UNITED STATES
SECURITIES AND EXCHANGE COMMISSION
WASHINGTON, D.C. 20549

FORM 8-K

CURRENT REPORT
Pursuant to Section 13 or 15(d)
of the Securities Exchange Act of 1934

Date of Report (Date of Earliest Event Reported): May 15, 2022

KemPharm, Inc.
(Exact Name of Registrant as Specified in Its Charter)

Delaware	001-36913	20-5894398
(State or Other Jurisdiction of Incorporation)	(Commission File Number)	(IRS Employer Identification No.)

1180 Celebration Boulevard, Suite 103, Celebration, FL	34747
(Address of Principal Executive Offices)	(Zip Code)

Registrant's Telephone Number, Including Area Code: (321) 939-3416

(Former Name or Former Address, if Changed Since Last Report)

Check the appropriate box below if the Form 8-K filing is intended to simultaneously satisfy the filing obligation of the registrant under any of the following provisions (see General Instruction A.2. below):

☐ Written communications pursuant to Rule 425 under the Securities Act (17 CFR 230.425)

☐ Soliciting material pursuant to Rule 14a-12 under the Exchange Act (17 CFR 240.14a-12)

☐ Pre-commencement communications pursuant to Rule 14d-2(b) under the Exchange Act (17 CFR 240.14d-2(b))

☐ Pre-commencement communications pursuant to Rule 13e-4(c) under the Exchange Act (17 CFR 240.13e-4(c))

Securities registered pursuant to Section 12(b) of the Act:

Title of each class	Trading Symbol(s)	Name of each exchange on which registered
Common Stock	**KMPH**	**The Nasdaq Stock Market LLC (Nasdaq Global Select Market)**

Indicate by check mark whether the registrant is an emerging growth company as defined in Rule 405 of the Securities Act of 1933 (§ 230.405 of this chapter) or Rule 12b-2 of the Securities Exchange Act of 1934 (§ 240.12b-2 of this chapter).

Emerging growth company ☐

If an emerging growth company, indicate by check mark if the registrant has elected not to use the extended transition period for complying with any new or revised financial accounting standards provided pursuant to Section 13(a) of the Exchange Act. ☐

Item 1.01. Entry into a Material Definitive Agreement.

Entry into Asset Purchase Agreement

On May 15, 2022, KemPharm, Inc. ("KemPharm" or the "Company"), a Delaware corporation and KemPharm Denmark A/S (the "Buyer"), a newly formed Danish company and wholly-owned subsidiary of KemPharm, entered into an Asset Purchase Agreement (the "Purchase Agreement") with Orphazyme A/S in restructuring, a Danish public limited liability company ("Orphazyme"). Under the terms of the Purchase Agreement, at closing the Buyer will purchase all of the assets and operations of Orphazyme related to arimoclomol and will settle all of Orphazyme's actual outstanding liabilities to its creditors with a cash payment of USD$12.8 million (the "Asset Purchase"). The Company expects that the cash payment will be financed by a revolving line of credit secured by KemPharm's balance sheet. In addition, the Buyer has agreed to assume an estimated reserve liability of USD$5.2 million related to revenue generated from Orphazyme's Early Access Program in France. The transaction is expected to close on or before June 1, 2022, subject to customary closing conditions and approval by Orphazyme's creditors and the Danish bankruptcy court.

The foregoing description of the Purchase Agreement does not purport to be complete and is qualified in its entirety by reference to the Purchase Agreement, which is filed as Exhibit 10.1 hereto, and is incorporated by reference into this Current Report on Form 8-K.

Item 7.01. Regulation FD Disclosure.

On May 15, 2022, the Company issued a press release announcing the entry into the Purchase Agreement. A copy of the press release is attached to this Current Report on Form 8-K as Exhibit 99.1 hereto.

In addition, the Company plans to provide supplemental information regarding the Asset Purchase in a conference call and live audio webcast with a slide presentation scheduled for May 16, 2022, at 8:30 a.m., EDT. A copy of the slide presentation is attached as Exhibit 99.2 hereto.

The information furnished pursuant to Item 7.01 of this Current Report on Form 8-K, including Exhibits 99.1 and 99.2, shall not be deemed "filed" for purposes of Section 18 of the Securities Exchange Act of 1934, as amended (the "Exchange Act"), or otherwise subject to the liability of such section, nor shall it be incorporated by reference into future filings by KemPharm under the Securities Act of 1933, as amended, or under the Exchange Act, unless the Company expressly sets forth in such future filing that such information is to be considered "filed" or incorporated by reference therein.

Item 9.01. Financial Statements and Exhibits.

(d) Exhibits

Exhibit No.	Description
10.1*	Asset Purchase Agreement by and among KemPharm, Inc., KemPharm Denmark A/S and Orphazyme A/S in restructuring, dated May 15, 2022.
99.1	Press release dated May 15, 2022.
99.2	Presentation dated May 16, 2022.
104	Cover Page Interactive Data File (embedded within the Inline XBRL document)

* Schedules have been omitted pursuant to Item 601(a)(5) of Regulation S-K. A copy of any omitted schedule will be furnished to the Securities and Exchange Commission upon request; provided, however, that the parties may request confidential treatment pursuant to Rule 24b-2 of the Exchange Act for any document so furnished.

SIGNATURES

Pursuant to the requirements of the Securities Exchange Act of 1934, the registrant has duly caused this report to be signed on its behalf by the undersigned hereunto duly authorized.

KemPharm, Inc.

Date: May 16, 2022

By: /s/ R. LaDuane Clifton
R. LaDuane Clifton, CPA
Chief Financial Officer, Secretary and Treasurer

Asset Purchase Agreement
regarding certain assets in Orphazyme A/S in restructuring

Gorrissen Federspiel

This Asset Purchase Agreement (the "**Agreement**") is on this day entered into between:

Seller: Orphazyme A/S in restructuring, CVR no. 32 26 63 55, a company incorporated and registered under the laws of Denmark having its registered address at Ole Maaløes Vej 3, 2200 København, Denmark (the "**Seller**"); and

Buyer: KemPharm Denmark A/S, CVR no. 43259326, a company incorporated and registered under the laws of Denmark having its registered address at C/O Moalem Weitemeyer Advokatpartnersels Amaliegade 3, 1256 Copenhagen, Denmark (the "**Buyer**"); and

Parent: KemPharm Inc, a company incorporated and registered under the laws of the state of Delaware having its principal executive office at 1180 Celebration Blvd., Suite 103, Celebration, Florida 34747, USA of Delaware having its principal executive office at 1180 Celebration (the "**Parent**"), as owner of the total share capital in the Buyer.

The Seller and the Buyer are collectively referred to as the "**Parties**" and/or each a "**Party**".

WHEREAS

(i) the Seller has agreed to sell, assign and transfer to the Buyer and the Buyer has agreed to purchase, accept assignment and assume from the Seller the Business (as defined below), excluding the Retained Assets, upon the terms and subject to the conditions herein set forth;

(ii) the Seller is a late-stage biopharmaceutical company developing Arimoclomol for Niemann-Pick disease type C (NPC);

(iii) Arimoclomol is an investigational drug candidate that amplifies the production of heat shock proteins (HSPs), which can rescue defective misfolded proteins and improve the function of lysosomes;

(iii) the Buyer has been informed that the Seller is in restructuring pursuant to the Danish Bankruptcy Act (in Danish: "konkursloven");

(iv) the purpose of this Agreement and the intention of the Parties are that the Buyer shall take over all relevant Assets of the Seller and – if possible and accepted by the relevant third parties – continue all early access programs on Arimoclomol and all processes of approval of Arimoclomol for commercial sale; and

(v) on 4 September 2019 the Seller has granted a floating charge covering certain of the Assets (the "**Floating Charge**") (in Danish "virksomhedspant") to Kreos Capital VI (UK) Limited ("**Kreos**") of EUR 9,000,000.

(vi) the Parent guarantees certain obligations of the Buyer under this Agreement.

The Parties have entered into the following Agreement regarding the purchase of the Business:

1 Certain Definitions

For the purpose of this business transfer agreement, unless the context otherwise requires or separately defined herein, the term

1.1 "Business" means the Seller's business activities as defined in this Agreement, including activities within the field of research, development, production, marketing, sale and/or licencing of medicinal products for the treatment of various diseases, including Lysosomal Storage Disease (LSD), neuromuscular diseases and related diseases, and other associated activities.

1.2 "Closing" means the completion of the sale and purchase of the Business and the other transactions provided for herein in accordance with Clause 9;

1.3 "Closing Date" means the date on which Closing occurs;

1.4 "EMA" means the European Medicines Agency.

1.5 "FDA" means the US Food and Drug Administration.

1.6 "French Revenue Liabilities" means any obligation to pay back revenue or other liability of the Seller and/or its Subsidiaries under the French Early Access Compassionate Use Program (ATU) relating to any sale performed under the French Early Access Compassionate Use Program performed both prior to the Closing Date by the Buyer and after the Closing Date by the Seller. The obligation relating to the time prior to the Closing Date is estimated to be maximum of $5,200,000.

1.7 "Office Locations" means Ole Maaløes Vej 3, 2200 København, Denmark;

1.8 "Restructuring Proposal" means a restructuring proposal issued pursuant to section 13 b of the Danish Bankruptcy Act;

1.9 "Signing" means the signing of this Agreement on the date of this Agreement;

1.10 "Subsidiaries" means Orphazyme Schweiz GmbH and Orphazyme US Inc; and

1.11 "Transaction" means the actions and agreements contemplated by this Agreement.

2 Sale and Purchase of assets and assumption of liabilities and obligations

2.1 Subject to the terms and conditions of this Agreement, on the Closing Date, the Seller sells, assigns and transfers to the Buyer, and the Buyer purchases, accepts assignment and assumes from the Seller all of the Seller's right, title and interest in and to all assets free and clear of any charges, pledges and similar third party rights other than the Retained Assets of the Seller as the same exists on the Closing Date (the "**Assets**"), including, but not limited to the following assets and rights:

2.1.1 Fixed Tangible Assets: All fixed tangible assets owned by the Seller and relating to the Business as of the Closing Date, including machinery, operating equipment, installations, fixtures, fittings, office equipment and furniture, IT hardware (e.g. desktops, laptops, printers, servers, etc.), spare parts, tools, consumables and fittings, and including the fixed tangible assets specified in **Schedule 2.1.1** (the "**Fixed Tangible Assets**").

2.1.2 Inventory: All inventory of the Business owned by the Seller and which are located at the Office Locations, with suppliers, warehouses or in transit as of the Closing Date, including raw materials, ancillary materials, work in progress (including work in progress on behalf of third parties), finished goods and goods for sale, and including the inventory specified in **Schedule 2.1.2** (the "**Inventory**").

2.1.3 Records: Documentation and information of the Seller concerning the Business as of the Closing Date, including databases, customer and supplier lists, files, correspondence and other documentation, and including the documentation and information concerning the Business as of the Closing Date located at the locations specified in **Schedule 2.1.3** (the "**Records**"), save for as set out in clause 2.2.2. For the avoidance of doubt, the Seller is allowed to keep a copy of the Records.

2.1.4 Intellectual Property: All intellectual property rights related to NME Diaoxazimes, NME Oximes, NME Pyridines (jointly "**Discovery Compounds**") and Arimoclomol, or in any other way relating to the Business, including trademarks, show-how, know-how, patents, copyrights, rights in business names, trade names, websites, other domain names, social media pages/accounts, telephone numbers (if assignable), historical and planned product developments for products forming part of the Business, trade secrets and know-how, and including the intellectual property specified in **Schedule 2.1.4** (the "**Intellectual Property**"). For the purpose of evidencing the transfer of the Intellectual Property from the Seller to the Buyer at public authorities such as the USPTO and/or EPO, the Seller is on the Closing Date obligated to executive and deliver to the Buyer the trademark and patent assignments as specified in clause 9.2.2.

2.1.4.1 The Buyer is, to a level consistent with the customary practices in the biotechnology and pharmaceutical industries, obliged to continue commercially reasonable efforts on developing Discovery Compounds for the purpose of having the drug approved and commercialised. If such efforts cannot be evidenced by the Buyer towards Seller no later than 48 months after Closing, the Buyer is obliged to reassign to the Seller all intellectual property rights relating to the Discovery Compounds, which were taken over from the Seller. To the extent the Buyer after Closing generates any new intellectual property rights related to Discovery Compounds, the Buyer retains full ownership to such new intellectual property, and such new intellectual property shall not be assigned to Seller. The Seller (or a thirdparty designated by the Seller) shall, however, for a period of 48 months after Closing be entitled to license such new intellectual property rights related to Discovery Compounds from the Buyer on reasonable terms negotiated in good faith between the parties at such relevant point in time.

2.1.5 Goodwill and Reputation: The goodwill and reputation related to the Business, but not the name/trademark "Orphazyme". The Seller acknowledges that the Buyer on a need to have basis and solely for the purpose of communication with patients, physicians, treatment centers, and for the purpose of distribution of Arimoclomol under early access programmes is entitled to use the name and trademark "Orphazyme" for a period of 24 month after Closing without the Seller being entitled to receive any royalty payment for such use, provided that such use is permitted under applicable law including regulatory requirements. The name may not be used by the Buyer in a way that could have an adverse effect for the Seller and the Buyer is liable towards the Seller for any unauthorized use of the name and/or any discredit caused by the Buyer's use of the name. When/if using the name "Orphazyme" the Buyer shall clearly state that the Buyer does not represent or act on behalf of the Seller.

2.1.6 FDA and EMA applications on approval of Arimoclomol and patients early access program: To the extent possible, which is not guaranteed by the Seller in any way, the Buyer shall take over and continue all patients early access programs for the Seller worldwide and shall continue the ongoing approval processes at the FDA and EMA, subject to applicable consents and permissions. With respect to the FDA application, the Parties acknowledge that both Parties and their respective counsels, consultants and employees shall corporate on regulatory matters pertaining to Arimoclomol prior to the meeting with the FDA on 14 July 2022. This cooperation includes to the extent necessary the sharing of data between the Parties, including confidential and privileged information. All costs for the Parties' consultants and counsels assisting in this respect after Closing, on request of the Buyer, must be paid by the Buyer.

2.1.7 Permits: All of the Seller's, if any, transferable permits, licenses, approvals, certifications, accreditations and other authorizations relating to the Business and/or the property located at the Office Locations, including those specified in **Schedule 2.1.7** to the extent assignable (the "**Permits**").

2.1.8 The Assets mentioned in this clause 2.1 shall not be regarded as complete if the Parties before or after the Closing Date become aware of further assets which are not described in this Agreement, but which nevertheless form part of the Business. In that case such further assets shall on or after the Closing Date without any notice from the Buyer be transferred and delivered to the Buyer without this triggering further payments to the Seller, unless otherwise stated in the Agreement or agreed in writing between the Parties.

2.1.9 The Seller shall use all reasonable commercial efforts to assist the Buyer with the Business transfer in order for the Buyer to operate the Business immediately after the Closing Date as previously carried out by the Seller prior to Closing. The Parties both acknowledge the importance of start working closely together immediately after Signing on important matters, including but not limited to resubmissions. In addition, the Buyer acknowledges that Employees working with legal, finance and bookkeeping to a reasonable extent will have to assist the Seller in paying obligations of the Seller, communication with suppliers, preparation of annual report, disclosures etc. in expectedly the first 2 months after Closing. The Seller shall not compensate the Buyer for this financially, unless the extent exceeds what

can reasonably be expected of the Buyer. Similarly, the CEO of the Seller will assist the Buyer to the same extent for the first 2 months after Closing without any compensation from the Buyer. If further assistance is needed from the Seller's CEO, the Buyer and the CEO will have to discuss entering into a consultancy agreement. The Seller's CEO will not act as CEO for the Buyer during the term of his employment with the Seller.

2.1.9.1 The Seller shall, subject to any legal, regulatory or listing requirements of the Seller, after Closing procure that all references and data related to any of the transferred Assets will be removed from the Seller's website and similar public platforms/medias. The Seller shall also after Closing procure that all traffic on the Seller's website related to the early access programs in a reasonable way is redirected to the Buyer's website. However, Seller is not to remove issued company announcements and annual reports (even though information on transferred Assets appear from these) or other information required to be kept publicly available under any laws or regulations or listing requirement applicable to the Seller.

2.2 Notwithstanding the generality of Clause 2.1, the following assets etc. of the Seller (the "**Retained Assets**") shall not be transferred from the Seller to the Buyer. All costs, risks, liabilities and expenses related to the Business, attributable to or accrued before or on Closing shall, with the qualifications and limitations provided for in this Agreement, be for the account of the Seller and all costs, risks, liabilities and expenses related to the Business due, attributable to or accrued after Closing shall be for the account of the Buyer, provided that, for the avoidance of doubt, that Employee Related Liabilities shall, as of Closing, be for the account of the Buyer subject to the terms set out in clause 3:

2.2.1 The Seller's Good Distribution Practice (GDP) licence/permit(s) issued by the Danish Medicines Agency (in Danish "Lægemiddelstyrelsen"), Certificate No: DK GDP 10000177.

2.2.2 Bookkeeping and other documents, including the entire ERP-system relevant for the Seller to continue operating an active company and/or to perform solvent liquidation and/or to comply with applicable law, remains with the Seller; provided that the Seller will give the Buyer access to information necessary for the Buyer to continue the Business and early access programs and the application processes with the FDA and EMA. The e-mail domain and accounts "@ophazyme.com" and all e-mails send to and from any e-mail under this domain is not to be transferred to the Seller, unless these e-mails are stored in the cloud database to be transferred to the Seller, where all data relevant for the early access programs, studies and approval processes with EMA and FDA is store. The Seller will disclose any such information to the Buyer, provided that such disclosure shall be subject to clause 12.2 on confidentiality and the Buyer is obliged not to use the information in any way that could have an adverse effect for the Seller, including but not limited to the Seller's defense in law suits and Seller's compliance with stock exchange regulation.

2.2.3 The Seller's receivables for deliveries performed under the French Early Access Compassionate Use Program prior to the Closing Date are not transferred to the Buyer. Hereby, the Seller shall (from Clinigen Ireland Ltd) retain rights to all revenue for any sale under the French Early Access Compassionate Use Program performed before (but not including) the Closing Date (to be used to repay Kreos). On the contrary, revenue and receivables for sale performed after (and including) the Closing Date is to be received by the Buyer (provided that the Buyer is allowed to continue the French Early Access Compassionate Use Program), which the Seller does not guarantee in any way.

2.2.4 Any other receivable, refund amount including but not limited to VAT returns, tax credit payments and tax deficits, or other claim for money of the Seller existing on the Closing Date, including but not limited to any amount of the Seller retained by or in other way owned by Clinigen Ireland Ltd. to the Seller relating to deliveries performed under the French Early Access Compassionate Use Program prior to the Closing Date.

2.2.5 Cash and money in bank accounts of the Seller.

2.2.6 The Seller's shares in Orphazyme Schweiz GmbH and Orphazyme US Inc. However, post Closing the Parties will discuss if some contracts are to be transferred from Orphazyme US Inc to the Buyer.

2.2.7 Internet and e-mail-domains/accounts including the name "Orphazyme". However, as for Employees, the Seller will keep their e-mail-accounts active for the first 4 months after Closing and hereby allow the Employees to use the e-mail accounts when working for the Buyer, until the Buyer has set up new e-mail-accounts for the Employees provided that Employee's signatures clearly state that the Buyer and not the Seller is the sender.

2.2.8 It is emphasised by the Seller that the Seller's lab room at Ole Maaløes Vej 3 and many office working stations are to be vacated before Closing. The lab equipment in the lap room and office equipment at vacated office stations are not transferred to the Buyer, as the Sellers will sell these assets to third party buyers prior to Closing when vacating the premises, which causes no adjustment of the Purchase Price. However, it is emphasised that the content of the Seller's freezer in the basement of Ole Maaløes Vej 3 and at any other thirdparty's storage facility shall also be transferred to the Buyer provided that the content relates to the transferred Business and Assets.

2.2.9 The laptops, telephones and other equipment specifically listed in **Schedule 2.2.9** and the telephone number/subscription of the Seller's CEO.

2.3 Subject to the terms and conditions of this Agreement, on the Closing Date, the Seller assigns and transfers to the Buyer, and the Buyer accepts assignment and assumes from the Seller the following liabilities and obligations of the Seller (the "**Assumed Liabilities**"):

2.3.1 Employee Related Liabilities: The Seller's liabilities and obligations related to the Employees as described in Clause 3 (the "**Employee Related Liabilities**");

2.3.2 The French Revenue Liabilities: The Buyer expects from a commercial perspective to have to satisfy all the Seller's French Revenue Liabilities to enable the Buyer to continue French Early Access Compassionate Use Program. On this basis, it has been a requirement of the Buyer to take over from the Seller and pay the French Revenue Liabilities directly to the receiver. The French Revenue Liabilities are expected to amount to a maximum of $5,200,000 per Closing. However, no adjustment of the Purchase Price shall be made, if the French Revenue Liabilities turns out to be of a higher or lower amount (or not payable at all).

2.3.3 All Seller's obligations under asset purchase agreement between CytRx Corporation and Seller of May 13, 2011 (**Schedule 2.3.3**), including milestone payments to CytRx Corporation pursuant to section 2.7 therein.

2.3.4 The Seller's obligation to perform a potential milestone payment of USD 1,000,000 to Hyman, Phelps & McNamara, P.C. in case of FDA Approval as defined in letter of July 7, 2021 (**Schedule 2.3.4**).

2.3.5 All rights and obligations under the lease agreement regarding Ole Maaløes Vej 3, 2200 København, Denmark, as the lease agreement is assigned from the Seller to the Buyer in all aspects. The Seller shall prior to or on the Closing Date deliver evidence that the landlord has approved the assignment of the lease agreement to the Buyer.

2.3.6 All rights and obligations of the Seller under any agreement relating to patients early access programs, studies, developments and approval of Arimoclomol and/or Discovery Compounds (including but not limited to leasing agreements, consultancy agreements, studies, licenses), and including those specified in **Schedule 2.3.6** (the "**Contracts**"), provided that (i) the Buyer has not in writing to the Seller declined to accept the assignment of a specific Contract no later than 28 June 2022 (any such declined Contracts shall be retained by the Seller) and (ii) that the agreements are assignable according to their terms and/or clause 14 c (2) of the Danish Bankruptcy Act or that the relevant contracting parties accepts assignment. The Buyer and the Seller shall both exercise reasonable efforts to ensure that the Contracts accepted by the Buyer are assigned and relevant consents are obtained and hereby that the Seller is released from any obligation of such Contract as per the Closing Date in accordance with Clause 2.5. However, the Buyer shall in good faith handle the dialogue, negotiation etc. with relevant contracting parties as the Buyer from Closing will have taken over the Employees from the Seller, whereby the Seller will have limited resources to assist in relation to such matters. However, the board and management of the Seller will assist by signing necessary documents etc. as set out in Clause 2.5.

2.4 Notwithstanding Clauses 2.1 and 2.3, the Buyer only acquires – except from sensitive personally information related to the Employees – non sensitive personal information, and no sensitive patient information is transferred to Buyer. The Parties are aware that the transfer of the Assets set out in this Agreement may constitute a transfer pursuant to the Danish Data Protection Act. The Parties agree that the transfer of the Assets can take place with title in the Danish Data Protection Act since there will only be transferred non-personally sensitive identity information. In the case there has been transferred personally sensitive information to the Buyer, and such transfer cannot take place with title in the Danish Data Protection Act or other relevant statutory law, then the Buyer shall not be authorised to use such sensitive personal information and shall be obliged to promptly delete such information as the time where the Buyer becomes aware hereof.

2.5 The ownership of, title to and risk of the Assets shall pass from the Seller to the Buyer on the Closing Date, and the Buyer shall accept from the Seller assignment of and assume the Assumed Liabilities on the Closing Date. Hereby, from (and including) the Closing Date all

obligations under the Assumed Liabilities rests with the Buyer. The Seller shall, however, remain liable for any actual or potential liability or obligation under the Assumed Liabilities relating to the period prior to the Closing Date, except from Employee Related Liabilities which are regulated by Clause 3.3. In addition, earnings on the Assets until the Closing Date belongs to the Seller (regardless if paid after the Closing Date), while earnings on the Assets (including and) after the Closing Date will belong to the Buyer. The Seller undertakes to assist to allow for transfer of any of the Assets and the Assumed Liabilities, including signing all required transfer documents and forms to effect the transfer. The payment of fees and registration duties shall be paid by the Buyer exclusively. The Buyer will handle the formalities and potential negations in relation to transfer of Assets and contracts as the Buyer will have taken over Seller's Employees.

2.6 The Assets shall be transferred to the Buyer without any charges or pledges. The Seller informs and warrants that the Seller will satisfy all of the Seller's obligations towards Kreos under the Floating Charge from the Purchase Price, meaning that the Buyer will take over the Assets free of Kreos' Floating Charge. As for potential liens in Inventory, the Seller will pay for storage and freight up to the Closing Date, while the Buyer will be obliged to pay for storage etc. after the Closing Date and/or to negotiate own terms and pay relevant costs for continues storage or release of the Inventory free from liens etc.

3 Employees

3.1 On the Closing Date, the Buyer shall assume the Seller's employment rights and duties towards the employees specified in **Schedule 3.1** (the "**Employees**"). The Employees will transfer in accordance with the Danish Business Transfer Act (in Danish: "*Lov om lønmodtageres retsstilling ved virksomhedsoverdragelse*").

3.2 In respect of any Employee listed in Schedule 3.1 not transferred pursuant to the provisions under the Danish Act on Transfers of Undertakings, the Parties shall in good faith cooperate to facilitate the transfer of such Employee(s) from the Seller to the Buyer, such transfer ultimately being subject to the acceptance by such Employee.

3.3 The employment relationships of the Employees shall be transferred to the Buyer on the same employment terms and conditions as applicable to the Employees at the Closing Date. As concerns the direct relation to the Employees, all obligations towards the Employees shall be taken over and assumed by the Buyer, including but not limited to earned salary, holiday and pensions pay accrued – but not settled – before the Closing Date to the extent that such obligations concern the period after the initiation of the restructuring process (in Danish: *"indledningen af rekonstruktionsbehandlingen"*). Notwithstanding aforesaid, the Seller shall reimburse the Buyer all incurred Employee costs to the extent that these costs concern – in whole or in part - the period before the Closing Date. Moreover, any retention bonus and bonus for a successful transfer of the Business offered to the Employees by the Seller from 11 March to the Closing Date is to be paid by the Seller (or reimbursed from the Seller to the Buyer if paid by the Buyer). Hence, the only obligation of the Seller towards the Employees after the Closing Date is to pay such bonus, as all other obligations rest with the Buyer and causes no reimbursement of the Seller towards the Buyer.

3.4 The Buyer has been informed that the Employees may be subject to the Seller's Long Term Incentive Program, which will terminate as of the Closing Date. The Buyer acknowledges that the Buyer after the Closing Date will seek to replace the Long Term Incentive Program with an incentive program on terms negotiated between the relevant Employees and the Buyer.

3.5 The Seller shall retain all liability in relation to employees of the Seller that are not listed in Schedule 3.1, including dismissed employees, save to the extent that the Buyer is liable for a dismissal executed in contravention of section 3(1) of the Danish Act on the Transfer of Undertakings ("Virksomhedsoverdragelsesloven").

4 Purchase Price

4.1 The Purchase Price for the Assets (and taking into account the Assumed Liabilities) has been determined at $ 12.8 million (the "**Purchase Price**"). On the Closing Date, the Purchase Price shall be paid by way of a payment of $12.8 million in cash transferred by the Buyer to a bank account designated by the Seller prior to Closing.

4.2 The purchase price shall be paid in the currency of USD.

4.3 In accordance with section 45 (2) of the Danish Act on Depreciation (in Danish "Afskrivningslovens § 45, stk. 2") the allocation of the Purchase Price on the Assets appears from **Schedule 4.3**.

5 Representations and Warranties

5.1 The Buyer is aware that the Seller is in restructuring pursuant to the Danish Bankruptcy Act. In that connection the Buyer has accepted that the Buyer cannot direct any claims or remedies towards the Seller in the event of any breach or lack of conformity, except for any defect in title relating to the Assets subject, however, to the disclosed material in the virtual data room, cf. Clause 6. The Parties agree that this has been taken into consideration when determining the Purchase Price.

5.2 The Seller does not undertake any representations or warranties, neither directly, implied nor written or oral, in respect of the Assets, which are transferred on an as-is basis, or in connection with this Agreement.

5.2.1 Notwithstanding Clause 5.2 , the Seller represents and warrants that to the best of the Seller's knowledge (understood as the actual knowledge of the CEO and the general counsel of the Seller as per the date of Signing) without having carried out inquiries other than a review carried out by the Seller's external IP advisor, the information contained in Schedule 2.1.4 represents a full and complete list of the Seller's Intellectual Property related to the Business as transferred to the Buyer as part of the Assets. However, it is emphasised by Seller that some of the intellectual property rights listed in schedule 2.4.1 have expired.

5.3 If the Buyer notwithstanding the provisions of this Clause 5 is entitled to direct a claim or remedy towards the Seller, the Parties agree that such claim or remedy only can be made in the form of a claim for a proportional reduction of the Purchase Price by deduction of the value of the Asset in question under this Agreement from the purchase price and subject to the rules under Danish law on pro rata reduction (in Danish: "forholdsmæssigt afslag"). Such claims are in the aggregate maximized to a sum corresponding to the Purchase Price and as effectively paid to the Seller.

5.4 Any and all claims pursuant to or arising out of this Agreement will lapse, if the Buyer does not take documented legal actions in respect of such claims and notifies the Seller hereof in writing within 6 months of the Closing Date.

5.5 The remedy in this Clause 5 is the exclusive remedy available to the Buyer. In particular, the Buyer is not entitled to cancel or rescind the Agreement or to claim damages.

6 Due diligence

6.1 The Buyer has been given the opportunity to conduct a due diligence investigation from the virtual data room Gamora at the data site Datasite.com.

6.2 The Parties agree that all material in the data room is considered disclosed and to the Buyer's knowledge as of the Closing Date to the extent that the matter was disclosed in a way which a prudent buyer should reasonable be expected to understand the repercussions of. The Buyer confirms that the Seller has answered all questions raised by the Buyer during the diligence investigation and that such answers to the Buyer's knowledge have been adequate and sufficient. The Buyer further confirms as per Signing not to be aware of any matter which could constitute a breach under this Agreement.

7 Covenants between Signing and Closing

7.1 Conduct of Business

7.1.1 During the period from Signing until Closing the Seller shall:

7.1.1.1 Operate the Business in the ordinary course of business consistent with past practice and in accordance with current legislation;

7.1.1.2 Not without the prior written consent of the Buyer terminate or materially amend any terms in the Contracts, including renegotiating or prolonging such agreements with terms less attractive to the Business than applicable as per Signing;

7.1.1.3 Not without the prior written consent of the Buyer materially change the employment terms or terminate the employment of or make redundant any of the Employees; and

7.1.1.4 Not without the prior written consent of the Buyer dispose of any of the Assets existing as of the Signing Date.

7.1.1.5 Procure that any annuities, prosecution matters and any other intellectual activity, that needs to be effectuated to prevent any harm, damage or loss of rights with respect to the Intellectual Property listed in schedule 2.1.4 is duly handled by the Seller.

8 Conditions to Closing

8.1 The Closing shall not occur as set forth in Clause 9 unless the restructuring proceedings of the Seller has been completed on one of following two ways:

(a) The Restructuring Proposal has been approved by a simple majority (measured by amounts) of the creditors of the Seller attending and able to vote on the Restructuring Proposal at a creditor's meeting, cf. Section 13 d of the Danish Bankruptcy Act (konkurslovens § 13 d) and the subsequent affirmation of the Restructuring Proposal by the Danish Bankruptcy Court, cf. Section 13 e of the Danish Bankruptcy Act (konkurslovens § 13 e), or

(b) The bankruptcy court has formally determined that the restructuring proceedings are terminated pursuant to section 15(1)(i) of the Danish Bankruptcy Act, allowing the restructuring proceedings to stop if the Seller is no longer insolvent.

8.2 If the conditions in Clause 8.1 has not been satisfied at Closing, none of the Parties are obliged by this Agreement and the Parties shall not have any claims of any kind towards each other based on this Agreement (other than as set forth in Clause 9.4), and this Agreement shall terminate. However, the Parties will in this situation in good faith seek to negotiate on potential alternative solutions.

9 Closing

9.1 Closing shall take place at 0.01 (CET), on 1 June 2022, or at such other place, date or time as may be agreed between the Parties.

9.2 On the Closing Date, the Seller shall deliver evidence to the Buyer that the landlord has approved the assignment of the lease agreement to the Buyer, cf. clause 2.3.5.

9.2.1 On the Closing Date, the Seller shall deliver a USB-stick to the Buyer with a copy of the virtual data room, together with written confirmation that the virtual data room has been closed and that no new documentation has been included in the virtual data room after Signing.

9.2.2 On Closing Date, the Seller shall deliver to the Buyer executed trademark and patent assignments in the form attached as **Schedule 9.2.2(a)** and **Schedule 9.2.2(b)**. The executed trademark and patent assignments shall be deliver by an international delivery service to Buyer's legal intellectual property representative, McAndrews, Held & Malloy, 500 West Madison Street, 34th Floor, Chicago, Illinois 60661, United States to the attention of Mr. Troy Groetken.

9.3 On the Closing Date, immediately following receipt by the Seller of the Purchase Price, the Seller shall satisfy all of the Seller's obligations towards Kreos under the Floating Charge, meaning that the Buyer shall take over the Assets free of Kreos' Floating Charge.

9.4 In case of any termination of this Agreement pursuant to Clause 8.2, this Agreement (other than Clauses 10 (Expenses), 12 (Confidentiality) and 19 (Governing Law and Venue)) shall terminate. Nothing in this Clause 9.4 shall be deemed to release either Party from any liability for any breach by such Party of the terms of this Agreement prior to termination.

10 Financial Statements

10.1 After Closing, the Seller shall deliver to the Buyer as promptly as practicable, (i) final audited financial statements for the year ended December 31, 2021, (ii) unaudited financial statements as of March 31, 2022, taking into account adjustments required by Regulation S-X (which shall have been reviewed by Seller's independent accountants in accordance with the Statement on Auditing Standards No. 100) (the "Financial Statements").

10.2 The Financial Statements shall be prepared in accordance with IFRS applied on a consistent basis throughout the periods covered thereby and the books and records of Seller, in a manner consistent with the preparation of the historical financial statements for the equivalent periods, including with respect to (i) footnote disclosure (except, in the case of interim financial statements, such exceptions and omitted footnotes as are customary in the preparation of interim financial statements) and (ii) the audit or review, as applicable, of Seller's independent accountants.

10.3 The Seller agrees to consent to the inclusion of any financial statements or information obtained by the Buyer in any filings by the Buyer with the SEC or any other securities regulatory authority or exchange.

10.4 The Seller agrees that it will use reasonable best efforts to cause its independent accountants to provide to the Buyer with a consent for the use of its report relating to any financial statements or information obtained by the Buyer pursuant to this section in any SEC filings by or on behalf of the Buyer.

10.5 The Seller shall reasonably cooperate with the Buyer, and shall cause its affiliates to reasonably cooperate with the Buyer, with respect to any financial statements (including pro forma financial statements) that the Buyer deems reasonably necessary in order to make any filing required by applicable Law or stock exchange requirements, including filings with the SEC.

10.6 The Seller's obligations under this clause 10 are in all aspects subject to restrictions and limitations of applicable law and stock exchange regulation of the Seller. The principles of clause 2.2.2 last sentence equally apply to information disclosed pursuant to this clause 10.

10.7 Information etc. pursuant to this clause 10 are disclosed from the Seller to the Buyer free of charge, if such information in any case are to be prepared by the Seller to comply with applicable law and stock exchange regulation of the Seller. To the extent the Seller is to prepare additional information to satisfy the terms of this clause 10, the Buyer will be obliged to compensate the Seller for any reasonable related costs.

11 Expenses

11.1 Each Party shall bear its own costs and expenses, including but not limited to fees to legal, financial and other advisors and representatives, in relation to the negotiation, preparation, execution and carrying into effect of this Agreement and other agreements referred to hereby.

12 Confidentiality

12.1 Pursuant to Section 13 b of the Danish Bankruptcy Act the terms of this Agreement will to a certain extent be disclosed to the creditors of the Seller in the Restructuring Proposal (unless the restructuring process is terminated pursuant to Clause 8.1(b) without presenting a Restructuring Proposal to the creditors). Such information shall not be considered confidential information. The Seller shall provide the Buyer with a copy of the Restructuring Proposal at the same time as such proposal is sent to the creditors of the Seller to allow the Buyer immediately to send out a stock exchange announcement, if and as required.

12.2 Except from information disclosed in the Restructuring Proposal, the Parties agree that this Agreement and all information related to the transfer is to be handled in confidence and shall not be disclosed to any third party without the prior written consent of the other Party, cf. however Clause 13, unless required by law or applicable stock exchange regulation.

13 VAT

13.1 The Parties agree that the Asset Transfer is a transfer of an existing business, and hence VAT exempt. Consequently, the Seller may notify the transfer to the Danish tax authorities, stating the name and address of the Buyer and the purchase price for the Assets in accordance with Section 1(5) of the Danish VAT Act (in Danish: “momsloven”).

13.2 If the Danish Tax Authority finds that the transfer is subject to VAT, the Buyer pays VAT on the transfer.

13.3 The Buyer takes over any VAT adjustment obligations (in Danish: “momsreguleringsforpligtelser”) on the Assets without this giving rise to a deduction in the purchase price and signs all documents necessary in relation hereto.

14 Severability

14.1 Should one or more of the provisions of this Agreement cease to apply or be modified as a result of invalidity, voidability or for other reason, this shall not affect the validity of the remaining provisions of this Agreement.

14.2 If one or more of the provisions of this Agreement are held to be contrary to Danish Law, the Parties agree that such provision(s) shall, to the extent possible, be modified and shall apply with such contents as may be validly agreed (seeking to maintain as much of the original intentions as possible) and that the remaining provisions of this Agreement shall still apply.

15 No assignment

15.1 Except as otherwise specifically set forth in this Agreement, this Agreement and any right or obligation hereunder may not be assigned, in whole or in part, by any Party without the consent of the other Party.

16 Waiver and amendments

16.1 This Agreement may be amended and the terms hereof may be waived only by written instrument signed by the Parties or in the case of a waiver, by the Party waiving its rights under this Agreement.

17 Headings

17.1 The headings inserted are for convenience and reference only and shall not be used to construe or interpret this Agreement.

18 Interpretation

18.1 The Parties have participated jointly in the negotiation and drafting of this Agreement. In the event of any ambiguity or question of intent or interpretation arises, this Agreement shall be construed as if drafted jointly by the Parties and no presumption or burden of proof shall arise favouring or disfavouring any Party by virtue of the authorship of any of the provisions of this Agreement.

19 Guarantee from Parent

19.1 The Parent, as a primary obligor, guarantees in all aspects the Buyer's obligations under clauses 2.3, 3, 4 and 13, but not other obligations of the Buyer under this Agreement.

20 Governing Law and Venue

20.1 This Agreement shall be governed by, and shall be construed in accordance with, the laws of Denmark, excluding its rules on choice of law.

20.2 Any dispute in connection with this Agreement shall be finally settled by arbitration in accordance with the Rules of Procedure of the Danish Institute of Arbitration (Danish Arbitration) and the Danish Arbitration Act (in Danish: voldgiftsloven). The place of the arbitration shall be Copenhagen and the language of the arbitration shall be English. The arbitration tribunal shall be composed of three arbitrators. The Buyer and the Seller shall each appoint one arbitrator. The Danish Institute of Arbitration shall appoint the third arbitrator, who shall be the chairman of the arbitration tribunal.

20.3 The arbitration award and the arbitration proceedings are confidential, and the Parties must not make any statements to the public as to the dispute, its outcome or the arbitration proceedings, save for as provided in Clause 12.

-oOo-

[Signatures on separate pages]

For Orphazyme A/S in restructuring as the Seller:

/s/ Anders Fink Vadsholt
Name: Anders Vadsholt

/s/ Georges Gemayel
Name: Georges Gemayel

Approved by the reconstructor:

/s/ John Sommer Schmidt
Name: John Sommer Schmidt

Gorrissen Federspiel

For KemPharm Denmark A/S as the Buyer:

/s/ Travis C. Mickle
Name: Travis C. Mickle, Ph.D.

/s/ R. LaDuane Clifton
Name: R. LaDuane Clifton, CPA

For KemPharm Inc as Parent pursuant to clause 19:

/s/ Travis C. Mickle
Name: Travis C. Mickle, Ph.D.

/s/ R. LaDuane Clifton
Name: R. LaDuane Clifton, CPA

KemPharm Announces Strategic Acquisition of Arimoclomol from Orphazyme, Expanding its Rare CNS Diseases Pipeline

Arimoclomol is an NDA-stage, revenue-generating investigational drug candidate being developed for the treatment of Niemann-Pick disease type C (NPC), a rare progressive neurodegenerative disease

KemPharm plans to refile the New Drug Application (NDA) for arimoclomol in NPC with the U.S. Food and Drug Administration (FDA) as early as the First Quarter of 2023

Conference call and live audio webcast with slide presentation is scheduled for tomorrow, May 16, 2022, at 8:30 a.m., EDT

Celebration, FL – May 15, 2022 – KemPharm, Inc. (NasdaqGS: KMPH) (KemPharm, or the Company), a specialty pharmaceutical company focused on the discovery and development of novel treatments for rare central nervous system (CNS) diseases, today announced a definitive agreement with Orphazyme A/S (in reconstruction) (ORPHA.CO; ORPH) (Orphazyme) to acquire arimoclomol, an orally-delivered, first-in-class heat shock protein (HSP) amplifier being developed as a treatment for Niemann-Pick disease type C (NPC). NPC is a rare progressive neurodegenerative disease that impacts children, adolescents, and adults, and is characterized by an inability of the body to transport cholesterol and lipids inside of cells, which leads to the abnormal accumulation of these substances within various tissues of the body, including the brain. Arimoclomol is currently being made available to NPC patients in the U.S., France and Germany under Orphazyme's Early Access Programs (EAP).

Under the terms of the agreement, KemPharm will purchase substantially all of the assets and operations of Orphazyme, including arimoclomol, for a cash payment of USD $12.8 million. The Company expects to finance the cash payment with a revolving line of credit secured by KemPharm's balance sheet. KemPharm intends to retain the majority of Orphazyme's current employees. In addition, KemPharm has agreed to assume an estimated reserve liability equal to approximately USD $5.2 million, which is an estimated future rebate due to the French regulatory authorities based on the revenue generated from the EAP in France. For the year ending December 31, 2022, the EAP is expected to generate at least USD $12 million in revenue based upon enrollment in France as of March 2022. The EAP is expected to remain in place until arimoclomol becomes commercially available in each of the current EAP markets. The transaction is expected to close on or before June 1, 2022, subject to customary closing conditions and approval by Orphazyme's creditors and the Danish bankruptcy court. Canaccord Genuity LLC acted as a strategic advisor to KemPharm for the transaction.

"This strategic acquisition of arimoclomol is a transformative event that significantly expands our rare CNS disease development pipeline, bringing to KemPharm an NDA-stage, revenue-generating product upon which we intend to build commercial capabilities that allow KemPharm to create and retain value for the benefit of shareholders," stated Richard Pascoe, Executive Chairman of KemPharm. "Moreover, the financial structure of the acquisition combined with the revenue currently being generated by arimoclomol from the early access program in France affords us the opportunity to acquire the asset in a capital efficient manner that has the potential to create positive cash flow, while incurring no shareholder dilution."

Arimoclomol is administered orally and has been studied in ten Phase 1, four Phase 2, and three pivotal Phase 2/3 trials. Arimoclomol has received Orphan Drug Designation (ODD) for NPC in the United States and the European Union. Arimoclomol has received Fast-Track Designation (FTD), Breakthrough Therapy Designation (BTD), and Rare Pediatric Disease Designation (RPDD) from the FDA for NPC. If approved in the U.S., arimoclomol would also be eligible to receive a Pediatric Priority Review Voucher. On June 17, 2021, Orphazyme received a Complete Response Letter (CRL) from the FDA regarding its NDA for arimoclomol for the treatment of NPC. Orphazyme also withdrew its European Marketing Authorisation Application (MAA) for arimoclomol for the treatment of NPC ahead of a final vote and opinion by the Committee for Medicinal Products for Human Use (CHMP).

“The acquisition of arimoclomol aligns perfectly with our strategy to build KemPharm’s value via the advancement and commercialization of novel treatments that address rare CNS conditions, including our lead clinical candidate, KP1077 in idiopathic hypersomnia,” stated Travis Mickle, Ph.D., President and Chief Executive Officer of KemPharm. “We have carefully evaluated the CRL issued by the FDA and the minutes from the subsequent Type A meeting, as well as the data that has been generated from the development work performed to date. We believe the efficacy signal for arimoclomol in NPC is convincing and that there is a viable regulatory path that could enable a successful NDA resubmission. KemPharm has significant experience with challenging regulatory situations, having successfully led or participated in three FDA product approvals, two of which followed an initial CRL. We welcome the opportunity to work with the FDA on the resubmission of the NDA for arimoclomol in NPC, which we expect to file as early as the first quarter of 2023.”

NPC is a rare progressive lysosomal storage disorder characterized by an inability of the body to transport cholesterol and lipids inside of cells. This leads to dysfunction in organs such as the brain, spleen and liver. NPC can range from a fatal disorder within the first few months after birth (neonatal period) to a late onset, chronic progressive disorder that remains undiagnosed well into adulthood. Disease progression is irreversible in all patients, and loss of neuro-cognitive function adversely impacts their daily life. The mean age of death is 13 years (Bianconi, 2019), and there are no approved treatments for NPC in the United States.

“NPC is an ultra-rare, inherited neurodegenerative disease that affects people of all ages from infancy to adulthood, and leads to progressive impairment of mobility, cognition, speech, and swallowing, culminating in premature death,” said Marc Patterson, MD, Professor of Neurology, Pediatrics and Medical Genetics at Mayo Clinic. “Therapies to treat NPC are desperately needed, and there is hope that a treatment such as arimoclomol could provide a solution to patients around the world who are living daily with the disease. It is encouraging that there is an opportunity to continue the regulatory process for arimoclomol with the FDA.”

Conference Call Information:

KemPharm will host a conference call and live audio webcast with a slide presentation today at 8:30 a.m., EDT. Interested participants and investors may access the conference call by dialing either:

- (833) 793-7231 (U.S.)
- (614) 999-1675 (international)
- Conference ID: 7880862

An audio webcast with slide presentation will be accessible via the Investor Relations section of the Company's website, http://investors.kempharm.com/. An archive of the webcast and presentation will be available for 90 days beginning at approximately 9:30 a.m. EDT, on May 16, 2022.

About KemPharm:

KemPharm is a specialty pharmaceutical company focused on the discovery and development of novel treatments for rare central nervous system (CNS) diseases through its proprietary LAT® (Ligand Activated Therapy) platform technology. KemPharm utilizes its proprietary LAT® platform technology to generate improved prodrug versions of FDA-approved drugs as well as to generate prodrug versions of existing compounds that may have applications for new disease indications. KemPharm's prodrug product candidate pipeline is focused on the high need areas of idiopathic hypersomnia (IH) and other CNS/rare diseases. In addition, the U.S. Food and Drug Administration (FDA) has approved AZSTARYS®, a new once-daily treatment for ADHD in patients age six years and older containing KemPharm's prodrug, serdexmethylphenidate (SDX), which is being commercialized by Corium, Inc. in the U.S., and APADAZ®, an immediate-release combination product containing benzhydrocodone, KemPharm's prodrug of hydrocodone, and acetaminophen, which is being commercialized by KVK-Tech, Inc. in the U.S. For more information on KemPharm and its pipeline of prodrug product candidates visit www.kempharm.com or connect with us on Twitter, LinkedIn, Facebook and YouTube.

Caution Concerning Forward Looking Statements:

This press release may contain forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995. Forward-looking statements include all statements that do not relate solely to historical or current facts, including without limitation and which can be identified by the use of words such as "may," "will," "expect," "project," "estimate," "anticipate," "plan," "believe," "potential," "should," "continue," "could," "intend," "target," "predict," or the negative versions of those words or other comparable words or expressions, although not all forward-looking statements contain these identifying words or expressions. Forward-looking statements are not guarantees of future actions or performance. These forward-looking statements include statements regarding the expected closing of KemPharm's acquisition of arimoclomol, including the timing and financing thereof, the acquisition's impact on KemPharm's operations and financial results, the expected revenue from the EAP and the timing or results of an NDA resubmission for arimoclomol. These forward-looking statements are based on information currently available to KemPharm and its current plans or expectations and are subject to a number of known and unknown uncertainties, risks and other important factors that may cause our actual results, performance or achievements to be materially different from any future results, performance or achievements expressed or implied by the forward-looking statements. These and other important factors are described in detail in the "Risk Factors" section of KemPharm's Annual Report on Form 10-K for the year ended December 31, 2021, as updated by the Quarterly Report on Form 10-Q for the three months ended March 31, 2022, and KemPharm's other filings with the Securities and Exchange Commission.

While we may elect to update such forward-looking statements at some point in the future, except as required by law, we disclaim any obligation to do so, even if subsequent events cause our views to change. Although we believe the expectations reflected in such forward-looking statements are reasonable, we can give no assurance that such expectations will prove to be correct. These forward-looking statements should not be relied upon as representing our views as of any date subsequent to the date of this press release.

This press release also may contain estimates and other statistical data made by independent parties and by us relating to market size and other data about our industry. This data involves a number of assumptions and limitations, and you are cautioned not to give undue weight to such estimates. In addition, projections, assumptions and estimates of our future performance and the future performance of the markets in which we operate are necessarily subject to a high degree of uncertainty and risk.

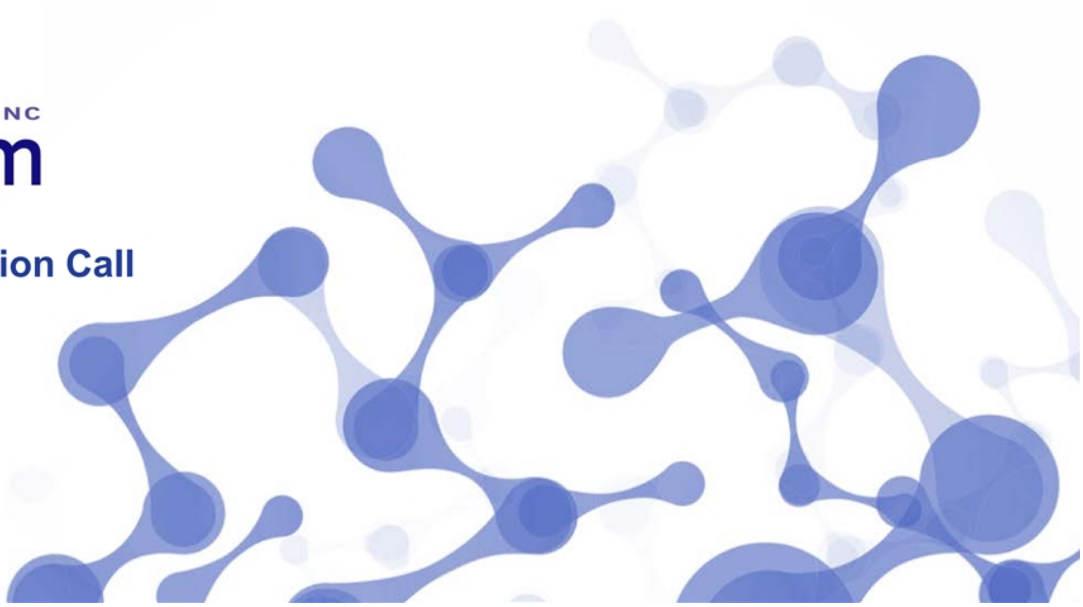
KemPharm Contacts:

Tiberend Strategic Advisors, Inc.
Jason Rando/Daniel Kontoh-Boateng
jrando@tiberend.com
dboateng@tiberend.com

KemPharm INC

Arimoclomol Acquisition Call

May 16, 2022



Cautionary Note Regarding Presentation Information

This presentation may contain forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995. Forward-looking statements include all statements that do not relate solely to historical or current facts, including without limitation and can be identified by the use of words such as “may,” “will,” “expect,” “project,” “estimate,” “anticipate,” “plan,” “believe,” “potential,” “should,” “continue,” “could,” “intend,” “target,” “predict,” or the negative versions of those words or other comparable words or expressions, although not all forward-looking statements contain these identifying words or expressions. Forward-looking statements are not guarantees of future actions or performance. These forward-looking statements include statements regarding the expected closing of KemPharm’s acquisition of arimoclomol, including the timing and financing thereof, the acquisition’s impact on KemPharm’s operations and financial results, the expected revenue from the EAP and the timing or results of an NDA resubmission for arimoclomol. These forward-looking statements are based on information currently available to KemPharm and its current plans or expectations and are subject to a number of known and unknown uncertainties, risks and other important factors that may cause our actual results, performance or achievements to be materially different from any future results, performance or achievements expressed or implied by the forward-looking statements. These and other important factors are described in detail in the “Risk Factors” section of KemPharm’s Annual Report on Form 10-K for the year ended December 31, 2021, as updated by the Quarterly Report on Form 10-Q for the three months ended March 31, 2022, and KemPharm’s other filings with the Securities and Exchange Commission.

While we may elect to update such forward-looking statements at some point in the future, except as required by law, we disclaim any obligation to do so, even if subsequent events cause our views to change. Although we believe the expectations reflected in such forward-looking statements are reasonable, we can give no assurance that such expectations will prove to be correct. These forward-looking statements should not be relied upon as representing our views as of any date subsequent to this presentation.

This presentation also may contain estimates and other statistical data made by independent parties and by us relating to market size and other data about our industry. This data involves a number of assumptions and limitations, and you are cautioned not to give undue weight to such estimates. In addition, projections, assumptions and estimates of our future performance and the future performance of the markets in which we operate are necessarily subject to a high degree of uncertainty and risk.

KEMPHARM VALUE PROPOSITION

Innovative pharmaceutical company discovering and developing novel treatments for rare CNS and neurodegenerative diseases

Focus on high-value areas with significant unmet needs with potential to internally commercialize

Two FDA approved and partnered medications, AZSTARYS® and APADAZ®, validate approach and regulatory expertise

Arimoclomol Acquisition - Expanding Pipeline Targeting Rare Diseases

Definitive agreement with Orphazyme A/S to acquire substantially all assets, including arimoclomol

- This acquisition is a significant expansion of KemPharm's development pipeline targeting rare diseases; allows KemPharm to leverage prior experience with challenging regulatory situations
  - Aligns with strategy to build value through the development and commercialization of novel treatments for rare diseases
- Arimoclomol is an NDA-stage, revenue-generating investigational drug candidate being developed for the treatment of Niemann-Pick disease type C (NPC), an ultra-rare progressive, disabling and fatal lysosomal storage disorder
  - Arimoclomol is currently available to NPC patients in the U.S., France, Germany and other European countries under early access programs (EAPs)
  - No approved treatments exist in the U.S. for NPC
- Favorable acquisition terms: "capital efficient" financial structure with potential for positive cash flow and no shareholder dilution
  - USD $12.8M cash payment for substantially all assets and operations of Orphazyme; French EAP is expected to generate revenue in excess of USD $12M in FY 2022

Definitive Agreement – Summary of Terms

- KemPharm will purchase substantially all of the assets and operations of Orphazyme, including arimoclomol, with a cash payment of USD $12.8 million
  - Cash payment will be financed with a revolving line of credit secured by KemPharm's balance sheet
- KemPharm will assume estimated reserve liabilities of USD $5.2 million which is an estimated future rebate due to the French regulatory authorities based on revenue generated from the EAP in France
  - **EAP for arimoclomol in France expected to generate revenue in excess of $12 million (USD) in FY 2022 based upon the actual Q1 2022 enrollment**
  - EAP expected to remain in place until arimoclomol becomes available commercially in France
- KemPharm intends to retain the majority of Orphazyme's current employees and continue operations through a new subsidiary in Denmark
- Transaction expected to close on or before June 1, 2022, subject to final approval by Orphazyme's creditors and the Danish bankruptcy court

Product Overview - Arimoclomol

Disease State, Market Overview and Regulatory Pathway

About Niemann-Pick Disease Type C (NPC) [1]

- **Ultra-rare progressive lysosomal storage disorder** characterized by an inability of the body to transport cellular cholesterol and lipids
  - Leads to dysfunction in organs such as the brain, spleen and liver
  - **Disease progression is irreversible in all patients and ultimately fatal**
  - Loss of neuro-cognitive function adversely impacts the daily lives of patients
- Most cases are detected during childhood and progress to cause life-threatening complications
  - NPC can range from a fatal disorder within the first few months after birth (neonatal period), to a late onset, chronic progressive disorder that remains undiagnosed well into adulthood
  - NPC is estimated to occur in 1 in 100,000-120,000 live births
  - **The mean age of death in NPC patients is 13 years[2]**
  - Estimated 1,800 patients in the U.S. and Europe
- **No approved treatments exist in the U.S. for NPC**
  - In Europe, there is only one treatment available, miglustat

Source: (1) https://rarediseases.org/
(2) Bianconi, 2019

Arimoclomol – Innovative Product for a High Unmet Need

- First-in-class, oral treatment intended for NPC
  - Capsule formulation designed to be swallowed whole, opened to allow contents to be mixed with soft foods/liquids or delivered through a gastric feeding tube
  - Nonclinical and clinical evidence demonstrated improved lysosomal and cellular function with arimoclomol treatment
- Studied in ten Phase 1, four Phase 2, and three Phase 2/3 trials in various rare diseases
  - Positive efficacy results from NPC trial (NPC-002)
  - Positive results from a Phase 2 trial in Gaucher's Disease (GD), a related lysosomal storage disorder
  - Safety data has been collected from more than 500 individuals for up to 5 years of treatment with no significant safety findings identified to date
- Received **Orphan Drug Designation** for NPC in the U.S. and EU; and **Fast-Track Designation**, **Breakthrough Therapy Designation**, and **Rare Pediatric Disease Designation** from the FDA for NPC
  - ***Eligible to receive Rare Pediatric Disease Priority Review Voucher if approved by the FDA***
  - Eligible to receive New Chemical Entity (NCE) and Orphan Drug Exclusivity
  - Patents and patent applications, if issued, could extend exclusivity through 2040

Overview of Regulatory Pathway in the U.S. – NDA Resubmission Process

- Orphazyme received a Complete Response Letter (CRL) from the FDA on Jun 17, 2021, regarding their NDA for arimoclomol for the treatment of NPC
  - The FDA identified three issues:
    1) Additional evidence needed to substantiate validity of the primary endpoint used in the single efficacy trial
    2) Required additional analysis related to how missing data is handled for statistical analysis
    3) Required additional support and data related to confirmatory evidence of efficacy
  - **The FDA did not request additional efficacy data in the CRL.**
- Type A End-of-Review Meeting was held on Oct 13, 2021:
  - *FDA agreed* to allow a reanalysis of the primary endpoint removing the cognition domain
  - *FDA agreed* to a rescoring and a reassessment of the swallowing domain including a qualitative study to further validate that domain
  - *FDA agreed* to further discussion of how best to handle missing data; no consensus was obtained at the meeting
  - Additional confirmatory evidence was provided to the FDA at the meeting; further confirmation from FDA as to the impact of those studies will be important

FDA Agreed with Proposed Plan to Strengthen Primary Endpoint and Analysis

1) Conduct a new analysis of the original 5-domain NPCCSS primary endpoint by removing the cognition domain (ambulation, swallowing, speech and fine motor skills remain) to form a new 4-domain NPCCSS endpoint
   - ✓ FDA confirmed that this is acceptable
2) Conduct a qualitative study to assess the validity and robustness of the swallowing domain, the scoring used in the trial and the clinical relevance
   - ✓ FDA confirmed this is acceptable and has reviewed and provided comments on the protocol
   - ✓ Study complete and should address the FDA's issue
3) Use the log ratio transformed analysis to address the FDA's concerns regarding missing data
   - No direct commitment from the FDA
   - Still needs discussed in detail and alternatives should be explored

Ultimately, using the prespecified analysis in the SAP with the original 5-domain NPCCSS endpoint met statistical significance and study *NPC-002 WAS SUCCESSFUL*.

- Agency's methodology was post-hoc and **NOT** prespecified; other statistical methods may be more appropriate **including the prespecified analysis originally agreed upon with the FDA**
- Missing data discussion is a common issue for sponsors with the FDA, especially in rare diseases

Modification of Primary Endpoint with FDA Agreement Demonstrates Improved Statistical Significance and Treatment Effect

Analysis	Full Population[1]		Migulstat Subgroup[1]	
	Treatment Effect[2]	p-value	Treatment Effect[2]	p-value
Prespecified 5-domain NPCCSS	-1.4	0.0456	-2.06	0.0060
Prespecified 4-domain NPCCSS	-1.54	0.0193	ND	ND
FDA post-hoc 5-domain NPCCSS	-1.24	0.1093	-2.23	0.0031
FDA post-hoc 4-domain NPCCSS	-1.33	0.0602	-2.23	0.0018
4-domain NPCCSS and potential swallowing rescore[3] (FDA post-hoc)	-1.70 (-1.48)	0.0150 (0.0470)	ND	ND

1. Analysis conducted as full population and migulstat subgroup. Miglustat was used as the Standard of Care treatment in roughly 80% of all patients including placebo. 5-domain NPCCSS is based on a scale from 0 to 25 broken up in five groups of 0 to 5 with zero considered normal and 5 the worst symptom for each domain. Domains include ambulation, speech, swallowing, fine motor skills and cognition. The 4-domain analysis removes cognition for a total potential score of 20.
2. Treatment effect as measured as a change from baseline. A decrease in score is considered an improvement and a change in score of one (or negative one) is considered clinically meaningful.
3. Analysis discussed with FDA at Type A meeting but not agreed upon as swallowing analysis and potential rescoring was not complete.

ND designates not determined

Bolstering Confirmatory Evidence Addresses Another Key Issue Raised in CRL

- **Since the CRL and Type A meeting, additional data has already been generated**
  - Numerous studies and additional analysis has been conducted to address this issue
  - Primary focus was to confirm or elucidate the mechanism of action, the potential beneficial effect of migulstat, and to further support the clinical data
  - Some of this new data includes:
    - ✓ Biomarkers
    - ✓ Results from open-label extension arms
    - ✓ Data from expanded access programs
    - ✓ In vitro studies
    - ✓ In vivo models of NPC
- **Totality of the evidence provided appears to support the clinical outcome**
  - Now that there is a sizable amount of new data available, FDA input will be sought to verify it is sufficient to address their issue

Path to Resubmission and Approval Appears Straightforward

- Current plan to address the major issues related to the CRL appear addressable
  - ✓ Primary endpoint work has concluded
  - ✓ Additional analyses have been conducted
  - ✓ Confirmatory evidence has been significantly augmented
- KemPharm has significant experience with challenging regulatory situations, including two FDA product approvals that followed initial CRLs and dealing with the FDA and statistical issues that can occur in clinical trials
  - Based on our experience and assessment of regulatory situation, we believe there is a viable pathway that could enable a successful NDA resubmission and subsequent approval for arimoclomol in NPC
    - Path may include additional non-clinical or clinical studies, though it is not expected that any long-term or efficacy trials would be needed
    - Federal Dispute Resolution Request (FDRR) may be utilized, if necessary
    - An advisory committee (ad com) may also be required by FDA after resubmission
- **We expect to resubmit the NDA for arimoclomol in NPC as early as Q1 2023**

Commercial Opportunity

Potential to Transform KemPharm's Business Model

Arimoclomol Market Opportunity Is Compelling, Even With Ultra-Rare Status

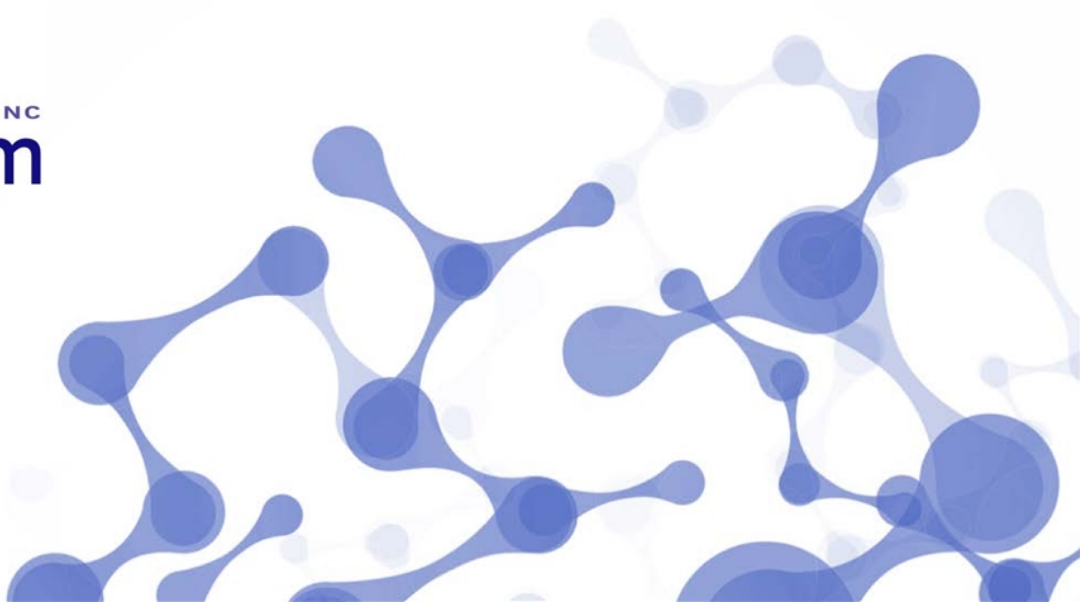
- Upon approval, KemPharm would currently be eligible to receive a Rare Pediatric Disease Priority Review Voucher
  - Last voucher sold for $110M, which was within the typical range of approx. $100M/voucher
  - Program could eventually end, making these vouchers more scarce
- Arimoclomol is already generating revenue through the French EAP system
  - 34 patients in the French EAP as of Mar 31, 2022
  - French program is the only system that reimburses for treatment prior to formal approval; rate is set by the Sponsor
  - Program typically remains in place while therapy is moving towards a marketing application and potential French and/or European approval
- **Global EAP programs represent the potential first adopters of arimoclomol post-approval**
  - Currently there are 151 global participants, with enrollment rising
  - As of Mar 31, 2022: 67 patients in U.S., 41 patients Germany, 34 patients in France, and 9 patients in other countries (Denmark, Switzerland and UK)

Arimoclomol Provides KemPharm the Opportunity to Commercialize and Retain Full Market Value

- Arimoclomol is an NDA-stage, revenue-generating product upon which we intend to build commercial capabilities that fit with the goal of allowing KemPharm to create and retain value for the benefit of shareholders
- **Arimoclomol represents an opportunity for KemPharm to launch with a small, focused commercialization effort that can be foundation for future rare products, including KP1077**
  - Typically, ultra-rare disease commercial teams are less than 20 individuals which can be expanded as additional products are approved
  - Lower marketing spend since population is well defined and physicians are usually primarily in treatment centers
  - Patient advocacy groups and relationships with treatment centers are also key drivers
  - Establishes a commercial platform that can be leveraged with other products, including KP1077
- **Arimoclomol represents a global market opportunity**
  - Current patients enrolled in the EAPs in U.S., France, Germany, and other European countries expected to transition directly to commercial once approved within each market
  - Partnerships/licensing opportunities may be available in other markets (Japan, China, others)

Full Pipeline of Product Candidates with Substantial Milestones in 2022 and Beyond

KemPharm INC



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