

Clear Strategic Direction Under Refreshed Board

April 2023

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Additional Information and Where to Find It

Zevra has filed with the Securities and Exchange Commission (the “SEC”) a definitive proxy statement on Schedule 14A, containing a form of WHITE proxy card, with respect to its solicitation of proxies for Zevra’s 2023 Annual Meeting of Stockholders. This communication is not a substitute for any proxy statement or other document that Zevra may file with the SEC in connection with any solicitation by Zevra.

INVESTORS AND SECURITY HOLDERS ARE URGED TO READ THE PROXY STATEMENT (INCLUDING ANY AMENDMENTS OR SUPPLEMENTS THERETO) FILED BY ZEVRA AND ANY OTHER RELEVANT DOCUMENTS FILED WITH THE SEC WHEN THEY BECOME AVAILABLE CAREFULLY AND IN THEIR ENTIRETY BECAUSE THEY WILL CONTAIN IMPORTANT INFORMATION ABOUT ANY SOLICITATION.

Investors and security holders may obtain copies of these documents and other documents filed with the SEC by Zevra free of charge through the website maintained by the SEC at www.sec.gov. Copies of the documents filed by Zevra are also available free of charge by accessing Zevra’s website at www.zevra.com.

Participants in the Solicitation

This communication is neither a solicitation of a proxy or consent nor a substitute for any proxy statement or other filings that may be made with the SEC. Nonetheless, Zevra, its directors and executive officers and other members of management and employees may be deemed to be participants in the solicitation of proxies with respect to a solicitation by Zevra. Information about Zevra’s executive officers and directors is available in Zevra’s definitive proxy statement for the 2023 Annual Meeting of Stockholders, which was filed with the SEC on March 15, 2023. The definitive proxy statement is available free of charge at the SEC’s website at www.sec.gov. Copies of the documents filed by Zevra are also available free of charge by accessing Zevra’s website at www.zevra.com.

Zevra Therapeutics: Value Creation Strategy



- We are transforming Zevra into a commercially driven rare disease therapeutics company, that incorporates the best elements of our existing prodrug platform
 - Following the approval and licensing of AZSTARYS® in March 2021, we immediately began a strategic review of the entire business to determine how to best create value for shareholders
 - After carefully reviewing the future potential of relying exclusively on a prodrug outlicensing strategy, we actively began to build a diversified pipeline that can be directly commercialized
- We have been strengthening our Board and leadership team, which are highly qualified to continue driving Zevra's transformation
- Our existing cash runway will fund the Company through all clinical, regulatory and commercial milestones into 2026

Our Extensive Engagement with Daniel Mangless

- We've engaged with Mangless over the course of more than 3 years on the company's strategy and performance
- After learning about Mangless' intention to nominate directors, we made multiple good-faith attempts to reach a mutually agreeable settlement – but he has refused to engage constructively
- Mangless fails to acknowledge that we have already addressed – and continue to address – Board refreshment and are focusing on a strategy that will drive real growth
- Our Board has undertaken a comprehensive director refreshment process over the past 18 months, resulting in three new highly qualified, independent Board members
- The Mangless nominees are highly unqualified to serve on the Board, and he offers no credible plan for growth other than to continue on the course that Zevra already pursued prior to making the strategic decision to focus on rare disease

Section I

We are confident in our strategy, which leverages our strengths and provides a focused pathway for Zevra to develop and commercialize its products in-house



Zevra is Positioned for Success



Following AZSTARYS® approval and licensing, we undertook a strategic process to enhance our long-term potential by focusing on therapeutics for rare diseases to commercialize on our own

- Numerous opportunities to create value for our shareholders as we execute our business plans
- Robust pipeline of promising rare disease product candidates with multiple value-creating milestones expected in 2023
 - Arimoclomol and KP1077 currently in development
- Accomplished Board of Directors with the right experience and skillsets to execute our strategy in rare disease therapeutics
- Our strong balance sheet coupled with ongoing and growing revenue from our licensed products and arimoclomol EAP funds our business without requiring further dilution
- Launch of Zevra brand marks the next key step in our evolution into a commercially focused rare disease therapeutics company

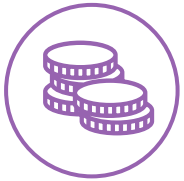
Significant Value-Creation Opportunity in Rare Disease Therapeutics for Patients and Shareholders



Unique partnerships with patient communities



Longer market exclusivity and less generic competition



Lower cost of R&D



Regulatory and financial incentives



Shorter development timelines and smaller studies



Small patient population served by specialist clinicians can be addressed with in-house commercial team

Multiple Potential Growth Catalysts in 2023 - 2024



This is an exciting time in the growth of Zevra, with strong prospects for a promising future

ARIMOCLOMOL

- Potential to re-file NDA as early as Q3 2023
- Anticipate ongoing quarterly revenue from French EAP reimbursements

KP1077 DEVELOPMENT PROGRAM

- Expect to file IND in Q2 2023 for narcolepsy
- Interim data from Phase 2 clinical trial expected as early as Q3 2023
- Top-line data expected by EOY 2023
- Potential to initiate Phase 3 trial in IH and narcolepsy following Phase 2 trial results

AZSTARYS®

- Potential to realize sales milestones and continued royalty revenue from AZSTARYS®
- Adds capital flexibility and potential to further extend cash runway

Supported by strong balance sheet to continue driving Zevra's transformation into 2026

✔ Supports development efforts and other pipeline expansion activities

✔ Available capital expected to extend cash runway into 2026

Arimoclomol – Innovative Product for an 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 significantly improved lysosomal and cellular function



EXTENSIVELY RESEARCHED

- Studied in ten Phase 1, four Phase 2, and three Phase 2/3 trials
- No significant safety findings identified to date (500+ patients treated)
- Positive efficacy results from NPC trial (NPC-002) and Phase 2 trial in Gaucher Disease (GD), both of which are lysosomal storage disorders



BENEFICIAL REGULATORY POSITIONING

- Orphan Drug Designation for NPC in U.S. and EU
- 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 FDA

Arimocloamol – Expanding Pipeline Targeting Rare Diseases

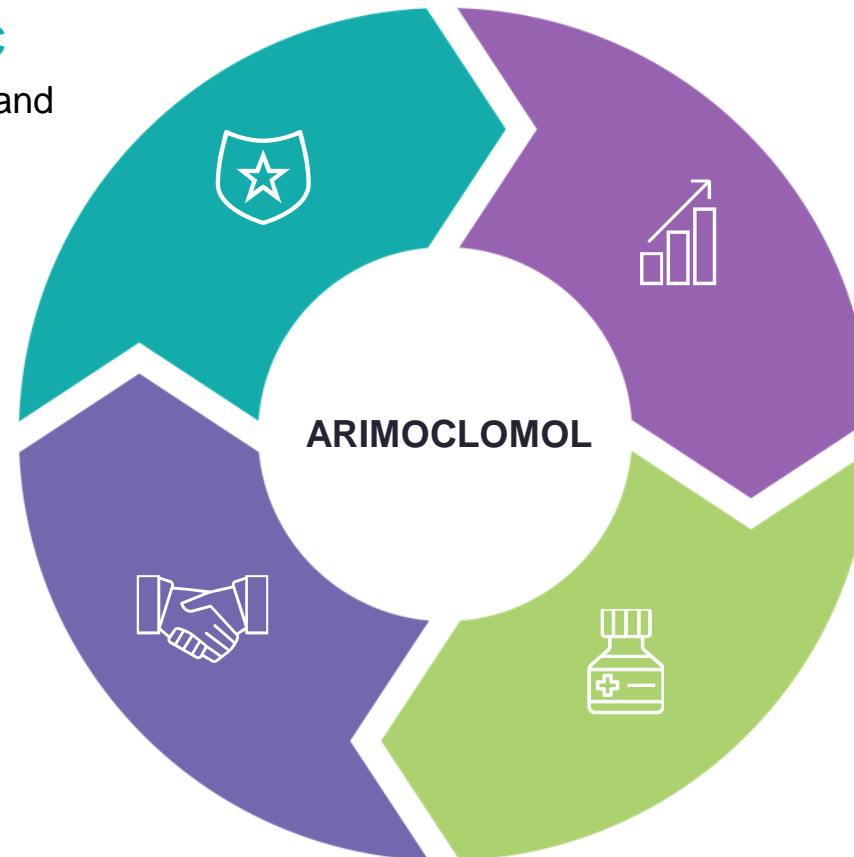
Aligns with strategy to build value through the development and commercialization of novel treatments for rare diseases

Niemann-Pick disease type C

- Ultra-rare progressive, disabling and fatal lysosomal storage disorder
- 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



High Upside Opportunity

- NDA-stage investigational drug candidate
- Zevra has expertise in NDA resubmissions following CRLs

Early Access Programs

- Available to NPC patients in the U.S., France, Germany and other European countries
- French EAP expected to generate annual net revenue of ~\$8M (gross revenue of ~\$12M)

Path to Resubmission and Approval Progressing



Zevra has significant experience with challenging regulatory situations, including two FDA product approvals that followed initial CRLs

RECENT ACTIVITIES

- Continuing to have an ongoing collaborative dialogue and periodic meetings with the FDA
 - Intended to ensure an optimal NDA data package that addresses all issues in the CRL
- Advancing activities to bolster arimoclomol NDA with confirmatory evidence for resubmission to the FDA
 - Working to analyze and process the new data generated since the CRL
 - This includes data obtained from a 4-year arimoclomol safety study and safety data from other clinical trials with arimoclomol

REGULATORY OUTLOOK

- Throughout this process, no new issues or concerns have been raised by the FDA
 - No new efficacy trial has been proposed by FDA
- We believe there is a viable pathway to enable a successful NDA resubmission and subsequent approval for arimoclomol in NPC
 - Path may include, if necessary, additional non-clinical or clinical studies, a Federal Dispute Resolution Request (FDRR) and/or an advisory committee (ad com) requested by either FDA or Zevra

Zevra expects to resubmit the NDA for arimoclomol in NPC as early as Q3 2023

Near-Term Opportunity to Commercialize Arimoclomol and Retain Full Market Value

Launch arimoclomol with a small, focused commercialization effort which can be foundation for future rare disease products, including KP1077



1 Small & nimble commercial team

2 Lower marketing spend

3 Patient advocacy relationships support adoption

4 Market entry through U.S. and E.U. EAPs

5 Commercial opportunities outside the U.S.

KP1077: Multiple Clinical Programs Targeting Rare Sleep Indications

KP1077 Represents a Potential “Portfolio in a Pill” Opportunity

IDIOPATHIC HYPERSOMNIA

- Lead KP1077 indication
- Investigational New Drug (IND) application cleared by FDA
- Ongoing Phase 2 clinical trial initiated in December of 2022
- Interim data from Phase 2 clinical trial expected as early as Q3 of 2023
- Top-line data expected by EOY 2023

NARCOLEPSY

- Second KP1077 indication would allow Zevra to address two rare sleep indications that are underserved by currently available medications
- Evaluate the potential to initiate narcolepsy Phase 3 trial based on IH Phase 2 results
 - Seek to leverage key data points from IH program to expedite narcolepsy development

KP1077 – Product Candidate Overview

Zevra is advancing KP1077 as a potential therapeutic treatment for Idiopathic Hypersomnia (IH)

Serdexmethylphenidate

- 100% SDX with multiple dosing options
- SDX granted Orphan Drug Designation for IH by FDA
- SDX designated C-IV by DEA

Regulatory & IP Advantages

- Orphan Drug Designation in IH
- Eligible for Fast-Track and Breakthrough Therapy designation
- Solid IP through 2037 and potentially beyond



Dosing Addresses Symptoms

- Dosed either 1x daily at bedtime or 2x daily at bedtime and at waking
- Potential to address primary IH symptoms: sleep inertia and brain fog

Improved Safety & Tolerability

- Greater tolerability and lower cardiovascular effects could allow for higher, more effective dosing (i.e. greater efficacy)
- No DDI potential with hormonal contraceptives; antidepressants

Phase 2 Clinical Trial Investigating KP1077 for the Treatment of IH

Multi-center, dose-optimizing, double-blind, placebo-controlled, randomized-withdrawal study to evaluate safety and efficacy of KP1077, as well as to assess the symptoms and severity of “brain fog”

PHASE 2 TRIAL (N=48)

Part 1:

- Five-week open-label titration phase
- Patients optimized to one of the four doses of SDX (80, 160, 240, or 320 mg/day)

Part 2:

- Two-week randomized, double-blind, withdrawal phase
- 2/3 receive active; 1/3 placebo
- 50% receive single daily dose; 50% receive half daily dose upon awakening and at bedtime

PRIMARY ENDPOINT

- Safety and tolerability of SDX

MAJOR SECONDARY ENDPOINT

- Change in Epworth Sleepiness Scale (ESS) total score

ADDITIONAL EXPLORATORY ENDPOINTS

- Patient Global Impression of Severity (PGI-S)
- Clinical Global Impression of Severity (CGI-S)
- Change in total score on the Idiopathic Hypersomnia Severity Scale (IHSS)
- New scale to assess the symptoms and severity of “Brain Fog”

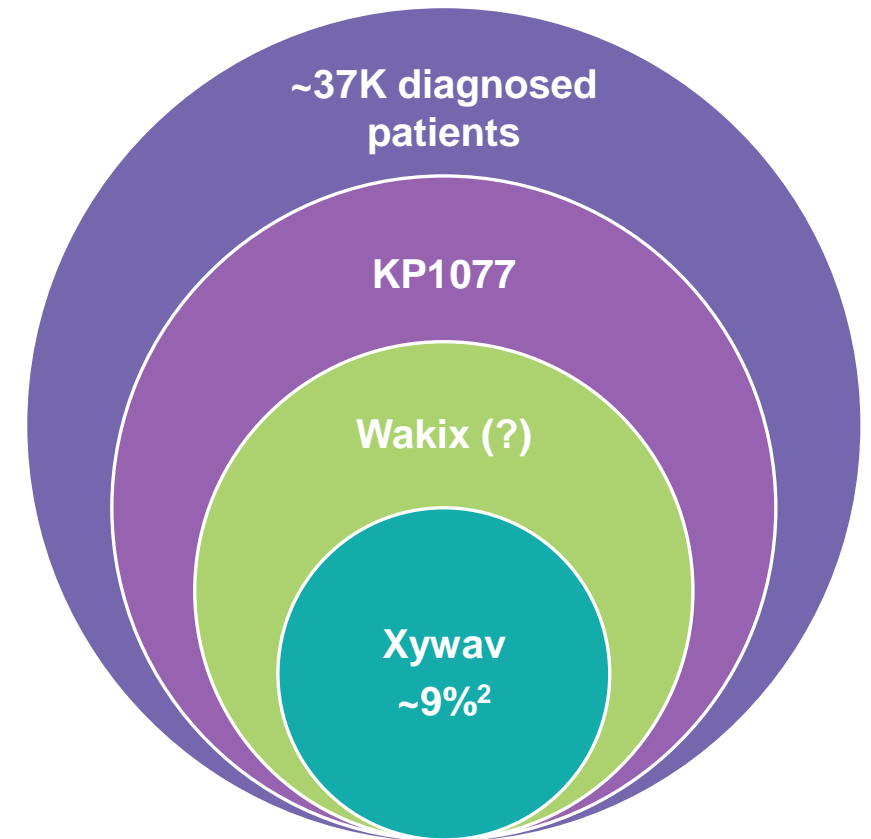
KP1077 Could Capture a Large Share of the IH Market Share

Competitive Products in IH

- Xywav® received FDA approval in August 2021 as the first therapy for IH
 - According to analysts, Xywav projected sales are ~\$300 million for IH by the end of 2025¹
- Wakix® currently enrolling patients in a Phase 3 clinical trial in IH

KP 1077 Potential Differentiation

- **MOA and improved efficacy of KP1077:** positioned as a monotherapy and combination use with oxybate (Xyrem, Xywav or others)
- **KP1077 safety profile:** Schedule IV, lack of drug-drug-interaction with hormonal contraceptives which is an issue with modafinil, reduced risk of adverse events compared to current off-label IH therapies
- **Xywav barriers to uptake:** clinical trial discontinuation rate of ~11% due to treatment emergent adverse events, boxed warning for CNS depression, abuse and misuse potential, REMS program, negative stigma associated with GHB²
- **Wakix barriers to uptake:** DDI, especially with antidepressants and antihistamines



Sources: (1) <https://investor.jazzpharma.com/investors/events-presentations>

(2) <https://www.reuters.com/business/healthcare-pharmaceuticals/us-fda-approves-jazz-pharmas-drug-excessive-daytime-sleepiness-2021-08-12/>

Section III

Our Board and leadership team are highly qualified to continue driving Zevra's transformation

Zevra's Board is Committed to Board Refreshment



In parallel with our strategic review, the Board initiated an assessment designed to support Zevra's shift to commercializing rare disease products

- Advised by leading executive search firms Korn Ferry and Caldwell Partners to support director refreshment process
- Identified critical skillsets and diversity needs among Board and leadership aligned with strategic priorities of developing and commercializing rare disease therapeutics
- Appointed three new independent directors to Board over last 18 months

New Board Appointments in Last 18 Months Bring Diverse Backgrounds and Skills



Tamara A. (Seymour) Favorito

*Board Member since August 2021
Audit Committee Chair
Compensation Committee Chair*



Christopher A. Posner

*Board Member since November 2022
Audit Committee
Compensation Committee*



Wendy L. Dixon, Ph.D.

*Board Member effective following
2023 Annual Meeting*

Biotech & Pharma Experience

30 years, including 20 years as a CFO

25 years

40+ years

Experience & Accomplishments

Extensive experience leading multiple private and public financings and M&A transactions, and leading the finance, investor relations, human resources, administration and managed care functions

Currently serves on the Board of Directors of Artelo Biosciences, Inc. and Kintara Therapeutics, Inc.

Executed multiple product launches that drove shareholder value

Leadership roles at Bristol Myers Squibb, Pfizer, Wyeth Pharmaceuticals and Endo International

Currently CEO and President of Cara Therapeutics, a commercial stage biotech company

Senior leadership roles at Bristol Myers Squibb, Merck, West Pharmaceuticals, Osteotech, Centocor and GlaxoSmithKline

Currently serves on the Board of Directors of Arivinas, Inc., Black Diamond Therapeutics, Inc., and Iovance Biotherapeutics, Inc.

Zevra's Three Director Candidates Have Deep Industry Experience and Expertise



Richard W. Pascoe

*CEO
Board Member since 2014*



Christopher A. Posner

*Board Member since 2022
Audit Committee
Compensation Committee*



David S. Tierney, M.D.

*Board Member since 2015
Compensation Committee
Nominating & Corporate Governance Committee*

Biotech & Pharma Experience

30+ years

25 years

30+ years

Experience & Accomplishments

Significant strategic, commercial and business development expertise

Executed multiple product launches that drove shareholder value

10+ product approvals as executive or Board member, including two successfully launched orphan drugs in U.S.

Led companies through strategic shifts, value-creating partnerships and commercial launches

Leadership roles at Bristol-Myers Squibb, Pfizer, Wyeth Pharmaceuticals and Endo International

Medical doctor, trained in internal medicine

Supported Zevra in obtaining two FDA approvals and building pipeline of rare disease product candidates

Currently CEO and President of Cara Therapeutics, a commercial stage biotech company

Currently CEO of Aramis Biosciences and on Board of Catalyst Pharmaceuticals, a rare disease company

Section IV

The Mangless Nominees have almost no relevant experience,
have communicated no strategy for Zevra and
have rejected all attempts to resolve proxy contest

The Mangless Nominees Are Highly Unqualified



Electing any of his nominees would diminish the overall quality of, and experience represented on, the Board

ZEVRA BOARD SKILLS & EXPERTISE	NUMBER OF ZEVRA DIRECTORS	John B. Bode	Douglas W. Calder	Corey M. Watton
Biotech / Pharmaceutical Experience	● ● ● ● ● ● ●	✗ No pharmaceutical or health care experience	✗ No public company Board experience	✗ No relevant industry or public company Board experience
Drug Development	● ● ● ● ● ● ●	✗ Held senior executive roles at companies with serious financial difficulties, including FISION and Tribune Publishing (formerly NYSE: TPUB)	✗ Claimed “financial expertise and extensive leadership experience” appears to be overstated, as first 11 years in pharmaceutical industry primarily in investor and media relations	✗ Finance experience at medical staffing and in-home senior care companies is wholly unrelated to Zevra
Medical and Scientific	● ● ● ● ● ● ●	✗ Despite claim of having “no financial relationship” with his nominees, Mangless is a significant shareholder in FISION Corporation (OTC: FSSN) — with market cap of ~\$1M — where Bode is interim CEO and Board member	✗ History with troubled companies, including	✗ Personal connection to fellow nominee Bode
Product Management	● ● ● ● ● ● ●		<ul style="list-style-type: none"> Wind-down at the Vaccine & Gene Therapy Institute of Florida 	<ul style="list-style-type: none"> Bode’s sister is director of finance at Fusion Medical Staffing, where Watton is CFO
Commercial and Marketing Operations	● ● ● ● ● ● ●		<ul style="list-style-type: none"> Chapter 11 bankruptcy at BioVest International 	<ul style="list-style-type: none"> From 2014 to 2020, Bode’s sister was director of finance at Home Instead Senior Care, where Watton was CFO during most of her tenure
Business Development and M&A Transactions	● ● ● ● ● ● ●		<ul style="list-style-type: none"> Liquidation of assets at Viragen 	
Finance	● ● ● ● ● ● ●			
Public Company Management and Board Service	● ● ● ● ● ● ●			

Mangless Rejected Our Attempts to Find Common Ground

- Zevra has proposed multiple paths forward to avoid a costly, distracting proxy contest, including ultimately extending a settlement offer with the following terms:
 - Appoint one of the Mangless Nominees, as selected by Mangless, to the Board, subject to the Board’s normal onboarding process;
 - Offer the Mangless Director membership on the Company’s Nominating and Corporate Governance Committee;
 - Appoint another new independent director after the 2023 Annual Meeting, and the Mangless Director would take part in selecting such candidate as part of the Company’s Nominating and Corporate Governance Committee; and
 - Introduce Mr. Mangless to the two newest directors to discuss the Company’s business and how the Board is addressing Mr. Mangless’ concerns
- Mangless ignored this offer and has rejected a dialogue with management, including with Dr. Mickle – whose relationship with Zevra is part of Mangless’ claimed strategic plans

1.26.23	1.28.23	2.1.23	2.3.23	2.9.23	2.14.23	2.16.23	2.20.23	2.24.23
Mangless provided notice of his intent to nominate the three Mangless nominees	CEO emailed Mangless to schedule teleconference with Mangless, CEO and Chairman	CEO and Chairman met with Mangless via teleconference When asked, Mr. Mangless indicated that he did not have any specific concerns with the Company’s business or its operations, nor did he offer any substantive new ideas or alternative strategies for consideration by the Board	CEO sent Mangless a text requesting a call to propose initial settlement offer Mangless responded informing the CEO he was not willing to have a call and requesting offer in writing Chairman emailed Mangless with the Offer	Mangless rejected the Offer He stated that if the Board appointed all three of his director nominees and Dr. Mickle agreed to stay with the Co, he might be willing to consider agreeing to a withdrawal of his director nominations and proposal	Dr. Mickle sent a text to Mangless asking to arrange a call No response	Dr. Mickle emailed Mangless to propose a call with himself, Mangless and the Chairman No response	The Chairman emailed Mangless to follow up on Mickle’s email with an updated settlement offer (outlined above) No response CEO called Mangless No response	CEO emailed Mangless noting effort to engage in a constructive discussion Mangless responded, noting that he would go forward with his nominations and all communications with him should be through his attorneys

Setting the Record Straight

Assertions vs. the Facts

<p>✗ “Mr. Mangless strongly believes that the Board must be refreshed to ensure that the interests of the stockholders are appropriately represented in the boardroom” <i>Mangless Definitive Proxy - Page 3</i></p>	<p>✓ Zevra has appointed three highly qualified independent directors in the last 18 months as part of the Company’s ongoing commitment to Board refreshment</p> <p>✓ Much of our strategic direction is the direct result of the Board and management team’s open communications with shareholders and their constructive input toward achieving our mutual goal of enhancing value</p>
<p>✗ “Zevra’s core technology could be leveraged to develop its existing pipeline and expand it beyond the existing opportunities to potentially license pipeline products and technology to other pharmaceutical companies looking to enhance their existing portfolios” <i>Mangless Definitive Proxy - Page 3</i></p>	<p>✓ The Board and management thoughtfully evaluated the best path forward for Zevra and determined that focusing exclusively on a pipeline of product candidates that would eventually compete in heavily generic markets would likely fail to deliver a significant return on investment to our shareholders.</p> <p>✓ Our decision to focus on rare diseases positions us to field a small, nimble commercial team that we believe can be highly effective and drive better return on capital than our results to date with an outlicensing model</p>
<p>✗ “The founder and chief architect of the company’s scientific successes is being completely sidelined to the detriment of the long-term enterprise value of the Company...Renew the relationship with the Zevra’s founder with a continued focus on developing the Company’s pipeline from the LAT platform” <i>Mangless Definitive Proxy - Page 8</i></p>	<p>✓ Following the 2023 Annual Meeting, Dr. Mickle will continue to support Zevra’s drug development and regulatory approval activities as a scientific advisor</p> <p>✓ “I fully support Zevra’s strategy to evolve into a commercial organization focused on developing transformational, patient focused therapies for rare diseases with limited or no treatment options. This is an exciting time for the Company with much opportunity ahead. Proxy battles can be costly and distracting, and as such, I intend to vote in favor of the Zevra slate.” – Travis C. Mickle, Ph.D., Zevra President & Board Member; Co-Founder</p>

Value Creation Underway at Zevra Therapeutics



- ✓ **Confident in our thoughtfully developed strategy to focus on rare diseases**
- ✓ **Refreshed and highly qualified Board and leadership team**
- ✓ **Advancing our late-stage clinical pipeline through arimoclomol and KP1077**
- ✓ **Solid financial position to fund development plans into 2026 with numerous market opportunities**
- ✗ **Mangless nominees have almost NO relevant or public company Board experience**
- ✗ **They have communicated NO new strategy for Zevra**
- ✗ **Electing the Mangless nominees would severely diminish the overall quality of, and experience represented on, the Zevra Board**

THANK YOU

