Zevra Therapeutics to Acquire Acer Therapeutics, Expanding its Rare Disease Portfolio and Adding Commercial Product

August 31, 2023

Proposed acquisition of Acer for $15M in Zevra stock plus Contingent Value Rights (CVRs) and Zevra's purchase of Acer's secured debt in capital efficient structure

Zevra to assume commercialization efforts of OLPRUVA™, recently approved for the treatment of urea cycle disorders (UCDs)

FDA-approved commercial asset expected to increase and diversify Zevra's revenues

OLPRUVA™commercial operations provide Zevra scale and cost synergies to complement the potential launch of arimoclomol

Adds EDSIVO™, a Phase 3 program for vascular Ehlers-Danlos Syndrome (vEDS), to Zevra's rare disease clinical pipeline

Zevra to discuss details during conference call today, at 8:30 a.m. ET

CELEBRATION, Fla. and NEWTON, Mass., Aug. 31, 2023 (GLOBE NEWSWIRE) -- Zevra Therapeutics, Inc. (NasdaqGS: ZVRA) ("Zevra"), a rare disease company melding science, data and patient need to create transformational therapies for diseases with limited or no treatment options, and Acer Therapeutics Inc. (Nasdaq: ACER) ("Acer"), a pharmaceutical company focused on development and commercialization of therapies for rare and life-threatening diseases, today announced the companies have entered into a definitive agreement pursuant to which Zevra would acquire Acer in a merger transaction having a total potential value for Acer stockholders of up to $91 million, consisting of (i) approximately 2.96 million shares of Zevra common stock valued at $15 million, or 0.121 shares of Zevra's common stock per share of Acer common stock based on the volume weighted average trading price (VWAP) of shares of Zevra's common stock during the 20 consecutive trading days ending on the trading date prior to today, and (ii) up to an additional $76 million in a series of potential cash payments pursuant to non-transferable Contingent Value Rights (CVRs) upon achievement of certain commercial and regulatory milestones for Acer's OLPRUVA (sodium phenylbutyrate) and Acer's EDSIVO (celiprol) within specified time periods. Certain additional cash payments are also possible pursuant to the CVRs with respect to milestones involving Acer's early-stage program ACER-2820 (emetine), as described further below. Zevra has also purchased Acer's secured debt at a discount from Nantahala Capital (Nantahala) through a series of transactions in capital efficient structure. In addition, Zevra has agreed to provide Acer with a bridge loan facility for up to $16.5 million, subject to certain terms and conditions. Both companies are deeply committed to developing and commercializing treatments for rare diseases with a strong focus on patients and remain dedicated to supporting communities with little or no existing therapeutic options. The merger is expected to expand Zevra's rare disease portfolio, as well as increase and diversify its revenues with the addition of a U.S. commercial asset, OLPRUVA, indicated for the treatment of UCDs. The transaction is subject to certain customary closing conditions, including, but not limited to, approval by Acer's stockholders.

"We believe that Acer's portfolio of rare disease programs, including the recent U.S. commercial approval of OLPRUVA for UCDs, is a perfect strategic fit for Zevra and creates significant opportunity for us to positively impact patient lives while creating shareholder value," said Joshua Schafer, Chief Commercialization Officer and Executive Vice President of Business Development of Zevra Therapeutics. "We are excited about Acer's clinical programs and are confident in the potential of OLPRUVA to bring UCD patients a more convenient and cost-effective treatment option over current therapies. Acer would bring unique rare disease operations and capabilities that would serve as a foundation to support the commercialization of Zevra's pipeline as it advances."

Chris Schelling, Acer Therapeutics' Chief Executive Officer and Founder, added, "Following years of product development and commitment to rare disease communities, culminating in the FDA approval of OLPRUVA, we are pleased to see our assets, pipeline and team positioned to unite under the Zevra umbrella. We look forward to working with the Zevra team to ensure a smooth transition as we work together on behalf of patients."

"This merger would support Zevra's vision of becoming a leading rare disease company bringing life-changing therapies to patients with a significant unmet need," said Christal Mickle, Zevra's interim Chief Executive Officer and Chief Development Officer. "The commercial launch of OLPRUVA in the U.S. requires a small, highly-focused commercial team, which is complementary to what we intend to build for arimoclomol, our product candidate for the treatment of Niemann-Pick disease Type C (NPC). We believe there is potential to realize significant synergies across our commercial organizations as both UCDs and NPC are metabolic related conditions and there is overlap among those physicians that treat both disorders."

Financial Details and Terms of the Transactions:
The transactions, which have been approved by the Boards of Directors of both companies, are currently anticipated to close in the fourth quarter of 2023, subject to Acer stockholder approval, as well as other customary closing conditions. The merger is

expected to accelerate Zevra's pathway to becoming a commercial-stage company by adding OLPRUVA, an FDA-approved asset, which is expected to add to Zevra's revenue. There are potential synergies to be realized by combining Acer's operations with Zevra's capabilities in preparation for the potential launch of arimoclomol. In addition, Zevra expects to acquire significant net operating loss (NOL) tax assets as part of this merger, providing potential tax savings against future earnings.

Under the terms of the definitive agreement, at closing, Zevra would issue 0.121 of a share of Zevra's common stock in respect of each share of Acer's common stock. In addition, Acer stockholders of record as of immediately prior to the effective time of the merger would receive non-transferable CVRs entitling the holders to receive up to $34 million in cash upon the achievement of certain commercial milestones for OLPRUVA, and up to an additional $42 million in cash upon the achievement of certain regulatory milestones for OLPRUVA and EDSIVO.

Approximately 2.96 million shares of Zevra common stock to be issued in the merger is calculated by dividing $15.0 million by the VWAP of Zevra's shares of common stock during the 20 consecutive trading days through yesterday, which was $5.0667. The 20-day trailing VWAP value represents a discount of approximately 2% to yesterday's closing share price for Acer.

The non-transferable CVRs will entitle the Acer stockholders of record to receive up to $34 million in cash upon the achievement of certain commercial milestones for OLPRUVA, and an additional $42 million in cash upon the achievement of certain regulatory milestones for other development programs. The proposed transactions also include non-transferable CVRs for ACER-2820, Acer's early phase emetine program.

Based on the number of Zevra shares issued and outstanding as of June 30, 2023, together with the equity issued to Nantahala as part of the debt acquisition as described below, the aggregate number of shares issuable to Acer stockholders in the merger is expected to represent approximately 7.6% of the issued and outstanding common stock of Zevra following the merger.

To provide for a smooth transition and uninterrupted operations, and subject to certain conditions, Zevra has extended a bridge loan facility to Acer of up to $16.5 million to provide additional working capital to, among other things, support the commercial launch of OLPRUVA until the expected closing of the merger transaction, and to provide the $10 million payment to Acer's termination of the 2021 collaboration and license agreement by and between Acer and Relief Therapeutics, and Acer's related entry into an exclusive license agreement with Relief for the development and commercialization rights for OLPRUVA in geographical Europe.

Additionally, Zevra has purchased Acer's secured debt from Nantahala representing an aggregate of principal, accrued interest other fees and premiums of approximately $35.3 million, for a total of $28.5 million to be paid using a combination of $12 million in cash financed from Zevra's existing margin line of credit facility, $5 million in a new promissory note held by Nantahala, with a three-year maturity and bearing interest initially at 9% per annum (increasing to 12% per annum if the note remains outstanding six months after issuance), and $11.5 million in Zevra's common stock based on the 20-day trailing VWAP calculation described above, or approximately 2.27 million shares, or approximately 5.8% of the issued and outstanding common stock of Zevra following the merger.

Bryan Cave Leighton Paisner LLP served as legal advisor to Zevra and Canaccord Genuity LLC served as exclusive financial advisor to Zevra for the transactions. Pillsbury Winthrop Shaw Pittman LLP served as legal advisor to Acer, and William Blair & Company, LLC served as exclusive financial advisor to Acer.

Conference Call Information:
Zevra will host a conference call and live audio webcast with a slide presentation today, August 31, 2023, at 8:30 a.m. ET, to discuss details of the acquisition agreement with Acer.

The audio webcast with a slide presentation will be accessible via the Investor Relations section of the Company's website, http://investors.zevra.com/. An archive of the webcast and presentation will be available for 90 days beginning at approximately 9:30 a.m. ET, on August 31, 2023.

Additionally, interested participants and investors may access the conference call by dialing either:

- (800) 245-3047 (U.S.)
- +1 (203) 518-9765 (International)
- Conference ID: ZevraUpdate

About Urea Cycle Disorders:
UCDs are a group of rare, genetic disorders that can cause harmful ammonia to build up in the blood, potentially resulting in brain damage and neurocognitive impairments, if ammonia levels are not controlled.[i] Any increase in ammonia over time is serious. Therefore, it is important to adhere to any dietary protein restrictions and have alternative medication options to help control ammonia levels.

About OLPRUVA:
ACER-001 (sodium phenylbutyrate) was approved for the treatment of certain UCDs in December 2022 and has recently been marketed under the brand name, OLPRUVA. OLPRUVA (sodium phenylbutyrate) for oral suspension is a prescription medicine used along with certain therapy, including changes in diet, for the long-term management of adults and children weighing 44

pounds (20 kg) or greater and with a body surface area (BSA) of 1.2 m^2 or greater, with urea cycle disorders (UCDs), involving deficiencies of carbamylphosphate synthetase (CPS), ornithine transcarbamylase (OTC), or argininosuccinic acid synthetase (AS). Please see Important Safety Information and full Prescribing Information, including Patient Information.

Important Safety Information:

Certain medicines may increase the level of ammonia in your blood or cause serious side effects when taken during treatment with OLPRUVA. Tell your doctor about all the medicines you or your child takes especially if you or your child takes corticosteroids, valproic acid, haloperidol, and/or probenecid.

OLPRUVA can cause serious side effects, including: 1) nervous system problems (neurotoxicity). Symptoms include sleepiness, tiredness, lightheadedness, vomiting, nausea, headache, confusion, 2) low potassium levels in your blood (hypokalemia) and 3) conditions related to swelling (edema). OLPRUVA contains salt (sodium), which can cause swelling from salt and water retention. Tell your doctor right away if you or your child get any of these symptoms. Your doctor may do certain blood tests to check for side effects during treatment with OLPRUVA. If you have certain medical conditions such as heart, liver or kidney problems, are pregnant/planning to get pregnant or breast-feeding, your doctor will decide if OLPRUVA is right for you.

The most common side effects of OLPRUVA include absent or irregular menstrual periods, decreased appetite, body odor, bad taste or avoiding foods you ate prior to getting sick (taste aversion). These are not all of the possible side effects of OLPRUVA. Call your doctor for medical advice about side effects. You may report side effects to FDA at 1-800-FDA-1088.

For additional information, please see Important Safety Information and full Prescribing Information, including Patient Information and discuss with your doctor.

About Niemann-Pick disease type C (NPC):
Niemann-Pick disease type C (NPC) is an ultra-rare and progressive, neurodegenerative lysosomal storage disorder characterized by an inability of the body to transport cholesterol and other lipids within the cell, leading to an accumulation of these substances in various tissue areas, including brain tissue. The disease is caused by mutations in the NPC1 or NPC2 genes which are responsible for making lysosomal proteins and is an autosomal recessive trait. Both children and adults can be affected by NPC with varying clinical presentations. Those living with NPC lose independence due to physical and cognitive limitations, with key neurological impairments presenting in speech, cognition, swallowing, ambulation, and fine motor skills. Disease progression is irreversible and can be fatal within months or take years to be diagnosed and advance in severity.

About Arimoclomol:
Arimoclomol, Zevra's orally-delivered, first-in-class investigational product candidate for the treatment of NPC, has been granted orphan drug designation, Fast Track designation, Breakthrough Therapy designation and rare pediatric disease designation for NPC by the FDA, and orphan medicinal product designation for the treatment of NPC by the European Medicines Agency (EMA). The arimoclomol NDA is currently being prepared for resubmission to the FDA.

About Zevra Therapeutics:
Zevra Therapeutics is a rare disease company melding science, data, and patient need to create transformational therapies for diseases with limited or no treatment options. With unique, data-driven clinical, regulatory, and commercialization strategies, the Company is overcoming complex drug development challenges to bring much-needed therapies to patients. With both regulatory and clinical stage product candidates, the Company is building its commercial capability to make new therapies available to the rare disease community.

Early access programs are made available by Zevra Therapeutics and its affiliates and are subject to the Company's Early Access Program (EAP) policy as published on its website at zevra.com. Participation in these programs is subject to the laws and regulations of each jurisdiction under which each respective program is operated. Eligibility for participation in any such program is at the treating physician's discretion.

About Acer Therapeutics:
Acer is a pharmaceutical company focused on the acquisition, development and commercialization of therapies for serious rare and life-threatening diseases with significant unmet medical needs. In the U.S., OLPRUVA (sodium phenylbutyrate) is approved for the treatment of urea cycle disorders (UCDs) involving deficiencies of carbamylphosphate synthetase (CPS), ornithine transcarbamylase (OTC), or argininosuccinic acid synthetase (AS). Acer is also advancing a pipeline of investigational product candidates for rare and life-threatening diseases, including: OLPRUVA (sodium phenylbutyrate) for treatment of various disorders, including Maple Syrup Urine Disease (MSUD); and EDSIVO (celiprolol) for treatment of vascular Ehlers-Danlos syndrome (vEDS) in patients with a confirmed type III collagen (COL3A1) mutation. For more information, visit www.acertx.com.

Cautionary Note 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, 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 without limitation statements regarding the proposed merger transaction, its timing and its consummation, the anticipated financial performance of

Zevra and Acer related thereto, including the anticipated closing of, benefits of, and synergies related to the proposed merger transaction, potential strategic implications as a result of the proposed merger transaction, Zevra's intention to provide bridge financing to Acer and the availability of such financing to Acer, Zevra's path to profitability, Zevra's strategic and product development objectives, including with respect to becoming a leading, commercially-focused rare disease company, Zevra's plans to build out commercial teams for products or product candidates, Zevra's commercial infrastructure investments and the impact of the proposed transactions on them, Zevra's industry, plans, goals and expectations concerning market position, future operations and other financial and operating information, and the potential for achievement of the milestones that would trigger cash payments pursuant to the CVRs that would be issued to the Acer stockholders in the merger. Forward-looking statements are based on information currently available to Zevra and Acer and their respective current plans or expectations, and are subject to several known and unknown uncertainties, risks, and other important factors that may cause actual results, performance, or achievements to be materially different from any future results, performance, or achievements expressed or implied by the forward-looking statements, including, but not limited to, uncertainties involving the following: the potential timing of the consummation of the proposed merger transaction and the ability of the parties to consummate the proposed merger; the satisfaction of the conditions precedent to consummation of the proposed merger, including the approval of Acer's stockholders; the ability to obtain required regulatory approvals at all or in a timely manner; any litigation related to the proposed transactions; disruption of Acer's or Zevra's current plans and operations as a result of the proposed transactions; the ability of Acer or Zevra to retain and hire key personnel; competitive responses to the proposed transactions; unexpected costs, charges or expenses resulting from the proposed transactions; the ability of Zevra to successfully integrate Acer's operations, products, product candidates and technology; the ability of Zevra to implement its plans, forecasts and other expectations with respect to Acer's business after the completion of the proposed transactions and realize additional opportunities for growth and innovation; the ability of Zevra to realize the anticipated synergies and related benefits from the proposed transactions in the anticipated amounts or within the anticipated timeframes or at all; and the ability to maintain relationships with Zevra's and Acer's respective employees, customers, other business partners and governmental authorities. These and other important factors are described in detail in the "Risk Factors" section of Zevra's and Acer's Annual Reports on Form 10-K for the year ended December 31, 2022, as updated in Zevra's and Acer's Quarterly Reports on Form 10-Q for the quarter ended June 30, 2023, and Zevra's and Acer's other filings with the Securities and Exchange Commission. While either Zevra or Acer may elect to update such forward-looking statements at some point in the future, each party disclaims any obligation to do so, except as required by law, even if subsequent events cause their respective views to change. Although each party believes the expectations reflected in such forward-looking statements are reasonable, neither party can assure that such expectations will prove correct. These forward-looking statements should not be relied upon as representing the views of either Acer as of any date after the date of this press release.

Important Additional Information Regarding the Transactions Will Be Filed With the SEC

In connection with the proposed transactions, Zevra and Acer will file relevant materials with the SEC, including a Zevra registration statement on Form S-4 that will include a proxy statement of Acer and will also constitute a prospectus of Zevra, and a definitive proxy statement will be mailed to stockholders of Acer. INVESTORS AND SECURITY HOLDERS OF ZEVRA AND ACER ARE URGED TO READ THE PROXY STATEMENT/PROSPECTUS THAT WILL BE INCLUDED IN THE REGISTRATION STATEMENT ON FORM S-4, AND OTHER RELEVANT DOCUMENTS FILED OR TO BE FILED WITH THE SEC IN CONNECTION WITH THE PROPOSED TRANSACTIONS OR INCORPORATED BY REFERENCE IN THE PROXY STATEMENT/PROSPECTUS (IF ANY) CAREFULLY AND IN THEIR ENTIRETY WHEN THEY BECOME AVAILABLE BECAUSE THEY WILL CONTAIN IMPORTANT INFORMATION ABOUT THE PROPOSED TRANSACTIONS, THE PARTIES TO THE PROPOSED TRANSACTIONS AND THE RISKS ASSOCIATED WITH THE PROPOSED TRANSACTIONS. Investors and security holders will be able to obtain, without charge, a copy of the registration statement, the proxy statement/prospectus and other relevant documents filed with the SEC (when available) from the SEC's website at www.sec.gov. Copies of the documents filed with the SEC by Zevra will be available free of charge on Zevra's investor relations website at investors.zevra.com under the tab "SEC Filings." Copies of the documents filed with the SEC by Acer will be available free of charge on Acer's investor relations website at www.acertx.com/investor-relations under the tab "SEC Filings."

Participants in the Solicitation

Zevra, Acer and certain of their directors, executive officers and other members of management may be deemed to be participants in the solicitation of proxies with respect to the proposed transactions. Information regarding the persons who may, under the rules of the SEC, be deemed participants in the solicitation of the stockholders of Acer in connection with the proposed transactions, including a description of their direct or indirect interests, by security holdings or otherwise, will be set forth in the proxy statement/prospectus when it is filed with the SEC. Information regarding Zevra's directors and executive officers is contained in Zevra's definitive proxy statement, which was filed with the SEC on March 15, 2023, the definitive proxy statement filed by Daniel J. Mangless, together with the other participants named therein, which was filed with the SEC on March 17, 2023, and Zevra's Current Reports on Form 8-K, filed with the SEC on March 30, 2023, May 8, 2023, May 15, 2023, and August 7, 2023. Information regarding Acer's directors and executive officers is contained in Acer's definitive proxy statement, which was filed with the SEC on April 14, 2023. Security holders and investors may obtain additional information regarding the interests of such persons, which may be different than those of Zevra's security holders generally, by reading the proxy statement/prospectus and other relevant documents regarding the transactions, which will be filed with the SEC. You may obtain these documents (when they become available) free of charge through the website maintained by the SEC at www.sec.gov and Zevra's or Acer's investor relations websites as described above.

No Offer or Solicitation

This communication is not intended to and does not constitute an offer to sell or the solicitation of an offer to subscribe for or buy or an invitation to purchase or subscribe for any securities or the solicitation of any vote or approval in any jurisdiction pursuant to the proposed transactions or otherwise, nor shall there be any sale, issuance or transfer of securities in any jurisdiction in

contravention of applicable law. This communication does not constitute a prospectus or prospectus equivalent document. No offering of securities shall be made except by means of a prospectus meeting the requirements of Section 10 of the Securities Act. In connection with the proposed transactions, Zevra will file a registration statement on Form S-4 that will include a proxy statement of Acer and will also constitute a prospectus of Zevra. INVESTORS AND SECURITY HOLDERS OF ZEVRA AND ACER ARE URGED TO READ THE PROXY STATEMENT/PROSPECTUS AND OTHER DOCUMENTS THAT WILL BE FILED WITH THE SEC CAREFULLY AND IN THEIR ENTIRETY WHEN THEY BECOME AVAILABLE BECAUSE THEY WILL CONTAIN IMPORTANT INFORMATION.

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[i] Ah Mew N, et al. Urea cycle disorders overview [updated June 22, 2017]. In: Adam MP, Ardinger HH, Pagon RA, et al, eds. GeneReviews® [Internet]. University of Washington; 1993-2022. Accessed March 20, 2022.

ZEVRA THERAPEUTICS

Source: Zevra Therapeutics; Acer Therapeutics Inc.