "Calendar Quarter" means the respective periods of three (3) consecutive calendar months ending on March 31, June 30, September 30 and December 31.

1.12 "Calendar Year" means a period of twelve (12) consecutive months ending on December 31.

1.13 "Claims" means all Third Party demands, claims, actions, proceedings, orders, findings and verdicts (in contract, tort or otherwise), as well as losses of any type, damages and legal costs resulting therefrom, including, without limitation, any product liability or substantially equivalent claims.

1.14 "Combination Product" means:

(i) a Product that contains both a Compound and one or more other Active Ingredients that are not Compounds;

(ii) a product consisting of one or more separate products packaged together with a Product in a single package or as a unit; or

(iii) a drug, device, test, kit or biological product packaged separately that is sold as a unit with a Product.

1.15 "Commencement" means, with respect to a clinical trial of any Compound or Product, the [***].

1.16 "Commercially Reasonable Efforts" means: (a) where applied to carrying out specific tasks and obligations of a Party under this Agreement other than development, manufacture or commercialization of a Product, expending reasonable, diligent, good faith efforts and resources to accomplish such task or obligation as a similarly situated pharmaceutical or biotechnology company (on its own or acting through any of its Affiliates, sublicensees or subcontractors) would normally use to accomplish a similar task or obligation under similar circumstances; and (b) where applied to the development, manufacture or commercialization of a Product, those reasonable efforts and resources customarily used by such Party with respect to a similar pharmaceutical product Controlled by such Party, which product is at a similar stage in its development or product life and is of similar market potential in the applicable market taking into account efficacy, safety profile, labeling, the then-current and expected competition in the applicable market, the likely timing of entry into the market, the expected extent and speed of market penetration, the patent and other proprietary position of the Product, the likelihood of regulatory approval given the regulatory structure involved, the profitability of the Product, including the cost of manufacture, royalties payable to licensors of patent or other intellectual property rights, alternative products and other relevant factors and other scientific, clinical or commercial factors. With respect to subpart (b) of this definition, Commercially Reasonable Efforts shall be determined on a market-by-market and indication-by-indication basis for a particular Product, and it is anticipated that the level of effort shall be different for different markets and different indications, and shall change over time, reflecting changes in the status of the Product and the market(s) and indication(s) involved.

1.17 "Compound" means any of the following: (a) Triheptanoin; (b) [***], and in each case of (a) and (b), including any [***] thereof.

1.18 "Confidential Information" means all proprietary Know-How, unpublished patent applications and other information and data of a financial, commercial, business, operational or technical nature which: (a) the disclosing Party or any of its Affiliates has supplied or otherwise made available to the other Party or any of its Affiliates in connection with this Agreement, whether prior to or during the Term and whether made available orally, by observation, in writing or in electronic form; or (b) the receiving Party has learned from the disclosing Party in the course of this Agreement, in each case including information comprising or relating to concepts, discoveries, inventions, data, designs or formulae in relation to this Agreement.

1.19 "Control" or **"Controlled"** means, with respect to any Know-How, molecule, material, Patents, other intellectual property, or any proprietary or trade secret information, the legal authority or right (whether by ownership, license or otherwise), as of the Effective Date or

during the Term, to: (i) grant ownership of or a license or sublicense to make, use, offer to sell, sell or import such molecule or material; (ii) grant ownership of or a license or a sublicense under such Know-How, Patents, or intellectual property; or (iii) otherwise disclose such proprietary or trade secret information, in each case without breaching the terms of any agreement with, obligation to or other arrangement with a Third Party, or misappropriating the proprietary or trade secret information of a Third Party; in each case as provided in this Agreement.

1.20 "Disclosing Party" is defined in Section 6.1(a).

1.21 "Dollar" or **"$"** means the legal tender of the United States.

1.22 "EMA" means the European Medicines Agency or any successor entity thereto performing substantially the same functions.

1.23 "Entity" means a partnership, limited partnership, limited liability partnership, corporation, limited liability company, business trust, joint stock company, trust, incorporated association, joint venture or similar entity or organization.

1.24 "European Union" means the European Union member states as then constituted; provided, as of the Effective Date, the European Union member states are Austria, Belgium, Bulgaria, Cyprus, the Czech Republic, Denmark, Estonia, Finland, France, Germany, Greece, Hungary, Ireland, Italy, Latvia, Lithuania, Luxembourg, Malta, the Netherlands, Poland, Portugal, Romania, Slovakia, Slovenia, Spain, Sweden, and the United Kingdom.

1.25 "Fatty Acid Oxidation Disorder" or **"FAOD"** means a group of inherited metabolic disorders associated with a specific enzyme defect in the fatty acid metabolic pathway and affecting utilization of dietary and stored fat. Fatty Acid Oxidation Disorders include, without limitation, disorders of the following enzymes: Carnitine-Acylcarnitine Translocase (CATR), Carnitine Palmitoyltransferase I and II (CPT I, CPT II), Very-Long Chain Acyl-CoA dehydrogenase (VLCAD), L-3-Hydroxy-Acyl-CoA Dehydrogenase (LCHAD), and Mitochondrial Trifunctional Protein (TFP).

1.26 "FD&C Act" means the federal Food, Drug and Cosmetic Act, as amended, and the regulations promulgated thereunder from time to time.

1.27 "FDA" means the United States Food and Drug Administration or any successor entity thereto performing substantially the same functions.

1.28 "Field" means [***].

1.29 "First Commercial Sale" means, with respect to any Product in any country or jurisdiction in the Licensed Territory, the first (1st) *bona fide* commercial sale by or on behalf of Ultragenyx, its Affiliates or sublicensees to a Third Party other than sublicensees for distribution, use or consumption of any such Product in such country or jurisdiction after the Regulatory Approvals and any applicable Pricing Approvals have been obtained for such Product in such country or jurisdiction.

1.30 "Generic Product" means, with respect to a particular Product, and on a country-by-country basis, any nutritional or pharmaceutical product, other than a Product, that contains the same Active Ingredient as such Product and that is commercialized by a Third Party, which

Third Party is not a licensee or sublicensee of Ultragenyx or its Affiliates, or any of their licensees or sublicensees, and has not obtained such pharmaceutical product from a chain of distribution including Ultragenyx or any of its Affiliates, licensees or sublicensees or further sublicensees. The term Generic Product does not include any Product licensed or produced by Ultragenyx or any of its Affiliates or sublicensees (i. e. an authorized generic product).

1.31 "Government Authority" means any federal, state, national, regional, provincial or local government, or political subdivision thereof, or any multinational organization or any authority, agency or commission entitled to exercise any administrative, executive, judicial, legislative, police, regulatory or taxing authority or power, any court or tribunal (or any department, bureau or division thereof, or any governmental arbitrator or arbitral body).

1.32 "IND" means an Investigational New Drug Application submitted to the FDA in conformance with applicable laws and regulations, or the foreign equivalent of any such application in any other country (such as a clinical trial application in the European Union).

1.33 "Indemnified Party" is defined in Section 9.4.

1.34 "Indemnifying Party" is defined in Section 9.4.

1.35 "IMD" is defined in Section 2.4.

1.36 "Know-How" means any and all tangible and intangible information and materials, including research and development data, regulatory submissions and correspondence, manufacturing information and processes, formulations, assays, cell lines, sequences, composition of matter, constructs, discoveries, improvements, modifications, processes, methods, protocols, formulas, utility, data (including physical, chemical, biological, toxicological, pharmacological, analytical, quality control, preclinical, clinical, and veterinary data), results, inventions, know-how and trade secrets, patentable or otherwise, and all other scientific, marketing, financial and commercial information or data, but excluding any of the foregoing to the extent described or claimed in any Patents.

1.37 "Law" means any federal, state, local, foreign or multinational law, statute, standard, ordinance, code, rule, regulation, resolution or promulgation, or any order by any Government Authority, or any license, franchise, permit or similar right granted under any of the foregoing, or any similar provision having the force or effect of law.

1.38 "Licensed Territory" means (a) as of the Effective Date, the United States, Canada and Mexico; or (b) if Ultragenyx exercises the Ultragenyx Option pursuant to Section 2.2(a), worldwide after the Licensed Territory is expanded pursuant to Section 2.2(d).

1.39 "Licensed Territory Product Infringement" is defined in Section 5.3(a).

1.40 "MAA" means a marketing approval application for Regulatory Approval of a Product that is filed with the EMA.

1.41 "Major European Market" means any of the following: [***].

1.42 "Major Market" means [***].

1.43 "Marketing Approval Application" means a BLA, NDA, MAA or similar application for Regulatory Approval that is filed with the applicable Regulatory Authority(ies) in any country or jurisdiction.

1.44 "Net Sales" means, with respect to any Product, the aggregate gross amount invoiced by Ultragenyx, any Affiliate, or sublicensee for sales of such Product to independent, unrelated Third Parties in *bona fide* arms' length transactions, less deductions for:

(a) the costs of packing, transportation, importation, postage, shipping and handling charges, and other charges, such as insurance and customs duties, relating thereto;

(b) any sales, excise or value added taxes imposed on or charged to the selling party and any other charges imposed by a Governmental Authority upon the sale of such Product and actually paid;

(c) trade, quantity, prompt settlement or similar discounts (including chargebacks and allowances) actually granted, allowed or incurred in connection with the sale of such Product that are customary in the trade;

(d) amounts repaid or credited on account of price adjustments, rejection, outdating, billing errors, recalls or return of such Product;

(e) bad debts that have been written off within twelve (12) months after the date of invoice; and

(f) rebates, reimbursements, fees or similar payments to (i) wholesalers and other distributors, pharmacies and other retailers, buying groups (including group purchasing organizations), health care insurance carriers, pharmacy benefit management companies, health maintenance organizations, Governmental Authorities, or other institutions or health care organizations; or (ii) to patients and other Third Parties arising in connection with any program applicable to a Product under which Ultragenyx, its Affiliates, or its sublicensees provides to low income, uninsured or other patients the opportunity to obtain Ultragenyx's pharmaceutical products at no cost or reduced cost.

Sales between Ultragenyx and its Affiliates and sublicensees shall be disregarded for purposes of calculating Net Sales, except if such purchaser is an end user.

If a Product is sold as part of a Combination Product, the Net Sales of such Product for the purpose of calculating royalties owed under this Agreement for sales of such Product under Section 4.4 or determining whether a sales-based milestone payment is due under Section 4.3(a), shall be determined as follows: first, Ultragenyx shall determine the actual Net Sales of such Combination Product (using the above provisions) and then such amount shall be multiplied by the fraction A/(A+B), where A is the invoice price of such Product, if sold separately, and B is the aggregate invoice price for an equivalent dose amount or unit of each other Active Ingredient, drug, device, test, kit or biological product in the Combination Product, if sold

separately. If any other Active Ingredient, drug, device, test, kit or biological product in the Combination Product is not sold separately, Net Sales shall be calculated by multiplying actual Net Sales of such Combination Product by a fraction A/C where A is the invoice price of such Product if sold separately, and C is the invoice price of the Combination Product. If neither the Product nor any other Active Ingredient, drug, device, test, kit or biological product in the Combination Product is sold separately, the adjustment to Net Sales shall be determined by the Parties in good faith to reasonably reflect the fair market value of the contribution of such Product in the Combination Product to the total fair market value of such Combination Product.

1.45 "Option Exercise Fee" is defined in Section 2.2(b).

1.46 "Option Exercise Notice" is defined in Section 2.2(b).

1.47 "Option Period" is defined in Section 2.2(a).

1.48 "Option Territory" means worldwide except for the United States, Canada and Mexico.

1.49 "Patent Counsel" is defined in Section 5.1(a).

1.50 "Patents" means all patents and patent applications and any patents issuing therefrom (which for the purpose of this Agreement shall be deemed to include certificates of invention and applications for certificates of invention), including all divisionals, continuations, substitutions, continuations-in-part, converted provisionals, continued prosecution applications, adjustments, re-examinations, reissues, additions, renewals, revalidations, extensions (including patent term extensions, and supplemental certificates and the like), registrations, pediatric exclusivity periods of any such patents and patent applications, and any and all foreign equivalents of the foregoing.

1.51 "Person" means any individual, Entity or Governmental Authority.

1.52 "Phase 3 Clinical Trial" means a human clinical trial, the principal purpose of which is to establish safety and efficacy in patients and which is designed and intended to serve as a pivotal study to support the filing of an NDA for the indication being studied, all in accordance with the trial protocol.

1.53 "Pricing Approvals" means, with respect to a Product in any country or jurisdiction, all pricing and reimbursement approvals for the Product from Government Authorities required by applicable Law or Governmental Authorities.

1.54 "Product" means any nutritional or pharmaceutical product, including all dosage forms and formulations, containing one or more Compound(s) as an Active Ingredient(s) (alone or as part of a Combination Product). Except when referred to in the Net Sales definition in describing how to calculate the Net Sales of Combination Products, all references to Product in this Agreement shall be deemed to include Combination Products. Two Products shall be deemed the same Product if they contain the same Compound as the Active Ingredient.

1.55 "Product Marks" is defined in Section 3.6(b).

1.56 "Proof of Concept Study" or "POC Study" means a human clinical trial of a Compound or Product conducted by Ultragenyx or any Affiliate to demonstrate preliminary clinical safety and efficacy with a small number of strictly selected patients.

1.57 "Receiving Party" is defined in Section 6.1(a).

1.58 "Regulatory Approval" means, with respect to a Product in any country or jurisdiction, the approvals by the applicable Regulatory Authority in such country or jurisdiction (other than Pricing Approvals) necessary for the commercialization of such Product.

1.59 "Regulatory Authority" means any applicable Government Authority responsible for granting Regulatory Approvals for Products, including the FDA, the EMA and any corresponding national or regional regulatory authorities.

1.60 "Regulatory Exclusivity" means, with respect to a Product, any market exclusivity granted by government or Regulatory Authority to exclude Third Parties from the development and/or commercialization of any Products containing as its Active Ingredient the same Compound as the Product.

1.61 "Regulatory Filings" means, with respect to the Compounds or Products, any submission to a Regulatory Authority of any appropriate regulatory application specific to Compounds or Products, and shall include any submission to a regulatory advisory board and any supplement or amendment thereto. **"Regulatory Filings"** includes any IND, NDA, BLA and any Marketing Approval Application.

1.62 "Retained Territory" means all countries of the world other than the Licensed Territory. For clarity, if Ultragenyx exercises the Ultragenyx Option pursuant to Section 2.2(a), there will be no countries, jurisdictions or territories in the Retained Territory.

1.63 "Retained Territory Product Infringement" is defined in Section 5.4(a).

1.64 "Royalty Report" means a written report or reports showing, with respect to a given Calendar Quarter, on a Product-by-Product basis: (a) the calculation of Net Sales for each such Product during such Calendar Quarter; (b) any applicable currency conversions; and (c) the royalties payable with respect to such Net Sales in United States Dollars.

1.65 "Royalty Term" has the meaning set forth in Section 4.4(b).

1.66 "Safety Data Exchange Agreement" has the meaning set forth in Section 3.4.

1.67 "Term" is defined in Section 7.1.

1.68 "Third Party" means any Person other than a Party or an Affiliate of a Party.

1.69 "Third Party License" is defined in Section 4.4(d).

1.70 "Third Party Patent Proceeding" is defined in Section 5.5.

1.71 "Ultra Orphan Indication" means, on a country-by-country basis, an indication other than FAOD for which a Product has been granted orphan drug exclusivity under Section 527 of the FD&C Act, or has been granted a corresponding exclusivity under the applicable Laws of another country or jurisdiction within the Licensed Territory, and which affects fewer than 20,000 people in the US.

1.72 "Ultragenyx Indemnitee" is defined in Section 9.1.

1.73 "Ultragenyx Option" is defined in Section 2.2(a).

1.74 "Ultragenyx Option Notice" is defined in Section 2.2(a).

1.75 "United States" or **"US"** means the United States of America including its territories and possessions.

1.76 "Valid Claim" means, with respect to any country a claim of any issued and unexpired patent included in the BRI Patents (as may be extended through supplementary protection certificate or patent term extension or the like) that has not been revoked, abandoned, held invalid, unpatentable or unenforceable by a patent office, court or other governmental agency of competent jurisdiction in a final and non-appealable judgment (or judgment from which no appeal was taken within the allowable time period) and which claim has not been disclaimed, denied or admitted to be invalid or unenforceable through reissue, re-examination, opposition or disclaimer or otherwise, or lost in an interference proceeding.

1.77 Interpretation. In this Agreement, unless otherwise specified:

(a) "includes" and "including" means respectively includes and including without limitation;

(b) words denoting the singular shall include the plural and vice versa and words denoting any gender shall include all genders;

(c) the word "or" shall not be deemed to be used in the exclusive sense and shall instead be used in the inclusive sense to mean "and/or";

(d) words such as "herein", "hereof', and "hereunder" refer to this Agreement as a whole and not merely to the particular provision in which such words appear; and

(e) the Exhibits and other attachments form part of the operative provision of this Agreement and references to this Agreement shall include references to the Exhibits and attachments.

Article 2

License

2.1 License to Ultragenyx.

(a) BRI hereby grants to Ultragenyx (i) an exclusive license, with the right to grant sublicenses in multiple tiers, under the BRI Technology to research, develop, make, have made, use, offer to sell, sell, have sold, import and export Compounds and Products in the Field in the Licensed Territory; and (ii) a non-exclusive license, with the right to grant sublicenses in multiple tiers, to use BRI Know-How relating to Products for research purposes, and for commercial purposes for exploitation of any BRI Patents not otherwise exclusively committed to a Third Party outside of the Licensed Territory. For clarity, the license granted to Ultragenyx under this Section 2.1(a) does not include the right for Ultragenyx to practice any BRI Patent or use any BRI Know-How to develop, make, use or sell compounds or products that are proprietary to BRI other than Compounds or Products.

(b) Ultragenyx may exercise its rights and perform its obligations under this Agreement by itself or through any of its Affiliates or Third Party sublicensees or contractors without the prior written consent of BRI. Ultragenyx shall remain responsible for all of its obligations under this Agreement that have been delegated, subcontracted or sublicensed to any of its Affiliates, sublicensees or contractors.

(c) After the expiration of the Royalty Term for a particular Product in a country within the Licensed Territory, the licenses granted to Ultragenyx by BRI under this Section 2.1 shall become fully paid-up, irrevocable and perpetual licenses for such Product in the Field in such country.

(d) All licenses granted to Ultragenyx under this Agreement are granted subject to (i) any limitations imposed by the terms of any government grant, government contract, or government cooperative agreement applicable to the BRI Patent Rights and (ii) applicable requirements of 35 U.S.C. Sections 200 et seq., as amended, and implementing regulations and policies.

2.2 Ultragenyx Option to Expand Licensed Territory.

(a) The Parties acknowledge and agree that, as of the Effective Date, the Licensed Territory is limited to the United States, Canada and Mexico as a result of the [***] Option. The [***] Option, if not exercised by [***], will expire on December 31, 2012. BRI shall promptly provide Ultragenyx with written notice if (i) the [***] Option lapses either as a result of (A) [***] not exercising such [***] Option; or (B) any waiver or early termination of the [***] Option or otherwise (the "[***] **Option Termination**"), and/or (ii) [***] exercising the [***] Option and the resulting license agreement between [***] and BRI subsequently terminating or expiring (such notice in each of (i) and (ii), the "**Ultragenyx Option Notice**"). BRI hereby grants to Ultragenyx an exclusive option, to expand the Licensed Territory to be worldwide (the "**Ultragenyx Option**"), exercisable in the case of (i) above at any time after Ultragenyx receives the Ultragenyx Option Notice and before June 30, 2013 and in the case of (ii) above within three (3) months after Ultragenyx receives the Ultragenyx Option Notice (the "**Option Period**"). If [***] exercises the [***] Option and subsequently obtains a license to develop, manufacture and commercialize Compound and Product in the Option Territory, BRI and Ultragenyx shall use reasonable efforts to approach [***] to discuss in good faith the coordination of such development, manufacture and commercialization of Compound and Product by [***] and Ultragenyx in separate territories, provided, that there is no obligation for any party to agree to any such coordination, except that neither [***] nor Ultragenyx can commercialize Compound or Product in the other party's territory.

(b) Ultragenyx may exercise the Ultragenyx Option by providing written notice to BRI (an "**Option Exercise Notice**") at any during the Option Period and by paying BRI the one-time option exercise fee of Seven Hundred Fifty Thousand Dollars ($750,000) (the "**Option Exercise Fee**").

(c) Within ten (10) days of the date of the Ultragenyx Option Notice, BRI shall provide Ultragenyx with a good faith estimate of the BRI Patent related costs and expenses that BRI anticipates incurring for the BRI Patents for the Option Territory from the date of the Ultragenyx Option Notice through the Option Expiration Date. Upon its receipt of Ultragenyx's Option Exercise Notice, BRI shall provide Ultragenyx with a final reasonably detailed accounting of all BRI Patent related costs and expenses incurred after the date of the Ultragenyx Option Notice until the date of receipt of such Option Exercise Notice. Within thirty (30) days of receipt of such final accounting, Ultragenyx shall reimburse BRI for any such reasonable BRI Patent related costs and expenses to the extent BRI has not been otherwise reimbursed for such costs and expenses.

(d) Upon BRI's receipt of the Option Exercise Notice and the Option Exercise Fee, the definition of Licensed Territory shall automatically be expanded to be worldwide.

2.3 BRI's Retained Rights. BRI retains the right to practice the BRI Technology outside the scope of the license granted to Ultragenyx in Section 2.1 (as may be expanded by Section 2.2), and the right to use the BRI Know-How for internal, not-for-profit research purposes. In addition, BRI shall have the right to continue to hold the IND for and conduct and complete the clinical trial that is being sponsored by BRI as of the Effective Date titled: "[***]" (the "**BRI Ongoing Study**") in accordance with the protocol existing as of the Effective Date; provided, that BRI may only (i) modify the protocol for the Ongoing Study, (ii) increase enrollment for the Ongoing Study or (iii) change the enrollment criteria for the Ongoing Study as follows: BRI shall discuss any such matters with Ultragenyx in good faith and BRI shall use best efforts to incorporate any of Ultragenyx's comments on such matters. During the Term, BRI shall have the right to conduct additional clinical trials using the Compound under the direct supervision of [***]; provided, that BRI shall first notify Ultragenyx of any such proposed clinical trial and discuss any opportunity for collaboration on such clinical trial between BRI and Ultragenyx and, in the event the Parties agree for BRI to conduct such clinical trial independently, BRI shall conduct such clinical trial pursuant to a protocol to be agreed upon by Ultragenyx and BRI.

2.4 Collaborative Research Agreement. The Parties shall negotiate in good faith the terms and conditions for a collaborative research agreement between Ultragenyx and BRI's Institute of Metabolic Disease ("**IMD**") regarding the Compound and such terms and conditions shall include (i) financial and technical support from Ultragenyx for the ongoing Phase 2 Clinical Trial in adult polyglucosan body disease and/or other development work at IMD and (ii) technical and scientific support from IMD to Ultragenyx in support of its development of the Compound and the Products. Neither Party shall be obligated to enter into such collaborative research agreement if the Ultragenyx and BRI cannot reach agreement on such terms and conditions after such good faith negotiations.

2.5 No Implied Licenses. Except as set forth herein, neither Party shall acquire any license or other intellectual property interest, by implication or otherwise, under any trademarks, Patents, Know-How or other intellectual property Controlled by the other Party.

2.6 Technology Transfer. In addition to BRI's obligation to provide information and documents related to BRI Know-How in accordance with Section 3.5(b), promptly after the Effective Date, BRI shall use best efforts, at no additional cost to Ultragenyx, to disclose and provide to Ultragenyx all BRI Know-How pertaining to the manufacture and development of any Compounds or Products that are Controlled by BRI as of the Effective Date, including all data from any and all clinical trials and preclinical studies, to the extent such BRI Know-How has not previously been provided to Ultragenyx. On a continuing basis during the Term, BRI shall, at no additional cost to Ultragenyx, disclose and provide to Ultragenyx additional BRI Know-How pertaining to the manufacture and development of any Compounds or Products that comes into existence or comes to BRI's attention after the Effective Date.

Article 3

Development, Manufacture and Commercialization

3.1 General. Subject to the terms and conditions of this Agreement, as between the Parties, Ultragenyx (on its own or acting through or together with any of its Affiliates, sublicensees or contractor manufacturers) shall have the sole and exclusive right to develop (including making Regulatory Filings and seeking Regulatory Approvals), manufacture and commercialize Compounds and Products in the Field in the Licensed Territory, at its sole discretion and at its cost and expense. The Parties acknowledge and agree that except as otherwise expressly provided in this Agreement, Ultragenyx shall have no obligation to disclose or share with BRI any preclinical or clinical data and information unless or until Ultragenyx exercises the Ultragenyx Option pursuant to Section 2.2(b).

3.2 Diligence.

(a) Ultragenyx (on its own or acting through any of its Affiliates, sublicensees or subcontractors) shall use Commercially Reasonable Efforts to develop and commercialize at least one (1) Product in FAOD and at least (1) Product in [***]. Specifically, Ultragenyx will use Commercially Reasonable Efforts to (a) [***]; (b) [***]; (c) [***]; and (d) within [***] after the Effective Date, perform at least one of the following: (i) [***]; or (ii) [***]; in each case of (a)-(d) above, provided that each such timeline shall be extended to account for any delay resulting from factors beyond Ultragenyx's reasonable control, including regulatory, medical, safety or efficacy delays.

(b) If Ultragenyx shall fail to achieve such milestones within such applicable time frame(s) (as such time frame(s) may be extended pursuant to 3.2(a) above), BRI may provide written notice to Ultragenyx and upon receipt of any such notice, the Parties shall discuss in good faith Ultragenyx's progress for the development of such Product in the

applicable jurisdiction and the Parties may agree on an amended timeline or a plan for Ultragenyx to continue its development of the Product. Each agreed upon [***] extension of the timeline shall be subject to the payment by Ultragenyx of an extension fee of [***]. For clarity, if a delay in timeline is due to new regulatory, development, or safety requirements or other similar actions by regulatory authorities or other government agencies, such timeline is not subject to the payment by Ultragenyx of an extension fee.

(c) For the purposes of this Section 3.2(c), all FAOD indications shall be deemed one "Indication Cluster" and all [***] shall be deemed another "Indication Cluster." On an Indication Cluster-by-Indication Cluster basis, in the event Ultragenyx fails to satisfy such new timeline or plan that is specific to a particular Indication Cluster, then upon written notice by BRI, the license granted to Ultragenyx pursuant to Section 2.1(a) shall become non-exclusive solely for such Indication Cluster, provided that all obligations for payment of royalties or milestones remain the same for such Indication Cluster and provided, that, if Ultragenyx subsequently obtains Regulatory Approval for the Product for such Indication Cluster, the license under Section 2.1(a) shall thereupon automatically convert back to an exclusive license for such Indication Cluster so long as BRI still maintains the right to grant an exclusive license to Ultragenyx at such time. In the event of the conversion of the license to non-exclusive, any sole and/or exclusive rights that Ultragenyx has under this Agreement with respect to such non-exclusive Indication Cluster, including without limitation such rights under Section 3.1, this Section 3.2, and Section 3.3 shall also automatically become non-exclusive. Further, on an Indication Cluster-by-Indication Cluster basis, Ultragenyx's rights regarding BRI Independent Studies under Sections 3.3 (i)-(iv), and Ultragenyx's rights under Sections 5.1(a), 5.3(b), Section 5.4(b), Section 5.5 (a) and (b), and Section 5.6, as well as BRI's corresponding obligations to Ultragenyx with respect to those Sections, in each case as such rights and obligations apply to any non-exclusive Indication Cluster, shall be suspended for so long as such license remains non-exclusive and shall be automatically reinstated if and when such license becomes exclusive pursuant to the foregoing. Failure to achieve such milestones shall not be a material breach of this Agreement. The remedy provided for pursuant to this Section 3.2 shall be BRI's sole and exclusive remedy for or relating to Ultragenyx's failure to achieve any milestone. If the license granted to Ultragenyx pursuant to Section 2.1(a) is converted to a non-exclusive license for a particular Indication Cluster pursuant to the terms of this Section 3.2, BRI shall promptly provide Ultragenyx with written notice of any Third Party license that BRI grants under the BRI Technology relating to Compounds or Products for such Indication Cluster.

3.3 Regulatory. As between the Parties, Ultragenyx (on its own or acting through or together with any of its Affiliates, sublicensees or contractor manufacturers) has the sole right to: (a) make all Regulatory Filings, submissions, reports, updates and supplements with any Regulatory Authority with respect to any Compound or Product in the Licensed Territory; (b) obtain, hold and maintain all INDs (other than the IND for the BRI Ongoing Study), Regulatory Approvals and Pricing Approvals in the Field in the Licensed Territory in the name of Ultragenyx or any of its Affiliates or sublicensees; and (c) conduct all meeting and discussions and handle all correspondence with any Regulatory Authority related to any Compound or Product in the Licensed Territory. For the BRI Ongoing Study and any additional study that BRI independently conducts pursuant to Section 2.3 ("**BRI Independent Studies**"): (i) BRI shall promptly provide Ultragenyx with copies of all material documents, information and

correspondence that are received from any Regulatory Authority; (ii) BRI shall provide Ultragenyx with copies of all material documents, information and correspondence that are planned for submission to any Regulatory Authority and BRI shall cooperate with and consider in good faith any input from Ultragenyx in preparing such submission; (iii) BRI shall provide Ultragenyx with reasonable advance notice of all meetings, conferences and discussions scheduled with any Regulatory Authority concerning such study(ies) and BRI shall consider in good faith any input from Ultragenyx in preparing for such meetings, conferences or discussions; and (iv) to the extent permitted by applicable Laws and by any obligations to Third Parties that (A) exist as of the Effective Date or (B) come into existence after the Effective Date, provided that BRI shall use Commercially Reasonable Efforts to obtain such rights for Ultragenyx in its negotiations with Third Parties after the Effective Date, Ultragenyx shall have the right to participate in any such meetings, conferences or discussions and BRI shall facilitate such participation. If either (x) [***] exercises the [***] Option or (y) Ultragenyx does not exercise the Ultragenyx Option, then, in each case, BRI and Ultragenyx shall each use reasonable efforts to facilitate global coordination of clinical data and safety data between Ultragenyx and [***] and/or any other applicable Third Party licensee of BRI in the Retained Territory in order for Ultragenyx, [***] and/or any Third Party licensee to comply with regulatory requirements.

3.4 Pharmacovigilance. If either (a) [***] exercises the [***] Option or (b) Ultragenyx does not exercise the Ultragenyx Option, then, in each case, BRI and Ultragenyx shall use reasonable efforts to facilitate BRI, Ultragenyx and [***] and/or any other applicable Third Party licensee of BRI in the Retained Territory entering into a safety data exchange agreement ("***Safety Data Exchange Agreement***") setting forth in detail the pharmacovigilance alert process and data exchange with respect to the Product to comply with all applicable legal obligations of Regulatory Authorities in the Licensed Territory and in the Retained Territory. For the Ongoing Study and any additional study that BRI independently conducts pursuant to Section 2.3, BRI shall cooperate with Ultragenyx and BRI shall transfer all safety data relating to the Product to Ultragenyx pursuant to a procedure to be agreed upon by the Parties; in the event of conversion of the license to non-exclusive, Ultragenyx shall also provide a reciprocal transfer of all its safety data to BRI.

3.5 Manufacture; Provision of Know-How.

(a) Subject to Section 3.5(b), Ultragenyx (on its own or acting through any of its Affiliates, sublicensees or subcontractors) shall be responsible for the manufacture and supply of Compounds and Products in the Licensed Territory for use by Ultragenyx, its Affiliates and sublicensees, at its cost and expense, itself or through one or more contract manufacturers or sublicensees. BRI shall use Commercially Reasonable Efforts to assist Ultragenyx in establishing a direct relationship with BRI's current supplier of Compounds.

(b) Promptly after the Effective Date, BRI shall provide Ultragenyx with all BRI Know-How that is necessary or reasonably useful to manufacture Compound or Product, including any and all reports and documentation from BRI's current or former supplier of Compounds that BRI does not have the ability to disclose to Ultragenyx under express or implied obligations of confidentiality.

3.6 Commercialization.

(a) Ultragenyx (on its own or acting through any of its Affiliates, sublicensees or subcontractors) shall book sales for the Products in the Licensed Territory and shall have sole control over pricing and other commercialization decisions with respect to the Products in the Licensed Territory.

(b) As between the Parties, Ultragenyx (on its own or acting through any of its Affiliates, sublicensees or subcontractors) shall have the right to brand commercialized Products using Ultragenyx trademarks and any other trademarks and trade names it determines appropriate for the Products, which may vary by country or within a country ("**Product Marks**"). Ultragenyx (on its own or acting through any of its Affiliates, sublicensees or subcontractors) shall own all rights in the Product Marks and shall be responsible for registration, maintenance, and defense of the Product Marks at its own costs and expense.

Article 4

Financial Provisions

4.1 Upfront Payment. In consideration for the license granted under this Agreement, Ultragenyx shall pay to BRI a one-time, non-refundable, non-creditable upfront payment of Two Hundred Fifty Thousand Dollars ($250,000) within thirty (30) days after the Effective Date.

4.2 Milestone Payments.

(a) Development and Regulatory Milestones. Ultragenyx shall pay to BRI the following one-time, non-refundable, non-creditable development and regulatory milestone payments upon the achievement of the corresponding milestone by Ultragenyx or any of its Affiliates or sublicensees:

Development and Regulatory Milestones	Payments
(i) [***]	$ [***]
(ii) [***]	$ [***]
(iii) [***]	$ [***]
(iv) [***]	$ [***]
(v) [***]	$ [***]
(vi) [***]	$ [***]
(vii) [***]	$ [***]
(viii) [***]	$ [***]
(ix) [***]	$ [***]
(x) [***]	$ [***]

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

(b) The Parties acknowledge and agree that the milestones set forth in Section 4.2(a)(ii), (vii), (viii), (ix) and (x) above will not be due if such Product triggering the milestone is in an indication that is not covered by a Valid Claim and if Ultragenyx has a financial obligation to pay a milestone pursuant to a Third Party License (as defined in Section 4.4(d) below) on such Product for such indication.

(c) The milestones set forth in Section 4.2(a) shall each be due after the first (1st) achievement of such milestone for the first Product to achieve such milestone by or on behalf of Ultragenyx or any of its Affiliates or sublicensees, and each such milestone payment shall be payable only once, regardless of how many Products or how many indications for which such milestone has been achieved.

(d) For clarity, the milestones set forth in sub-Section 4.2(a)(i), (ii), (v), (vi), (ix) and (x) above may only be due if Ultragenyx exercises the Ultragenyx Option pursuant to Section 2.2(b).

(e) Ultragenyx shall notify BRI in writing promptly upon each milestone event set forth in this Section 4.2 and BRI shall thereafter submit an invoice to Ultragenyx for the milestone payment corresponding to such milestone event as set forth in this Section 4.2. Ultragenyx shall make such applicable milestone payment within [***] days after receipt of such invoice from BRI.

4.3 Sales Milestone Payments.

(a) Ultragenyx shall pay to BRI the non-refundable, non-creditable sales milestone payments set forth below. The sales milestones shall each be due after the first achievement of such milestone for annual aggregate Net Sales of all Products by or on behalf of Ultragenyx or any of its Affiliates or sublicensees, and each such milestone payment shall be payable only once, regardless of how many Calendar Years during which such sales milestone have been reached.

Sales Milestones	Payments
First (1st) Calendar Year in which aggregate total Net Sales by Ultragenyx, its Affiliates and sublicensees for all Products throughout the Licensed Territory exceed $[***]	[***]
First (1st) Calendar Year in which aggregate total Net Sales by Ultragenyx, its Affiliates and sublicensees for all Products throughout the Licensed Territory exceed $[***]	$ [***]

(b) Ultragenyx shall notify BRI in writing within [***] days following the end of the Calendar Year during which any milestone event is achieved and BRI shall thereafter submit an invoice to Ultragenyx for the corresponding milestone payment as set forth in Section 4.4(a). Ultragenyx shall make such applicable milestone payment within [***] days following receipt of such invoice.

4.4 Royalty Payments.

(a) Royalty Rates for Products. Subject to the other terms of this Section 4.4, during the Royalty Term, Ultragenyx shall make quarterly royalty payments to BRI equal to [***] percent ([***]%) of the Net Sales of the Products in the Licensed Territory by Ultragenyx and any of its Affiliates or sublicensees.

(b) Royalty Term. On a Product-by-Product basis and country-by-country basis, Ultragenyx's royalty payment obligations under this Section 4.4 shall commence upon the First Commercial Sale of such Product in such country and expire upon the later of: (i) the expiration of the period of the first Regulatory Exclusivity granted by the applicable Regulatory Authority applicable to such Product in such country in connection with approval in such country for either FAOD or an Ultra Orphan Indication; or (ii) the expiration of the last-to-expire Valid Claim included in BRI Patents claiming the composition of matter of, or the method of making or using, such Product in such country in connection with approval in such country for FAOD or an Ultra Orphan Indication ("**Royalty Term**").

(c) Royalty Reduction for a Product Subject to Generic Competition. For a particular Product in a particular country, during any period during the Royalty Term if one (1) or more Generic Product(s) with respect to such Product is being sold in such country then the applicable royalty rate under this Section 4.4 shall be reduced by [***] percent ([***]%) for such Product in such country.

(d) Royalty Reduction for Third Party Payment Obligations. If Ultragenyx (or any of its Affiliates or sublicensees) enters into any agreement with a Third Party under which Ultragenyx obtains rights under any intellectual property (including any Patent or Know-How) Controlled by a Third Party, which is necessary to the use, development, manufacture, commercialization or import of any Compound or Product (including for the use or commercialization of any Compound or Product in a particular indication) (each, a "**Third Party License**"), Ultragenyx shall have the right to credit up to [***] percent ([***]%) of the amounts owed by Ultragenyx under any such Third Party License against Ultragenyx's royalty payments to BRI under this Section 4.4 for the same Product, on a country-by-country basis, provided that, by operation of this Section 4.4(d), Ultragenyx's royalty payment obligation to BRI shall not be reduced by more than [***] percent ([***]%) for the first such Third Party License or [***] percent ([***]%) for the second and any subsequent such Third Party License(s). For clarity, any applicable royalty reduction pursuant to Section 4.4(c) shall be in addition to any applicable royalty reduction pursuant to this Section 4.4(d) and such royalty reduction is not included within the aforementioned limits.

4.5 Reports; Payment of Royalty; Annual Reconciliation. During the Term, following the First Commercial Sale of a Product and on a Calendar Quarter basis, Ultragenyx shall furnish to BRI a Royalty Report. Reports shall be due within [***] days following the close of each Calendar Quarter. Royalties shown to have accrued by each Royalty Report shall be due and payable on the date such royalty report is due. Ultragenyx shall keep complete and accurate records in sufficient detail to enable the royalties payable hereunder to be determined.

4.6 Currency; Exchange Rate.

(a) All payments to be made by Ultragenyx to BRI under this Agreement shall be made in Dollars by bank wire transfer in immediately available funds to a bank account designated by written notice from BRI to Ultragenyx. In the case of sales outside the United States, the rate of exchange to be used in computing the monthly amount of currency equivalent in Dollars due BRI shall be made at the monthly rate of exchange published in *The Wall Street Journal* (U.S., Eastern Edition), prevailing on the last Business Day of the month preceding the month in which such sales are recorded by Ultragenyx.

(b) If pursuant to applicable Law or fiscal policy of a particular country, a remittance of royalties in the currency stipulated in this Section 4.6 is restricted or forbidden, Ultragenyx shall provide notice thereof to BRI, and payment of such royalty shall be made by the deposit thereof in local currency to the credit of BRI in a recognized banking institution designated by BRI or its Affiliates. When in any such country the applicable Law or fiscal policy would then allow the transmittal of such royalty payments, all royalties or other sums that Ultragenyx would have been under obligation to transmit but for the prohibition, shall promptly be transmitted to BRI to the extent allowable.

4.7 Late Payments. If a Party does not receive payment of any sum due to it on or before the due date therefor, simple interest shall thereafter accrue on the sum due to such Party from the due date until the date of payment at a per-annum rate of prime plus [***] percentage points or the maximum rate allowable by applicable Law, whichever is less.

4.8 Taxes.

(a) Taxes on Income. Each Party shall be solely responsible for the payment of all taxes imposed on its share of income arising directly or indirectly from the activities of the Parties under this Agreement.

(b) Tax Cooperation. The Parties agree to cooperate with one another and use reasonable efforts to avoid or reduce tax withholding or similar obligations in respect of royalties, milestone payments, and other payments made by Ultragenyx to BRI under this Agreement. To the extent Ultragenyx is required to deduct and withhold taxes on any payment to BRI, Ultragenyx shall pay the amounts of such taxes to the proper Governmental Authority in a timely manner, and the sum payable to BRI shall be decreased by the same amount. BRI shall provide Ultragenyx any tax forms that may be reasonably necessary in order for Ultragenyx to not withhold tax or to withhold tax at a reduced rate under an applicable bilateral income tax treaty. BRI shall use reasonable efforts to provide any such tax forms to Ultragenyx in advance of the due date. Each Party shall provide the other with reasonable assistance to enable the recovery, as permitted by Law, of withholding taxes or similar obligations resulting from payments made under this Agreement, such recovery to be for the benefit of BRI as the Party bearing such withholding tax under this Section 4.8(b).

4.9 Records and Audit Rights.

(a) Ultragenyx shall keep complete, true and accurate books and records in relation to this Agreement. Upon the written request of BRI and not more than [***] in each Calendar Year, Ultragenyx shall permit an independent certified public accounting firm selected by BRI, and reasonably acceptable to Ultragenyx, to have access during normal business hours to such of the records of Ultragenyx as may be reasonably necessary to verify the accuracy of the royalty reports hereunder for any Calendar Year ending not more than [***] prior to the date of such request. BRI shall treat all financial information subject to review under this Section 4.9 or under any sublicense agreement in accordance with the confidentiality and non-use provisions of this Agreement.

(b) Ultragenyx may require an accounting firm conducting an audit hereunder to sign a non-disclosure agreement to protect the confidentiality of Ultragenyx's Confidential Information before providing such accounting firm access to Ultragenyx's facilities, books or records. Upon completion of any audit hereunder, the accounting firm shall provide both Ultragenyx and BRI a written report disclosing whether the royalty reports submitted by Ultragenyx are correct or incorrect, whether the amounts paid are correct or incorrect, and in each case, the specific details concerning any discrepancies.

(c) BRI shall bear its internal expenses and the out-of-pocket costs for engaging such accounting firm in connection with performing such audits; provided, however, that if any such audit uncovers an underpayment of milestones payments or royalties by Ultragenyx that exceeds [***] percent ([***]%) of the total owed for such payment or payment period, as applicable, then Ultragenyx shall reimburse BRI for the expenses and costs for such audit.

(d) If such accounting firm identifies an underpayment by Ultragenyx during such period, Ultragenyx shall pay BRI the amount of the discrepancy within [***] days of the date BRI delivers to Ultragenyx such accounting firm's written report with the amount of underpayment accruing interest at the rate such forth in Section 4.7. If such accounting firm identifies an overpayment by Ultragenyx during such period, Ultragenyx shall, at its option, have the right to request a refund of such overpaid amount, or credit such overpaid amount against subsequent payment obligations to BRI, and BRI shall make such refund to Ultragenyx within [***] days of Ultragenyx's request if so requested. If Ultragenyx has no future payment obligations under this Agreement, then Ultragenyx may require BRI to refund such overpayment and BRI shall pay such overpaid amount to Ultragenyx within [***] days of Ultragenyx's request.

Article 5

Intellectual Property Rights

5.1 Patent Prosecution in the Licensed Territory.

(a) As between the Parties, Ultragenyx, acting through outside patent counsel of its choice ("**Patent Counsel**"), shall have the first right, but not the obligation, to take the lead in the preparation, filing, prosecution and maintenance of the BRI Patents in the Licensed Territory, at Ultragenyx's cost and expense. BRI shall cooperate with Ultragenyx in filing and prosecution of such BRI Patents in the Licensed Territory, including by providing Ultragenyx with data and other information as appropriate and executing all necessary paperwork. Within [***] days after the Effective Date, BRI shall provide to Ultragenyx those copies of all patent filings, and the correspondence between BRI and patent authorities, for BRI Patents in the Licensed Territory existing as of the Effective Date. Within [***] days after Ultragenyx exercises the Ultragenyx Option pursuant to Section 2.2(a), BRI shall provide Ultragenyx those copies of all patent filings, and the correspondence between BRI and patent authorities, for BRI Patents in the Licensed Territory existing as of the date of exercise of the Ultragenyx Option that have not already been provided to Ultragenyx pursuant to this Section 5.1(a). Ultragenyx will keep BRI reasonably informed of the status of the prosecution of such BRI Patents in the Licensed Territory. For the purpose of this Article 6, "prosecution" shall include any pre-grant and post-grant proceeding including patent interference proceeding, opposition proceeding and other similar proceedings, appeals or petitions to any Board of Appeals in the patent office, appeals to any court for any patent office decisions, reissues and reexamination proceedings, and applications for patent term extensions and the like.

(b) Ultragenyx will notify BRI of any decision not to file for, prosecute or maintain, or not to continue to pay the expenses of prosecution or maintenance of (collectively, "Patent Support"), any BRI Patents in the Licensed Territory. Ultragenyx will provide such notice at least [***] days prior to any filing or payment due date, or any other due date that requires action, in connection with such BRI Patent. In such event, BRI shall have the right, but not the obligation, to file for, or continue prosecution or maintenance of, such BRI Patent in the Licensed Territory, at its expense. In the event BRI does maintain such BRI Patent in the Licensed Territory and such BRI Patent in the Licensed Territory covers the applicable Product that triggers any royalty or milestone payment, Ultragenyx shall continue payment of the applicable royalties and milestones.

5.2 Patent Prosecution in the Retained Territory.

(a) If the Licensed Territory does not expand pursuant to Section 2.2(a), BRI shall use Commercially Reasonable Efforts to prosecute and maintain all of the patents and applications included within the BRI Patents in the Retained Territory. BRI shall: (i) keep Ultragenyx advised of the status of all communications and actual and prospective filings regarding such BRI Patents in the Retained Territory, (ii) give Ultragenyx a reasonable opportunity (but in no event less than [***] business days) to review and comment on any such communications and filings proposed to be sent to any patent authority, and (iii) incorporate all reasonable comments of Ultragenyx before making any such communication or filing related to such BRI Patents in the Retained Territory.

(b) Should BRI (and [***] if the [***] Option has been exercised and [***] has the right) decide that it is no longer interested in maintaining or prosecuting any BRI Patent in the Retained Territory, BRI shall promptly advise Ultragenyx thereof and, upon written notice by Ultragenyx, BRI agrees that Ultragenyx may prosecute and maintain such BRI Patent in its own name, and BRI shall execute all required documents in order to assign any such BRI Patent to Ultragenyx; provided, however, that any such assignment by BRI shall be subject to the [***] Option.

5.3 Patent Enforcement and Defense in the Licensed Territory.

(a) Each Party shall give the other Party written notice of any infringement by a Third Party of any BRI Patents through the development or commercialization of a Product in the Field in the Licensed Territory (a "**Licensed Territory Product Infringement**"), within [***] Business Days after such Licensed Territory Product Infringement comes to such Party's attention.

(b) Ultragenyx shall have the sole and exclusive right, but not the obligation, to bring and control any legal action in connection with such Licensed Territory Product Infringement in the Licensed Territory at its own expense and discretion as it reasonably determines appropriate. BRI shall have the right to be represented in any such action by counsel of its choice at its own expense. Should Ultragenyx decline to bring a legal action in connection with a Licensed Territory Product Infringement in the Licensed Territory, BRI shall have the right, but not the obligation, to bring such a legal action. In the event that the Licensed Territory Product Infringement involves a Generic Product, and Ultragenyx declines to bring a legal action, Ultragenyx shall not be permitted to reduce royalties due BRI pursuant to Section 4.4(c). For clarity, Ultragenyx's right to bring and control any legal action under this Section 5.3(b)shall not apply to a Licensed Territory Product Infringement in an Indication Cluster during the period of time in which Ultragenyx has anon-exclusive license.

(c) At the request of a party bringing a legal action under Section 5.2(b) (the "**Litigating Party**"), the other Party shall reasonably cooperate and provide any information or assistance in connection with any legal action under this Section 5.3, including executing reasonably appropriate documents, cooperating in discovery and joining as a party to the action if required, all at the Litigating Party's expense.

(d) Any recoveries resulting from such an action relating to a claim of Licensed Territory Product Infringement shall be first applied against payment of costs and expenses in connection with the action of the Party which initiated and prosecuted the action. The other Party shall then, to the extent possible, recover its costs and expenses incurred in connection with the action. Solely in the case in which Ultragenyx is the Litigating Party, any such recoveries in excess of such costs and expenses of the Parties shall be retained by Ultragenyx and shall be deemed Net Sales subject to Ultragenyx's royalty payment obligation to BRI under Section 4.4. In the event BRI is the Litigating Party, all recoveries in excess of such costs and expenses of the Parties shall be retained by BRI.

5.4 Patent Enforcement and Defense in the Retained Territory.

(a) If the Licensed Territory does not expand pursuant to Section 2.2(a), each Party shall give the other Party written notice of any infringement by a Third Party of any BRI Patents through the development or commercialization of a Product in the Field in the Retained Territory (a "**Retained Territory Product Infringement**"), within [***] Business Days after such Retained Territory Product Infringement comes to such Party's attention.

(b) As between the Parties, BRI shall have the right, but not the obligation, to bring and control any legal action in connection with such Retained Territory Product Infringement in the Retained Territory at its own expense. BRI shall, if it has brought or controls such legal action, use Commercially Reasonable Efforts to: (i) keep Ultragenyx advised of the status of all such legal actions, (ii) give Ultragenyx a reasonable opportunity to review and comment on any filings, motions, pleadings or other strategic decisions relating to such legal action, and (iii) incorporate all reasonable comments of Ultragenyx in conducting such legal action, and, in each case, BRI shall use Commercially Reasonable Efforts to require [***] or any other applicable Third Party licensee of BRI in the Retained Territory to do so.

5.5 Third Party Patent Proceedings. Each Party will notify the other Party in writing prior to challenging any Patents controlled by a Third Party that are necessary or reasonably useful to use, develop, manufacture, commercialize or import any Compound or Product. Such challenges include declaratory judgment actions, inter parties re-examinations, interferences, oppositions and other similar proceedings (collectively "**Third Party Patent Proceeding**").

(a) Except as the Parties otherwise agree, Ultragenyx shall have the first right to bring and control any legal action in connection with such Third Party Patent Proceeding in the Field in the Licensed Territory, at its own expense and discretion as it reasonably determines appropriate.

(b) At the request of Ultragenyx, BRI shall reasonably cooperate and provide any information or assistance in connection with any legal action under this Section 5.3, including executing reasonably appropriate documents, cooperating in discovery and joining as a party to the action if required, all at Ultragenyx's expense. Ultragenyx shall keep BRI reasonably informed of the status of such action.

5.6 Patent Extensions. The Parties shall cooperate in obtaining patent term restoration (under but not limited to Drug Price Competition and Patent Term Restoration Act), supplemental protection certificates or their equivalents, and patent term extensions with respect to the BRI Patents in any country or region in the Licensed Territory where applicable, provided, that Ultragenyx shall have the final decision making authority on the foregoing.

Article 6

Confidentiality; Publication

6.1 Duty of Confidence. Subject to the other provisions of this Article 6:

(a) all Confidential Information disclosed by or on behalf of a Party or its Affiliates ("**Disclosing Party**") under this Agreement, or in the course of contemplating a transaction under this Agreement prior to the execution of this Agreement, shall be maintained in confidence and otherwise safeguarded by the recipient Party and its Affiliates ("**Receiving Party**"), in the same manner and with the same protection as such Receiving Party maintains its own confidential information, but at least with reasonable protection;

(b) the Receiving Party may only use any such Confidential Information for the purposes of performing its obligations or exercising its rights under this Agreement; and

(c) the Receiving Party may disclose Confidential Information of the other Party to: (i) its Affiliates and sublicensees; and (ii) employees, directors, agents, contractors, consultants and advisers of the Party and its Affiliates, licensees and sublicensees, in each case to the extent reasonably necessary for the purposes of, and for those matters undertaken pursuant to, this Agreement; provided that such Persons are bound to maintain the confidentiality of the Confidential Information in a manner consistent with the confidentiality provisions of this Agreement. Notwithstanding the foregoing, the Parties acknowledge and agree that BRI may not disclose any Confidential Information of Ultragenyx to [***] or any other licensee or sublicensee of the Compound and/or Product in the Option Territory without Ultragenyx's prior written consent. Ultragenyx acknowledges that, pursuant to the [***] Option as amended on February 22, 2012, BRI is required to disclose preclinical and clinical data and information regarding regulatory submissions in the Licensed Territories to [***], and with respect to BRI Independent Studies, shall require no consent from Ultragenyx to do so.

6.2 Exceptions. The foregoing obligations as to particular Confidential Information of a Disclosing Party shall not apply to the extent that the Receiving Party can demonstrate that such Confidential Information:

(a) was known by the Receiving Party at the time of its receipt, and not through a prior disclosure by the Disclosing Party, as documented by the Receiving Party's written records;

(b) was in the public domain before its receipt from the Disclosing Party, or thereafter enters the public domain through no fault of the Receiving Party;

(c) is subsequently disclosed to the Receiving Party by a Third Party who is not under a direct or indirect obligation of confidentiality to the Disclosing Party; or

(d) is developed by the Receiving Party independently and without use of or reference to any Confidential Information received from the Disclosing Party, as documented by the Receiving Party's written records.

Any combination of features or disclosures shall not be deemed to fall within the foregoing exclusions merely because individual features are published or available to the general public or in the rightful possession of the Receiving Party unless the combination itself and principle of operation are published or available to the general public or in the rightful possession of the Receiving Party.

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

6.3 Authorized Disclosures. Notwithstanding the obligations set forth in Sections 6.2 and 6.5, a Party may disclose the other Party's Confidential Information (including this Agreement and the terms herein) solely to the extent:

(a) such disclosure: (i) is reasonably necessary for the filing or prosecuting Patents as contemplated by this Agreement; (ii) is reasonably necessary in connection with Regulatory Filings for Products; (iii) is reasonably necessary for the prosecuting or defending of legal actions, including litigation, as contemplated by this Agreement; or (iv) is made to any Third Party bound by written obligation of confidentiality and non-use similar to those set forth under this Article 6, to the extent otherwise necessary or appropriate in connection with the exercise of its rights or the performance of its obligations hereunder;

(b) such disclosure is reasonably necessary: (i) to such Party's directors, attorneys, independent accountants or financial advisors for the sole purpose of enabling such directors, attorneys, independent accountants or financial advisors to provide advice to the Receiving Party, provided that in each such case on the condition that such directors, attorneys, independent accountants and financial advisors are bound by confidentiality and non-use obligations substantially consistent with those contained in this Agreement; provided, however, that the term of confidentiality for such directors, attorneys, independent accountants and financial advisors shall be no less than [***]; or (ii) to actual or potential investors, sublicensees or acquirors solely for the purpose of evaluating an actual or potential investment, sublicense or acquisition; provided that in each such case on the condition that such actual or potential investors, sublicensees and acquirers are bound by confidentiality and non-use obligations substantially consistent with those contained in this Agreement; provided, however, that the term of confidentiality for such actual or potential investors and acquirers shall be no less than [***]; or

(c) such disclosure is required by judicial or administrative process, provided that in such event such Party shall promptly inform the other Party of such required disclosure and provide the other Party an opportunity to challenge or limit the disclosure obligations. Confidential Information that is disclosed by judicial or administrative process shall remain otherwise subject to the confidentiality and non-use provisions of this Article 7, and the Party disclosing Confidential Information pursuant to law or court order shall take all steps reasonably necessary, including seeking of confidential treatment or a protective order to ensure the continued confidential treatment of such Confidential Information.

6.4 Scientific Publications. Subject to Section 6.3, Ultragenyx or its sublicensee(s) shall have the sole right to make any public publication or presentation of any data regarding any Compound or Product, provided, however, that to the extent such data arises as a result of work performed by a BRI employee or contractor, or with financial support by BRI, BRI shall be provided with a copy of any proposed publication at least [***] days prior to submission for BRI's review and comment, and as appropriate in accordance with scientific journal standards, shall designate the appropriate BRI personnel as co-authors. However, such publication or presentation shall not include any Confidential Information of BRI without the prior written consent of BRI. Subject to Section 6.3, BRI shall make no public publication or presentation of any data regarding any Compound or Product without the prior written consent of Ultragenyx. Notwithstanding the foregoing, this Section 6.4 shall not apply to any publication or presentation of data relating to BRI Know-How or BRI Patents existing as of the Effective Date, provided that BRI shall provide Ultragenyx with a copy of any proposed publication or presentation at least [***] days prior to submission, for Ultragenyx's review and comment.

6.5 Publicity; Use of Names. Notwithstanding anything to the contrary in this Agreement, until the expiration of the [***] Option and Ultragenyx's receipt of the Ultragenyx Option Notice, the existence and the terms of this Agreement are each Party's Confidential Information and such shall be held in strict confidence and not disclosed by either Party, except with the prior express written permission of the other Party or as may be required by applicable Law. Subject to Sections 6.1, 6.2 and 6.3, no other disclosure of the existence or the terms of this Agreement may be made by either Party or its Affiliates except as provided in this Section 6.5, and no Party shall use the name, trademark, trade name or logo of the other Party, its Affiliates or their respective employees in any publicity, promotion, news release or disclosure relating to this Agreement or its subject matter, except as provided in this Section 6.5 or with the prior express written permission of the other Party, except as may be required by applicable Law.

(a) A Party may disclose this Agreement and its terms, and material developments or material information generated under this Agreement, in securities filings with the US Securities and Exchange Commission (or equivalent foreign agency) to the extent required by applicable Law after complying with the procedure set forth in this Section 6.5(a). In such event, the Party seeking such disclosure will prepare a draft confidential treatment request and proposed redacted version of this Agreement to request confidential treatment for this Agreement, and the other Party agrees to promptly (and in any event, no more than seven (7) days after receipt of such confidential treatment request and proposed redactions) give its input in a reasonable manner in order to allow the Party seeking disclosure to file its request within the time lines proscribed by applicable Law. The Party seeking such disclosure shall exercise Commercially Reasonable Efforts to obtain confidential treatment of this Agreement from the US Securities and Exchange Commission (or equivalent foreign agency) as represented by the redacted version reviewed by the other Party.

(b) The Parties agree that any news release or other public announcement relating to the terms and conditions of this Agreement or the performance hereunder shall not be made until after the earlier of (i) expiration or termination of the [***] Option and (ii) [***] exercise of the [***] Option. Any such news release, any further news release or other public disclosure that would disclose information other than that already in the public domain, shall first be reviewed and approved by both Parties (with such approval not to be unreasonably withheld or delayed).

(c) The Parties agree that after a disclosure pursuant to Section 6.5(b), a press release or other public announcement pursuant to Section 6.5(c) has been reviewed and approved by the other Party, the disclosing Party may make subsequent public disclosures reiterating such information without having to obtain the other Party's prior consent and approval.

Article 7

Term and Termination

7.1 Term. The term of this Agreement will commence upon the Effective Date and continue in full force and effect, on a Product-by-Product and country-country basis, until the expiration of the royalty obligations of Ultragenyx with respect to the applicable Product, unless earlier terminated as set forth in Section 7.2 (the "**Term**"). After the expiration of this Agreement for a particular Product in a particular country, Ultragenyx's license in such country shall become fully paid, royalty-free, perpetual and irrevocable.

7.2 Termination.

(a) Termination by Ultragenyx for Convenience. At any time, Ultragenyx may terminate this Agreement, in its entirety or on a Product-by-Product basis, by providing written notice of termination to BRI, which notice includes an effective date of termination at least ninety (90) days after the date of the notice.

(b) Termination for Material Breach. If either Party believes that the other is in breach of its material obligations hereunder (other than Ultragenyx's nonfulfillment of its obligations under Section 3.2, the remedy of which is set forth in Section 3.2), then the non-breaching Party may deliver notice of such breach to the other Party. The allegedly breaching Party shall have ninety (90) days from such notice to dispute or cure such breach. If the Party receiving notice of breach fails to cure, or fails to dispute, that breach within the period set forth above, then the Party originally delivering the notice of breach may terminate this Agreement effective on written notice of termination to the other Party, provided that such non-breaching Party delivers such notice of termination within sixty (60) days of the end of such one hundred ninety(90)-day cure period. If the allegedly breaching Party in good faith disputes such material breach or disputes the failure to cure or remedy such material breach and provides written notice of that dispute to the other Party within the period set forth above, the notifying Party may not terminate this Agreement until one independent industry expert mutually agreeable to both Parties has determined that the allegedly breaching Party is in material breach of this Agreement, and such breaching Party further fails to cure such breach within thirty (30) days after such determination by the independent industry expert (and such termination shall then be effective upon written notification from the notifying Party to the breaching Party).

(c) Termination for Bankruptcy. Either party may terminate this Agreement in the event the other Party ceases doing business as a going concern, makes an assignment for the benefit of creditors, shall not be paying its debts in the ordinary course, shall have become insolvent or shall, voluntarily or involuntarily, become party to insolvency proceeding, or commits any act of bankruptcy. Notwithstanding any other provision of this Agreement to the contrary, in the event that BRI becomes a debtor under the United States Bankruptcy Code (11 U.S.C. §101 et. seq. or any similar law in any other country (the "**Bankruptcy Code**")) and rejects this Agreement pursuant to Section 365 of the Bankruptcy Code (a "**Bankruptcy Rejection**"), (i) the license to the BRI Technology described under this Agreement shall be deemed fully retained by and vested in Ultragenyx as protected intellectual property rights under Section 365(n)(1)(B) of the Bankruptcy Code and further shall be deemed to exist immediately before the commencement of the bankruptcy case in which BRI is the debtor; and (ii) Ultragenyx shall have all of the rights afforded to non-debtor licensees under Section 365(n) of the Bankruptcy Code, subject to its continued compliance with all its obligations under this Agreement. All rights and licenses now or hereinafter granted by BRI to Ultragenyx under or pursuant to any section of this Agreement, including Section 2.1 are rights to "intellectual property" (as defined in the Bankruptcy Code).

7.3 Effect of Termination.

(a) Termination by Ultragenyx for Convenience; Termination by BRI for Breach or Bankruptcy. Upon termination of this Agreement by Ultragenyx pursuant to Section 7.2(a) or by BRI pursuant to Sections 7.2(b) or 7.2(c), the following consequences shall apply to the termination and shall be effective as of the effective date of such termination:

(i) Each Party shall pay all amounts then due and owing to the other Party as of the termination date;

(ii) All licenses and other rights granted to Ultragenyx under the BRI Technology will terminate, and Ultragenyx shall immediately discontinue sales of Product;

(iii) No later than thirty (30) days after the effective date of such termination, Ultragenyx shall return or cause to be returned to BRI all Confidential Information in tangible form received from BRI and all copies thereof and all materials substances or compositions delivered or provided by BRI; provided, however, that Ultragenyx may keep one copy of Confidential Information received from the other Party in its confidential files for record purposes;

(iv) No later than thirty (30) days after the effective date of termination, at BRI's option, the Parties shall negotiate in good faith the terms and conditions (including financial compensation to Ultragenyx) for a license under patent applications, patents, Know-How, and any other information and documentation Controlled by Ultragenyx as of the effective date of such termination necessary for BRI to continue development and commercialization of Products in the Licensed Territory, but solely for those indications, the use of which are claimed by a Valid Claim in the BRI Technology as of the date of such termination. If the Parties have failed to agree upon such terms of compensation within ninety (90) days of initiation of such negotiation, the matter shall be referred to three independent industry experts having expertise in intellectual property valuation and who are mutually agreeable to both Parties. The industry experts shall take into account all pertinent factors, including but not limited to each party's relative contribution to the Ultragenyx Technology, and within thirty (30) days make a joint recommendation of appropriate compensation for the license ("License Compensation"). BRI shall have the right, but not the obligation, to accept the license at the recommended License Compensation. If BRI does not accept the license at the recommended License Compensation, then Ultragenyx shall have no further obligation to negotiate with BRI regarding the terms and conditions for a license pursuant to this Section 7.3(a)(iv) and Ultragenyx shall have no obligation to grant BRI any such license. If BRI does accept the license at the recommended License Compensation, then Ultragenyx shall have the obligation to grant BRI the license at the recommended License Compensation; and

(v) At the written request of any of Ultragenyx's sublicensees under this Agreement, BRI shall negotiate in good faith with such sublicensee an agreement between BRI and such sublicensee under which such sublicensee will obtain a direct license under the BRI Technology. For clarity, under no circumstances shall BRI be required to grant a direct license to any sublicensee which does not compensate BRI at a level substantially equivalent to the compensation received by BRI under this Agreement with respect to the applicable technology, taking into consideration any costs associated with BRI's license to Ultragenyx Technology. Ultragenyx shall indemnify, defend and hold BRI Indemnitees harmless from and against any Claims brought by any sublicensee in connection with the termination of an Ultragenyx sublicense, or the inability of BRI and any sublicensee to negotiate a mutually

acceptable license to BRI Technology, provided that Ultragenyx shall have no indemnification obligation to BRI under this Section 7.3(a)(v) to the extent such Claim arises as a result of BRI's bad faith in negotiation with an Ultragenyx sublicensee.

(b) Termination by Ultragenyx for Breach or Bankruptcy. Upon termination of this Agreement by Ultragenyx pursuant to Sections 7.2(b) or 7.2(c), the following consequences shall apply to the termination and shall be effective as of the effective date of such termination:

(i) Each Party shall pay all amounts then due and owing to the other Party as of the termination date;

(ii) The licenses and other rights granted by BRI to Ultragenyx under the BRI Technology will remain in full force and effect as set forth in Sections 2.1; provided that Ultragenyx fulfills its payment obligations to BRI under Article 4 pursuant to the terms and conditions set forth in Article 4, further provided that, on a Product-by-Product and country by country basis, for any Product that is the subject of the underlying breach, milestone payments under Sections 4.2 and 4.3 and royalty payments under Section 4.4 with respect to such Product shall be reduced by [***] percent ([***]%);

(iii) No later than thirty (30) days after the effective date of such termination, BRI shall return or cause to be returned to Ultragenyx all Confidential Information in tangible form received from Ultragenyx and all copies thereof and all materials, substances or compositions delivered or provided by Ultragenyx; provided, however, that BRI may keep one copy of Confidential Information received from Ultragenyx in its confidential files for record purposes; and

(iv) In addition to the provisions set forth in Section 7.5, the provisions relating to BRI Patents in Sections 6.1, 6.2, 6.3 and 6.4 shall survive the termination of this Agreement for so long as the license to Ultragenyx survives under this Section 7.3(b).

7.4 Survival. Expiration or termination of this Agreement shall not relieve the Parties of any obligation accruing prior to such expiration or termination. Without limiting the foregoing, the provisions of Articles 1, 6, 9 (to the extent the Claims arise out of actions or omissions during the Term) and 10, and Sections 5.1, 7.3, 7.4 and 7.5, and any other provisions which by their nature are intended to survive, shall survive the expiration or termination of this Agreement.

7.5 Termination Not Sole Remedy. Termination is not the sole remedy under this Agreement and, whether or not termination is effected and notwithstanding anything contained in this Agreement to the contrary, all other remedies will remain available except as agreed to otherwise herein.

Article 8

Representations and Warranties

8.1 Representations and Warranties of Each Party. Each Party represents and warrants to the other Party as of the Effective Date that:

(a) it has the full right, power and authority to enter into this Agreement, to perform its obligations hereunder; and

(b) this Agreement has been duly executed by it and is legally binding upon it, enforceable in accordance with its terms, and does not conflict with any agreement, instrument or understanding, oral or written, to which it is a party or by which it may be bound, nor violate any material law or regulation of any court, governmental body or administrative or other agency having jurisdiction over it.

8.2 Representations and Warranties by BRI. BRI represents and warrants to Ultragenyx as of the Effective Date that:

(a) it has not previously assigned, transferred, conveyed or otherwise encumbered its right, title and interest in BRI Patents or BRI Know-How with respect to any of the Compounds or Products other than pursuant to the [***] Option;

(b) it has the right to grant the license and rights herein to Ultragenyx and it has not granted any license, right or interest in, to or under the BRI Patents or BRI Know-How to any Third Party with respect to any of the Compounds or Products other than pursuant to the [***] Option;

(c) to the best of its knowledge, it has provided to Ultragenyx all of the following information relating to the Compound or Product: all communications to and from Regulatory Authorities, all protocols and amendments for any human clinical studies, all safety reports sent to the FDA or other Regulatory Authorities, all Regulatory Filings filed with Regulatory Authorities, including all INDs filed with the FDA, and all clinical, non-clinical, and research study reports in its possession;

(d) the [***] Option does not grant [***] any license rights to BRI Know-How and BRI Patents with respect to the United States, Canada or Mexico;

(e) to the best of its knowledge, except as otherwise stated in this Section 8.2(e), the development, use, sale and import of Compounds or Products in the Licensed Territory do not infringe any valid intellectual property rights owned or possessed by any Third Party and do not breach any obligation of confidentiality or non-use owed by BRI to a Third Party, provided however, that the Parties acknowledge and agree that the following Third Parties may own or possess intellectual property rights covering the method of use of the Compound and/or Product: [***];

(f) there are no claims, judgments or settlements against or owed by BRI and to the best of BRI's knowledge, there are no pending or threatened claims or litigation, in each case relating to any Compounds or Products, or to the BRI Patents or BRI Know-How in the Licensed Territory;

(g) there is no provision in the [***] Option or the agreement to be entered into between BRI and [***] upon [***] exercise of the [***] Option that expressly addresses the performance of clinical trials by BRI or any other party in the Option Territory and during the Term BRI shall not extend the duration of the [***] Option, or amend the terms of such [***] Option or the agreement to be entered into between BRI and [***] upon [***] exercise of the [***] Option (other than financial), in each case without Ultragenyx's prior written consent; and

(h) the list of Patents contained in Exhibit A is a complete list of all BRI Patents.

8.3 No Other Warranties. EXCEPT AS EXPRESSLY STATED IN THIS ARTICLE 8, (A) NO REPRESENTATION, CONDITION OR WARRANTY WHATSOEVER IS MADE OR GIVEN BY OR ON BEHALF OF ULTRAGENYX OR BRI; AND (B) ALL OTHER CONDITIONS AND WARRANTIES WHETHER ARISING BY OPERATION OF LAW OR OTHERWISE ARE HEREBY EXPRESSLY EXCLUDED, INCLUDING ANY CONDITIONS AND WARRANTIES OF MERCHANTABILITY, FITNESS FOR A PARTICULAR PURPOSE OR NON-INFRINGEMENT.

Article 9

Indemnification; Liability

9.1 Indemnification by BRI. BRI shall indemnify and hold Ultragenyx and its Affiliates, and their respective officers, directors, agents and employees ("**Ultragenyx Indemnitees**") harmless from and against any Claims arising under or related to this Agreement against them to the extent arising or resulting from:

(a) the negligence or willful misconduct of any of the BRI Indemnitees; or

(b) the breach of any of the warranties or representations made by BRI to Ultragenyx under this Agreement; or

(c) any breach by BRI of its material obligations pursuant to this Agreement

except in each case, to the extent such Claims result from the material breach by any Ultragenyx Indemnitee of any covenant, representation, warranty or other agreement made by Ultragenyx in this Agreement or the negligence or willful misconduct of any Ultragenyx Indemnitee.

9.2 Indemnification by Ultragenyx. Ultragenyx shall indemnify and hold BRI, its Affiliates, and their respective officers, directors, agents and employees ("**BRI Indemnitees**") harmless from and against any Claims arising under or related to this Agreement against them to the extent arising or resulting from:

(a) the development, manufacture, packaging, use, sale or commercialization of the Compounds or Products, or use of the BRI Technology, by or on behalf Ultragenyx or any of its Affiliates, sublicensees or contractors in the Field in the Licensed Territory; or

(b) the negligence or willful misconduct of any of the Ultragenyx Indemnitees, sublicensees, or contractors; or

(c) the breach of any of the warranties or representations made by Ultragenyx to BRI under this Agreement;

(d) any representation made or warranty given by Ultragenyx, or any of its Affiliates, sublicensees or contractors with respect to Compounds, Products, BRI Patents or BRI Know-How;

(e) any infringement claims relating to Compounds or Products;

(f) any asserted violation of applicable Laws by any Ultragenyx Indemnitees, sublicensees or contractors; or

(g) any breach by Ultragenyx of its material obligations pursuant to this Agreement;

except in each case, to the extent such Claims result from the material breach by any BRI Indemnitee of any covenant, representation, warranty or other agreement made by BRI in this Agreement or the negligence or willful misconduct of any BRI Indemnitee.

9.3 Insurance. From and after the Effective Date, Ultragenyx shall maintain for a period of [***] after the expiration or termination of this Agreement, commercial product liability insurance (including contractual liability insurance and clinical trial insurance) with insurance carriers with, at least, an AM BEST rating of A- VII to cover the activities of Ultragenyx Indemnitees, for minimum limits of [***] dollars ($[***]) per claim and [***] dollars ($[***]) in the aggregate. Such insurance shall cover BRI Indemnitees as additional insureds. Ultragenyx shall furnish a certificate of insurance evidencing such coverage. Ultragenyx will provide thirty days' written notice to BRI of cancellation or material change in coverage. The minimum amounts of insurance coverage required herein shall not be construed as creating any limitation on the Ultragenyx's indemnity obligation under Section 9.2. of this Agreement. If any coverage is written on a Claims Made policy form, the Retroactive Date is to be the first date that the first Claims Made policy was effective and is not to be advanced during the term of the project. Additionally, if the coverage is written on a Claims Made form, the insurance policies will remain in full force and effect, or if canceled or non renewed Ultragenyx shall purchase an Extended Reporting Period "Tail Coverage", for a minimum period of [***] after the termination of this Agreement.

9.4 Indemnification Procedure. If either Party is seeking indemnification under Sections 9.1 or 9.2 (the "**Indemnified Party**"), it shall inform the other Party (the "**Indemnifying Party**") of the claim giving rise to the obligation to indemnify pursuant to such section as soon as reasonably practicable after receiving notice of the claim. The Indemnifying Party shall have the right to assume the defense of any such claim for which it is obligated to indemnify the Indemnified Party. The Indemnified Party shall cooperate with the Indemnifying Party and the Indemnifying Party's insurer as the Indemnifying Party may reasonably request, and at the Indemnifying Party's cost and expense. The Indemnified Party shall have the right to participate, at its own expense and with counsel of its choice, in the defense of any claim or suit that has been assumed by the Indemnifying Party. Neither Party shall have the obligation to indemnify the other Party in connection with any settlement made without the Indemnifying Party's written consent, which consent shall not be unreasonably withheld, conditioned or delayed. If the Parties cannot agree as to the application of Section 9.1 or 9.2 as to any claim, pending resolution of the dispute pursuant to Section 10.6, the Parties may conduct separate defenses of such claims, with each Party retaining the right to claim indemnification from the other Party in accordance with Section 9.1 or 9.2 upon resolution of the underlying claim.

9.5 Mitigation of Loss. Each Indemnified Party will take and will procure that its Affiliates take all such reasonable steps and action as are reasonably necessary or as the Indemnifying Party may reasonably require in order to mitigate any Claims (or potential losses or damages) under this Article 9. Nothing in this Agreement shall or shall be deemed to relieve any Party of any common law or other duty to mitigate any losses incurred by it.

9.6 Special, Indirect and Other Losses. NEITHER PARTY NOR ANY OF ITS AFFILIATES SHALL BE LIABLE IN CONTRACT, TORT, NEGLIGENCE, BREACH OF STATUTORY DUTY OR OTHERWISE FOR ANY CONSEQUENTIAL, SPECIAL OR PUNITIVE DAMAGES OR FOR LOSS OF PROFITS SUFFERED BY THE OTHER PARTY, EXCEPT TO THE EXTENT ANY SUCH DAMAGES ARE REQUIRED TO BE PAID TO A THIRD PARTY AS PART OF A CLAIM FOR WHICH A PARTY PROVIDES INDEMNIFICATION UNDER THIS ARTICLE 9.

Article 10

General Provisions

10.1 Force Majeure. Neither Party shall be held liable to the other Party nor be deemed to have defaulted under or breached this Agreement for failure or delay in performing any obligation under this Agreement to the extent such failure or delay is caused by or results from causes beyond the reasonable control of the affected Party, potentially including embargoes, war, acts of war (whether war be declared or not), acts of terrorism, insurrections, riots, civil commotions, strikes, lockouts or other labor disturbances, fire, floods, earthquakes or other acts of God, or acts, omissions or delays in acting by any Government Authority or the other Party or unavailability of materials related to the manufacture of Compounds or Products. The affected Party shall notify the other Party in writing of such force majeure circumstances as soon as reasonably practical, and shall promptly undertake and continue diligently all reasonable efforts necessary to cure such force majeure circumstances or to perform its obligations in spite of the ongoing circumstances. Notwithstanding the foregoing, neither Party shall be excused from making payments owed hereunder because of a force majeure affecting such Party unless such force majeure event affects the method of payment.

10.2 Assignment.

(a) This Agreement may not be assigned or otherwise transferred, nor may any right or obligation hereunder be assigned or transferred, by either Party without the prior written consent of the other Party. Notwithstanding the foregoing, either Party may, without consent of the other Party, assign this Agreement and its rights and obligations hereunder in whole or in part to an Affiliate of such Party, or to its successor in interest by way of merger, acquisition, or sale of all or substantially all of its assets to which this Agreement relates. Any attempted assignment not in accordance with this Section 10.2 shall be null and void and of no legal effect. Any permitted assignee shall assume all assigned obligations of its assignor under this Agreement. The terms and conditions of this Agreement shall be binding upon, and shall inure to the benefit of, the Parties and their respected successors and permitted assigns.

(b) Notwithstanding anything to the contrary in this Agreement, in the event that a Party undergoes a merger, acquisition, or sale of all or substantially all of its assets to which this Agreement relates, no intellectual property rights of the Third Party assignee, acquiror or successor of such Party or any Affiliate of such Third Party shall be included in the subject matter licensed hereunder, to the extent that such intellectual property rights were held by such Third Party prior to the merger, acquisition or sale, or are created outside of any activities under this Agreement by personnel who were not employees of the acquired Party at the time of the acquisition.

10.3 Severability. If any one or more of the provisions contained in this Agreement is held invalid, illegal or unenforceable in any respect, the validity, legality and enforceability of the remaining provisions contained herein shall not in any way be affected or impaired thereby, unless the absence of the invalidated provision(s) adversely affects the substantive rights of the Parties. The Parties shall in such an instance use their best efforts to replace the invalid, illegal or unenforceable provision(s) with valid, legal and enforceable provision(s) which, insofar as practical, implement the purposes of this Agreement.

10.4 Notices. All notices which are required or permitted hereunder shall be in writing and sufficient if delivered personally, sent by facsimile (and promptly confirmed by personal delivery, registered or certified mail or courier), sent by internationally recognized courier or sent by registered or certified mail, postage prepaid, return receipt requested, addressed as follows:

If to BRI:

Baylor Research Institute
3310 Live Oak Street, Suite 501
Dallas, TX 75204
Attn: Chief Operating Officer
Fax: (214) 820-4952

with a copy to:

Law Department
Baylor Health Care System
4005 Crutcher Street
Dallas, TX 75246
Attn: BRI Attorney
Fax: (214) 820-1535

If to Ultragenyx:

Ultragenyx Pharmaceutical Inc.
60 Leveroni Court
Novato, CA 94949
Attn: Tom Kassberg, Chief Business Officer
Fax: (415) 483-8820

with a copy to:

Cooley LLP
3175 Hanover Street
Palo Alto, CA 94304
Attn: Lila Hope, Esq.
Fax: (650) 849-7400

or to such other address(es) as the Party to whom notice is to be given may have furnished to the other Party in writing in accordance herewith. Any such notice shall be deemed to have been given: (a) when delivered if personally delivered or sent by facsimile on a Business Day (or if delivered or sent on a non-Business Day, then on the next Business Day); (b) on the Business Day after dispatch if sent by internationally recognized overnight courier; or (c) on the fifth (5th) Business Day following the date of mailing, if sent by mail.

10.5 Applicable Law. This Agreement shall be governed by and construed in accordance with the laws of the State of New York and the patent laws of the United States without reference to any rules of conflict of laws.

10.6 Dispute Resolution. The Parties shall negotiate in good faith and use reasonable efforts to settle any dispute, controversy or claim ("**Dispute**") arising from or related to this Agreement or the breach thereof. If any Dispute has not been resolved within [***] days of written notice detailing the nature of the Dispute by one Party to the other, the Parties shall each immediately refer such dispute to respective senior executives at the level of Senior Vice President or above for consideration and resolution. If the Dispute has not been resolved within [***] days of referral to such senior executives, either party may bring an action in a court of competent jurisdiction.

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

10.7 Compliance. Each Party agrees that in performing its obligations or exercising its rights under this Agreement: (a) it shall comply in all material respects with all applicable Laws; (b) it will not employ or engage any Person who has been debarred by any Regulatory Authority, or, to such Party's knowledge, is the subject of debarment proceedings by a Regulatory Authority; and (c) it will be primarily responsible for any activities performed on its behalf by an Affiliate, licensee, sublicensee, contractor or subcontractor.

10.8 Entire Agreement.

(a) This Agreement, together with the Exhibits, contains the entire understanding of the Parties with respect to the licenses granted hereunder. Any other express or implied agreements and understandings, negotiations, writings and commitments, either oral or written, in respect to the licenses granted hereunder are superseded by the terms of this Agreement. The Exhibits to this Agreement are incorporated herein by reference and shall be deemed a part of this Agreement.

(b) This Agreement supersedes the Mutual Nondisclosure Agreement, dated February 8th, 2012, between Ultragenyx and BRI (the "**Prior NDA**"). All Confidential Information disclosed by one Party to the other Party under the Prior NDA shall be deemed Confidential Information of such disclosing Party under this Agreement and shall be subject to the terms of this Agreement.

10.9 Amendments. This Agreement may be amended, or any term hereof modified, only by a written instrument duly executed by authorized representative(s) of both Parties.

10.10 Headings. The captions to the several Articles, Sections and subsections hereof are not a part of this Agreement, but are merely for convenience to assist in locating and reading the several Articles and Sections hereof.

10.11 Independent Contractors. It is expressly agreed that BRI and Ultragenyx shall be independent contractors and that the relationship between the two Parties shall not constitute a partnership, joint venture or agency. Neither BRI nor Ultragenyx shall have the authority to make any statements, representations or commitments of any kind, or to take any action, which shall be binding on the other Party, without the prior written consent of the other Party.

10.12 Waiver. The waiver by either Party of any right hereunder, or of any failure of the other Party to perform, or of any breach by the other Party, shall not be deemed a waiver of any other right hereunder or of any other breach by or failure of such other Party whether of a similar nature or otherwise.

10.13 Cumulative Remedies. No remedy referred to in this Agreement is intended to be exclusive, but each shall be cumulative and in addition to any other remedy referred to in this Agreement or otherwise available under law.

10.14 Waiver of Rule of Construction. The rule of construction that any ambiguity in this Agreement shall be construed against the drafting Party shall not apply.

10.15 Business Day Requirements. In the event that any notice or other action or omission is required to be taken by a Party under this Agreement on a day that is not a Business Day then such notice or other action or omission shall be deemed to be required to be taken on the next occurring Business Day.

10.16 Counterparts. This Agreement may be executed in counterparts by original signature, facsimile or PDF files, each of which shall be deemed an original, but all of which together shall constitute one and the same instrument.

[Remainder of page intentionally left blank; signature page follows.]

IN WITNESS WHEREOF, the Parties intending to be bound have caused this Agreement to be executed by their duly authorized representatives.

Baylor Research Institute

By: /s/ Jaime Walkawiak
Name: Jaime Walkawiak
Title: VP Research Operations

Ultragenyx Pharmaceutical Inc.

By: /s/ Emil Kakkis
Name: Emil Kakkis MD PhD
Title: Chief Executive Officer

Exhibit A

BRI Patents Existing as of the Effective Date

Case Number	Case Type	Country	Priority Case Number	Inventor Name	Status, Filing Date, App. Serial No. Pub No. & Date	Pat/Reg No., Issue/Reg Date	Title
[***]							

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

Exhibit 10.6

COLLABORATION AND LICENSE AGREEMENT

BETWEEN

NOBELPHARMA CO., LTD.

AND

ULTRAGENYX PHARMACEUTICALS, INC

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

COLLABORATION AND LICENSE AGREEMENT

This COLLABORATION AND LICENSE AGREEMENT (the "Agreement"), effective as of September 30, 2010 (the "Effective Date"), is made by and between Nobelpharma Co., Ltd., a Japanese corporation, having a principal place of business at Kyodo Bldg. (Horidome), 12-10 Nihonbashi-kobunacho, Chuo-Ku, Tokyo, Japan ("NPC"), and Ultragenyx Pharmaceuticals, Inc., a California corporation, having offices at 77 Digital Drive, Suite 210, Novato, CA 94949, U.S.A. ("UPI") (each a "Party," together the "Parties").

BACKGROUND

WHEREAS, NPC owns or controls certain patent and know-how rights with respect to the Compound (as defined below);

WHEREAS, NPC desires to collaborate with UPI on the research, development and commercialization of the Compound;

WHEREAS, UPI desires to obtain from NPC the licenses set forth herein, and NPC desires for the reasons described above to grant such licenses to UPI, all on the terms and conditions set forth in this Agreement

NOW, THEREFORE, for and in consideration of the covenants, conditions and undertakings hereinafter set forth, it is agreed by and between the Parties as follows:

ARTICLE 1

DEFINITIONS

As used herein, the following terms will have the meanings set forth below:

1.1 "AAA" means the American Arbitration Association.

1.2 “Affiliate” of a Party means any corporation or other business entity which during the Term of this Agreement controls, is controlled by or is under common control with such Party but only for so long as such entity controls, is controlled by, or is under common control with such Party. For the purposes of this definition, with respect to a particular entity “control” means the ownership directly or indirectly of fifty percent (50%) or more of the stock entitled to vote for the election of directors, and for non-stock organizations, of the equity interests entitled to control the management of such entity.

1.3 “Business Day” means a day other than Saturday, Sunday or any day on which the Bank of Japan or commercial banks located in New York, New York are authorized or obligated by applicable Laws to close.

1.4 “Commercialization” means activities directed to obtaining pricing and reimbursement approvals, marketing, promoting, distributing, importing or selling a Product. “Commercialize” means to engage in Commercialization.

1.5 “Commercially Reasonable Efforts” means efforts and resources normally used by a similarly situated therapeutic pharmaceutical company for a product owned by it or to which it has exclusive rights, which is of similar market potential at a similar stage in its development or product life, taking into account issues of safety and efficacy, product profile, the competitiveness of the marketplace, the proprietary position of the compound or product, the regulatory and reimbursement structure involved, the profitability of all applicable products, and other relevant commercial, scientific and other factors.

1.6 “Common Stock” means the common stock of UPI.

1.7 “Compound” means the compound identified as N-acetylneuraminic acid.

1.8 “Confidential Information” of a Party means any data and information of a confidential and proprietary nature (including, but not limited to, trade secrets, know-how, technical and business information, patent information, structures, models, techniques, formula, processes, compositions, compounds, apparatus, specifications, samples and inventions) of such Party previously received or to be received (regardless of whether orally, in writing, by e-mail or any other means) by the other Party in connection with the negotiation, execution or performance of this Agreement, but excludes any information: (i) that is already publicly known when this Agreement is executed or it is received by the other Party; or (ii) that becomes publicly known after the Effective Date without any fault of the other Party.

1.9 “Controlled” or “Controls” means the legal authority or right of a Party hereto (or any of its Affiliates), when used in reference to intellectual property, to grant a license or sublicense of intellectual property rights to another Party, or otherwise to disclose proprietary or trade secret information to such other Party, without breaching the terms of any agreement with a Third Party, or misappropriating the proprietary or trade secret information of a Third Party.

1.10 “Covered” or “Covering” means, with respect to a particular Valid Claim and a Product, that the manufacture, use, sale, offer for sale or importation of such Product, but for the licenses granted herein, would infringe such Valid Claim.

1.11 “Development” means non-clinical and clinical drug development activities related to the development and submission of information to a Regulatory Authority, including, without limitation, toxicology, pharmacology and other discovery and pre-clinical efforts, test method development and stability testing, formulation development, delivery system development, quality assurance and quality control development, statistical analysis, and clinical studies (including, without limitation, pre- and post-approval studies). Development specifically excludes regulatory activities directed to obtaining pricing and reimbursement approvals and all other Commercialization activities. “Develop” means to engage in Development.

1.12 “Development Plan” means the development plan to be prepared jointly by the Parties as soon as reasonably practicable after the execution of this Agreement and as may be revised from time to time in accordance with this Agreement. The Development Plan will set forth the Development activities, including the anticipated costs, on a summary basis. The Parties agree that there does not have to be absolute consensus between them with respect to the activities contained in the Development Plan. Provided that where there is a disagreement between the Parties or a consensus on a Development activity in the Development Plan cannot be reached, then the Development Plan will note such disagreement.

1.13 “Drug Substance” means a quantity of the Compound in bulk form.

1.14 “EMEA” means the European Medicines Agency, or any successor agency thereto.

1.15 “EU” means the European Union, which as of the Effective Date consists of Austria, Belgium, Bulgaria, Czech Republic, Denmark, Estonia, Finland, France, Germany, Greece, Hungary, Ireland, Italy, Latvia, Lithuania, Luxembourg, Malta, The Netherlands, Poland, Portugal, Romania, Slovakia, Slovenia, Spain, Sweden and the United Kingdom, and that certain portion of Cyprus included in such organization. In the event that the European Union adds one or more new member nations during the Term of this Agreement, the Parties will discuss in good faith whether to amend the definition of “EU” to include such nation(s).

1.16 “Europe” means the [***].

1.17 “FDA” means the United States Food and Drug Administration and any successor thereto having substantially the same functions.

1.18 “First Commercial Sale” means, with respect to a particular Product and Territory, the first bona fide commercial sale of such Product following Regulatory Approval to market such Product to a Third Party in such Territory by or under authority of a Party, its Affiliates or Sublicensees.

1.19 “First UPI Approval” means the first Regulatory Approval for a Product in the United States or Europe, whichever is the earlier.

1.20 “GAAP” means United States Generally Accepted Accounting Principles (as consistently applied by the applicable Party and its Affiliates).

1.21 “GMP” means the then-current good manufacturing practices required by: (a) the provisions of 21 C.F.R., parts 210 and 211 and all applicable rules, regulations, orders and guidances (as the same may from time to time be amended); (b) ICH, Guidance for Industry Q7a Good Manufacturing Practice Guidance for Active Pharmaceutical Ingredients (as the same may from time to time be amended); (c) the provisions of Chapter II of EC Commission Directive 91\356\EEC together with the Guide to Good Manufacturing Practice published by the EC Commission in 1992 (ISBN 92-826-3180-X) (as the same may from time to time be amended); and (d) any other applicable Laws, guidelines, regulations and industry standards, that apply to any manufacturing or processing activities hereunder, or the facilities in which any such activities are performed.

1.22 “IND” means an Investigational New Drug Application (as defined in the United States Food, Drug and Cosmetic Act, as amended, and the regulations promulgated thereunder), or any corresponding application, registration or certification with a Regulatory Body in any jurisdiction.

1.23 “Invention” shall mean any process, method, composition of matter, article of manufacture, discovery or finding, whether or not patentable.

1.24 “JCAA” means the Japan Commercial Arbitration Association.

1.25 “JHSF” means the Japan Health Sciences Foundation.

1.26 “Joint Know-How” shall mean any Know-How, resulting from the Agreement, obtained, developed or invented jointly by at least one employee of NPC or others acting on behalf of NPC and at least one employee of UPI or others acting on behalf of UPI, during the course of Development activities under the Agreement, where inventorship of an Invention, whether patentable or not, is to be determined in accordance with the patent laws of the United States.

1.27 “Joint Patent Rights” shall mean all Patent Rights that claim Joint Know-How.

1.28 “Know-How” means any protocols, formulas, data, Inventions, methods, proprietary information, processes, techniques, technology, materials (including biological or other materials) and trade secrets, patentable or otherwise, and any intellectual property rights (other than Patent Rights) therein.

1.29 “Korosho” means the Japanese Ministry of Health, Labour and Welfare, or any successor agency thereto.

1.30 “Laws” means all laws, statutes, rules, regulations, ordinances and other pronouncements having the effect of law of any federal, national, multinational, state, provincial, county, city or other political subdivision, agency or other body, domestic or foreign.

1.31 “Manufacture” and “Manufacturing” mean, with respect to a product or compound, the manufacturing, processing, formulating, packaging, labeling, storing and quality control testing of such product or compound.

1.32 “Net Sales” means the actual amounts invoiced for Products sold by a Person and its Affiliates and Sublicensees to a Third Party (excluding any sales among such Person and its Affiliates or Sublicensees where the Affiliate or Sublicensee is not itself the user of the Product) less the following amounts related to the Products: (a) credits, allowances, discounts, rebates, and chargebacks for spoiled, damaged, outdated, rejected, and returned Products, (b) freight and insurance costs incurred with respect to the shipment of the Products to customers, (c) duties, surcharges and other governmental charges, (d) sales, use, value-added, excise and other similar taxes (excluding income taxes), (e) cash, quantity, trade and similar discounts, rebates, allowances and other price reductions actually granted or paid by a Person and its Affiliates to the extent that such reductions relate to sales of the Products, and (f) actual uncollectible amounts. If a sale, transfer or other disposition with respect to a product is made for consideration other than cash or is not at arm’s length, then the Net Sales from such sale, transfer or other disposition shall be the arm’s length fair market value thereof. For purposes of this Agreement, “sale” means any transfer or other distribution or disposition, but shall not include

transfers or other distributions or dispositions of product, at no charge, for pre-clinical, clinical or regulatory purposes or in connection with patient assistance programs or other charitable purposes or to physicians or hospitals for promotional purposes. In the event that a Product is sold that includes more than one active ingredient, Net Sales for purposes of determining payments under this Agreement shall be limited to the portion of the Net Sales (determined in accordance with the preceding paragraph) allocated to the Compound rather than the other active ingredient(s), as determined by good faith negotiations between the Parties.

1.33 "NDA" means a New Drug Application (as defined in the United States Food, Drug and Cosmetic Act, as amended, and the regulations promulgated thereunder), or any corresponding application, registration or certification for Regulatory Approval of a Product with a Regulatory Authority in any jurisdiction.

1.34 "[***] Technology" means the technology developed based on studies conducted by [***] related to the [***].

1.35 "North America" means [***].

1.36 "NPC Know-How" means all Know-How that is (i) necessary or useful for the Development and/or Commercialization of the Compound and/or Products; and (ii) Controlled by NPC or its Affiliates as of the Effective Date or during the Term of this Agreement, excluding Joint Know-How.

1.37 "NPC Patent Rights" means any Patent Rights that are (i) necessary or useful for the Development and/or Commercialization of the Compound and/or Products; and (ii) Controlled by NPC or its Affiliates during the Term of this Agreement, excluding Joint Patent Rights.

1.38 “NPC Territory” means [***].

1.39 “Patent Rights” means any patents, patent applications, certificates of invention, or applications for certificates of invention and any supplemental protection certificates, together with any extensions, registrations, confirmations, reissues, substitutions, divisions, continuations or continuations-in-part, reexaminations or renewals thereof, and any foreign counterparts to any of the foregoing.

1.40 “Person” means any individual, firm, corporation, partnership, limited liability company, trust, business trust, joint venture company, governmental authority, association or other entity.

1.41 “Phase II Trials” means human clinical trials, the principal purpose of which is to evaluate both clinical efficacy and safety of an investigational product, and/or to obtain a preliminary evaluation of the dosage regimen of an investigational product, as more fully defined in 21 C.F.R. §312.21(b) or similar clinical study in a country other than the United States.

1.42 “Phase III Trials” means human clinical trials, the principal purpose of which is to establish substantial evidence of both safety and efficacy in patients with the disease or condition being studied, as more fully defined in 21 C.F.R. §312.21(c) or similar clinical study in a country other than the United States. Phase III Trials shall also include any other human clinical trial intended to serve as a pivotal trial to support the submission of an application for Regulatory Approval.

1.43 “Product” means a pharmaceutical preparation which incorporates the Compound as an active drug substance.

1.44 “Regulatory Approval” means approval of the Regulatory Authority in a country necessary for the marketing and sale of a Product in the applicable country. As used herein, “Regulatory Approval” shall not include pricing or reimbursement approval.

1.45 “Regulatory Authority” means the FDA, Korosho, Pharmaceuticals and Medical Devices Agency of Japan and any or all equivalent governmental or administrative authorities outside the United States or Japan whose approval is required for manufacture, marketing, promotion, sale or distribution of the Products.

1.46 “Sublicensee” means a Third Party expressly licensed by a Party to make, use, import, offer for sale or sell Product. The term “Sublicensee” shall not include distributors (i.e. a Third Party who purchases product from a Party for resale).

1.47 “Territory” means the NPC Territory or the UPI Territory, as the case may be.

1.48 “Third Party” means any Person or entity other than NPC and UPI, and their respective Affiliates.

1.49 “Trademark” means any word, name, symbol, color, designation or device or any combination thereof, including, without limitation, any trademark, trade dress, brand mark, house mark, trade name, brand name, logo, or business symbol (whether or not registered or registerable) used or displayed with respect to any Product.

1.50 "UPI Know-How" means all Know-How that is (i) necessary or useful for the Development and/or Commercialization of the Compound and/or Products; and (ii) Controlled by UPI or its Affiliates as of the Effective Date or during the Term of this Agreement, excluding Joint Know-How.

1.51 "UPI Patent Rights" means any Patent Rights that are (i) necessary or useful for the Development and/or Commercialization of the Compound and/or Products; and (ii) Controlled by UPI or its Affiliates during the Term of this Agreement, excluding Joint Patent Rights.

1.52 "UPI Territory" means [***].

1.53 "Valid Claim" means (i) a claim of an issued and unexpired patent (or the equivalent in a supplementary protection certificate), which has not lapsed or become abandoned or been declared invalid or unenforceable by a court of competent jurisdiction or an administrative agency from which no appeal can be or is taken or (ii) a claim of a pending patent application, filed in good faith, which claim shall not have been canceled, withdrawn, abandoned or rejected by an administrative agency from which no appeal can be taken; *provided* that no more than five (5) years has passed since the filing date for such patent application.

1.54 Additional Terms. In addition to the foregoing, the following terms shall have the meaning defined in the corresponding Section below:

Definition	Section Where Defined
Abandonment	9.3.3
Business Year	8.1.2
Competitive Product	5.4
Indemnification Claim	12.3
Indemnitee	12.3
Indemnitor	12.3
Infringement	9.5
Intellectual Properties	9.3.1
JDCC	2.1
JHSF License Agreement	3.1.1
JHSF Royalty	7.6.2
Losses and Claims	12.1
Supply Agreement	6.1
Term	13.1

ARTICLE 2

JOINT DEVELOPMENT AND COMMERCIALIZATION COMMITTEE

2.1 Joint Development and Commercialization Committee. The Parties agree to establish a joint Development and Commercialization committee ("JDCC"), which shall consist of no fewer than two (2) permanent members, with the JDCC having equal representation by each Party on the JDCC. Each Party may replace any or all of its representatives on the JDCC at any time upon written notice to the other Party. Such representatives shall include individuals within or designated by the senior management of each Party. JDCC representatives of each Party shall, individually or collectively, have expertise and/or responsibility in business to provide oversight and management of Development and Commercialization activities undertaken under this Agreement. Any member of the JDCC may designate a substitute to attend and perform the functions of that member at any meeting of the JDCC, and each member of the JDCC may invite such other non-members (subject to the confidentiality provisions set forth in Article 10) as deemed necessary to any meeting of the JDCC as nonvoting observers to help explore and resolve the issues before the JDCC.

2.1.1 Meetings. The JDCC shall meet [***] annually during the Term or more frequently as the Parties agree may be necessary or appropriate, or at such frequency as agreed by the respective committee members. Meetings of the JDCC shall alternate between the facilities of UPI in the United States (or such other location as specified by UPI), and the facilities of NPC in Japan (or such other location as specified by NPC), with the first such meeting to take place in Tokyo, Japan, provided that such meetings may also be held by video conference upon either Party's reasonable request. The JDCC members will otherwise communicate regularly by telephone, electronic mail, facsimile and/or video conference.

2.1.2 Responsibility; Decision Making. The JDCC shall perform the following functions: (i) exchange information concerning the overall strategy and timelines for the Development Plan; (ii) review and evaluate data and progress of the activities under the Development Plan; (iii) resolve disputes or disagreements between the Parties with respect to the Development Plan; (iv) ensure open communication between the Parties as relates to the Development Plan, including making arrangements for a third party service provider to provide for a secure electronic data room to share data, documentation, information and materials generated for or used in the research, development, production or other exploitation of the Compound and Products; (v) make amendments to the Development Plan then in effect; and (vi) taking such other actions as are specifically allocated to the JDCC under this Agreement. Any approval, determination or other action of the JDCC shall require agreement of the members of the JDCC, with each Party having one (1) vote. Action that may be taken at a meeting of the

JDCC also may be taken without a meeting if a written consent setting forth the action so taken is signed by all members of the JDCC. In the event the JDCC is unable to reach consensus on a particular matter within its jurisdiction, the matter shall be referred to executives of the Parties in accordance with Section 14.1, and if such referral does not resolve such matter, then the dispute shall be resolved via arbitration in accordance with Section 14.2.

2.2 Reports; Records; Transfer of Data and Documentation. Each Party shall cooperate fully with the other Party and shall provide the other Party with all data, documentation, information and materials generated or used by the Party in the research, development, production or other exploitation of the Compound and Products, in accordance with this Section 2.2. Each Party shall, following the end of [***] of each calendar year, report to the other Party in reasonable detail at the first meeting of the JDCC to be held after such [***] its Development and Commercialization activities in such Party's Territory (other than those under the Development Plan) during the [***] period until such [***]. Each Party agrees to provide the other Party with any preclinical data obtained during the course of each study specified in the Development Plan and any clinical data obtained during the course of its Development activities within a reasonable time period after the completion of each such study, to the extent a Party has the right to so provide such data. Any such data so provided shall be deemed the Confidential Information of both Parties, and each Party shall be authorized to use such data solely for the Development and Commercialization of Products in their respective Territory or as otherwise permitted under this Agreement.

ARTICLE 3

DEVELOPMENT AND COMMERCIALIZATION IN THE NPC TERRITORY

3.1 Development and Commercialization of Products. NPC shall have responsibility for Development and Commercialization of Products in the NPC Territory at its sole discretion and expense in accordance with this Article 3.

3.1.1 Commercially Reasonable Efforts. NPC shall use Commercially Reasonable Efforts (by itself or through contract service organizations or other permitted Sublicensees) to Develop and Commercialize the Product in the NPC Territory. Without limiting the foregoing, NPC shall use Commercially Reasonable Efforts to initiate, as defined in this Section 3.1.1, such clinical trials as maybe required by the relevant Regulatory Authority for a Product in Japan within [***] after the Effective Date, subject to no intervening action by such Regulatory Authority, *provided* that Nishino Technology is validly and effectively licensed to NPC under a license agreement with the JHSF (the "JHSF License Agreement") within a reasonable period of time after the Effective Date. In this Section 3.1.1, "initiate" shall mean the filing of an IND by NPC (including, but not limited to, an IND of investigator initiated trial sponsored by NPC) or, if applicable, such filing by referencing any existing IND of UPI.

3.1.2 Responsibilities. NPC shall be responsible for the strategy, plans and budgets for marketing and promotion of Products in the NPC Territory, including without limitation establishing and managing marketing, promotion, and distribution capabilities. NPC shall file and be the owner of all regulatory filings in the NPC Territory for Products, including all NDAs and Regulatory Approvals, unless otherwise agreed by the Parties.

3.2 Initial Document and Data Transfer. During the [***] period following the Effective Date NPC shall provide UPI with one (1) electronic or paper copy in English or Japanese of all documents, data or other information Controlled by NPC as of the Effective Date, to the extent (x) that such documents, data and information are (i) subject to the NPC Know-How license under Section 5.1; and (ii) reasonably necessary for UPI to obtain the Regulatory Approval of Products in the UPI Territory and (y) that NPC has the right to so provide. Such documentation shall not be used by UPI for any purpose other than Development, Manufacture and Commercialization of the Compound and Products in the UPI Territory in accordance with this Agreement. NPC shall be responsible for the cost of providing one (1) set of copies only. Any and all such materials delivered to UPI pursuant to this Section 3.2 are and shall remain the property of NPC, *provided* that all such material shall be treated as being Confidential Information of both Parties. The Parties agree that, consistent with the exclusive license granted under Section 5.1, during the term in which such license is in effect: (i) NPC shall not undertake any further Development activities in the UPI Territory; and (ii) NPC shall not engage or correspond with Regulatory Authorities in the UPI Territory without prior written approval of UPI.

3.3 Technical Assistance. Upon reasonable request by UPI, NPC shall reasonably cooperate with UPI to provide technical assistance with respect to Development of Products in the UPI Territory. Such cooperation shall include providing UPI with reasonable access by teleconference or in person at NPC's corporate and manufacturing facilities (subject to NPC's customary rules and restrictions with respect to site visits by non-NPC personnel) to NPC personnel involved in the research, manufacturing and development of the Compound and Products. Upon UPI's request, NPC may in its discretion dispatch NPC personnel to UPI in order to support Development of Products by UPI in the UPI Territory. All expenses for such dispatch including traveling and lodging expenses shall be equally shared by the Parties unless otherwise agreed between the Parties.

3.4 Trademarks. NPC shall have the right to select, own, maintain and enforce Trademarks for Products in the NPC Territory. In the event that UPI desires to use for Products in the UPI Territory one or more Product-specific Trademarks that are used in the NPC Territory and owned or Controlled by NPC, NPC shall, upon UPI's reasonable request, grant to UPI a royalty-free exclusive license, with the right to grant and authorize sublicenses, to use, display and enforce such Trademark(s) in connection with Products in the UPI Territory, *provided* that UPI will pay all registration and maintenance costs for such Trademarks and will agree to comply with the terms of a reasonable trademark policy provided by NPC to UPI in writing.

ARTICLE 4

DEVELOPMENT AND COMMERCIALIZATION IN THE UPI TERRITORY

4.1 Development and Commercialization of Products. UPI shall have responsibility for Development and Commercialization of Products in the UPI Territory at its sole discretion and expense in accordance with this Article 4.

4.1.1 Commercially Reasonable Efforts. UPI shall use Commercially Reasonable Efforts (by itself or through contract service organizations or other permitted Sublicensees) to Develop and Commercialize Products in the UPI Territory. Without limiting the foregoing, UPI shall use Commercially Reasonable Efforts to initiate, as defined in this Section 4.1.1, such clinical trials as maybe required by FDA (or EMEA) for a Product in North

America (or a major country in Europe) within [***] after the Effective Date, subject to no intervening FDA (or EMEA) action; provided that all documents, filings and other information reasonably necessary for UPI to achieve such deadline and Controlled by NPC shall be provided by NPC in accordance with terms of this agreement. In this Section 4.1.1, "initiate" means the filing of a separate IND by UPI or, if applicable, such filing by referencing any existing IND of NPC.

4.1.2 Responsibilities. UPI shall be responsible for the establishment and implementation of the strategy, plans and budgets for marketing and promotion of Products in the UPI Territory, including without limitation establishing and managing marketing, promotion, and distribution capabilities. UPI shall file and be the owner of all regulatory filings in the UPI Territory for Products, including all NDAs and Regulatory Approvals, unless otherwise agreed by the Parties. UPI agree that, consistent with the exclusive license granted under Section 5.2, during the term in which such license is in effect: (i) UPI shall not undertake any further Development activities in the NPC Territory; and (ii) UPI shall not engage or correspond with Regulatory Authorities in the NPC Territory without prior written approval of NPC.

4.2 Technical Assistance. Upon reasonable request by NPC, UPI shall reasonably cooperate with NPC to provide technical assistance with respect to Development of Products in the NPC Territory. Such cooperation shall include providing NPC with reasonable access by teleconference or in person at UPI's corporate and manufacturing facilities (subject to UPI's customary rules and restrictions with respect to site visits by non-UPI personnel) to UPI personnel involved in the research, manufacturing and development of the Compound and Products. Upon NPC's request, UPI may in its discretion dispatch UPI personnel to NPC in order to support Development of Products by NPC in the NPC Territory. All expenses for such dispatch including traveling and lodging expenses shall be equally shared by the Parties unless otherwise agreed between the Parties.

4.3 Trademarks. UPI shall have the right to select, own, maintain and enforce Trademarks for Products in the UPI Territory. In the event that NPC desires to use for Products in the NPC Territory one or more Product-specific Trademarks that are used outside the NPC Territory and owned or Controlled by UPI, UPI shall, upon NPC's reasonable request, grant to NPC a royalty-free exclusive license, with the right to grant and authorize sublicenses, to use, display and enforce such Trademark(s) in connection with Products in the NPC Territory, *provided* that NPC will pay all registration and maintenance costs for such Trademarks and will agree to comply with the terms of a reasonable trademark policy provided by UPI to NPC in writing.

ARTICLE 5

LICENSES AND COVENANTS NOT TO COMPETE

5.1 License to UPI. Subject to the terms and conditions of this Agreement, NPC hereby grants to UPI an exclusive license under the NPC Patent Rights and NPC Know-How, with the limited right to sublicense, solely as set forth in Section 5.3, to research, Develop, make and have made, use, import, offer for sale, sell and otherwise exploit and Commercialize Products in the UPI Territory.

5.2 License to NPC. Subject to the terms and conditions of this Agreement, UPI hereby grants to NPC an exclusive license under the UPI Patent Rights and UPI Know-How, with the limited right to sublicense solely as set forth in Section 5.3, to research, Develop, make and have made, use, import, offer for sale, sell and otherwise exploit and Commercialize Products in the NPC Territory.

5.3 License and Sublicense Grants. (a) Each Party shall have the right to grant sublicenses under the NPC Patent Rights, NPC Know-How, UPI Patent Rights and UPI Know How, as applicable, to contract service organizations and similar Third Parties to the extent reasonably necessary for such Party to exercise its rights or perform its obligations under Section 3.1 and 4.1 and the Supply Agreement. (b) Upon Commercialization, each Party shall have the right to sublicense the respective license granted in Section 5.1 and Section 5.2 to a bona fide collaboration partner; provided that such sublicensee shall not have the right to grant any further sublicenses. (c) Any sublicense granted under this Section 5.3 shall be subject to the following: (i) such sublicense agreement shall refer to this Agreement and shall be subordinated to and consistent with the terms and conditions of this Agreement; and (ii) the granting Party shall remain responsible for the performance of this Agreement (including without limitation obligations to participate in the JDCC and use Commercially Reasonable Efforts with respect to Development and Commercialization of the Product).

5.4 Covenant Not to Compete. Each Party hereby covenants that for a period of [***] after the Effective Date, it will not (by itself or through authorization of, or collaboration with, others) (i) conduct any Phase II Trials or Phase III Trials for purposes of seeking Regulatory Approval of a Competitive Product; or (ii) market, sell or promote any Competitive Product. For the purposes of this Section 5.4, "Competitive Product" means [***].

5.5 License on Completion of Term. On completion of the Term, solely due to the passage of time in accordance with Section 13.1, NPC shall grant to UPI an irrevocable, world-wide, fully-paid and royalty-free license under the NPC Patent Rights and NPC Know-How to research, Develop, Manufacture, use, import, offer for sale, sell and otherwise exploit Products in the UPI Territory, and UPI shall grant to NPC an irrevocable, world-wide, fully-paid and royalty-free license under the UPI Patent Rights and UPI Know-How to research, Develop, Manufacture, use, import, offer for sale, sell and otherwise exploit Products in the NPC Territory.

5.6 No Other Rights; No Implied Licenses. Other than the licenses expressly granted under this Agreement, no right, title or interest in NPC Patent Rights and NPC Know-How shall be granted, by implication, estoppel or otherwise, to UPI and no right, title or interest in UPI Patent Rights and UPI Know-How shall be granted, by implication, estoppel or otherwise, to NPC.

ARTICLE 6

SUPPLY

6.1 Manufacturing and Supply. The Parties shall negotiate in good faith and use Commercially Reasonable Efforts to enter into a definitive supply agreement setting forth the terms and conditions for Manufacture and supply to UPI by NPC of Products and/or Drug Substance (the "Supply Agreement") prior to the initiation of a Phase II Trial by UPI for the Product in the United States. Until and unless such Supply Agreement is entered into between the Parties, NPC shall use Commercially Reasonable Efforts to Manufacture and supply Products (and/or Drug Substance upon UPI's request) to UPI. It is understood and agreed between the Parties that under the Supply Agreement NPC shall not be obliged in any event, but shall use Commercially Reasonable Efforts, to Manufacture and supply the Products (and/or Drug Substance). The Supply Agreements shall at a minimum contain the terms and conditions that

are usual and customary in the pharmaceutical industry for a supply agreement of this nature and size, including without limitation that: (i) the Product or Drug Substance will be manufactured in compliance with GMP, and (ii) there shall be a reasonable quantity of inventory of the Product produced for storage in the UPI Territory. The size of such inventory will be agreed upon between the Parties and included in the Development Plan as well as in any plans for Commercialization.

6.2 Termination of Supply. If NPC desires to terminate the supply of Products and/or Drug Substance to UPI in accordance with this Agreement and the Supply Agreement, then NPC shall give UPI [***] written notice of termination if before the First UPI Approval in any major market country of the UPI Territory and [***] written notice of termination if after the First UPI Approval in any major market country of the UPI Territory. During the notice period, NPC must use Commercially Reasonable Efforts to supply the reasonable orders expected to maintain the successful development and commercialization of the Products. Upon the termination of the supply of Products and/or Drug Substance by NPC to UPI, NPC will then use Commercially Reasonable Efforts to obtain and provide to UPI, in a timely fashion, batch records, assays methods and manufacturing details required and customary for the technical transfer of production activities for Products and/or Drug Substance to a new facility of UPI's choice (with respect to such records, methods and details held or owned by a Third Party only to the extent that NPC is permitted to do so in writing by such Third Party), such that such UPI's new facility may be validated and activated for production of Products and/or Drug Substance.

If UPI desires to terminate the supply from NPC of Products and/or Drug Substance in accordance with this Agreement and the Supply Agreement, UPI must give NPC [***] written notice of termination if before the First UPI Approval in any major market country of the UPI

Territory and [***] written notice of termination if after the First UPI Approval in any major market country of the UPI Territory. NPC will provide sufficient technical information in a timely fashion to allow the institution of production of Products and/or Drug Substance at a new UPI manufacturing location at UPI's expense (with respect to such information held or owned by a Third Party only to the extent that NPC is permitted to do so in writing by such Third Party).

6.3 Notwithstanding the aforementioned, in case that UPI proceeds to Manufacture Products and/or Drug Substance independently from NPC, NPC has a right to purchase the Products and/or Drug Substance from UPI, and Supply Agreement shall define the terms and conditions, which are consistent with Article 6.1 and 6.2, for Manufacture and supply to NPC by UPI of the Products and/or Drug Substance. In the event that NPC exercises its rights under this section, UPI shall use Commercially Reasonable Efforts to Manufacture and supply Products (and/or Drug Substance upon NPC's request) to NPC.

ARTICLE 7

PAYMENTS

7.1 Upfront Payments and Equity Issuance.

7.1.1 Upfront License Fee. UPI has paid ¥10,000,000 to NPC as an upfront license fee per the terms of the Term Sheet for Sialic Acid Treatment dated May 28, 2010 entered into between NPC and UPI. Upon the Effective Date of this Agreement such fee shall become non-refundable.

7.1.2 Equity Issuance. As soon as reasonably practicable after the execution of this Agreement, UPI shall issue to NPC and NPC shall purchase 3,000 shares of common stock of UPI (representing approximately [***] percent ([***]%) of the outstanding capital of UPI) pursuant to stock purchase agreement consistent with UPI founder's stock purchase agreement which shall be disclosed to NPC upon the execution of this Agreement and containing such provisions and terms that are customary and usual for a stock purchase agreement with an investment of this size and nature with such reasonable modifications to be mutually agreed upon in good faith by the Parties. The purchase price of the UPI common stock shall be $0.01 per share. UPI shall ensure that such shares of UPI common stock when issued will be duly authorized, fully paid and non-assessable.

7.2 Development Milestones. UPI shall make non-refundable milestone payments to NPC in the amounts set forth below within [***] Business Days of the achievement of each of the milestones set forth therein:

1. Upon completion by NPC of formulation design of Products to be Commercialized in Japan but in no event before December 31, 2010: ¥20,000,000

2. Completion by NPC of teratogenicity study of Products using rats in Japan but in no event before March 31, 2011: ¥20,000,000

Notwithstanding the foregoing, the milestone payment set forth above shall be non-refundable only if NPC has not breached any material obligations hereunder at the time of the achievement of each milestone set forth above.

7.3 Approval Milestones. UPI shall make a nonrefundable milestone payment of ¥200,000,000 to NPC within [***] Business Days of [***].

7.4 Single Milestones. It is understood and agreed that the payments under these Sections 7.2 and 7.3 shall be due only once, upon the first instance of the milestone events described above. Accordingly, if for example UPI makes one or more of the payments due under Sections 7.2 and 7.3 with respect to achievement of the corresponding milestones, and then subsequently achieves the same development milestone again, UPI shall not be obligated to make any additional milestone payment.

7.5 Royalties.

7.5.1 Net Sales by UPI and its Affiliates. UPI shall pay to NPC a royalty of [***] percent ([***]%) on UPI's Net Sales of Products in the UPI Territory.

7.5.2 Net Sales by NPC and its Affiliates. NPC shall pay to UPI a royalty of [***] percent ([***]%) on NPC's Net Sales of Products in the NPC Territory (excluding Japan).

7.6 Additional Terms.

7.6.1 Royalty Term. The royalties due pursuant to Section 7.5 above shall be payable on a country-by-country and Product-by-Product basis commencing on the First Commercial Sale in the Party's Territory and continuing until the expiration of this Agreement.

7.6.2 Third Party Royalty Payments. If (i) UPI or its Affiliate or Sublicensee, as applicable, is legally required to obtain a license from any Third Party under Patent Rights or Know-How covering a Product in order to import, use or sell such Product; and (ii) UPI or its Affiliate or Sublicensee, as applicable, is legally required under such license to pay to such Third Party license fees, milestone payments, or royalties calculated on Net Sales of such Product, then the amount of UPI's royalty obligations under Section 7.5.1 hereof, shall be reduced by [***]

percent ([***]%) of the amount of such license fees, milestone payments or royalties paid to such Third Party, *provided* however, that the royalties payable in any calendar quarter under Section 7.5.1 hereof shall not be reduced in any event below [***] percent ([***]%) of the amounts set forth under Section 7.5.1. If (i) NPC or its Affiliate or Sublicensee, as applicable, is legally required to obtain a license from any Third Party under Patent Rights or Know-How covering a Product in order to import, use or sell such Product; and (ii) NPC or its Affiliate or Sublicensee, as applicable, is legally required under such license to pay to such Third Party license fees, milestone payments, or royalties calculated on Net Sales of such Product, then the amount of NPC's royalty obligations under Section 7.5.2 hereof shall be reduced by [***] percent ([***]%) of the amount of such license fees, milestone payments or royalties paid to such Third Party, *provided* however, that the royalties payable in any calendar quarter under Section 7.5.2 hereof shall not be reduced in any event below [***] percent ([***]%) of the amounts set forth under Section 7.5.2. UPI and NPC shall use Commercially Reasonable Efforts to minimize the amount of any of the foregoing payments owed to Third Parties. Prior to a Party committing to any payments to any Third Party under this Section 7.6.2, it shall provide the other Party with written notice of a potential need to obtain any Third Party license and the Parties shall discuss such potential need in good faith, *provided* that such discussions shall not unreasonably limit or delay such Party's reasonable judgment with respect thereto. Notwithstanding the foregoing, UPI and NPC shall equally share the royalty under the JHSF License Agreement (the "JHSF Royalty"), if any, for use of the Nishino Technology in the UPI Territory. NPC shall pay the license fee to the JHSF and use Commercially Reasonable Efforts to negotiate and obtain the HSTTC license for the Nishino Technology. Each Party shall bear the cost incurred in their respective Territory in connection with the prosecution and maintenance of the intellectual properties related to the Nishino Technology.

ARTICLE 8

PAYMENTS, BOOKS AND RECORDS

8.1.1 Reports; Payments. After the First Commercial Sale of a Product on which royalties are payable by a Party hereunder, such Party shall make and deliver to the other Party quarterly written reports within [***] days after the end of each calendar quarter during the Term stating the quantity, description, and aggregate Net Sales by country of each such Product sold by that Party and its Affiliates and Sublicensees during the calendar quarter. Within thirty [***] of the date of such report, the relevant Party shall pay to the other Party the royalties described above.

8.1.2 After the First Commercial Sale of a Product on which royalties are payable by UPI hereunder, UPI shall make and deliver to NPC annual written reports within [***] days after the end of each business year during the Term, which commences from April 1 and ends on March 31 of the following calendar year (the "Business Year"), stating the sales volume, gross sales, aggregate Net Sales and other items reasonably requested by NPC, by country of each such Product sold by UPI and its Affiliates and Sublicenses during the relevant Business Year. Notwithstanding the provisions of Article 10, NPC may disclose such annual reports to the JHSF subject to the JHSF's entering into a confidentiality agreement reasonably satisfactory to UPI. Further, UPI shall also promptly provide NPC with any information reasonably requested by NPC, and NPC may, notwithstanding the provisions of Article 10, disclose such information to the JHSF subject to the JHSF's entering into a confidentiality agreement reasonably satisfactory to UPI.

8.2 Payment Method. All payments due under this Agreement shall be made by bank wire transfer in immediately available funds to a bank account designated by the Party entitled to receive such payment. All payments hereunder shall be made in Japanese Yen. Any payments that are not paid on the date such payments are due under this Agreement shall bear interest to the extent permitted by applicable Law at a rate equal to the 3-month LIBOR rate at the close of business on the date such payment is due, plus an additional [***]%, calculated on the number of days such payment is delinquent.

8.3 Place of Royalty Payment; Currency Conversion. With respect to gross sales or Net Sales invoiced in a currency other than U.S. Dollars, such gross sales or Net Sales in the quarterly report or annual report as set forth in Section 8.1 above shall be expressed in the invoiced currency, together with the U.S. Dollar and Japanese Yen equivalent (and the royalty payment hereunder shall be made based on such Japanese Yen equivalent), calculated using the arithmetic average of the spot rates on the close of business on the last Business Day of each month of the relevant calendar quarter or the relevant Business Year, in which such gross sales or Net Sales were made. The "closing mid-point rates" found in the "dollar (or yen) spot forward against the dollar (or yen)" table published by The Financial Times or any other publication as agreed to by the Parties shall be used as the source of spot rates to calculate the average as defined in the preceding sentence.

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

8.4 Records; Inspection. Each Party shall keep, and shall ensure that its Affiliates keep, complete, true and accurate books of account and records for the purpose of determining the amounts payable under this Agreement. Such books and records shall be kept at the principal place of business of such Party, for at least [***] following the end of the calendar quarter during the Term to which they pertain. Such records will be open for inspection (i) by a public accounting firm designated by the auditing Party which is reasonably acceptable to the audited Party and subject to such accounting firm entering into a satisfactory confidentiality agreement, solely for the purpose of determining the payments to the auditing Party hereunder, or (ii) by the JHSF if the audited Party is UPI and upon a reasonable request by NPC, which is the auditing Party. Such inspections may be made no more than once each calendar year, at reasonable times and on reasonable advance notice. Inspections conducted under this Section 8.4 shall be at the expense of the auditing Party, unless an error in books of account and records of the audited Party producing an increase exceeding [***] percent ([***]%) of the amount payable under this Agreement stated for the period covered by the inspection is found in the course of such inspection, whereupon all reasonable costs relating to the inspection for such period shall be borne by the audited Party. Any unpaid or overpaid amounts that are discovered during the course of such inspection will be promptly paid or refunded by the appropriate Party, in each case together with interest noted in Section 8.2 thereon from the date such payments were due (if underpaid) or paid (if overpaid).

8.5 Withholding Taxes. Each Party shall pay any and all taxes levied on account of amounts payable to it under this Agreement. If applicable Laws or regulations require that taxes be withheld, the paying Party will (i) deduct those taxes from the remittable payment, (ii) timely pay the taxes to the proper authority, and (iii) send proof of payment to the other Party within [***] days following that payment.

ARTICLE 9

INTELLECTUAL PROPERTY

9.1 Ownership.

9.1.1 Ownership. All intellectual property right and other right, title, and interest in: (i) UPI Patent Rights and UPI Know-How shall be owned solely by UPI; and (ii) NPC Patent Rights and NPC Know-How shall be owned solely by NPC. All intellectual property right and other right, title and interest in Joint Patent Rights and Joint Know-How shall be owned jointly by UPI and NPC.

9.1.2 Disclosure of Joint Know-How. Each Party shall promptly disclose to the other all Inventions within the Joint Know-How.

9.1.3 Inventorship. In the event of any disagreement between the Parties regarding the inventorship of any Invention, the Parties shall subject the disagreement to the arbitration process contained in Section 14.2.

9.1.4 Execution of Documents. Each Party shall execute such documents and perform such acts as may be reasonably requested by the other Party to enable the other Party to use its interest in the Joint Know-How and Joint Patent Rights or to otherwise give effect to the provisions set forth in this Section 9.1.

9.2 Intellectual Property Right Application. If a Party intends to make application in such Party's Territory for patent(s), utility model(s) and/or other intellectual property right(s) concerning any Know-How obtained, developed or invented by such Party during the course of the Development activities under this Agreement (including, but not limited to, Joint Know-How), then such Party shall disclose the contents of such application to the other Party in advance of the filing for review and comment.

9.3.1 NPC Territory. As used herein, “prosecution” shall include interferences, reexaminations, reissues, oppositions, and the like. In the NPC Territory, NPC shall have the sole right to control the preparation, filing, prosecution and maintenance of NPC Patent Rights, NPC Know-How, UPI Patent Rights, UPI Know-How, Joint Patent Rights and Joint Know-How (collectively, the “Intellectual Properties”) using patent counsel of NPC’s choice. UPI shall have the right to review and comment on prosecution and patent applications prior to their filing.

9.3.2 UPI Territory. In the UPI Territory, UPI shall have the sole right to control the preparation, filing, prosecution and maintenance of the Intellectual Properties using patent counsel of UPI’s choice. NPC shall have the right of review and comment on prosecution and patent applications prior to their filing.

9.3.3 Abandonment. Each Party shall give timely notice to the other Party of any decision not to file applications for, or to cease prosecution and/or maintenance of, or not to continue to pay the expenses of prosecution and/or maintenance of, any Joint Patent Rights in their respective Territory (such decision, an “Abandonment”) and, in such case, shall permit the other Party, at its sole discretion and expense, to file or to continue prosecution or maintenance of such Joint Patent Rights in their own Territory. Any such patent rights or patent applications for which the option granted under this Section is exercised shall not be considered part of the Joint Patent Rights thereafter and the claims of such patent rights or patent applications shall not be subject to the license provisions of Article 5 unless otherwise agreed by the Parties.

9.3.4 Intellectual Property Costs. Unless otherwise provided for in this Agreement:

(a) in the UPI Territory, all costs associated with filing, prosecuting, issuing and maintaining UPI Patent Rights, NPC Patent Rights, UPI Know-How and NPC Know-How including interference, opposition, reexamination and reissue actions, shall be borne by UPI;

(b) in the NPC Territory, all costs associated with filing, prosecuting, issuing and maintaining NPC Patent Rights, UPI Patent Rights, NPC Know-How and UPI Know-How including interference, opposition, reexamination and reissue actions shall be borne by NPC; and

(c) all costs associated with filing, prosecuting, issuing and maintaining Joint Patent Rights and Joint Know-How, including interference, opposition, reexamination and reissue actions, shall be equally shared by the Parties.

9.4 Cooperation. Each Party will promptly provide to the other Party information reasonably requested by other Party that is necessary for the prosecution activities pursuant to Section 9.3 above. The Parties agree to cooperate and to take such actions as the Parties mutually agree are reasonable to maximize the protections available under relevant regulations such as the safe harbor provisions of 35 U.S.C. 103(c) for USA patents/patent applications.

9.5 Infringement. UPI and NPC shall promptly notify the other in writing of any alleged or threatened infringement of any of the Intellectual Properties or infringement of any Third Party's intellectual property by developing, making, having made, using, importing, selling or having sold Products or Drug Substance (collectively, the "Infringement"), of which they become aware.

9.5.1 UPI Territory. In the UPI Territory, UPI shall have the sole right to bring and control any action or proceeding with respect to Infringement by counsel of its own choice.

9.5.2 NPC Territory. In the NPC Territory, NPC shall have the sole right to bring and control any action or proceeding with respect to Infringement by counsel of its own choice.

9.5.3 Unless otherwise provided for in this Agreement:

(a) in the UPI Territory, all costs associated with the enforcement of UPI Patent Rights, NPC Patent Rights, UPI Know-How and NPC Know-How and defense against Infringement shall be borne by UPI;

(b) in the NPC Territory, all costs associated with the enforcement of UPI Patent Rights, NPC Patent Rights, UPI Know-How and NPC Know-How and defense against Infringement shall be borne by NPC; and

(c) all costs associated with the enforcement of Joint Patent Rights and Joint Know-How shall be equally shared by the Parties.

9.6 Patent Term Extension; Data Exclusivity. UPI shall use Commercially Reasonable Efforts to obtain patent term extensions, supplemental protection certificates or data exclusivity periods (such as those periods listed in the FDA's Orange Book or its equivalent with the EMEA), or their equivalents in any jurisdiction in Europe and North America, and NPC shall cooperate with UPI in doing so at UPI's expense, with respect to Products and NPC Patent Rights and UPI Patent Rights covering the Products. If elections with respect to obtaining such patent term extensions or data exclusivity periods are to be made, UPI shall have the right to make the election to seek patent term extension or data exclusivity periods, *provided* that such election will be made so as to maximize the period of marketing exclusivity for the Products.

ARTICLE 10

CONFIDENTIALITY

10.1 Confidentiality. During the Term of this Agreement and for a period of [***] following the expiration or earlier termination hereof, each Party shall maintain in confidence the Confidential Information of the other Party, shall not use or grant the use of the Confidential Information of the other Party except as expressly permitted hereby, and shall not disclose the Confidential Information of the other Party (in each case, irrespective of whether such Confidential Information is also the Confidential Information of such Party), except (i) on a need-to-know basis to such Party's directors, officers and employees, (ii) to such Party's consultants performing work contemplated by the Agreement, and to any subcontractor performing work for such Party hereunder, or (iii) to the extent such disclosure is reasonably necessary in connection with such Party's activities under rights and licenses expressly authorized by this Agreement. To the extent that disclosure to any person is authorized by this Agreement, prior to disclosure, a Party shall obtain written agreement of such person to hold in confidence and not disclose, use or grant the use of the Confidential Information of the other Party except as expressly permitted under this Agreement. Each Party shall notify the other Party promptly upon discovery of any unauthorized use or disclosure of the other Party's Confidential Information.

10.2 Permitted Use and Disclosures. The confidentiality obligations under this Article 10 shall not apply to the extent that a Party is required to disclose information by applicable Law, regulation or order of a governmental agency or a court of competent jurisdiction, including filings required by the Securities and Exchange Commission or any similar body, or any securities exchange; *provided*, however, that such Party shall provide written notice thereof to

the other Party (to the extent not prohibited by Law or court order), and consult with the other Party with respect to such disclosure and provide the other Party reasonable opportunity to object to any such disclosure or to request confidential treatment thereof. Notwithstanding the provisions of this Article 10, the Parties agree that: (i) either Party may, to the extent reasonably necessary, disclose Confidential Information of the other Party to any Regulatory Authority in connection with the Development of a Product which it has the right to Develop under this Agreement; (ii) to the extent they have not done so as of the Effective Date, the Parties shall agree upon a press release related to this Agreement. In addition, each Party will consider in good faith any request by the other Party for a public disclosure not otherwise permitted pursuant to this Article 10, with consent for such disclosure not to be unreasonably withheld, conditioned or delayed. In the event of any termination of this Agreement under Article 13, the Parties shall agree on an announcement of such termination; provided that the Parties shall use reasonable efforts to fashion such announcement so as to minimize any negative impact on either Party as a result of such announcement. Once a particular item of information has been publicly disclosed pursuant to this Article 10, further consent will not be needed for further disclosures thereof.

10.3 Additional Nondisclosure. Each of the Parties hereto agrees not to disclose the terms and conditions of this Agreement to any Third Party without the prior written consent of the other Party hereto, which consent shall not be unreasonably withheld, conditioned or delayed, except to (i) the JHSF subject to the JHSF's entering into a confidentiality agreement reasonably satisfactory to UPI, (ii) such Party's attorneys, advisors and actual collaborators on a need to know basis under circumstances that reasonably protect the confidentiality thereof, (iii) Inabata & Co., Ltd., Hisanaga & Co., Ltd. and Development Bank of Japan Inc. (collectively, the "NPC's Shareholders") as long as the NPC's Shareholders maintain ownership of NPC's stock

and are subject to substantially similar terms and conditions of confidentiality obligations of this Agreement, (iv) banks, potential investors and other financial institutions from which NPC or UPI are receiving, will receive or plan to be receiving financing from provided they are subject to confidentiality obligations with substantially similar terms and conditions to the confidentiality obligations of this Agreement, or (v) others to the extent required by Law (and with appropriate requests made for confidential treatment), including disclosures associated with filings required to be made by Law with the Securities and Exchange Commission, any similar body, or any national securities exchange.

10.4 Publication. Any manuscript prepared by UPI or NPC (the "Disclosing Party" in this Section 10.4; for the purpose of this Section 10.4, any manuscript prepared by UPI's or NPC's Affiliate shall be deemed as manuscript prepared by UPI or NPC, respectively) on subject matter in connection with this Agreement to be published or publicly disclosed pursuant to this Article 10, shall be subject to the prior review of the other Party (the "Receiving Party" in this Section 10.4) at least [***] days prior to such publication or disclosure and may not be published or publicly disclosed without the prior written consent of the Receiving Party, which consent shall not be unreasonably withheld, conditioned or delayed. Further, to avoid loss of patent rights as a result of premature public disclosure of patentable information, the Receiving Party shall notify the Disclosing Party in writing within [***] Business Days after receipt of such manuscript of whether the Receiving Party desires to file a patent application on any invention disclosed in such manuscript. In the event that the Receiving Party desires to file such a patent application, the Disclosing Party shall withhold publication or disclosure of such manuscript until the earlier of (i) a patent application is filed thereon, or (ii) the Parties determine after consultation that no patentable invention exists after receipt by the Disclosing Party of the

Receiving Party's written notice of the Receiving Party's desire to file such patent application. Further, if such manuscript contains the information of the Receiving Party that is subject to use and nondisclosure restrictions under this Article 10, the Disclosing Party agrees to remove, upon request of the Receiving Party, such information from the proposed publication or disclosure. The determination of authorship and final editorial control over any publications related to any clinical study or research will be the responsibility of the Party that funded such clinical study or research.

ARTICLE 11

REPRESENTATIONS AND WARRANTIES

11.1 Mutual Representations and Warranties. Each Party represents and warrants to the other Party as of the Effective Date that: (i) it has all requisite corporate power and authority to enter into this Agreement and to perform its obligations under this Agreement; (ii) execution of this Agreement and the performance by such Party of its obligations hereunder have been duly authorized; (iii) this Agreement is legally binding and enforceable on each Party in accordance with its terms; (iv) execution and delivery of this Agreement and the performance of such Party's obligations hereunder do not conflict with or violate any applicable Laws and do not conflict with, or constitute a breach or default under, any contractual obligation of such Party; and (v) all necessary consents, approvals and authorizations of all governmental authorities and other Persons required to be obtained by such Party in connection with entering into this Agreement have been obtained.

11.2 Representations and Warranties of NPC. NPC represents and warrants to UPI that:

11.2.1 Intellectual Property Rights. As of the Effective Date: (i) NPC has not previously assigned, transferred, conveyed or otherwise encumbered its right, title and interest in the NPC Patent Rights or NPC Know-How in the UPI Territory in a manner inconsistent with the rights, options and licenses granted to UPI herein or NPC's obligations to Manufacture and supply Drug Substance and/or Product pursuant to Article 6, including without limitation by authorizing any Third Party to Manufacture, use or sell the Compound or any Products in any country in the UPI Territory; and (ii) NPC is not aware of any facts or circumstances that would cause the UPI Territory to be invalid or unenforceable, and all necessary fees and other actions required in order to maintain the NPC Patent Rights have been paid or performed to date. In particular, as of the Effective Date NPC owns all right, title and interest in and to the NPC Patent Rights in the UPI Territory, and has not granted any right or license under the NPC Patent Rights authorizing any Third Party to Manufacture, use or sell the Compound in any country the UPI Territory that are inconsistent with the terms of this Agreement.

11.2.2 Disclosure. As of the Effective Date, there is no matter within the knowledge of NPC which NPC has intentionally, knowingly, or negligently failed to disclose, which by itself or in connection with any other matters not disclosed by NPC (i) is material to the evaluation of the Compound and/or Products; and (ii) would materially adversely affect the further Development or Commercialization of any Product.

11.3 Representations and Warranties of UPI. UPI represents and warrants to NPC that:

11.3.1 Intellectual Property Rights. As of the Effective Date: (i) UPI has not previously assigned, transferred, conveyed or otherwise encumbered its right, title and interest in the[UPI Patent Rights or] UPI Know-How in the NPC Territory in a manner inconsistent with

the rights, options and licenses granted to NPC herein, including without limitation by authorizing any Third Party to Manufacture, use or sell the Compound or any Products in any country in the NPC Territory; and (ii) UPI is not aware of any facts or circumstances that would cause the NPC Territory to be invalid or unenforceable, and all necessary fees and other actions required in order to maintain the UPI Patent Rights have been paid or performed to date. In particular, as of the Effective Date UPI owns all right, title and interest in and to the UPI Patent Rights in the NPC Territory, and has not granted any right or license under the UPI Patent Rights authorizing any Third Party to Manufacture, use or sell the Compound in any country the NPC Territory that are inconsistent with the terms of this Agreement.

11.3.2 Disclosure. As of the Effective Date, there is no matter within the knowledge of UPI which UPI has intentionally, knowingly, or negligently failed to disclose, which by itself or in connection with any other matters not disclosed by UPI (i) is material to the evaluation of the Compound and/or Products; and (ii) would materially adversely affect the further Development or Commercialization of any Product.

ARTICLE 12

INDEMNIFICATION; PHARMACOVIGILANCE

12.1 UPI Indemnity. UPI shall indemnify, defend and hold harmless NPC and its Affiliates, and their respective officers, directors, employees, agents, licensors, and their respective successors, heirs and assigns and representatives, from and against any and all claims, threatened claims, damages, losses, suits, proceedings, liabilities, costs (including, without limitation, reasonable legal expenses, costs of litigation and reasonable attorney's fees) or judgments, whether for money or equitable relief, of any kind, arising out of any claim, action,

lawsuit or other proceeding brought by a Third Party ("Losses and Claims") arising out of or relating, directly or indirectly, to: (i) the Development or Commercialization of the Compound or any Product in the UPI Territory by UPI, its Affiliates or Sublicensees; (ii) the Commercialization of the Compound or any Product in Europe by UPI or its Affiliates; (iii) breach by UPI of Article 11 or any other provision of this Agreement; or (iv) the negligence, recklessness or willful misconduct of UPI; *except* where such Losses and Claims are attributable to a failure by NPC to comply with applicable Law or the negligence, recklessness or willful misconduct of NPC.

12.2 NPC Indemnity. NPC shall indemnify, defend and hold harmless UPI and its Affiliates, and their respective officers, directors, employees, agents, licensors, and their respective successors, heirs and assigns and representatives, from and against any and all Losses and Claims arising out of or relating, directly or indirectly, to: (i) the Development or Commercialization of the Compound or any Product in the NPC Territory by NPC or its Affiliates or Sublicensees; (ii) breach by NPC of Article 11 or any other provision of this Agreement; or (iii) the negligence, recklessness or willful misconduct of NPC; *except where* such Losses and Claims are attributable to a failure by UPI to comply with applicable Law or the negligence, recklessness or willful misconduct of UPI.

12.3 Indemnification Procedure. A claim to which indemnification applies under Section 12.1 or Section 12.2 shall be referred to herein as an "Indemnification Claim". If any Person or Persons (collectively, the "Indemnitee") intends to claim indemnification under this Article 12, the Indemnitee shall notify the other Party (the "Indemnitor") in writing promptly upon becoming aware of any claim that may be an Indemnification Claim (it being understood and agreed, however, that the failure by an Indemnitee to give such notice shall not relieve the

Indemnitor of its indemnification obligation under this Agreement except and only to the extent that the Indemnitor is actually prejudiced as a result of such failure to give notice). The Indemnitor shall have the right to assume and control the defense of the Indemnification Claim at its own expense with counsel selected by the Indemnitor and reasonably acceptable to the Indemnitee, provided, however, that an Indemnitee shall have the right to retain its own counsel, with the fees and expenses to be paid by the Indemnitee, if representation of such Indemnitee by the counsel retained by the Indemnitor would be inappropriate due to actual or potential differing interests between such Indemnitee and any other Party represented by such counsel in such proceedings. If the Indemnitor does not assume the defense of the Indemnification Claim as aforesaid, the Indemnitee may defend the Indemnification Claim but shall have no obligation to do so. The Indemnitee shall not settle or compromise the Indemnification Claim without the prior written consent of the Indemnitor, and the Indemnitor shall not settle or compromise the Indemnification Claim in any manner which would have an adverse effect on the Indemnitee's interests (including without limitation any rights under this Agreement or the scope or enforceability of the NPC Patents Rights or NPC Know-How), without the prior written consent of the Indemnitee, which consent, in each case, shall not be unreasonably withheld or delayed. The Indemnitee shall reasonably cooperate with the Indemnitor at the Indemnitor's expense and shall make available to the Indemnitor all pertinent information under the control of the Indemnitee, which information shall be subject to Article 10.

12.4 Insurance. Within [***] months after the Effective Date, each Party shall at its own expense procure and maintain during the Term and for a period of [***] years thereafter an insurance policy/policies, adequate to cover its obligations hereunder and which is/are consistent with normal business practices of prudent companies similarly situated. Each Party's insurance

coverage shall include without limitation [***]. Such insurance shall not be construed to create a limit of the insuring Party's liability with respect to its indemnification obligations under this Article 12. Each Party shall provide the other Party with a certificate of insurance or other evidence thereof upon request. Each Party shall provide the other Party with written notice at least [***] days prior to the cancellation, non-renewal or a material change in such insurance or self-insurance which materially adversely affects the rights of the other Party hereunder.

12.5 Global Pharmacovigilance. NPC and UPI shall negotiate in good faith and use reasonable efforts to enter into an agreement regarding the pharmacovigilance of Products within a reasonable period of time after the Effective Date.

ARTICLE 13

TERM AND TERMINATION

13.1 Term. The Term of this Agreement shall commence on the Effective Date, and shall continue in full force and effect on a country-by-country until the date of the first launch of a generic product of the Product in a country, unless earlier terminated as provided in this Article 13 (the "Term").

13.2 Termination for Breach. Subject to Section 13.5, either Party to this Agreement may terminate this Agreement in the event the other Party hereto shall have materially breached or defaulted in the performance of any of its material obligations hereunder (other than failure to use Commercially Reasonable Efforts (by itself or through contract service organizations or other permitted Third Party licensees or sublicensees) to Develop or Commercialize a Product under Section 3.1.1 or Section 4.1.1), and such breach or default shall have continued for thirty

(30) days after written notice thereof was provided to the breaching Party by the non-breaching Party. Any termination shall become effective at the end of such thirty (30) day period unless the breaching Party has cured any such breach or default prior to the expiration of the thirty (30) day period.

13.3 Failure to Develop or Commercialize Products. Five (5) years after the Effective Date or two (2) years after the first Regulatory Approval in a major market country, whichever is the earlier, the Parties shall discuss in good faith on how to deal with the Development and Commercialization of Products in countries where Products have not been commercially sold regardless of whether the Parties have performed their obligations under Section 3.1.1 or 4.1.1.

13.4 Termination For Bankruptcy. Either Party hereto shall have the right to terminate this Agreement forthwith by written notice to the other Party (i) if the other Party is declared insolvent or bankrupt by a court of competent jurisdiction, (ii) if a voluntary or involuntary petition in bankruptcy is filed in any court of competent jurisdiction against the other Party and such petition is not dismissed within thirty (30) days after filing, (iii) if the other Party shall make or execute an assignment of substantially all of its assets for the benefit of creditors, or (iv) substantially all of the assets of such other Party are seized or attached and not released within thirty (30) days thereafter.

13.5 Disputed Breach. If either Party disputes in good faith the existence or materiality of a breach specified in a notice provided by the other Party pursuant to Section 13.2, and the allegedly breaching Party provides notice to the other Party of such dispute within the applicable thirty (30) day cure period, the other Party shall not have the right to terminate this Agreement unless and until the existence of such material breach or failure has been determined in

accordance with Section 14.2 and the allegedly breaching Party fails to cure such breach within thirty (30) days following such determination (except to the extent such breach involves the failure to make a payment when due, which breach must be cured within ten (10) days following such determination). It is understood and acknowledged that while such a dispute is pending all of the terms and conditions of this Agreement shall remain in effect and the Parties shall continue to perform all of their respective obligations hereunder. The Parties further agree that any payments that are made by one Party to the other Party pursuant to this Agreement pending resolution of the dispute shall be promptly refunded if an arbitrator determines pursuant to Section 14.2 that such payments are to be refunded by one Party to the other Party.

13.7 Effect of Termination. Upon expiration or termination of this Agreement for any reason, the rights and obligations of the Parties shall be as set forth in this Section 13.7.

13.7.1 License on Termination for UPI Breach or Bankruptcy. Upon termination of this Agreement by NPC pursuant to Section 13.2 or 13.4 hereof, UPI shall grant to NPC an irrevocable, world-wide, fully-paid and royalty-free license under the UPI Patent Rights and UPI Know-How to research, Develop, Manufacture, use, import, offer for sale, sell and otherwise exploit Products in the NPC Territory.

13.7.2 License on Termination for NPC Breach or Bankruptcy. Upon termination of this Agreement by UPI pursuant to Section 13.2 or 13.4, NPC shall grant to UPI an irrevocable, world-wide, fully-paid and royalty-free license under the NPC Patent Rights and NPC Know-How to research, Develop, Manufacture, use, import, offer for sale, sell and otherwise exploit Products in the UPI Territory.

13.7.3 Survival. The following provisions shall survive termination or expiration of this Agreement for any reason: Sections 5.5, 9.1, 13.7, 15.1 through 15.4, and 15.7 through 15.12, and Articles 1, 8, 10, 12 and 14. Expiration or termination of this Agreement for any reason shall not relieve the Parties of any liability or obligation accruing on or prior to such expiration or termination, or which is attributable to a period prior to such expiration or termination, nor preclude either Party from pursuing any rights and remedies it may have hereunder or at law or in equity which accrued or are based upon any event occurring prior to such expiration or termination. Unless otherwise set forth in this Agreement, any other rights, obligations and provisions hereunder shall terminate and cease to have effect upon expiration or termination of this Agreement.

ARTICLE 14

DISPUTE RESOLUTION

14.1 Escalation to Senior Executives. Other than pursuit of equitable relief as provided in Section 14.4, in the event of a dispute or matter of significant concern arises between the Parties, then at the request of either Party, the matter shall be escalated to the CEO of UPI and the senior executive at NPC with responsibility for pharmaceutical products and authority to make a decision on behalf of the respective Party with respect to such matter. Upon such request, such senior executives shall make themselves reasonably available to meet, and shall meet either by telephone or if specifically requested, in person, to attempt to resolve such matter, and shall thereafter continue to use good faith efforts to attempt to resolve such matter unless it becomes clear that the matter cannot be resolved by mutual agreement. Thereafter, either Party may initiate arbitration pursuant to Section 14.2 below upon written notice to the other Party.

14.2 Arbitration of Disputes. Other than pursuit of equitable relief as provided in Section 14.4, the Parties shall resolve disputes in accordance with this Section 14.2. Any disputes, controversies or differences that may arise under or in relation to this Agreement and which cannot be settled under Section 14.1 shall be settled by arbitration. The arbitration shall be conducted in San Francisco, California in accordance with the applicable rules of the AAA if UPI is the respondent in such dispute or in Tokyo in accordance with the applicable rules of the JCAA if NPC is the respondent in such dispute. Any arbitration conducted under this Section 14.2 shall be conducted in English. The determination of the arbitrator as to the resolution of any dispute shall be binding and conclusive upon all Parties.

14.4 Injunctive Relief. This Article 14 shall not be construed to prohibit either Party from seeking preliminary or permanent injunctive relief, restraining order or decree of specific performance in any court of competent jurisdiction to the extent not prohibited by this Agreement. For avoidance of doubt, any such equitable remedies provided under this Article 14 shall be cumulative and not exclusive and are in addition to any other remedies, which either Party may have under this Agreement or applicable Law.

ARTICLE 15

MISCELLANEOUS

15.1 Governing Laws. This Agreement and any dispute arising from the construction, performance or breach hereof shall be governed by and construed, and enforced in accordance with, the laws of the state of New York, without reference to conflicts of laws principles.

15.2 Waiver. It is agreed that no waiver by either Party hereto of any breach or default of any of the covenants or agreements herein set forth shall be deemed a waiver as to any subsequent and/or similar breach or default.

15.3 Assignment. This Agreement shall not be assignable by either Party without the written consent of the other Party hereto, except either Party may assign this Agreement without such consent to its Affiliates, or to an entity that acquires all or substantially all of the business or assets of such Party related to this Agreement whether by merger, reorganization, acquisition, sale, or otherwise; *provided*, however, that the assignee shall agree in writing to be bound by the terms and conditions of this Agreement.

15.4 Independent Contractors. Both Parties are independent contractors under this Agreement. Nothing herein contained shall be deemed to create an employment, agency, joint venture or partnership relationship between the Parties or any of their agents or employees, or any other legal arrangement that would impose liability upon one Party for the act or failure to act of the other Party. Neither Party shall have any express or implied power to enter into any contracts or commitments or to incur any liabilities in the name of, or on behalf of, the other Party, or to bind the other Party in any respect whatsoever.

15.5 Compliance with Laws. In exercising their rights under this license, the Parties shall fully comply in all material respects with the requirements of any and all applicable Laws, regulations, rules and orders of any governmental body having jurisdiction over the exercise of rights under this license including, without limitation, those applicable to the discovery, development, manufacture, distribution, import and export and sale of Products pursuant to this Agreement.

15.6 Patent Marking. Each Party agrees to mark, and to make sure that its Sublicensees mark, all Products sold by or under authority of such Party pursuant to this Agreement in accordance with the applicable statute or regulations relating to patent marking in the country or countries of manufacture and sale thereof.

15.7 Notices. All notices, requests and other communications hereunder shall be in writing and shall be personally delivered or by registered or certified mail, return receipt requested, postage prepaid, in each case to the respective address specified below, or such other address as may be specified in writing to the other Parties hereto and shall be deemed to have been given upon receipt:

UPI: Ultragenyx Pharmaceutical Inc.
77 Digital Drive, Suite 210
Novato, CA 94949 USA
Attn: Chief Executive Officer

With Copies to (for information purposes only):

Wright Legal Advisory, LLC,
3213 W, Wheeler St, Suite 307
Seattle, WA 91899
Attn: Managing Member

NPC: Nobelpharma Co., Ltd.
Kyodo Bldg. (Horidome)
12-10 Nihonbashi-kobunacho, Chuo-Ku
Tokyo 103-0024, Japan
Attention: Managing Director and CEO

15.8 Severability. In the event that any provision of this Agreement becomes or is declared by a court of competent jurisdiction to be illegal, unenforceable or void, this Agreement shall continue in full force and effect to the fullest extent permitted by law without said provision, and the Parties shall amend the Agreement to the extent feasible to lawfully include the substance of the excluded term to as fully as possible realize the intent of the Parties and their commercial bargain.

15.9 Advice of Counsel. UPI and NPC have each consulted counsel of their choice regarding this Agreement, and each acknowledges and agrees that this Agreement shall not be deemed to have been drafted by one Party or another and will be construed accordingly.

15.10 Complete Agreement. This Agreement constitutes the entire agreement, both written and oral, between the Parties with respect to the subject matter hereof, and all prior agreements respecting the subject matter hereof, either written or oral, express or implied, shall be abrogated, canceled, and are null and void and of no effect. No amendment or change hereof or addition hereto shall be effective or binding on either of the Parties hereto unless reduced to writing and executed by the respective duly authorized representatives of UPI and NPC.

15.11 Headings. The captions to the several Sections and Articles hereof are not a part of this Agreement, but are included merely for convenience of reference and shall not affect its meaning or interpretation.

15.12 Counterparts. This Agreement may be executed in counterparts, each of which shall be deemed to be an original and all of which together shall be deemed to be one and the same agreement.

[remainder of page intentionally left blank]

IN WITNESS WHEREOF, the Parties hereto have caused this Agreement to be duly executed by their authorized representatives and delivered in duplicate originals as of the Effective Date.

Nobelpharma Co., Ltd.

/s/ Jin Shiomura
Name: Jin Shiomura
Title: Managing Director and CEO

Ultragenyx Pharmaceutical Inc.

/s/ Emil Kakkis
Name: Emil Kakkis
Title: CEO and President

LICENSE AGREEMENT

Effective as of September 1, 2012 (the "Effective Date"), St. Jude Children's Research Hospital, having a principal place of business at ("INSTITUTION"), and Ultragenyx Pharmaceutical, Inc., a Delaware corporation having a principal place of business at 60 Leveroni Court, Novato, CA ("LICENSEE"), agree as follows:

1. BACKGROUND

1.1. INSTITUTION has an assignment of certain Technology, as hereinafter defined, from the laboratory of Dr. Alessandra D'Azzo.

1.2. INSTITUTION desires to have the Technology perfected and marketed at the earliest possible time in order that products resulting therefrom may be available for public use and benefit.

1.3. LICENSEE desires a license under said Technology to develop, manufacture, have made, use, and sell product(s) incorporating the Technology.

2. DEFINITIONS

2.1. "Affiliate" means any corporation or other entity that is directly or indirectly controlling, controlled by or under common control with LICENSEE. For the purpose of this definition, "control" shall mean the direct or indirect beneficial ownership of at least fifty percent (50%) of the outstanding shares or other voting rights of the subject entity to elect directors, or if not meeting the preceding, any entity owned or controlled by or owning or controlling at the maximum control or ownership right permitted in the country where such entity exists.

2.2. "Licensed Field" means the use of PPCA protein to treat, prevent and/or diagnose galactosialidosis and other Monogenetic Disorders.

2.3. "Licensed Know-How" means scientific, clinical or other information, knowledge and know-how and materials developed in laboratory of Dr. Alessandra D'Azzo as of the Effective Date and for a period of [***] years thereafter that are directed to the Technology, including but not limited to data, reports, and materials from preclinical studies, cell lines, or other specific assay reagents for assays or enzyme production processes, and access to the mouse models for purposes of development of enzyme replacement therapy for galactosialidosis and other materials related to the development and implementation of related products.

2.4. "Licensed Patent(s)" means (i) U.S. provisional application nos. [***] and any future applications that claim priority to any of the foregoing provisional applications, (ii) all divisions, substitutions and continuations of any of the preceding, (iii) all foreign patent applications corresponding to or claiming, priority from any of the preceding, and (iv) all U.S. and foreign patents issuing on any of the preceding, including patents of addition, reexaminations, reissues and extensions.

2.5. "Monogenetic Disorders" means diseases caused by a mutation in a single gene. For the avoidance of doubt, this term does not include Alzheimer's Disease or any form of cancer.

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

2.6. "Net Sales" means the gross revenue received by LICENSEE and/or its Affiliates or Sublicensees from sales of Products to third parties, less the following items but only insofar as they are included in such gross revenue: (i) import, export, value added, excise and sales taxes, tariffs, and custom duties; (ii) rebates, refunds, and credits for any rejected or returned Products or because of retroactive price reductions, rebates or chargebacks; (iii) charges for packaging, shipping and insurance; and (iv) trade, cash or quantity discounts or rebates to the extent actually granted (including Medicaid and other government-mandated rebates).

2.7. "Orphan Drug Product" means a Product for a specific indication for the which the FDA has granted "Orphan-drug exclusive approval" as defined in 21 CFR Part 316 or which has been granted Orphan Drug exclusivity under similar regulations in the European Union or Japan.

2.8. "Product" means any PPCA protein product discovered, developed, manufactured and/or commercialized by or on behalf of LICENSEE or its Affiliates for which LICENSEE or its Affiliates incorporates and uses any Licensed Know-How in its regulatory filings to support its regulatory approval.

2.9. "Retained Field" means all uses of PPCA outside the Licensed Field.

2.10. "Technology" means the technology developed in the laboratory of Dr. Alessandra D'Azzo as of the Effective Date relating to the use of Protective Protein/Cathepsin A ("PPCA") protein to treat, prevent and/or diagnose galactosialidosis and other Monogenetic Disorders.

3. GRANT

3.1. INSTITUTION hereby grants and LICENSEE hereby accepts a worldwide, exclusive license, with the right to grant sublicense under multiple tiers, under the Licensed Know-How to make, have made, import, use, sell and offer for sale and otherwise commercialize and exploit Products in the Licensed Field, and practice any method, process or procedure within the Licensed Know-How and/or Licensed Patents in connection therewith.

3.2. Notwithstanding Section 3.1, INSTITUTION shall retain the nontransferable right to practice the Licensed Patents for its internal, academic, non-commercial research.

3.3. INSTITUTION hereby grants LICENSEE a right of first negotiation to obtain an exclusive license under the Licensed Patents, together with an exclusive license under the Licensed Know-How in the Retained Field. INSTITUTION shall promptly notify LICENSEE in writing when it is ready to negotiate such a license and in any case prior to INSTITUTION presenting any third party with the opportunity to obtain such a license under such Licensed Patents or Licensed Know-How. LICENSEE shall notify INSTITUTION within [***] days after receiving such notification as to whether it desires to obtain such right. In the event LICENSEE notifies INSTITUTION in writing of its desire to obtain such right, then INSTITUTION shall negotiate in good faith with LICENSEE on an exclusive basis for a period of [***] months to agree upon the terms and conditions under which LICENSEE shall obtain such right. In the event LICENSEE does not notify INSTITUTION of its desire within such [***] day period, or if LICENSEE so notifies INSTITUTION, but INSTITUTION and LICENSEE do not reach

agreement on such terms and conditions within such [***] months period despite good faith negotiations, then INSTITUTION shall have the right to negotiate with any third party with respect to such grant of right, provided that INSTITUTION shall not grant any third party such right on terms and conditions, taken as a whole, that are less favorable to INSTITUTION than those last offered by LICENSEE.

4. GOVERNMENT RIGHTS

The rights for which a license is granted hereunder shall be subject to all of the terms, conditions and limitations of Title 35 United States Code Sections 200 through 212 (the "Bayh-Dole Act") to the extent that such rights constitute a "subject invention" as that term is defined in the Bayh-Dole Act.

5. DILIGENCE. LICENSEE shall use commercially reasonable efforts to develop and commercialize at least one (1) Product, consistent with sound and reasonable business practices and judgments, provided that LICENSEE's interest in any other products designed to treat or prevent Galactosialidosis shall not be taken into account when considering the commercial reasonableness of LICENSEE's efforts with respect to Products. LICENSEE shall submit to INSTITUTION a written progress report within [***] days of January 1 each year after the Effective Date describing all activities conducted during the previous year to diligently develop and commercialize Products. Any efforts of LICENSEE's Affiliates and sublicensees shall be considered efforts of LICENSEE for the sole purpose of determining LICENSEE's compliance with its obligation under this Section 5.

6. PAYMENTS

6.1. LICENSEE agrees to pay to INSTITUTION a nonrefundable, license issue fee of Ten Thousand Dollars ($10,000.00) within [***] days of the Effective Date.

6.2. Subject to Section 6.4, LICENSEE shall pay to INSTITUTION royalties equal to [***] percent ([***]%) of Net Sales of Products sold by LICENSEE or its Affiliates or Sublicensees which are Orphan Drug Products, but only on a Product-by-Product and country-by-country basis for so long as, and only in the country where, such Product has market exclusivity as an Orphan Drug Product.

6.3. In the event that an Orphan Drug Product under this Agreement is sold in a combination product containing other active components, then Net Sales on the combination product shall be calculated using one of the following methods:

(a) By multiplying the net selling price of the combination product by the fraction A/(A+B) where A is the gross selling price, during the royalty-paying period being considered, of such Product sold separately, and B is the gross selling price, during the royalty period in question, of the other active components sold separately; or

(b) In the event that no such separate sales are made of such Product, Net Sales on the combination product for royalty determination shall be as reasonably allocated between such Product and the other active components, based on their relative importance and proprietary protection, as agreed by the parties. If the parties fail to reach agreement such allocation shall be submitted to binding arbitration.

6.4. No more than one royalty payment shall be due with respect to a sale of a particular Product. No royalty shall be payable under Section 6.2 with respect to sales of Products among LICENSEE and its Affiliates or Sublicensees or with respect to Products distributed without charge for use in research and/or development, in clinical trials or as promotional samples or otherwise distributed without charge to third parties.

6.5. For the purpose of determining amounts payable under this Agreement, any Net Sales denominated in currencies other than U.S. dollars shall be converted into U.S. dollars according to LICENSEE's reasonable standard internal conversion procedures, including LICENSEE's standard internal rates and conversion schedule. All payments to INSTITUTION shall be made in U.S. dollars.

7. REPORTS, PAYMENTS AND ACCOUNTING

7.1. Beginning with the first sale of an Orphan Drug Product, LICENSEE shall make written reports of royalty payments due, if any, to INSTITUTION within [***] days of [***] and [***]. This report shall state the number, description, and aggregate Net Sales of the applicable Product(s) received by LICENSEE during the previous completed calendar half year, and resulting calculations of royalty payments due INSTITUTION pursuant to Sections 6.2 through 6.5 for such completed calendar half year. Concurrent with the submission of each such report, LICENSEE shall pay INSTITUTION any royalties due for the [***] covered by such report.

7.2. LICENSEE agrees to keep and maintain records for a period of [***] years showing the sale, use and other disposition of Orphan Drug Products sold or otherwise disposed of under the license herein granted. Such records will include sufficient detail to enable the royalties payable hereunder by LICENSEE to be determined. LICENSEE further agrees to permit its books and records to be examined by an independent certified public accountant selected by INSTITUTION and acceptable to LICENSEE once per calendar year during the term of this Agreement, for the sole purpose of verifying the reports and payments made by LICENSEE. Such examination shall be made at LICENSEE'S place of business during ordinary business hours with at least [***] days prior written notice. Such examination is to be at the expense of INSTITUTION except in the event that the results of the audit reveal an under reporting of payments due INSTITUTION of [***] percent ([***]%) or more, then the audit costs shall be paid by LICENSEE within [***] days of notice by INSTITUTION to LICENSEE.

8. NEGATION OF WARRANTIES

8.1. Nothing in this Agreement is or shall be construed as:

(a) A warranty or representation by INSTITUTION as to the validity or scope of any Licensed Patent(s);

(b) A warranty or representation that anything made, used, sold, or otherwise disposed of under any license granted in this Agreement is or will be free from infringement of patents, copyrights, and other rights of third parties;

(c) An obligation to bring or prosecute actions or suits against third parties for infringement except to the extent and in the circumstances described in Article 13;

(d) Granting by implication, estoppel, or otherwise any licenses or rights under patents or other rights of INSTITUTION or other persons other than to the Licensed Patent(s), regardless of whether such patents or other rights are dominant or subordinate to any Licensed Patent(s); or

(e) An obligation to furnish any technology or technological information, except as expressly set forth in this Agreement.

8.2. Except as expressly set forth in this Agreement, INSTITUTION MAKES NO REPRESENTATIONS AND EXTENDS NO WARRANTIES OF ANY KIND, EITHER EXPRESS OR IMPLIED. THERE ARE NO EXPRESS OR IMPLIED WARRANTIES OF MERCHANTABILITY OR FITNESS FOR A PARTICULAR PURPOSE.

9. REPRESENTATIONS AND WARRANTIES

9.1. INSTITUTION represents and warrants that it owns all right, title and interest in and to the Technology, Licensed Know-How and Licensed Patents, subject to the license set forth in Section 3.1.

9.2. INSTITUTION represents and warrants that it has not granted any third party right or interest in any of the Technology, Licensed Know-How or Licensed Patents that is inconsistent with the rights granted to LICENSEE herein and will not grant any third party such a right during the term of this Agreement.

9.3. INSTITUTION represents and warrants that it has the power to enter into this Agreement and the right to grant the rights granted herein to LICENSEE.

9.4. LICENSEE represents and warrants that it has the power to enter into this Agreement and meet its obligations under this Agreement.

9.5. INSTITUTION represents and warrants that Licensed Patents do not claim the use of Products in the Licensed Field.

10. INDEMNITY

10.1. LICENSEE agrees to indemnify, hold harmless, and defend INSTITUTION and its respective trustees, officers, employees, students, and agents (the “Indemnitees”) against any and all liability, damage, loss or expense incurred by or imposed on the Indemnitees or any one of them, arising out of third party claims arising out of the manufacture, use, sale, or other disposition of Product(s) by LICENSEE or its Affiliates or Sublicensee(s), or the exercise of the license granted herein, except to the extent arising out of the gross negligence or willful misconduct of any Indemnitee.

10.2. NEITHER PARTY SHALL BE LIABLE TO THE OTHER PARTY FOR ANY INDIRECT, SPECIAL, CONSEQUENTIAL, PUNITIVE OR OTHER DAMAGES WHATSOEVER, WHETHER GROUNDED IN TORT (INCLUDING NEGLIGENCE), STRICT LIABILITY, CONTRACT OR OTHERWISE.

11. INSTITUTION NAMES AND MARKS

11.1. LICENSEE agrees not to identify INSTITUTION or use the name of any INSTITUTION faculty member, employee, or student or any trademark, service mark, trade name, or symbol of INSTITUTION or that is associated with INSTITUTION in any promotional advertising or other promotional materials to be disseminated to the public without INSTITUTION's prior written consent, which consent shall not be unreasonably withheld. INSTITUTION and LICENSEE agree that reports in scientific literature and presentations of research and development work are not considered promotional materials. Promotional materials shall also not include disclosures required under any laws or government regulations or by the rules of any stock exchange of any country.

11.2. Notwithstanding Section 11.1, LICENSEE may publicly disclose, on its website or otherwise, that it has obtained from INSTITUTION an exclusive license under the Licensed Know-How.

12. TERM AND TERMINATION

12.1. This Agreement shall be effective as of the Effective Date and, unless earlier terminated in accordance with Sections 12.2 or 12.3, shall expire upon expiration of the last payment obligation of LICENSEE under Section 6.2. After the expiration of this Agreement, the license granted to LICENSEE hereunder shall become fully-paid, royalty-free, perpetual and irrevocable.

12.2. LICENSEE may terminate this Agreement as a whole or solely with respect to any country by giving INSTITUTION notice in writing at least thirty (30) days in advance of the effective date of termination selected by LICENSEE.

12.3. INSTITUTION may terminate this Agreement if LICENSEE is in material breach of any provision hereof and LICENSEE fails to remedy any such breach within sixty (60) days after receipt of written notice thereof by INSTITUTION. Upon any such termination, (i) LICENSEE and its Affiliates shall have six (6) months to complete the manufacture of any Products that then are work in progress and to sell their inventory of Products, provided LICENSEE pays the applicable royalties in accordance with Section 6.2, and (ii) INSTITUTION shall accept an assignment by LICENSEE of any sublicenses granted by LICENSEE to third parties, and any sublicense so assigned shall remain in full force and effect.

12.4. Surviving any termination are:

(a) LICENSEE's obligation to pay royalties accrued prior to termination;

(b) Any cause of action or claim of LICENSEE or INSTITUTION, accrued, because of any breach or default by the other party; and

(c) The provisions of Articles 7, 8, 10, 14 and 16.

13. ASSIGNMENT

Neither party may assign this Agreement or any part hereof without the express written consent of the other, which consent shall not be unreasonably withheld; provided, however, LICENSEE may assign this Agreement or any portion hereof to an Affiliate or to a successor of all or substantially all its assets, stock or business relating hereto without the written consent of INSTITUTION and shall provide INSTITUTION notice of any such assignment.

14. ARBITRATION

14.1. Any controversy arising under or related to this Agreement, and any disputed claim by either party against the other under this Agreement excluding any dispute relating to patent validity or infringement arising under this Agreement, shall be settled by arbitration in accordance with the JAMS rules.

14.2. Upon request by either party, arbitration will be initiated by a third party arbitrator mutually agreed upon in writing by LICENSEE and INSTITUTION within [***] days of such arbitration request. Judgment upon the award rendered by the arbitrator shall be final and nonappealable and may be entered in a court having jurisdiction thereof. The parties agree that any provision of applicable law notwithstanding, they will not request and the arbitrators shall have no authority to award punitive or exemplary damages against any party. The costs of the arbitration, including administrative fees and fees of the arbitrators shall be shared equally by the parties. Each party shall bear the cost of its own attorneys' fees and expert fees.

14.3. Any arbitration shall be held at a location mutually agreed upon by the parties.

14.4. The parties shall maintain the confidential nature of the arbitration proceeding and the Award, including the Hearing, except as may be necessary to prepare for or conduct the arbitration hearing on the merits, or except as may be necessary in connection with a court application for a preliminary remedy, a judicial challenge to an Award or its enforcement, or unless otherwise required by law or judicial decision.

15. NOTICES

All notices under this Agreement shall be deemed to have been fully given when done in writing and deposited in the United States mail, registered or certified, or overnight deliver service (e.g., DHL, Federal Express) and addressed as follows:

To INSTITUTION:

St. Jude Children's Research Hospital
262 Danny Thomas Place
Memphis, TN 38105
Attention: Technology Licensing Director

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

To LICENSEE:	Ultragenyx Pharmaceutical Inc., 60 Leveroni Court, Novato, CA 94949; Attention: Chief Executive Officer

Either party may change its address upon written notice to the other party.

16. CONFIDENTIALITY

INSTITUTION shall maintain this Agreement and the reports and any information provided by LICENSEE to INSTITUTION pursuant to Sections 5 and 7 in confidence and not disclose such information or reports to any third party, except as required by law and disclosed after notice, to LICENSEE and after requesting confidential treatment and a protective order, if available.

17. WAIVER

None of the terms of this Agreement can be waived except by the written consent of the party waiving compliance.

18. APPLICABLE LAW

This Agreement shall be governed by the laws of the State of New York, without reference to principles of conflicts of laws.

19. ENTIRE AGREEMENT

This Agreement constitutes the entire Agreement between LICENSEE and INSTITUTION and supersedes all prior communications, understandings and agreements with respect to the subject matter of this Agreement. This Agreement may not be amended except with a written agreement signed by LICENSEE and INSTITUTION.

IN WITNESS WHEREOF the parties have executed this Agreement effective as of the Effective Date Set forth above.

Ultragenyz Pharmaceutical, Inc.	St. Jude Children's Research Hospital
By: /s/ Tom Kassberg	By: /s/ J. Scott Elmer
Name: Tom Kassberg	Name: J. Scott Elmer
Title: CBO	Title: Director, Office of Technology Licensing

Exhibit 10.8

EXCLUSIVE LICENSE AGREEMENT

This License Agreement (the "**Agreement**") is made and entered into as of November 22, 2010, (the "**Effective Date**") by and between Saint Louis University ("**SLU**"), having an address at 3700 West Pine Mall, Fusz Memorial Hall, Second Floor, Saint Louis, Missouri 63108, and Ultragenyx Pharmaceutical Inc. ("**LICENSEE**"), having an address at 77 Digital Drive, Suite 210, Novato, California 94949.

WHEREAS, SLU is the owner by assignment of certain Patent Rights and Technology, as defined below, which SLU owns or controls, and the SLU has the right to grant licenses under such Patent Rights and Technology;

WHEREAS, SLU desires to have the Technology utilized in the public interest;

WHEREAS, LICENSEE has represented to SLU, to induce SLU to enter into this Agreement, that LICENSEE is engaged in the research and development of Beta-glucuronidase for commercial purposes.

NOW, THEREFORE, in consideration of the above premises and the mutual covenants contained herein, the parties hereby agree as follows:

1. DEFINITIONS

1.1. The term "**Affiliate**" means any person or entity which directly or indirectly owns or controls LICENSEE, or which is controlled by or under common control with LICENSEE. For purposes of this definition, "control" means the ownership by LICENSEE, directly or indirectly, of fifty percent (50%) or more of the outstanding equity securities of a corporation entitled to vote in the election of directors or the direct or indirect ownership by a person or entity of fifty percent (50%) or more of the outstanding equity securities of LICENSEE entitled to vote in the election of LICENSEE'S directors.

1.2. The term "**Confidential Information**" shall mean all ideas and information of any kind that are held in confidence by one Party and transferred, disclosed or made available by such Party to a receiving Party and are identified at the time of disclosure as being proprietary or confidential. The obligations in this Agreement with respect to Confidential Information shall not apply to any portion of the Confidential Information that the receiving Party can demonstrate by legally sufficient evidence (i) now or hereafter, through no act or failure to act on the part of the receiving Party, is or becomes public; (ii) is known to the receiving Party or one of its

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

Affiliates at the time such Party receives such Confidential Information from the disclosing Party; (iii) is hereafter furnished to the receiving Party by an unrelated Third Party without violating any agreement with the disclosing Party; or (iv) is independently developed by the receiving Party or one of its Affiliates without use of any Confidential Information received from the other Party. LICENSEE, prior to disclosure of information believed by LICENSEE to fall into parts (i)-(iv) of this Section, shall provide written notice to SLU with sufficient time so that SLU can make an independent assessment as to whether the information is or is not Confidential Information.

1.3. The term "**Fair Market Value**" mean the cash consideration which LICENSEE or its Sublicensee would realize from an unaffiliated, unrelated buyer in an arm's length sale of an identical item sold in the same quantity and at the same time and place of the transaction.

1.4. The term "**FDA**" means the United States Food and Drug Administration.

1.5. The term "**Field of Use**" means use of the beta-glucuronidase product candidate as a treatment for human disease.

1.6. The term "**First Commercial Sale**" means the first invoiced sale of a Licensed Product to a Third Party by LICENSEE following the receipt of any Regulatory Approval required for the sale of such Licensed Product.

1.7. The term "**Licensed Product**" means any service, product, process, or part thereof the manufacture, use, importation, sale or offer for sale of which in the absence of the license granted hereunder, would infringe a Valid Claim or during the period where any product has a license to Technology granted hereunder has market exclusivity as an Orphan Drug.

1.8. The term "**Net Sales**" means the gross sales of the Licensed Product invoiced by LICENSEE, its Affiliates or Sublicensees to a Third Party, less the following items:

(a) reasonable freight, packaging, shipping, storage, and insurance costs, if separately stated;

(b) amounts repaid or credited by reason of rejections, defects or returns or because of retroactive price reductions;

(c) rebates (price reductions, rebates to social and welfare systems, charge backs or reserves for chargebacks, cash sales incentives, government mandated rebates and similar types of rebates, e.g., Pharmaceutical Price Regulation Scheme and Medicaid and discounts for quantity purchases, cash payments, and for wholesalers and distributors).

No deductions shall be made for commissions paid to individuals whether they are with independent sales agencies or regularly employed by LICENSEE or Sublicensees and on its payroll, or for cost of collections.

1.9. The term "**Non-Commercial Research Purposes**" shall mean use of Patent Rights and Technology for academic research or other not-for-profit scholarly purposes which are undertaken at a non-profit or governmental institution that does not use the Patent Rights and Technology in the production or manufacture of products for sale or the performance of services for a fee.

1.10. The term "**Orphan Drug**" shall mean a Licensed Product for a specific indication for which the FDA has granted "Orphan-drug exclusive approval" as defined in 21 CFR Paid 316 or which be granted Orphan Drug exclusivity under similar regulations in the European Union or Japan.

1.11. The term "**Patent Costs**" means out-of-pocket expenses incurred by SLU in connection with the preparation, filing, prosecution, maintenance, and interference proceedings of patent applications and patents, including the fees and expenses of attorneys and patent agents, foreign and domestic, filing fees, maintenance fees, and annuity fees, but excluding costs associated with any patent infringement actions.

1.12. The term "**Patent Rights**" means the patent applications and patents listed in Exhibit A, including all related international filings, reissues, reexaminations, divisions, or continuations thereof and patent applications and any inventions, whether patented or not, which SLU owns or controls throughout the world, covering or related to the beta-glucuronidase product candidate**.**

1.13. The term "**Party**" means an individual, partnership, limited partnership, joint venture, trustee, trust, corporation, company, unlimited or limited liability company, unincorporated organization or other entity or a government, state or agency or political subdivision thereof.

1.14. "**Parties**" means SLU and LICENSEE.

1.15. The term "**Regulatory Approval**" means any approvals (including pricing and reimbursement approvals), licenses, registrations or authorizations of any national or international or local regulatory agency, department, bureau or other governmental entity, necessary for the manufacture and/or sale of a Licensed Product in a regulatory jurisdiction within the Territory.

1.16. The term "**Sale Transaction**" means the merger of LICENSEE with and into a Third Party, or the sale to, or other acquisition by, a Third Party, of all or substantially all of the assets of the business of LICENSEE in which the Patent Rights and Technology forms part, or all or substantially all, of the equity interests in LICENSEE, but excluding, for greater certainty, the sublicensing by LICENSEE to a Third Party of any of the rights granted to LICENSEE hereunder.

1.17. The term "**Sublicensee**" means any Party to which LICENSEE has sublicensed the rights granted to it under Section 2 below and in accordance with the terms thereof, but the term "Sublicensee" shall not include: (i) any Affiliate of LICENSEE or (ii) any Party who manufactures a Licensed Product for LICENSEE or an Affiliate of LICENSEE but does not sell such Licensed Products other than to LICENSEE or an Affiliate of LICENSEE.

1.18. The term "**Sublicensing Revenue**" means all upfront, milestone and royalty payments and other consideration received by LICENSEE from a Sublicensee in consideration for the sublicensing by LICENSEE of all or part of the rights granted to it under Section 3 below and in accordance with the terms thereof, except for direct reimbursement of research expenditures actually incurred and excluding any research and development grants. Any non-cash consideration so received by LICENSEE from Sublicensees shall be valued at its Fair Market Value as of the date of receipt.

1.19. The term "**Technology**" means scientific information, knowledge and know-how and materials related to SLU's beta-glucuronidase product candidate for the treatment of [***].

1.20. The term "**Term**" has the meaning ascribed to it in Section 11.1.

1.21. The term "**Territory**" means World Wide.

1.22. The term "**Third Party**" means any Party other than LICENSEE, its Affiliates, Distributors, Sub-Distributors, or SLU.

1.23. The term "**Valid Claim**" means a claim in any unexpired patent or patent application listed in Exhibit A or contained in the Patent Rights that has not been disclaimed, revoked or held invalid or unenforceable by a final unappealable decision of a court or government agency of competent jurisdiction.

2. GRANT OF RIGHTS

2.1. License to Patent Rights and Technology. Subject to the terms and conditions hereof, SLU hereby grants to LICENSEE, who accepts, an exclusive license under the Patent Rights and the Technology in the Field of Use in the Territory, to develop, have developed, to make, have made, to use, have used, to import, have imported, to offer for sale, sell and have sold any Licensed Product. The license granted under this section shall remain in effect throughout the Term and shall, subject to Section 2, be sublicensable by LICENSEE and/or its Sublicensees through multiple tiers.

2.2. Affiliates and Distributors. LICENSEE may exercise its rights and delegate its obligations under this Agreement through and to its Affiliates, distributors and sub-distributors through multiple tiers. Such exercise shall not constitute a sublicense of the rights granted to it hereunder.

2.3. Retained Rights. The exclusivity of the above license is subject to the retained rights of SLU defined by:

(a) make, use and further develop the Patent Rights and Technology for its own educational, research and patient care purposes;

(b) grant to others non-exclusive licenses to make and use for Non-Commercial Research Purposes the subject matter described and claimed in Patent Rights and Technology;

(c) grant licenses to Third Parties outside the Field of Use expressly granted herein.

2.4. Federal Government Rights Reserved. Notwithstanding the exclusive license granted herein, the Federal Government shall receive all the rights to the Patent Rights and Technology as required by law or regulation to be reserved to the government. The Parties agree that the Federal Government is hereby granted a non-exclusive, non-transferable, irrevocable, royalty free license to practice or have practiced on its behalf throughout the world the Patent Rights and Technology. All rights granted in this Agreement are expressly granted subject to the rights of the Federal Government and such rights are specifically reserved to the Federal Government by this Agreement.

2.5. Consulting Agreements. In the event LICENSEE desires to have a consulting agreement with any SLU faculty member(s), any such consulting agreement will be separate and apart from this Agreement, and in accord with SLU policy and procedures.

2.6. Sponsored Research Agreements. In the event LICENSEE desires to fund research at SLU, such research agreements shall be separate and apart from this Agreement, and in accord with SLU policy and procedures.

3. SUBLICENSING

3.1. The right of the LICENSEE to sublicense, in whole or in part, the rights and obligations contained in this Agreement is prohibited without prior written consent from SLU, Should SLU grant to LICENSEE in writing the right to sublicense, in whole or in part the rights and obligations contained in this Agreement, any sublicense will be subject to the following:

(a) LICENSEE shall execute a written sublicense with each Sublicensee which shall be subject to LICENSEE’s rights and obligations under the terms of this Agreement. LICENSEE shall cause any such sublicense

agreement to contain terms that are at least as protective of the Patent Rights and the Technology and Confidential Information of SLU as the terms set forth in this Agreement, and that also include no provisions that would be in violation of the license grant set forth in this Agreement. Any such sublicense agreement shall further: (i) prohibit Sublicensee's further sublicense of the rights delivered hereunder; (ii) name SLU as an intended third party beneficiary of the obligations of Sublicensee without imposition of obligation or liability on the part of SLU to the Sublicensee; and (iii) bear signature from SLU indicating SLU's review and approval of the sublicense agreement. To the extent that any terms, conditions or limitations of any sublicense agreement are inconsistent with this Agreement, those terms, conditions and limitations are null and void against SLU, even through SLU has approved the sublicense in writing.

(b) LICENSEE shall be solely responsible for the enforcement of the terms of any sublicense and for collection of payment due thereunder.

(c) SLU shall be entitled to request a separate accounting from LICENSEE of any Sublicensing Revenue on a yearly or less frequent basis.

(d) Upon termination of this Agreement as provided hereunder, SLU will stand in the place of LICENSEE with respect to the Sublicensee for a period of [***] days during which time SLU will negotiate with Sublicensee in good faith and under reasonable terms and conditions to execute a new license between Sublicensee and SLU. If no new license is completed within the [***] day period, the sublicense will terminate.

4. PAYMENTS

4.1. License Issue Fee. LICENSEE shall pay to SLU a one-time, sign-on fee in the amount of Ten Thousand United States Dollars (USD $10,000.00) (the "License Issue Fee"), payable within [***] days after the Effective Date.

4.2. Milestone Payments. LICENSEE shall provide SLU with written notice within [***] days of its achievement of each of the milestone events set forth below. Within [***] days after delivering each such notice, LICENSEE shall pay to SLU the amounts set forth below:

Milestone	Amount
Approval of a glucuronidase-based enzyme therapy for treatment for MPS VII	$100,000

4.3. Royalty Payments. Subject to section 9.4, beginning after the first $[***] of cumulative worldwide sales of the product and during the Term. LICENSEE shall pay to SLU a royalty of:

4.3.1 [***] percent ([***]%) on Net Sales in any country or region, other than the United States, Japan and the European Union, where there is a Valid Claim covering a Licensed Product.

4.3.2 [***] ([***]%) on Net Sales in the United States, Japan and the European Union where there is a Valid Claim covering a Licensed Product or during the period where the Licensed Product has market exclusivity as an Orphan Drug.

Royalty payments shall be made in accordance with Section 4.5.

4.4. Additional expenses. Each party shall bear its own legal and professional expenses in connection with the negotiation and execution of this license agreement.

4.5. Reporting and Payment Terms. Commencing on the First Commercial Sale, LICENSEE agrees to deliver to SLU, within [***] days after each [***]-month period, CFO (or equivalent) signed payment of royalties owed and a report showing the information on which provided payments are calculated, including a breakdown of Net Sales of each Licensed Product on a country-by-country basis and to accompany each such report with the payment shown to be due thereby.

4.6. Currency Conversion. If any currency conversion is required in connection with any payments to SLU hereunder, Net Sales shall first be calculated in the relevant foreign currency and then converted to U.S. Dollars against the currency in question on the rate of exchange applicable using the currency exchange rates quoted by *Bloomberg Professional*, a

service of Bloomberg L.P., during the period of such Net Sales, or in the event *Bloomberg Professional* is not available, then *International Financial Statistics* (publisher, International Monetary Fund) during the royalty period of such Net Sales, for the currency of the country in which the sale is made at the average rate of exchange during the royalty period of such Net Sale

4.7. Royalty Stacking. If LICENSEE pays if obliged to pay royalties to a Third Party for patents necessary to the manufacture, use or sale of a Licensed Product(s), then LICENSEE may credit [***] percent ([***]%) of the Third Party royalties paid against royalties otherwise due to SLU for the Net Sales; provided, however that the royalties paid to SLU for the Net Sales shall not be less than [***] percent ([***]%) of those otherwise due. Such reduction of royalties allowed hereunder shall apply on an annual basis with no carryover of Third Party royalty balance from one calendar year to the following calendar year.

4.8. Development Assistance. SLU shall provide LICENSEE with all information related to the Patent Rights and Technology as may be known or possessed by SLU and may be reasonably necessary for LICENSEE to exploit the license granted in this agreement. SLU shall provide LICENSEE with reasonable technical assistance in connection with such transfer of the information related to the Patent Rights and Technology. Technical assistance after completion of the license is not to exceed [***] business hours of work, and LICENSEE shall pay SLU for any necessary technical assistance at a nominal rate of $[***] per hour, plus approved expenses. These expenses may include a single trip for two people to LICENSEE or an agent of LICENSEE to assist in technology transfer of the production process.

5. BEST EFFORTS

LICENSEE shall use its best efforts to (a) develop a Licensed Product and bring the Licensed Product to Market as soon as practicable, consistent with sound and reasonable business practices and judgment; and (b) LICENSEE shall market, promote, manufacture and sell the Licensed Product, whether it be itself or through Sublicensees, throughout the term of this Agreement. LICENSEE's failure to perform in accordance with this Section shall be grounds for SLU to terminate or render this license non-exclusive. In making this determination, SLU shall take into account the normal course of such programs conducted with sound and reasonable business practices and judgment and shall take into account the reports provided hereunder by LICENSEE.

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

6. INTELLECTUAL PROPERTY

6.1. Each Party shall own and retain all right, title and interest in all intellectual property that: (a) was created, discovered, developed or conceived and reduced to practice by such Party prior to the Effective Date; or (b) was created, discovered, developed or conceived and reduced to practice by such Party after the Effective Date but is unrelated to the subject matter of this Agreement.

6.2. LICENSEE (for itself and Sublicensees) acknowledges and agrees that SLU is and shall remain (as to LICENSEE) the owner of the Patent Rights and the Technology, and that LICENSEE (including its Affiliates and Sublicensees) has no rights in or to the Patent Rights or the Technology except as provided hereunder.

6.3. As between SLU and LICENSEE, any invention that is solely conceived under this Agreement by SLU, including any improvements and enhancements related to the Patent Rights or the Technology, whether patentable or not, shall be owned by SLU, including all intellectual property rights therein. SLU shall license any such improvements (to the extent it is contractually permitted to do so), to LICENSEE under the terms of this Agreement and free of any additional royalty expense. In the event SLU is not contractually permitted to Licenses any such Improvements to LICENSEE, SLU shall offer LICENSEE an option to obtain exclusive license rights to any improvements developed by SLU individually or jointly with others (to the extent contractually permitted to do so). The parties will negotiate in good faith for a period of time not to exceed [***] months to establish the terms of a separate license agreement related to any improvements and enhancements related to the Patent Rights or the Technology. In addition, any invention that is solely conceived under this Agreement by LICENSEE or its Affiliates Sublicensees, including any improvements and enhancements related to the Patent Rights or the Technology, whether patentable or not, shall be owned by LICENSEE, including all intellectual property rights therein.

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

7. DISCLAIMERS

7.1. SLU Representations. SLU represents and warrants that:

(a) it has full authority to enter into this Agreement;

(b) it is not a Party to or bound by any contract or any other obligation whatsoever that limits or impairs its ability to enter into this Agreement;

(c) it has and shall continue during the Term to have the full right and legal capacity to grant the rights granted to LICENSEE hereunder;

(d) it is not aware of any information or fact that would render any of the Patent Rights invalid or unenforceable and that it has exclusive title to and ownership of all Patent Rights and Technology;

(e) neither SLU nor any of its faculty, professors, students, staff or employees received funding from any Third Party to create, discover, develop or conceive and reduce to practice the Patent Rights and/or the Technology, which funding would entitle Third Party to any rights whatsoever to the results of any such funding; and

(f) the patents and patent applications listed in Exhibit A constitute all of the Patent Rights owned or Controlled by SLU in connection with the Technology (as used in this Section, “Controlled” means the ability of SLU to grant a license or sublicense, other than as a result of this Agreement and without violating the terms of any agreement or other arrangement with any Third Party).

7.2. LICENSEE Representations. LICENSEE hereby represents and warrants that:

(a) it has full authority to enter into this Agreement; and

(b) it is not a Party to or bound by any contract or any other obligation whatsoever that limits or impairs its ability to enter into this Agreement or to perform its obligations under this Agreement.

7.3. Warranty Disclaimer. SLU MAKES NO REPRESENTATIONS AND EXTENDS NO WARRANTIES OF ANY KIND, EITHER EXPRESSES OR IMPLIED. Nothing in this Agreement is or shall be construed as:

(a) a warranty or representation by SLU as to the validity or scope of any Patent Rights, issued or pending;

(b) a warranty or representation that anything made, used, sold or otherwise disposed of under any license granted in this Agreement is or will be free from infringement of patents, copyrights and other rights of Third Parties;

(c) a warranty of merchantability or fitness for a particular purpose;

(d) an obligation to bring or prosecute actions or suits against Third Parties for infringement, except to the extent and in the circumstances described in Section 9.3;

(e) a grant by implication, estoppel, or otherwise of any licenses under patent applications or patents of SLU or Third Parties other than as provided in Section 2 hereof; or

(f) a warranty of the usefulness, accuracy or safety of the Licensed Products.

7.4. Additional Disclaimer of Liability. SLU ADDITIONALLY DISCLAIMS ALL OBLIGATIONS AND LIABILITIES ON THE PART OF SLU FOR INDIRECT, PUNITIVE OR CONSEQUENTIAL DAMAGES, ATTORNEYS' FEES, EXPERTS' FEES AND COURT COSTS (EVEN IF SLU HAS BEEN ADVISED OF THE POSSIBILITY OF SUCH DAMAGES, FEES, OR COSTS) ARISING OUT OF OR IN CONNECTION WITH THE MANUFACTURE, USE, OR SALE OF THE PATENT RIGHTS AND TECHNOLOGIES LICENSED UNDER THIS AGREEMENT. LICENSEE, AFFILIATES AND SUBLICENSEE(S) ASSUME ALL RESONSIBILITY AND LIABILITY FOR LOSS OR DAMAGE CAUSED BY A PRODUCT MANUFACTURED, USED, OR SOLD BY LICENSEE, AFFILIATES AND SUBLICENSEE(S) WHICH IS A LICENSED PRODUCT(S) AS DEFINDED IN THIS AGREEMENT.

8. INDEMNIFICATION AND INSURANCE

8.1. Indemnification by LICENSEE. LICENSEE agrees to indemnify, hold harmless and defend SLU, its trustees, officers, employees and agents, against any and all liability and/or damages with respect to any claims, suits, demands, judgments or causes of action (collectively "SLU Losses") arising out of (a) the development, manufacture, storage, sale or other distribution, or any other use of Licensed Products or Patent Rights, or exercise of rights granted hereunder, by LICENSEE or Sublicensees, distributors, agents or representatives; and/or (b) the use by end-users and other Third Parties of Licensed Products. In the event any such claims, demands or actions are made, LICENSEE shall defend SLU at LICENSEE's sole expense by counsel selected by LICENSEE. No settlement, consent judgment or other voluntary final disposition may be entered into without the prior written consent of SLU, which consent shall not be unreasonably withheld.

8.2. Insurance. During the Term of this Agreement, LICENSEE shall procure and maintain occurrence-based comprehensive general liability insurance in amounts not less than $[***] per claim and $[***] annual aggregate and name SLU, its trustees, officers, employees and agents as an additional insured. Such comprehensive general liability insurance shall provide (i) [***] and (ii) [***]. If LICENSEE elects to self-insure all or part of the limits described above, such self-insurance program must be reasonably acceptable to SLU. LICENSEE agrees that no amount greater than the sum of $[***] shall be deductible under LICENSEE's primary coverage for SLU and LICENSEE against any claims or suits arising from alleged defects in Licensed Products. The minimum amounts of insurance coverage required shall not be construed to create or limit LICENSEE's liability with respect to its indemnification under this Agreement.

At the time as any Licensed Product is first used in commerce or in humans, LICENSEE shall provide SLU with a certificate or certificates of insurance evidencing that SLU has been named as an additional insured Party, along with its trustees, officers, employees and agents, and evidencing that the insurer(s) is/are required to notify SLU in writing at least [***] days in advance of any termination of the policy or certificate, or any modification that would cause LICENSEE no longer to be in compliance with the provisions of this Section, or would cause the representation and warranties set forth above in this Section no longer to be true, such

written notification to specify the reason for such termination, the nature of the proposed modification, as the case may be. It is expressly agreed by the Parties that the provisions of this Section regarding insurance shall in no way limit LICENSEE's indemnity obligation, except to the extent that LICENSEE's insurer(s) actually pays SLU amounts for which SLU is entitled to be indemnified under this Agreement, nor shall SLU have any obligation to pursue any insurer as a precondition to its rights to be indemnified by LICENSEE. If LICENSEE does not obtain replacement insurance within such [***] day period specified above, SLU shall have the right to terminate this Agreement effective at the end of such [***] day period without notice or any additional waiting periods.

9. PROSECUTION, MAINTENANCE AND ENFORCEMENT OF PATENT RIGHTS

9.1. Prosecution and Maintenance.

(a) SLU shall have full control over filing, prosecution and maintenance of the patent applications and patents contained in the Patent Rights. SLU will keep LICENSEE advised of the status of patent prosecution by promptly providing LICENSEE with copies of official communications about the patent applications and patents contained in the Patent Rights in sufficient time to allow for review and comment by LICENSEE. LICENSEE shall have reasonable opportunities to advise and cooperate with SLU in such filing, prosecution and maintenance.

9.2. Patent Costs.

(a) All Patent Costs shall be the responsibility of LICENSEE, whether incurred prior to or after the Effective Date of this Agreement. LICENSEE shall reimburse SLU for such Patent Costs incurred by and billed to the SLU prior to the Effective Date of this Agreement will be payable to SLU within 30 days after the Effective Date of this Agreement. Patent Costs incurred after the Effective Date of this Agreement, or Patent Costs incurred before, but billed to SLU after the Effective Date of this Agreement, shall be paid by LICENSEE within [***] days of the receipt of an invoice from SLU. Payments pursuant to this Section 9.2(a) are not creditable against royalties.

(b) LICENSEE may elect to surrender a portion or all of its license rights under the Patent Rights in any country by providing to SLU written notice of such intent at least [***] days contained in the Patent Rights, as set forth at Section 9.1, upon the expiration of the [***] day notice period (or such longer period specified in LICENSEE's notice). In the event LICENSEE elects to surrender any or all of license rights under the Patent Rights, such applicable patent(s) and/or patent application(s) shall be excluded from the definition of "Patent Rights" and from the scope of the license granted under this Agreement, and all rights relating thereto shall revert to SLU, and may thereafter be freely licensed by SLU to Third Parties.

9.3. Enforcement of Patent Rights.

(a) In the event LICENSEE or SLU becomes aware of any actual or potential infringement of any Patent Rights, that Party shall promptly notify the other and the Parties shall discuss the most appropriate action to take. SLU and LICENSEE will cooperate with each other to attempt to terminate such infringement without litigation.

(b) If attempts to abate such infringement are unsuccessful, LICENSEE shall consult with SLU and shall consider the views of SLU regarding the advisability of the proposed action and its effect on the public interest. LICENSEE may (without being required to) bring an action at its own expense, in which event SLU shall cooperate with LICENSEE as reasonably requested, at LICENSEE's expense. No settlement, consent judgment or other voluntary final disposition of the action may be entered into without the prior written consent of SLU, which consent shall not be unreasonably withheld. To the extent LICENSEE's recoveries from such infringement action exceed LICENSEE's expenses, LICENSEE agrees to pay SLU [***] percent ([***]%) of such excess recoveries; provided that any of LICENSEE's expenses not recovered shall be credited towards any future royalties, annual maintenance fees, milestone payments, the Minimum Annual Revenue or any other payments, as provided in Section 3. For greater certainty, such of LICENSEE's expenses in excess of recoveries shall not be credited towards any future Patent Costs, as provided in Section 9.2.

(c) If required by law, SLU shall permit any action under this Section to be brought in its name, including being joined as a Party-plaintiff, provided that LICENSEE shall hold SLU harmless from, and indemnify SLU against, any costs, expenses, or liability that SLU incurs in connection with such action.

(d) In the event that LICENSEE elects not to institute or prosecute any suit to enjoin or recover damages from any infringer, then SLU alone may, in its sole discretion and at its expense, initiate and conduct an infringement action and any settlement or award which may be obtained shall solely belong to SLU.

9.4. Third Party Infringement Claims.

(a) In the event any Licensed Product becomes the subject of a claim for patent or other proprietary right infringement anywhere in the world by virtue of the incorporation of the Patents Rights therein (a "Third Party Infringement Claim"), the Parties shall promptly give notice to the other and meet to consider the Third Party Infringement Claim and the appropriate course of action and LICENSEE shall remit any payments due to SLU under this Agreement in escrow from the date of such Third Party Infringement Claim until final resolution of such claim (the "Period"). LICENSEE shall have the right, but not the obligation, to conduct the defense at its own expense of any such Third Party Infringement Claim brought against LICENSEE and/or SLU, in which event SLU shall cooperate with LICENSEE as reasonably requested, at LICENSEE's expense. In no event may a settlement, consent judgment or other voluntary final disposition of the Third Party Infringement Claim be entered into without the prior written consent of SLU, which consent shall not be unreasonably withheld.

(b) In the event a Third Party Infringement Claim is dismissed by a final unappealable decision, monies held in escrow pending such final unappealable decision ("the Monies") will be remitted to SLU as entire fulfillment of LICENSEE's financial obligations hereunder during the Period.

(c) In the event a Third Party Infringement Claim is upheld, whether by a Court or a settlement, an amount equal to the Monies minus [***] percent ([***]%) of the consideration actually paid to the Third Party with respect to such claim will be remitted to SLU as entire fulfillment of LICENSEE's obligations hereunder during the Period. The balance of the Monies will be remitted to LICENSEE. Thereafter, LICENSEE shall pay to SLU a royalty of [***] percent ([***]%) on Net Sales in the country where such Third Party Infringement Claim was brought.

10. REPORTING, VERIFICATION AND PAYMENT

10.1. Books and Records. LICENSEE agrees to keep proper records of scientific research and keep records of the latest [***] years of Net Sales of Licensed Products in accordance with generally accepted accounting practices. Such records shall include all information necessary for the accurate determination of royalty payments, Sublicensing Revenue and milestone achievement.

10.2. Audit. SLU, at its own expense, shall have the right, [***] per calendar year, and upon [***] days prior written notice, to have a certified public accountant, selected by SLU and reasonably acceptable to LICENSEE, inspect and audit in the location(s) where such records are maintained, the records of LICENSEE during usual business hours for the sole purpose of, and only to the extent necessary for, determining the correctness of payments due hereunder by LICENSEE, with all information disclosed being deemed Confidential Information of LICENSEE. Such audit shall be completed within [***] business days, subject to extension by the auditor if the auditor reasonably determines in good faith that data or information it requires is not available and identifies the data or information required. Results of such audit shall be made available to the Parties. If the audit reflects an underpayment of amounts due to SLU pursuant to this Agreement, such underpayment shall be promptly remitted to SLU by LICENSEE. If the underpayment is equal to or greater than [***] ([***]%) percent of the amount that was otherwise due, SLU shall be entitled to have LICENSEE pay the reasonable out-of-pocket costs incurred by SLU to retain such independent certified public accountant to conduct such review. Any underpayment is subject to the late payment fee of Section 10.5.

10.3. Foreign Payments. Royalties based on Net Sales in any foreign country shall be payable to SLU in United States Dollars. Dollar amounts shall be calculated using the foreign exchange rate, as published by the Wall Street Journal, in effect for such foreign currency on the last business day of each quarter for which a report is required. Where royalties are due for Net Sales in a country where, for reasons of currency, tax or other regulations, transfer of foreign currency out of such country is prohibited, LICENSEE has the right to place SLU's royalties in a

bank account in such country in the name of and under the sole control of SLU; provided, however, that the bank selected be reasonably acceptable to SLU and that LICENSEE inform SLU of the location, account number, amount and currency of money deposited therein. After SLU has been so notified, those monies shall be considered as royalties duly paid to SLU and will be completely controlled by SLU.

10.4. Taxes. Taxes imposed by any foreign or United States governmental agency on any payments to be made to SLU by LICENSEE hereunder shall be paid by LICENSEE without deduction from any payment due to SLU.

10.5. Late Payments. Late payments shall be subject to a charge of [***] percent ([***]%) per month compounded. LICENSEE shall indemnify SLU for all attorneys' fees and costs incurred by SLU in obtaining a full payment of late payments owed to SLU. The payment of such late charges shall not prevent SLU from exercising any other rights it may have as a consequence of the lateness of any payment.

11. TERM AND TERMINATION

11.1. Term. Unless earlier terminated under this Section 11.2, this Agreement shall become effective as of the date of this Agreement and expire the latest date of when the Orphan Drug exclusively expires in the United States, Japan or the European Union, or until the last patent based on technology licensed hereunder shall terminate, lapse or invalidation of the last remaining Valid Claim in the Territory, at which time the licenses granted to LICENSEE under this Agreement to snake, have made, to use, have used, to import, have imported, to offer for sale, sell and have sold any Licensed Product shall be fully paid-up. For greater certainty, SLU acknowledges and agrees that during the Term, no royalties shall be due by LICENSEE to SLU for Net Sales in countries where there is no Valid Claim Covering a Licensed Product in such countries.

11.2. Termination.

(a) Termination by LICENSEE. LICENSEE may terminate this Agreement by giving a ninety (90) written notice to SLU. Further:

(i) LICENSEE shall pay all amounts due and owing as of the effective date of termination; and

(ii) LICENSEE shall submit a report of the type described in Section 10.1.

(b) Termination by SLU. SLU may terminate this Agreement if LICENSEE:

(i) materially breaches this Agreement in a manner that can be cured (including but not limited to failure to pay annual maintenance fees, milestones, royalties or reports thereof) and such breach remains uncured for ninety (90) days following written notice of breach, such notice stating the nature of the defaults claimed by SLU;

(ii) is subject to a petition for relief under any bankruptcy legislation, or makes an assignment for the benefit of creditors, or is subject to the appointment of a receiver for all or a substantial part of LICENSEE's assets, and such petition, assignment or appointment prevents LICENSEE (as a legal or as a practical matter) from performing its obligations under this Agreement, or such petition, assignment or appointment is not otherwise dismissed or vacated within ninety (90) days; or

(iii) files a claim against SLU related to the Licensed Product, Patent Rights and/or Technology.

11.3. Consequences of Expiration or Termination.

(a) In the event of expiration of this Agreement or termination of the Agreement for any reason whatsoever:

(i) neither Party shall be discharged from any liability or obligation to the other that became due or payable prior to the effective date of such expiration or termination;

(ii) SLU is under no obligation to refund any payments made by LICENSEE to SLU prior to the effective date of such termination or expiration;

(iii) the rights and obligations of the Parties under Sections 4.1, 6, 7, 8, 10.1, 10.2, 11.3, 12 and 13 shall survive any expiration or termination of this Agreement; and

(iv) in any sublicense, SLU will stand in the place of LICENSEE with respect to the Sublicensee for a period of ninety (90) days during which time SLU will negotiate in good faith under reasonable terms and conditions with Sublicensee to execute a new license between Sublicensee and SLU, and if no new license is completed within the ninety (90) day period, the sublicense will terminate.

(b) In the event of termination of the Agreement:

(i) if LICENSEE or its Sublicensees then possess Licensed Product in inventory, have started the manufacture thereof or have accepted orders therefore, LICENSEE or its Sublicensees shall have the right, for up to one hundred twenty (120) days following date of termination, to sell their inventories thereof, complete the manufacture thereof and market such fully manufactured Licensed Product, in order to fulfill such accepted orders, subject to the obligation of LICENSEE to pay SLU the royalty payments therefore as provided in Section 3 of this Agreement;

(ii) subject to Section 11.3(b)(i), LICENSEE shall discontinue the manufacture, use, marketing, offering for sale and sale of Licensed Products; and

(iii) LICENSEE shall provide a final report of the type described in Section 10.1, including any allowable post-termination sales, by no later than thirty (30) days following the expiry of the 120-day period referred to in Section 11.3(b)(i).

12. CONFIDENTIAL INFORMATION

Each of SLU and LICENSEE shall hold in confidence any Confidential Information (including trade secrets) disclosed by the other or otherwise obtained by such Party from the other Party as a result of this Agreement, and each of SLU and LICENSEE shall protect the confidentiality thereof with the same degree of care that it exercises with respect to its own information of a like nature, but in no event less than reasonable care. LICENSEE shall have the right to provide Confidential Information to its Affiliates and Sublicensees, subject to the confidentiality obligations imposed by this Section. Without the prior written consent of the disclosing Party, a receiving Party shall not use, disclose, or distribute any Confidential Information, in whole or in part, except as required to perform such Party's obligations under this Agreement or in exercise or furtherance of its rights under this Agreement. Access to the disclosing Party's Confidential Information shall be restricted to the receiving Party's employees, agents and consultants, who, in each case, need to have access to carry out a permitted use and are bound in writing to maintain the use and confidentiality restrictions of such Confidential Information. The obligations set forth in this Section shall survive any termination or expiration of this Agreement in perpetuity (with respect to trade secrets and confidential financial information) and for the length of the life of the patent (with respect to all other Confidential Information).

All Confidential Information communicated by SLU to LICENSEE, including, without limitation, information contained in patent applications, shall be received in strict confidence by LICENSEE, used only for the purposes of this Agreement and not disclosed by LICENSEE or its agents or employees ("Representatives") without the prior written consent of SLU, unless such information is required to be disclosed by law, provided that LICENSEE shall first give notice to SLU of such disclosure and shall have made a reasonable effort to maintain the confidentiality of such information. Nothing contained herein shall prevent LICENSEE from disclosing Confidential Information to Sublicensees so long as such Sublicensees agree to be bound by these confidentiality provisions.

13. CHOICE OF LAW; DISPUTE RESOLUTION

13.1. Governing Law/Venue. This Agreement is made in accordance with and shall be governed and construed in accordance with the laws of the State of Missouri. The Parties agree that any action to interpret and/or enforce the terms of this Agreement shall be brought in the Circuit Court for St. Louis City or the United States District Court for the Eastern District of Missouri, and the Parties consent to the jurisdiction and venue of those courts.

13.2. Amicable Resolution. The Parties shall attempt to settle any controversy between them amicably. To this end, a senior executive from each Party shall consult and negotiate to reach a solution. The Parties agree that the period of amicable resolution shall toll any otherwise applicable statute of limitations. However, nothing in this clause shall preclude any Party from commencing mediation if said negotiations do not result in a signed written settlement agreement within thirty (30) days after written notice that these amicable resolution negotiations have commenced.

13.3. Mediation. If a controversy arises out of or relates to this Agreement, or the breach thereof, and if the controversy cannot be settled through amicable resolution, the Parties agree to try in good faith to settle the controversy by mediation. The Party seeking mediation shall propose [***] mediators, each of whom shall be a lawyer licensed to practice by the state of Missouri, having practiced actively in the field of commercial law for at least [***] years, to the other Party who shall select the mediator from the list. The Parties shall split the cost of the mediator equally. The Parties agree that the period of mediation shall toll any otherwise applicable statute of limitations. However, nothing in this clause shall preclude any Party from commencing other courses of remedy, including but not limited to if said negotiations do not result in a signed written settlement agreement within [***] days after written notice that amicable resolution negotiations have commenced.

13.4. Costs. The Party prevailing on substantially all of its claims shall be entitled to recover its costs, including attorneys' fees, , as well as for any ancillary proceeding.

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

13.5. Survival. The provisions of this Section shall survive expiration or termination of this Agreement.

14. NOTICES; PAYMENT INFORMATION

Except as otherwise provided, payments to be made hereunder to SLU shall be made by wiring the required amount to SLU's bank in accordance with SLU's instructions or by mailing or sending by commercial courier checks to SLU's address. Except as otherwise provided, notices and reports provided for herein shall effectively be given by mailing the same by certified or registered mail or by delivery by commercial courier, in each case properly addressed with charges prepaid. For the purposes of making payments and giving notices, the addresses of the Parties are as follows:

Saint Louis University
3700 West Pine Mall
Fusz Memorial Hall, Second Floor
St Louis, MO 63108
Attn: Director, Office of Innovation and Intellectual Property

Ultragenyx Pharmaceutical Inc.
77 Digital Drive
Suite 210
Novato, CA 94949
Attn: Chief Executive Officer

or to such subsequent addresses as either Party may furnish the other by giving notice thereof as provided in this Section 14.

15. MISCELLANEOUS

15.1. Assignment. Any attempt to assign or transfer this Agreement or any portion thereof in violation of this Section shall be void. This Agreement and all of the provisions hereof shall be binding upon and inure to the benefit of the Parties hereto and their respective successors and permitted assigns, but neither this Agreement nor any of the rights, interests or obligations hereunder shall be assigned by any Party hereto without the prior written consent of the other

Party, nor is this Agreement intended to confer upon any other Party except the Parties hereto any rights or remedies hereunder; provided, however, that LICENSEE may assign any or all of its rights and interests and delegate its obligations hereunder to any of its Affiliates (whether present or future) (provided that no such assignment to an Affiliate shall relieve assignor of its obligations hereunder); and provided further, that LICENSEE may assign its rights and interests and delegate its obligations hereunder (i) in connection with a Sale Transaction or (ii) as collateral to any of LICENSEE's lenders or any of LICENSEE's (or its Affiliate's) lenders. This Agreement shall inure to the benefit of, and be binding upon, the Parties hereto and their successors and permitted assigns. For greater certainty, LICENSEE shall not be required to make any payment whatsoever to SLU for the cash or non-cash proceeds received by LICENSEE or the holders of LICENSEE's equity (as and when received) with respect to any Sale Transaction.

15.2. Headings. The headings used in this Agreement are for convenience of reference only and are not intended to be a part of or to affect the meaning or interpretation of this Agreement.

15.3. Amendment. No amendment or modification hereof shall be valid or binding upon the Parties unless made in writing and signed by the Parties.

15.4. Force Majeure. Any delays in performance by any Party under this Agreement (other than the payment of monies due) shall not be considered a breach of this Agreement if and to the extent caused by occurrences beyond the reasonable control of the Party affected, including but not limited to, acts of god, embargoes, governmental restrictions, strikes or other concerted acts of workers, fire, flood, explosion, riots, wars, civil disorder, rebellion or sabotage. The Party suffering such occurrence shall immediately notify the other Party and any time for performance hereunder shall be extended by the actual time of delay caused by the occurrence.

15.5. Independent Contractors. In making and performing this Agreement, SLU and LICENSEE act and shall act at all times as independent contractors and nothing contained in this Agreement shall be construed or implied to create an agency, partnership or employer and employee relationship between SLU and LICENSEE. At no time shall one Party make commitments or incur any charges or expenses for or in the name of the other Party except as specifically provided herein.

15.6. Use of SLU's Name. Except as otherwise provided herein or required by law, LICENSEE will not originate any publication, news release or other public announcement, written or oral, whether in the public press or otherwise, relating to this Agreement or to the performance hereunder, without the prior written approval of SLU, which approval will not be unreasonably withheld. Such planned publication, news release or other public announcement shall be provided at least fourteen (14) days in advance for approval by SLU. SLU agrees that LICENSEE may make known in promotional and technical literature that the Patent Rights and Technology were developed at SLU; provided, however, that such use shall not state or imply that SLU has any relationship with LICENSEE other than as licensor.

15.7. Markings. LICENSEE shall comply with all applicable United States and foreign statutes related to the marking of Licensed Product(s) and Licensed Product(s) packaging with patent pending, patent number(s), copyrights or other intellectual property notices and legends required to maintain the intellectual property rights licensed in this Agreement.

15.8. Entity Status. If LICENSEE or a Sublicensee does not qualify, or loses qualification, as a small entity as provided by the United States Patent and Trademark Office, LICENSEE must notify SLU immediately.

15.9. Publication. LICENSEE agrees that SLU (including its employees) shall have a right to publish on the Technology in scholarly journals in accordance with its general policies and academic mission, and that this Agreement shall not restrict, in any fashion, SLU's right to publish on the Technology provided however that SLU shall provide LICENSEE with advance notice of any publication.

15.10. Severability. If any term, condition or provision of this Agreement is held to be unenforceable by a court having proper jurisdiction for any reason, it shall, if possible, be interpreted rather than voided, in order to achieve the intent of the Parties to this Agreement to the extent possible. In any event, all other terms, conditions and provisions of this Agreement shall be deemed valid and enforceable to the full extent of the law.

15.11. Compliance with Law. LICENSEE shall comply with and shall insure that any Affiliate or Sublicensee complies with all government statutes and regulations that relate to Licensed Products, including, but not limited to, FDA statutes and regulations and the Export Administration Act of 1979 (50 App. U.S.C. §2401 *et seq*.), as amended, and the regulations promulgated thereunder, and any applicable similar laws and regulations of any other country. Without limiting the generality of the foregoing, LICENSEE agrees that all Licensed Products used or sold in the United States shall be manufactured substantially in the United States to the extent required by Federal law.

15.12. Export Control Regulations. The Technology is subject to, and LICENSEE agrees to comply in all respects with U.S. export controls under the Export Administration Regulations (15 C.F.R. Part 734 et seq.) and U.S. economic sanctions and embargoes codified in 31 C.F.R. Chapter V. LICENSEE agrees that LICENSEE bears sole responsibility for understanding and complying with current U.S. trade controls laws and regulations as applicable to its activities subject to this Agreement. LICENSEE agrees not to sell any goods, services, or technologies subject to this Agreement, or to release or disclose or re-export the same: (i) to any destination prohibited by U.S. law, including any destination subject to U.S. economic embargo; (ii) to any end-user prohibited by U.S. law, including any person or entity listed on the U.S. government's Specially Designated Nationals list, Denied Parties List, Debarred Persons List, Unverified List, or Entities List; (iii) to any foreign national in the U.S. or abroad without prior license if required; or (iv) to any user, for any use, or to any destination without prior license if required.

15.13. Violation of Laws and Reformation. The Parties hereby agree that neither Party intends to violate any public policy, statutory or common law, rule, regulation, treaty or decision of any government agency or executive body thereof of any country or community or association of countries, and that if any word, sentence, paragraph or clause or combination thereof of this Agreement is found, by a court or executive body with judicial powers having jurisdiction over this Agreement or any of the Parties hereto, in a final, unappealable order to be in violation of any such provision in any country or community or association of countries, such words, sentences, paragraphs or clauses or combination shall be inoperative in such country or community or association of countries, and the remainder of this Agreement shall remain binding

upon the Parties hereto. In lieu of such inoperative words, sentences, paragraphs or clauses, or combination of clauses, there will be added automatically as part of this Agreement, a valid, enforceable and operative provision as close to the original language as may be possible which preserves the economic benefits to the Parties.

15.14. Waiver. None of the terms, covenants, and conditions of this Agreement can be waived except by the written consent of the Party waiving compliance. Waiver of one term, covenant or condition, shall not be construed as waiver of any other term, covenant or condition.

15.15. Entire Agreement. This Agreement and Exhibits attached hereto contain the entire agreement and understanding between the Parties with respect to the subject matter hereof, and merges all prior discussions, representations and negotiations with respect to the subject matter of this Agreement.

15.16. Counterparts. This Agreement may be executed in counterparts, each of which shall be deemed an original and all of which when taken together shall be deemed but one instrument.

IN WITNESS WHEREOF, the parties hereto have executed this Agreement by their duly authorized officers or representatives.

SAINT LOUIS UNIVERSITY

By: /s/ Raymond C. Tait
Title: Vice President, Research

ULTRAGENYX PHARMACEUTICAL, INC.

By: /s/ Emil D. Kakkis
Title: President and Chief Executive Officer

SUPPLY AGREEMENT

between

CREMER OLEO GmbH & **Co KG,** Glockengiesserwall 3, 20095 Hamburg,

Germany

— hereinafter referred to as Cremer —

and

Ultragenyx Pharmaceutical Inc, 60 Leveroni Court, Suite 200, Novato, California 94949, United States of America

— hereinafter referred to as Ultragenyx —

- each party also referred to as a "Party" and jointly as the "Parties" –

Preamble

Whereas, Cremer is a producer of oleo chemical products;

Whereas, Ultragenyx is a biotechnology company committed to bringing life-enhancing therapeutics for patients with rare and ultra-rare genetic diseases, also known as orphan diseases, to market;

Whereas, the Parties desire that Cremer supplies to Ultragenyx the product Triheptanoin (hereinafter also referred to as the "**Product**") in bulk form pursuant to the terms and conditions of this Agreement;

Whereas, Ultragenyx intends to process the Product into a pharmaceutical product in the meaning of Sec. 2 German Pharmaceuticals Act (Arzneimittelgesetz —AMG) and to market the processed Product in the Field (as defined below) (hereinafter referred to as the "Purpose"); and

Whereas, Ultragenyx intends to obtain regulatory approval for the processed Product as a pharmaceutical product in the meaning of Sec. 2 AMG.

Now therefore, the Parties hereto agree as follows:

Article 1 Supply of Product

1) Subject to the terms and conditions set forth in this Agreement Cremer shall supply Ultragenyx with the Product free from defect and meeting the product specification attached to this Agreement as **Annex A** (the "Product Specifications").

2) Cremer shall supply Ultragenyx exclusively with the Product worldwide. The aforesaid exclusivity is limited to [***] (collectively, the "Field"). Cremer may supply the Product to other customers outside of the Field.

3) Ultragenyx shall purchase the Product exclusively from Cremer.

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

Article 2 Orders and Delivery

1) The Product will be ordered by Ultragenyx through purchase orders. Purchase orders shall be submitted in any written or electronic form or by facsimile, setting out the quantity of Product required and the date for delivery. Cremer shall give its order confirmation in writing stating the quantity and Price (as defined below). Cremer shall not be obliged to deliver the Product in the absence of a written order confirmation given to Ultragenyx.

2) Delivery of the Product in bulk form by Cremer shall be EXW (Incoterms 2010), unless otherwise agreed in writing by the Parties.

3) Within [***] days of execution of this Agreement, Cremer shall deliver to Ultragenyx the Master Batch Record for the Product for Ultragenyx to review.

4) All Product shall be delivered with the applicable certificate of analysis and batch records for the Product delivered and an invoice for the quantity of Product delivered.

5) If Ultragenyx obtains regulatory approval for the processed Product, the Parties shall enter into a separate commercial supply agreement for the Product that sets forth the forecasting and ordering mechanism for commercial supply of the Product, enablement of the manufacturing process in the event of a failure to supply, the term of such commercial supply agreement and other customary terms and conditions.

Article 3 Prices and payment

1) The prices payable by Ultragenyx to Cremer for the Product (the “Price”) shall be agreed [***] every contract year; provided, that the Price may not increase more than the [***] for such period or [***]%, whichever is higher. At the date of signing the Parties agree on a Price of €[***] per kilogram for the Product.

2) If the parties cannot agree on a price for the Product by the beginning of a following contract year, Cremer may refuse to deliver the Product to Ultragenyx until the Parties agreed on a respective price.

3) Payments shall be made by Ultragenyx in Euro and within [***] days after receipt of a proper invoice.

4) Transfer of title with respect to any Product shall be subject to full payment and settlement of all claims Cremer may have against Ultragenyx in connection with the execution of this Agreement.

Article 4 Specification; Warranties; Cremer’s Liability; Indemnification

1) The Parties assume that the Product constitutes an active pharmaceutical ingredient in the meaning of Sec. 4 para. 19 AMG. Ultragenyx shall process the Product into a pharmaceutical product in the meaning of Sec. 2 AMG and market the processed Product as a pharmaceutical product in the meaning of Sec 2 AMG and to perform clinical trials. Cremer does not participate in the processing, manufacturing and marketing of the respective pharmaceutical product or in the clinical trials.

2) Upon execution of this Agreement and any purchase order, Cremer shall provide Ultragenyx with following documentation regarding the Product: Certificate of Analysis and the applicants' part of the Drug Master File once compiled.

3) Cremer represents and warrants that all quantities of Product delivered under the Agreement were manufactured in accordance with GMP. The Product shall be free from defects if it is within the specifications according to Annex A.

4) Cremer represents and warrants that it has not received any written notice from a third party alleging that the manufacture, use or sale of the Product infringes intellectual property rights of a third party.

5) Ultragenyx will perform final release of the Product. Ultragenyx may rely on the documentation provided by Cremer and Ultragenyx will not need to independently test the Product unless Ultragenyx determines such independent testing is necessary. In the event that the Product fails to conform to the Product Specifications, and/or GMP, Ultragenyx may reject the Product by giving written notice to Cremer within [***] days after receipt of the Product and all documentation (except such [***] day period will not apply for any latent defect). Within [***] days following receipt of the rejected and returned Product from Ultragenyx, Cremer will, at Ultragenyx's choice, replace such quantity of Product with Product conforming to the Product Specifications, and GMP or refund Ultragenyx the Price paid for such Product.

6) Cremer does not warrant or represent that the Product is effective in a pharmaceutical way within the meaning of Sec. 4 para. 19 AMG. Cremer does not warrant or represent that the Product is safe in a pharmaceutical and pharmacological way. Cremer does not warrant or represent that the Product is suitable for the intended Purpose by Ultragenyx. Cremer is not a pharmaceutical manufacturer within the meaning of Sec. 4 para. 18 AMG. Cremer's liability in connection with the Purpose and the processing and marketing of a pharmaceutical product is excluded. No. 9 below applies.

7) Except for a claim arising out of Cremer's intentional misconduct or gross negligence under this Agreement, in the event of legal proceedings being instituted against Cremer by a third party arising out of Ultragenyx's development, processing and commercialization of the Product, Ultragenyx shall indemnify and keep indemnified Cremer in full against all damages, losses, injuries, costs and expenses in connection with such legal proceedings. Cremer will inform Ultragenyx about any legal proceedings being instituted against Cremer without delay. Ultragenyx shall control the respective legal proceedings but shall not settle any claim that admits fault on behalf of Cremer without Cremer's consent (not be unreasonably withheld).

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.

8) In the event of legal proceedings being instituted against Ultragenyx by a third party arising out of Cremer's intentional misconduct or gross negligence under this Agreement, Cremer shall indemnify and keep indemnified Ultragenyx in full against all damages, losses, injuries, costs and expenses in connection with such legal proceedings. Ultragenyx will inform Cremer about any legal proceedings being instituted against Ultragenyx without delay. Cremer shall control the respective legal proceedings but shall not settle any claim without Ultragenyx's consent (not be unreasonably withheld).

9) Cremer's liability arising from this Agreement is limited to intentional misconduct or gross negligence. This limitation of liability does not apply to the injury of the life, body or health of a person, to claims according to the Product Liability Act (Produkthaftungsgesetz) or any other coercive legal liability claims.

10) NEITHER PARTY MAY CLAIM AND NEITHER PARTY IS LIABLE FOR CLAIMS FOR INDIRECT DAMAGES AND LOSSES, SUCH AS SPECIAL OR CONSEQUENTIAL LOSS OR DAMAGE, ANY LOSS OF ACTUAL OR ANTICIPATED PROFIT, OR REVENUE, ANTICIPATED SAVINGS OR BUSINESS OR DAMAGE TO GOODWILL OR BRAND EQUITY, ARE EXCLUDED.

Article 5 Term and Termination

1) This Agreement shall become effective on the date of its execution and shall remain in force for three years (the "Initial Term"). Thereafter, the Agreement shall be automatically renewed for additional two year periods (each a "Renewal Term", the Initial Term and all Renewal Terms, the "Term") unless either Party notifies the other Party of its intention not to renew in writing at least three calendar months before the expiration of the then current Term.

2) If a Party materially breaches an obligation under this Agreement and does not cure such breach within sixty (60) days of receiving notice of such breach from the non-breaching Party, the non-breaching Party may terminate this Agreement immediately upon written notice to the breaching Party.

3) Every termination has to be in writing.

Article 6 General Terms and Conditions

The application of General Terms and Conditions of any Party is excluded.

Article 7 Product Development

At the request and expense of Ultragenyx, Cremer shall perform development work for Ultragenyx to develop new formulations of the Product. All such work shall be performed pursuant to a statement of work (including a budget) to be agreed upon by the Parties and attached as an annex to this Agreement (each, a "Statement of Work"). In the event that in the course of performing a Statement of Work new Product know-how and intellectual property rights may result, can be created or have been created the Parties will enter into a separate Agreement in order to define the rights and duties regarding the aforesaid know how and intellectual property rights.

Article 8 Invalidity

In the event that any individual clauses of these terms and conditions are, or shall become, invalid, this shall not affect the validity of the remaining clauses. An invalid condition shall be deemed to have been replaced by such provision which is legally valid and corresponds nearest to the economic purpose of the clause originally deemed invalid.

Article 9 Applicable Law; Modifications; Annexes; Miscellaneous

1) The laws of the Federal Republic of Germany shall apply to the Agreement and any legal relations thereof, especially any purchase order, between Cremer and Ultragenyx shall be governed by that law. The law of the United Nations Conventions of the formation of Agreements for the international sale of goods (CISG) is excluded. Exclusive place of Jurisdiction is Hamburg, Germany.

2) No addition or modification to this Agreement shall be valid unless made in writing and signed by the Parties.

3) The Annex attached to this Agreement form an integral part of the Agreement.

4) This Agreement, including the Annexes and any Statement of Work, constitutes the entire agreement between the Parties concerning the subject matter hereof and supersedes all written or oral prior agreements or understandings with respect thereto except the Confidentiality Agreement between the parties dated September 26th, 2012. This Agreement shall be binding upon and shall inure to the benefit of the Parties hereto, their successors and assigns.

5) All waivers must be in writing and signed by the Party to be charged. Any waiver or failure to enforce any provision of this Agreement on one occasion will not be deemed a waiver of any other provision or of such provision on any other occasion.

6) Each Party must deliver all notices, consents, and approvals required or permitted under this Agreement in writing to the other Party at the address specified above, by personal delivery, by certified or registered mail (postage prepaid and return receipt requested), by a nationally-recognized overnight carrier, or by facsimile transmission with electronic confirmation of transmission. Notice will be effective upon receipt or refusal of delivery. Each Party may change its address for receipt of notice by giving notice of such change to the other Party.

7) This Agreement may be executed in counterparts by original signature, facsimile or PDF files, each of which shall be deemed an original, but all of which together shall constitute one and the same instrument.

Hamburg, November 19th, 2012

/s/ Thomas Kassberg
Ultragenyx Pharmaceutical Inc

/s/ Dr. R. Stephan
CREMER OLEO GmbH & Co KG

CREMER OLEO GmbH & Co. KG
Postfach 10 11 20, D-20007 Hamburg
Tel: 040/320 11-0, Telefax 320 11-400

Annex A — Specification of the Product
Trihepatanoin (Heptansäuretriglycerid)

No	Test	EP method	Limits
1	[***]	[***],	[***]
2	[***]	[***]	[***]
3	[***]	[***]	[***]
4	[***]	[***]	[***]
5	[***]	[***]	[***]
6	[***]	[***]	[***]
7	[***]	[***]	[***]
8	[***]	[***]	[***]
9	[***]	[***]	[***]
10	[***]	[***]	[***]
11	[***]	[***]	[***]
12	[***]		[***]
13	[***]	[***]	[***]
18	[***]	[***]	[***]

[***] Certain information in this document has been omitted and filed separately with the Securities and Exchange Commission. Confidential treatment has been requested with respect to the omitted portions.