

# **Zevra Investor Call: *Acer Acquisition***

*August 31, 2023*



# Cautionary Note Regarding Forward-Looking Statements



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 promise and potential impact of our preclinical or clinical trial data or Acer Therapeutics Inc.’s (“Acer’s”) clinical trial data, including without limitation the timing and results of any clinical trials or readouts, the consummation and timing of the transaction, our anticipated financial performance, including anticipated closing of and synergies related to the transaction, our industry, business strategy, plans, goals and expectations concerning our market position, future operations, the timing or results of any Investigational New Drug applications and NDA submissions, including the resubmission of the NDA for arimoclomol, communications with the FDA, the potential uses or benefits of arimoclomol, KP1077, SDX or any other product candidates for any specific disease indication or at any dosage, the potential benefits of any of Zevra’s product candidates, the success or timing of the launch or commercialization of AZSTARYS® or any other products or related sales milestones, the sufficiency of cash to fund operations, our plans or ability to seek funding, our plans with respect to our share repurchase program, and our strategic and product development objectives. These forward-looking statements are based on information currently available to Zevra 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 expressed or implied by the forward-looking statements, including, but not limited to, the following: uncertainties as to the timing of the consummation of the proposed transactions and the ability of the parties to consummate the proposed transactions; the satisfaction of the conditions precedent to consummation of the proposed transactions, 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 transaction; disruption of Acer’s or Zevra’s current plans and operations as a result of the proposed transaction; the ability of Acer or Zevra to retain and hire key personnel; competitive responses to the proposed transaction; unexpected costs, charges or expenses resulting from the proposed transaction; 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 transaction and realize additional opportunities for growth and innovation; the ability of Zevra to realize the anticipated synergies and related benefits from the proposed transaction 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 discussed under the caption “Risk Factors” in our Annual Report on Form 10-K filed with the SEC on March 7, 2023, as updated by our Quarterly Report on Form 10-Q filed with the SEC on August 14, 2023, and in our other filings with the SEC could cause actual results to differ materially from those indicated by the forward-looking statements made herein.

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.

# Important Additional Information Regarding Transactions to be Filed with the SEC



In connection with the proposed transaction(s), Zevra Therapeutics, Inc. (Zevra) and Acer Therapeutics, Inc. (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](http://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](http://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](http://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](http://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.



# OLPRUVA™ Important Safety Information

**OLPRUVA [ol proo vah] (sodium phenylbutyrate) for oral suspension**

**This summary does not include all information about OLPRUVA and is not meant to take the place of discussions with your healthcare provider about your or your child's treatment. Please read this important information carefully and discuss any questions about OLPRUVA with your healthcare provider.**

## **What is OLPRUVA?**

- OLPRUVA 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<sup>2</sup> or greater, with urea cycle disorders (UCDs), involving deficiencies of carbamylphosphate synthetase (CPS), ornithine transcarbamylase (OTC) or argininosuccinic acid synthetase (AS).
- Episodes of rapid increase of ammonia in the blood (acute hyperammonemia) may happen in people during treatment with OLPRUVA. OLPRUVA is not for the treatment of acute hyperammonemia, which can be life-threatening and requires emergency medical treatment.
- OLPRUVA is not approved in children weighing less than 44 pounds (20 kg) or in children weighing 44 pounds (20 kg) or greater with a BSA of less than 1.2 m<sup>2</sup>.

**Before taking OLPRUVA, tell your or your child's healthcare provider about all your medical conditions, including if you:**

- have heart problems
- have kidney or liver problems
- are pregnant or plan to become pregnant. It is not known if OLPRUVA will harm your unborn baby. If you become pregnant during treatment with OLPRUVA, call Acer Therapeutics Inc. at 1-833-657-7882 to report the pregnancy.
- are breastfeeding or plan to breastfeed. It is not known if OLPRUVA passes into your breast milk. Talk to your doctor about the best way to feed your

baby if you take OLPRUVA.

**Tell your healthcare provider about all the medicines you or your child take**, including prescription and over-the-counter medicines, vitamins, and herbal supplements.

**Certain medicines may increase the level of ammonia in your blood or cause serious side effects when taken during treatment with OLPRUVA.**

**Especially tell your healthcare provider if you or your child take:**

- corticosteroids
- valproic acid
- haloperidol
- probenecid

Know the medicines you take. Keep a list of them to show your or your child's healthcare provider and pharmacist when you get a new medicine.

**Keep OLPRUVA and all medicines out of the reach of children.**

# OLPRUVA™ Important Safety Information (continued)



## How should I or my child take OLPRUVA?

Read the detailed Instructions for Use that comes with OLPRUVA for information about the right way to prepare and take a dose of OLPRUVA.

- Take OLPRUVA exactly as prescribed by your healthcare provider.
- Your healthcare provider may change your dose if needed. Do not change your dose unless your healthcare provider tells you to.
- Your healthcare provider will prescribe OLPRUVA based on your or your child's weight.
- Take your OLPRUVA dose with food.
- If you miss a dose of OLPRUVA, take it as soon as possible that same day.
- Do not** give or take OLPRUVA through a gastrostomy or nasogastric tube.
- If you take too much OLPRUVA, call your healthcare provider or go to the nearest hospital emergency room right away.

## What are the possible side effects of OLPRUVA?

OLPRUVA can cause serious side effects, including:

- Nervous system problems (neurotoxicity).** Call your healthcare provider right away if you or your child get any of the following symptoms during treatment with OLPRUVA:
  - sleepiness
  - nausea
  - tiredness
  - headache
  - lightheadedness
  - confusion
  - vomiting

- Low potassium levels in your blood (hypokalemia).** Your healthcare provider will monitor your blood potassium levels during treatment with OLPRUVA and treat if needed.

- Conditions related to swelling (edema).** OLPRUVA contains salt (sodium), which can cause swelling from salt and water retention. Your healthcare provider will decide if OLPRUVA is right for you if you have certain medical conditions that cause edema, such as heart failure, liver problems or kidney problems.

The most common side effects of OLPRUVA include:

- absent or irregular menstrual periods
- body odor
- decreased appetite
- bad taste or avoiding foods that you ate prior to getting sick (taste aversion)

Your healthcare provider may do certain blood tests to check you or your child for side effects during treatment with OLPRUVA.

These are not all of the possible side effects of OLPRUVA. Call your doctor for medical advice about side effects. You are encouraged to report negative side effects of prescription drugs to the FDA. Visit [www.fda.gov/MedWatch](http://www.fda.gov/MedWatch) or call 1-800-FDA-1088.

For additional Important Safety Information see full [Prescribing Information](#) and [Patient Information](#), including [Instructions for Use](#), and discuss with your doctor.

# Executing on Zevra's Strategy to Become a Leading Rare Disease Therapeutics Company

Adding a complementary rare disease portfolio, accelerating revenue potential and cost synergies

## Deal Consideration

- ✓ Zevra to acquire Acer Therapeutics in a stock exchange transaction for \$0.61 per share, or \$15M of Zevra's common stock, approximately 2% discount to Acer's last closing price
- ✓ Contingent value rights (CVRs) of up to \$76M based on achieving future commercial and regulatory milestones, as well as potential payments related to preclinical program
- ✓ Purchase of \$35.3M in Acer senior secured debt at a discount through a capital efficient structure
- ✓ Bridge loan facility of up to \$16.5M to ensure ongoing OLPRUVA™ commercial efforts and operations

## Portfolio Value

- ✓ OLPRUVA is approved in the U.S. as a nitrogen-binding agent indicated as adjunctive therapy to standard of care, which includes dietary management, for the chronic management of adult and pediatric patients with Urea Cycle Disorders (UCDs) involving deficiencies of CPS, OTC, or AS enzymes. UCDs are a group of rare genetic disorders impacting the removal of ammonia from the bloodstream
- ✓ EDSIVO™, Phase 3 program for the treatment of vascular Ehlers-Danlos syndrome (vEDS), a rare genetic disorder impacting blood vessels, expands Zevra's rare disease development pipeline
- ✓ Additional rare disease capabilities and expertise





## Strategic Fit

- ✓ Highly complementary portfolio of therapies addressing rare diseases
- ✓ Significant synergies and potential cost savings across commercial organization
- ✓ Accelerates Zevra's commercial platform and capabilities to support arimoclomol launch, if approved
- ✓ Augments workforce and functional capabilities
- ✓ OLPRUVA expected to increase and diversify Zevra's revenues

# Acer Acquisition Accelerates Zevra's Growth into Commercial Organization and Expands Development Pipeline

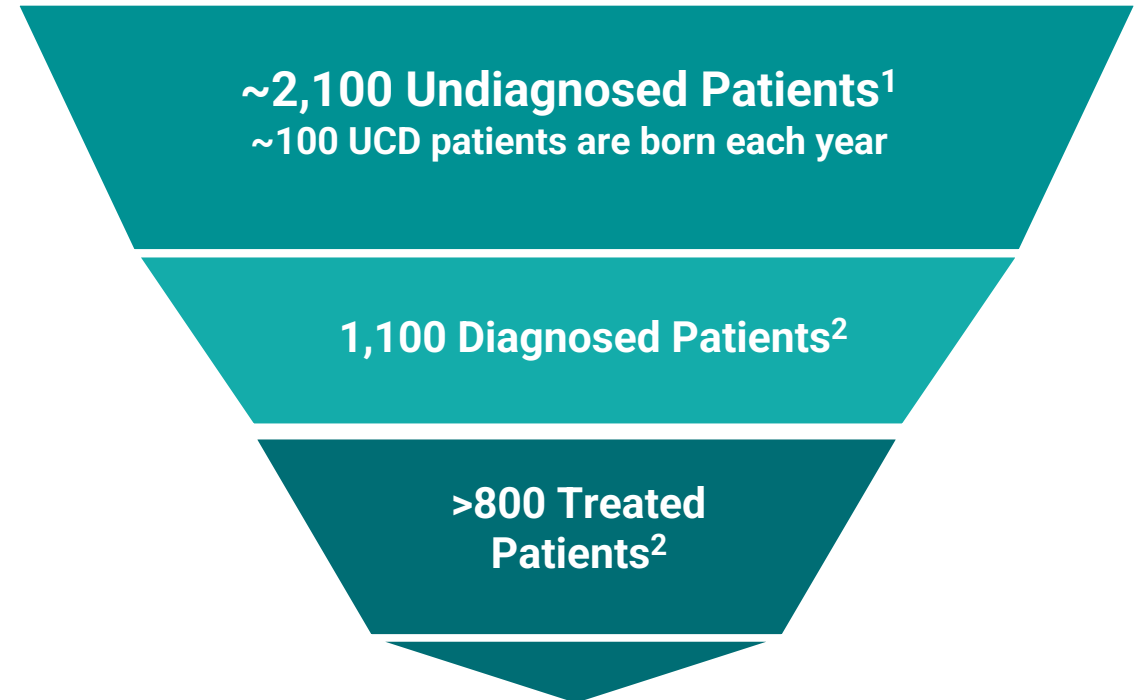
PHASE 1	PHASE 2	PHASE 3	NDA	FDA APPROVED	NEXT MILESTONE
Arimoclomol Nieman Pick Type C					NDA Resubmission Q4 2023
KP1077 Idiopathic Hypersomnia					Interim P2 Analysis Q3 2023
KP1077 Narcolepsy					Trial ongoing
AZSTARYS® <i>serdexmethylphenidate and dexamethylphenidate</i> Attention Deficit/Hyperactivity Disorder					On track to receive next sales milestone
OLPRUVA™ <i>sodium phenylbutyrate for oral suspension</i> Urea Cycle Disorder					Approved and commercially available in US
EDSIVO™ <i>celiprolol</i> Vascular Ehlers-Danlos Syndrome					Phase 3 trial ongoing

## Combined Portfolio

- 
 Highly complementary portfolio targeting rare diseases
- 
 Upcoming milestones and catalysts
- 
 Robust pipeline with clinical and commercial assets
- 
 Overlap of treating physicians

# Urea Cycle Disorders (UCDs)

- UCDs are a group of rare, genetic disorders caused by mutations that result in a deficiency of one of the six enzymes or two transporters of the urea cycle, including carbamylphosphate synthetase (CPS), ornithine transcarbamylase (OTC) or argininosuccinic acid synthetase (AS)
- These enzymes are responsible for removing ammonia from the bloodstream
- Elevated ammonia levels in both symptomatic and asymptomatic patients can be neurotoxic leading to neurocognitive damage, among other symptoms



Current treatments:  
Phenylbutyrate

1. <https://www.drugs.com/slideshow/top-10-most-expensive-drugs-1274>

2. HealthVerify Payer Claims data analysis 2021



# OLPRUVA™ Has Potential to Address Unmet Needs In Treatment of UCDs



## UCDs: An area of high unmet need

- Currently, phenylbutyrate is standard of care for patients with UCDs
- Palatability, odor, preparation, route of administration and packaging are the most important attributes for treatment adherence among patients with UCDs<sup>1</sup>
- Adherence can also be affected when patients skip their mid-day dose, because existing treatments are inconvenient to take to work or school

## OLPRUVA: FDA approved, novel formulation of phenylbutyrate with convenient, single-dose packaging

- Potential to improve compliance through more convenient administration and a formulation designed for palatability
- Dual-coated formulation delays release in water for up to 5 minutes, rapidly dissolves in acid environments (e.g., stomach)
- Patent protection through 2036

## Multiple opportunities for potential indication expansion for OLPRUVA

- Ongoing investigations in UCDs to determine feasibility to enhance administration flexibility options and improve bioavailability via pre-meal administration
- Other investigational life cycle indications include Maple Syrup Urine disease, a liver disorder

# OLPRUVA™ is a Novel Formulation of Phenylbutyrate with Convenient, Single-Dose Packaging

Phenylbutyrate Formulations				
	OLPRUVA	Raviciti®	Buphenyl®	Pheburane®
Efficacy and Safety in UCs	✓	✓	✓	✓
Formulation	Dual-coated oral pellets	Clear, oily liquid	Powder or Tablets	Single-coated oral pellets
Palatability	Tasteless up to 5 minutes	Tasteless	Bitter Taste	Up to 10 seconds
Packaging	Single-dose envelopes	Glass vials with syringe for each dose	Tub of powder or pills	Large bottle of pellets
Portability	++	+	-	+
Administration	Mix with water and Mix-Aid	Meter dose into syringe from glass vial	Measure powder and mix with water or take up to 40 tablets per day	Pour directly into mouth or sprinkle on each bite of applesauce or carrot puree

# OLPRUVA™ Designed to Address Patient Pain Points

## Palatable

Dual-coating formulation designed for palatability

## Pre-Measured

Convenient individual dose packets support dosing accuracy

## Portable

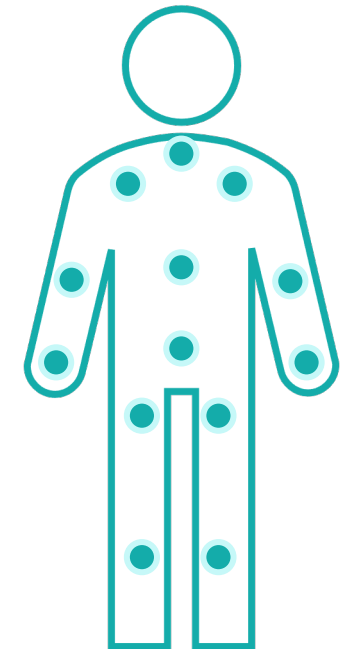
Discrete single-dose envelopes are easily carried when on the go



# Vascular Ehlers-Danlos Syndrome (vEDS)

Acer's pipeline includes EDSIVO™ (celiprolol) an investigational treatment for vEDS in patients with a confirmed type III collagen (COL3A1) mutation

- Inherited, genetic disorder caused by mutations in the genes responsible for the structure, production, or processing of collagen
- Spectrum disorder, vEDS is the most serious form
- Causes abnormal fragility in blood vessels, which can give rise to aneurysms, abnormal connections between blood vessels known as arteriovenous fistulas, arterial dissections, and spontaneous vascular ruptures, all of which can be potentially life-threatening
- Estimated total COL3A1-positive vEDS patient prevalence in the U.S. could be as high as 7,500 patients
- Full enrollment in ongoing registration enabling Phase 3 trial for EDSIVO expected in 2024



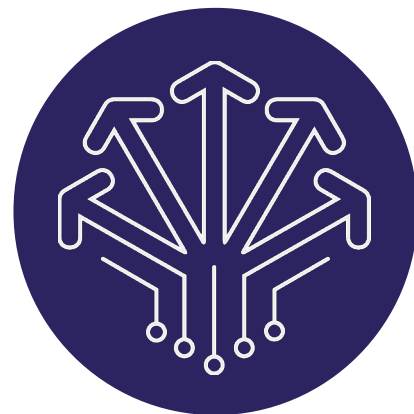


# Zevra + Acer = Expected Synergies

Complementary portfolios have the potential to provide scale and efficiency



Prescribing  
Physicians/  
Specialty  
Pharmacies



Patient Services Across  
Indications



Data Management  
Systems



Workforce &  
Functional Capabilities



Existing Commercial  
Contracts and Sales  
Organization

Anticipated synergies between UCD and NPC indications allow for "nimble team approach"

# Capital Efficient Transaction Structure

Series of transactions designed to add value, mitigate risk and capture opportunities

## Debt Purchase

- Purchased Acer's secured debt of \$35.3M from Nantahala Capital
- Paid \$28.5M in consideration as follows:
  - \$12M in cash from Zevra's existing margin loan facility
  - \$5M note held by Nantahala
  - \$11.5M of Zevra's common stock based on the 20-day VWAP as of Aug 29, 2023 (\$5.0667 per share)

**Debt purchase intended to support Acer merger transaction, mitigating risk and providing security**



## Acer Acquisition

- Each share of Acer stock will receive \$0.61 per share, or \$15M in Zevra common stock, which is ~2% discount to Acer's last closing price
- Up to \$76M in CVRs to Acer's shareholders providing potential future cash payments based on specified sales and regulatory milestones
- Bridge loan facility of \$16.5M to provide \$10M for termination of Acer's agreement with Relief, and to support ongoing OLPRUVA activities through closing

**Total deal value of up to \$91M, with potential to increase and diversify Zevra's revenue immediately, if approved by Acer shareholders**

# Zevra Positioned for Success in Rare Disease

Combined company will operate under the name Zevra Therapeutics, Inc.



## Continued Strategic Focus on Rare Disease

- Focus on delivering therapies addressing significant unmet needs in rare disease
- Leveraging unique insights and capabilities to develop rare disease therapies
- Strong relationships with rare disease community – including KOLs and advocates



## Differentiated Portfolio In Rare Disease

- Arimoclomol for Niemann-Pick disease Type C (NPC); NDA filing expected in Q4 2023
- KP1077 for idiopathic hypersomnia (IH) and narcolepsy; interim data expected by end of Q3 2023, topline data H1 2024
- OLPRUVA to treat urea cycle disorders (UCDs); FDA approved product
- EDSIVO for vascular Ehlers-Danlos syndrome (vEDS); complete trial enrollment expected in 2024



## Creating Value

- Strong balance sheet supports investment in Zevra's portfolio and ongoing operations
- Increased and diversified revenue from multiple products
- Commercial capabilities to provide access to treatments for patients with rare diseases
- Significant near-term catalysts to increase shareholder value