

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

FORM 10-Q

(Mark One)

☒ QUARTERLY REPORT PURSUANT TO SECTION 13 OR 15(d) OF THE SECURITIES EXCHANGE ACT OF 1934

For the quarterly period ended June 30, 2024

OR

☐ TRANSITION REPORT PURSUANT TO SECTION 13 OR 15(d) OF THE SECURITIES EXCHANGE ACT OF 1934

For the Transition Period From _____ to _____

Commission File Number: 001-36913

Zevra Therapeutics, Inc.

(Exact Name of Registrant as Specified in Its Charter)

Delaware
(State or Other Jurisdiction of Incorporation or Organization)

20-5894398
(I.R.S. Employer Identification No.)

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

34747
(Zip Code)

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

(Former Name, Former Address, and Former Fiscal Year if Changed Since Last Report)

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

Title of each class	Trading Symbol	Name of each exchange on which registered
Common Stock, \$0.0001 par value per share	ZVRA	The Nasdaq Stock Market LLC (Nasdaq Global Select Market)

Indicate by check mark whether the registrant: (1) has filed all reports required to be filed by Section 13 or 15(d) of the Securities Exchange Act of 1934 during the preceding 12 months (or for such shorter period that the registrant was required to file such reports), and (2) has been subject to such filing requirements for the past 90 days. Yes ☒ No ☐

Indicate by check mark whether the registrant has submitted electronically every Interactive Data File required to be submitted pursuant to Rule 405 of Regulation S-T during the preceding 12 months (or for such shorter period that the registrant was required to submit such files). Yes ☒ No ☐

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

Large accelerated filer ☐ Accelerated filer ☐
Non-accelerated filer ☒ Smaller reporting company ☒
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. ☐

Indicate by check mark whether the registrant is a shell company (as defined in Rule 12b-2 of the Exchange Act). Yes ☐ No ☒

As of August 12, 2024, the registrant had 52,617,789 shares of common stock outstanding.

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SPECIAL NOTE REGARDING FORWARD-LOOKING STATEMENTS

This Quarterly Report on Form 10-Q, including the section entitled "Management's Discussion and Analysis of Financial Condition and Results of Operations," contains forward-looking statements regarding future events and our future results that are subject to the safe harbors created under the Securities Act of 1933, as amended, or the Securities Act, and the Securities Exchange Act of 1934, as amended, or the Exchange Act. Forward-looking statements relate to future events or our future financial performance. We generally identify forward-looking statements by terminology such as "may," "will," "would," "should," "expects," "plans," "anticipates," "could," "intends," "target," "projects," "contemplates," "believes," "estimates," "predicts," "assume," "intend," "potential," "continue" or other similar words or the negative of these terms. We have based these forward-looking statements largely on our current expectations about future events and financial trends that we believe may affect our business, financial condition and results of operations. The outcome of the events described in these forward-looking statements is subject to risks, uncertainties and other factors described in Part II, Item 1A. "Risk Factors" and elsewhere in this Quarterly Report on Form 10-Q and Part I, Item 1A. "Risk Factors" of our Annual Report on Form 10-K for the fiscal year ended December 31, 2023, filed with the SEC on April 1, 2024. Accordingly, you should not place undue reliance upon these forward-looking statements. We cannot assure you that the events and circumstances reflected in the forward-looking statements will be achieved or occur, the timing of events and circumstances and actual results could differ materially from those anticipated in the forward-looking statements. Forward-looking statements contained in this report include, but are not limited to, statements about:

- our ability to integrate Acer (as defined below) into our business successfully or realize the anticipated synergies and related benefits of the Merger (as defined below);
- the progress of, outcome or and timing of any regulatory approval for any of our product candidates and the expected amount or timing of any payment related thereto under any of our collaboration agreements;
- our ability to continue as a going concern;
- the progress of, timing of and expected amount of expenses associated with our research, development and commercialization activities;
- our ability to raise additional funds on commercially reasonable terms, or at all, in order to support our continued operations;
- the sufficiency of our cash resources to fund our operating expenses and capital investment requirements for any period;
- the expected timing of our clinical trials for our product candidates and the availability of data and results of those trials;
- our expectations regarding federal, state and foreign regulatory requirements;
- the potential therapeutic benefits and effectiveness of our products and product candidates;
- the size and characteristics of the markets that may be addressed by our products and product candidates;
- our intention to seek to establish, and the potential benefits to us from, any strategic collaborations or partnerships for the development or sale of our products and product candidates; if approved;
- our expectations as to future financial performance, expense levels and liquidity sources;
- the timing of commercializing our products and product candidates, if approved;
- senior leadership and board member transitions and refreshments; and
- other factors discussed elsewhere in this report.

The forward-looking statements made in this report relate only to events as of the date on which the statements are made. We have included or made reference to important factors in the cautionary statements included in this report, particularly in the section entitled "Risk Factors" where we make reference to Part I, Item 1A. "Risk Factors" of our Annual Report on Form 10-K for the fiscal year ended December 31, 2023, filed with the SEC on April 1, 2024, that we believe could cause actual results or events to differ materially from the forward-looking statements that we make. Our forward-looking statements do not reflect the potential impact of any future acquisitions, mergers, dispositions, joint ventures or investments we may make. Except as required by law, we do not assume any intent to update any forward-looking statements after the date on which the statement is made, whether as a result of new information, future events or circumstances or otherwise.

NOTE REGARDING COMPANY REFERENCE

Unless the context otherwise requires, we use the terms "Zevra," "Company," "we," "us" and "our" in this Quarterly Report on Form 10-Q to refer to Zevra Therapeutics, Inc., formerly known as KemPharm, Inc. prior to February 21, 2023. We have proprietary rights to a number of trademarks used in this Quarterly Report on Form 10-Q that are important to our business, including LAT[®] and the Zevra logo. All other trademarks, trade names and service marks appearing in this Quarterly Report on Form 10-Q are the property of their respective owners. Solely for convenience, the trademarks and trade names in this Quarterly Report on Form 10-Q are referred to without the[®] and[™] symbols, but such references should not be construed as any indicator that their respective owners will not assert, to the fullest extent under applicable law, their rights thereto.

On August 30, 2023, the Company and Aspen Z Merger Sub, Inc., an indirect wholly-owned subsidiary of Zevra ("Merger Sub") entered into an Agreement and Plan of Merger (the "Merger Agreement") with Acer Therapeutics Inc. ("Acer"). On November 17, 2023 (the "Closing Date"), we completed the acquisition of Acer. Pursuant to the Merger Agreement, on the Closing Date, Merger Sub was merged with and into Acer (the "Merger"), with Acer continuing as the surviving entity and as a wholly-owned subsidiary of Zevra.

PART I — FINANCIAL INFORMATION

ITEM 1. UNAUDITED CONDENSED CONSOLIDATED FINANCIAL STATEMENTS

ZEVRA THERAPEUTICS, INC.
UNAUDITED CONDENSED CONSOLIDATED BALANCE SHEETS
(in thousands, except share and par value amounts)

	June 30, 2024	December 31, 2023
Assets		
Current assets:		
Cash and cash equivalents	\$ 39,260	\$ 43,049
Securities at fair value	9,998	24,688
Accounts and other receivables	8,947	17,377
Prepaid expenses and other current assets	2,686	1,824
Total current assets	60,891	86,938
Inventories	10,198	9,841
Property and equipment, net	678	736
Operating lease right-of-use assets	911	790
Goodwill	4,701	4,701
Intangible assets, net	66,154	69,227
Other long-term assets	875	94
Total assets	\$ 144,408	\$ 172,327
Liabilities and stockholders' equity		
Current liabilities:		
Accounts payable and accrued expenses	\$ 20,452	\$ 28,403
Line of credit payable	—	37,700
Current portion of operating lease liabilities	596	543
Current portion of discount and rebate liabilities	6,768	4,550
Other current liabilities	2,633	2,524
Total current liabilities	30,449	73,720
Long-term debt	58,328	5,066
Warrant liability	7,856	16,100
Operating lease liabilities, less current portion	544	456
Discount and rebate liabilities, less current portion	8,115	7,663
Other long-term liabilities	6,638	7,458
Total liabilities	111,930	110,463
Commitments and contingencies (Note D)		
Stockholders' equity:		
Preferred stock:		
Undesignated preferred stock, \$0.0001 par value, 10,000,000 shares authorized, no shares issued or outstanding as of June 30, 2024, or December 31, 2023	—	—
Common stock, \$0.0001 par value, 250,000,000 shares authorized, 43,567,156 shares issued and 41,991,464 shares outstanding as of June 30, 2024; 43,110,360 shares issued and 41,534,668 shares outstanding as of December 31, 2023	4	4
Additional paid-in capital	479,361	472,664
Treasury stock, at cost	(10,983)	(10,983)
Accumulated deficit	(436,325)	(399,778)
Accumulated other comprehensive income (loss)	421	(43)
Total stockholders' equity	32,478	61,864
Total liabilities and stockholders' equity	\$ 144,408	\$ 172,327

See accompanying notes to unaudited condensed consolidated financial statements.

ZEVRA THERAPEUTICS, INC.
UNAUDITED CONDENSED CONSOLIDATED STATEMENTS OF OPERATIONS
(in thousands, except share and per share amounts)

	Three months ended June 30,		Six months ended June 30,	
	2024	2023	2024	2023
Revenue, net	\$ 4,449	\$ 8,470	\$ 7,874	\$ 11,646
Cost of product revenue (excluding \$1,546 and \$3,074 in intangible asset amortization for the three and six months ended June 30, 2024, respectively shown separately below)	3,573	677	3,748	802
Intangible asset amortization	1,546	—	3,074	—
Operating expenses:				
Research and development	10,521	7,433	22,798	16,088
Selling, general and administrative	12,604	6,612	22,535	13,839
Total operating expenses	23,125	14,045	45,333	29,927
Loss from operations	(23,795)	(6,252)	(44,281)	(19,083)
Other income (expense):				
Interest expense	(2,110)	(197)	(2,845)	(379)
Fair value adjustment related to warrant and CVR liability	5,779	2,118	9,406	575
Fair value adjustment related to investments	1	131	(26)	327
Interest and other income (expense), net	270	1,553	1,199	2,593
Total other income	3,940	3,605	7,734	3,116
Loss before income taxes	(19,855)	(2,647)	(36,547)	(15,967)
Income tax benefit	(70)	74	—	177
Net loss	\$ (19,925)	\$ (2,573)	\$ (36,547)	\$ (15,790)
Basic and diluted net loss per share of common stock:				
Net loss	\$ (0.48)	\$ (0.08)	\$ (0.87)	\$ (0.46)
Weighted average number of shares of common stock outstanding:				
Basic and diluted	41,899,087	33,898,233	41,839,582	34,180,818

See accompanying notes to unaudited condensed consolidated financial statements.

ZEVRA THERAPEUTICS, INC.
UNAUDITED CONDENSED CONSOLIDATED STATEMENTS OF COMPREHENSIVE LOSS
(in thousands)

	Three months ended June 30,		Six months ended June 30,	
	2024	2023	2024	2023
Net loss	\$ (19,925)	\$ (2,573)	\$ (36,547)	\$ (15,790)
Other comprehensive loss:				
Foreign currency translation adjustment	280	(162)	464	(338)
Other comprehensive loss	280	(162)	464	(338)
Comprehensive loss	<u>\$ (19,645)</u>	<u>\$ (2,735)</u>	<u>\$ (36,083)</u>	<u>\$ (16,128)</u>

See accompanying notes to unaudited condensed consolidated financial statements.

ZEVRA THERAPEUTICS, INC.
UNAUDITED CONDENSED CONSOLIDATED STATEMENTS OF CHANGES IN STOCKHOLDERS' EQUITY
(in thousands)

	Common Stock	Additional Paid-in Capital	Treasury Stock, at cost	Accumulated Deficit	Other Comprehensive Income (loss)	Total Stockholders' Equity
Balance as of January 1, 2024	\$ 4	\$ 472,664	\$ (10,983)	\$ (399,778)	\$ (43)	\$ 61,864
Net loss	—	—	—	(16,622)	—	(16,622)
Stock-based compensation expense	—	2,119	—	—	—	2,119
Issuance of common stock in exchange for consulting services	—	56	—	—	—	56
Issuance of common stock as part of the Employee Stock Purchase Plan	—	71	—	—	—	71
Issuance of common stock for options exercised	—	1,146	—	—	—	1,146
Other comprehensive loss	—	—	—	—	184	184
Balance as of March 31, 2024	\$ 4	\$ 476,056	\$ (10,983)	\$ (416,400)	\$ 141	\$ 48,818
Net loss	—	—	—	(19,925)	—	(19,925)
Stock-based compensation expense	—	2,632	—	—	—	2,632
Issuance of common stock in exchange for consulting services	—	193	—	—	—	193
Issuance of common stock as part of the Employee Stock Purchase Plan	—	480	—	—	—	480
Other comprehensive loss	—	—	—	—	280	280
Balance as of June 30, 2024	<u>\$ 4</u>	<u>\$ 479,361</u>	<u>\$ (10,983)</u>	<u>\$ (436,325)</u>	<u>\$ 421</u>	<u>\$ 32,478</u>

See accompanying notes to unaudited condensed consolidated financial statements.

ZEVRA THERAPEUTICS, INC.
UNAUDITED CONDENSED CONSOLIDATED STATEMENTS OF CHANGES IN STOCKHOLDERS' EQUITY, CONTINUED
(in thousands)

	Common Stock	Additional Paid-in Capital	Treasury Stock, at cost	Accumulated Deficit	Other Comprehensive Income	Total Stockholders' Equity
Balance as of January 1, 2023	\$ 3	\$ 436,269	\$ (7,536)	\$ (353,729)	\$ 113	\$ 75,120
Net loss	—	—	—	(13,217)	—	(13,217)
Stock-based compensation expense	—	945	—	—	—	945
Shares repurchased as part of the Share Repurchase Program	—	—	(3,447)	—	—	(3,447)
Issuance of common stock in exchange for consulting services	—	42	—	—	—	42
Other comprehensive loss	—	—	—	—	(176)	(176)
Balance as of March 31, 2023	\$ 3	\$ 437,256	\$ (10,983)	\$ (366,946)	\$ (63)	\$ 59,267
Net income	—	—	—	(2,573)	—	(2,573)
Stock-based compensation expense	—	2,151	—	—	—	2,151
Issuance of common stock in exchange for consulting services	—	25	—	—	—	25
Issuance of common stock as part of the Employee Stock Purchase Plan	—	165	—	—	—	165
Other comprehensive loss	—	—	—	—	(162)	(162)
Balance as of June 30, 2023	\$ 3	\$ 439,597	\$ (10,983)	\$ (369,519)	\$ (225)	\$ 58,873

See accompanying notes to unaudited condensed consolidated financial statements.

ZEVRA THERAPEUTICS, INC.
UNAUDITED CONDENSED CONSOLIDATED STATEMENTS OF CASH FLOWS
(in thousands)

	Six months ended June 30,	
	2024	2023
Cash flows from operating activities:		
Net loss	\$ (36,547)	\$ (15,790)
Adjustments to reconcile net loss to net cash used in operating activities:		
Stock-based compensation expense	4,751	3,096
Inventory obsolescence charge	3,219	—
Non-cash interest expense	710	—
Depreciation and amortization expense	3,124	157
Fair value adjustment related to warrant and CVR liability	(9,406)	(481)
Fair value adjustment related to investments	26	(327)
Consulting fees paid in common stock	249	67
Loss (gain) on foreign currency exchange	188	(138)
Change in assets and liabilities:		
Accounts and other receivables	8,430	(5,734)
Prepaid expenses and other assets	(862)	(335)
Inventories	(3,576)	125
Operating lease right-of-use assets	298	161
Accounts payable and accrued expenses	(8,602)	2,947
Discount and rebate liability	2,670	3,097
Operating lease liabilities	(278)	(216)
Other liabilities	332	622
Net cash used in operating activities	(35,274)	(12,749)
Cash flows from investing activities:		
Purchases of property and equipment	—	(52)
Purchases of investments	(129)	(17,467)
Maturities of investments	14,793	34,000
Net cash provided by investing activities	14,664	16,481
Cash flows from financing activities:		
Proceeds from issuance of debt, net of lender fees	58,990	12,800
Payment of third-party debt issuance costs	(2,091)	—
Repayment of debt	(42,700)	(13,007)
Proceeds from insurance financing arrangements	1,082	1,256
Proceeds from Employee Stock Purchase Plan	551	166
Payments of principal on insurance financing arrangements	(431)	(564)
Payments to repurchase shares as part of the Share Repurchase Program	—	(3,447)
Proceeds from issuance of stock	1,146	—
Repayment of principal on finance lease liabilities	—	(3)
Net cash provided by (used in) financing activities	16,547	(2,799)
Effect of exchange rate changes on cash and cash equivalents	274	(203)
Net (decrease) increase in cash and cash equivalents	(3,789)	730
Cash and cash equivalents, beginning of period	43,049	65,466
Cash and cash equivalents, end of period	\$ 39,260	\$ 66,196
Supplemental cash flow information:		
Cash paid for interest	\$ 2,135	\$ 261

See accompanying notes to unaudited condensed consolidated financial statements.

ZEVRA THERAPEUTICS, INC.
NOTES TO UNAUDITED CONDENSED CONSOLIDATED FINANCIAL STATEMENTS

A. Description of Business, Basis of Presentation, and Significant Transactions

Organization

Zevra Therapeutics, Inc. (the "Company") is a rare disease company combining science, data and patient needs to create transformational therapies for diseases with limited or no treatment options. The Company has a diverse portfolio of products and product candidates, which includes preclinical development programs, clinical stage pipeline and commercial stage assets. The Company's pipeline includes arimoclomol, an orally-delivered, first-in-class investigational product candidate being developed for Niemann-Pick disease type C ("NPC"), which has been granted orphan drug designation, Fast-track designation, Breakthrough Therapy designation and rare pediatric disease designation for the treatment of NPC by the U.S. Food and Drug Administration ("FDA") and orphan medical product designation for the treatment of NPC by the European Medicines Agency ("EMA"). The arimoclomol New Drug Application ("NDA") for NPC was resubmitted to the FDA on December 21, 2023. On August 2, 2024, the FDA convened a meeting with the recently formed Genetic Metabolic Diseases Advisory Committee ("GeMDAC") to review the NDA for arimoclomol at which the GeMDAC voted favorably that the data supports that arimoclomol is effective in the treatment of patients with NPC. On August 9, 2024, the Company received the first round of arimoclomol labeling comments from the FDA and is evaluating the feedback. The FDA has assigned a PDUFA date of September 21, 2024. KP1077 is the Company's lead clinical development product candidate which is being developed as a treatment for idiopathic hypersomnia ("IH"), a rare neurological sleep disorder, and narcolepsy. KP1077 is comprised solely of serdexmethylphenidate ("SDX"), the Company's proprietary prodrug of d-methylphenidate ("d-MPH"). The FDA has granted KP1077 orphan drug designation for the treatment of IH. OLPRUVA® (sodium phenylbutyrate) for oral suspension is approved by the FDA for the treatment of urea cycle disorders ("UCDs"). The Company also has a pipeline of investigational product candidates, including celirolol for the treatment of vascular Ehlers-Danlos syndrome in patients with a confirmed type III collagen mutation.

The Company changed its name from KemPharm, Inc. to Zevra Therapeutics, Inc. effective as of February 21, 2023. On March 1, 2023, following its name change, the Company's common stock began trading on the Nasdaq Global Select Market under the ticker symbol "ZVRA".

Going Concern

The accompanying unaudited condensed consolidated financial statements have been prepared on a going concern basis, which contemplates the realization of assets and the satisfaction of liabilities in the normal course of business. During the six months ended June 30, 2024, and 2023, the Company incurred a net loss of \$36.5 million and \$15.8 million, respectively, and as of June 30, 2024, has an accumulated deficit of \$436.3 million. The Company has sustained operating losses for the majority of its corporate history and expects to continue to incur operating losses and negative operating cash flows until revenues reach a level sufficient to support ongoing operations.

The Company's ability to continue operating as a going concern is contingent upon its ability to generate revenue from approved products or obtain product candidate regulatory approvals, which would generate revenue, milestones, and cash flow sufficient to support ongoing operations and the satisfaction of financial covenants. These factors raise substantial doubt as to the Company's ability to continue as a going concern for at least one year from the date the unaudited condensed consolidated financial statements are being issued. The unaudited condensed consolidated financial statements do not include any adjustments that might result from the outcome of these uncertainties.

The Company's liquidity needs will be largely determined by the success of operations through the progression of its products and product candidates in the future. The Company also may consider other sources to fund operations including: (1) out-licensing rights to certain of its technologies and product candidates, pursuant to which the Company would receive cash royalties and milestones; (2) raising additional capital through equity or debt financings or from other sources; (3) obtaining product candidate regulatory approvals, which would generate revenue, milestones and cash flow; (4) reducing spending on one or more research and development programs, including by discontinuing development; and/or (5) restructuring operations to change its overhead structure. The Company is in the early stages of its commercialization effort for OLPRUVA and does not yet have a substantial basis to project future earnings, and its other sources of revenue are not sufficient to sustain its present activities on their own. Accordingly, the Company's ability to continue as a going concern may require it to obtain additional financing to fund its operations.

Basis of Presentation

The Company prepared the unaudited condensed consolidated financial statements in accordance with U.S. GAAP and the rules and regulations of the Securities and Exchange Commission ("SEC") and, in the Company's opinion, reflect all adjustments, including normal recurring items that are necessary.

Merger

On August 30, 2023, the Company and Aspen Z Merger Sub, Inc., a wholly-owned subsidiary of Zevra ("Merger Sub"), entered into an Agreement and Plan of Merger (the "Merger Agreement") with Acer Therapeutics Inc. ("Acer"), a pharmaceutical company focused on development and commercialization of therapies for rare and life-threatening diseases. On November 17, 2023 (the "Closing Date"), the Company completed the acquisition of Acer. Pursuant to the Merger Agreement, on the Closing Date, Merger Sub was merged with and into Acer (the "Merger"), with Acer continuing as the surviving entity and as a wholly-owned subsidiary of Zevra. In connection therewith, Zevra also purchased Acer's secured debt from Nantahala Capital Management, LLC ("NCM"), certain of its affiliates and certain other parties (collectively with NCM, "Nantahala") through a series of transactions and Zevra agreed to provide Acer with a bridge loan facility for up to \$18.0 million ("Bridge Loan"), subject to certain terms and conditions. The Merger expanded Zevra's rare disease portfolio, as well as increased and diversified its revenues with the addition of a U.S. commercial asset, OLPRUVA, indicated for the treatment of UCDs. See Note L for further discussion related to the Merger.

Registration Statements on Form S-3

In connection with the Merger, Zevra and Nantahala concurrently entered into a registration rights agreement, pursuant to which Zevra agreed to file a resale registration statement with respect to the resale of the Zevra common stock issuable to Nantahala. On February 5, 2024, Zevra filed a registration statement on Form S-3 (File No. 333-276856) registering an aggregate of 2,269,721 shares of Zevra's common stock. On April 5, 2024, we filed an amendment to the registration statement on Form S-3 (File No. 333-250945) covering the issuance of the shares of our common stock issuable upon the exercise of warrants issued in connection with the Merger (Note L) and remaining unexercised as of the date of the amendment, which was declared effective on April 8, 2024.

On June 4, 2024, the Company filed a registration statement on Form S-3 (File No. 333-279941) (the "June 2024 Registration Statement") under which the Company may sell securities, including as may be issuable upon conversion, redemption, repurchase, exchange or exercise of securities, in one or more offerings up to a total aggregate offering price of \$350.0 million, \$75.0 million of which was allocated to the sale of the shares of common stock issuable under the 2024 ATM Agreement (as described further below). The registration statement was declared effective on June 13, 2024.

August 2024 Offering

On August 8, 2024, the Company entered into an underwriting agreement (the "Underwriting Agreement") with Cantor Fitzgerald & Co. and William Blair & Company, L.L.C., as representatives of the several underwriters named therein (collectively, the "Underwriters"), in connection with the offering, issuance and sale by the Company of 9,230,770 shares of the Company's common stock at a public offering price of \$ 6.50 per share, pursuant to the June 2024 Registration Statement and a related prospectus supplement dated August 8, 2024 filed with the SEC (the "August 2024 Offering"). Under the terms of the Underwriting Agreement, the Company also granted the Underwriters an option exercisable for 30 days to purchase up to an additional 1,384,615 shares of its Common Stock at the public offering price, less underwriting discounts and commissions, which the Underwriters exercised in full on August 9, 2024. The August 2024 Offering closed on August 12, 2024. Total shares issued were 10,615,385. Net proceeds from the offering were approximately \$64.5 million, after deducting underwriting discounts and commissions and estimated offering expenses payable by the Company. The Company intends to use the net proceeds of the offering to support the pre-commercial launch activities for arimoclomol, continued commercial support for OLPRUVA and the continued development of celirolol and KP1077 through potential NDA filings and other general corporate purposes.

Based on the planned use of proceeds, the Company believes that the net proceeds from the August 2024 Offering and its existing cash and cash equivalents will be sufficient to fund its operating expenses and capital expenditure requirements into the first quarter of 2027, subject to continuing compliance with the Company's debt covenants.

Entry into 2024 ATM Agreement

On July 12, 2024, the Company entered into an equity distribution agreement (the "2024 ATM Agreement") with Citizens JMP Securities LLC ("Citizens JMP") under which the Company may offer and sell, from time to time at its sole discretion, shares of its common stock having an aggregate offering price of up to \$75.0 million through Citizens JMP as its sales agent. The issuance and sale, if any, of common stock by the Company under the 2024 ATM Agreement will be made pursuant to the June 2024 Registration Statement, the accompanying prospectus, and the related prospectus supplement dated July 12, 2024. Citizens JMP may sell the common stock by any method permitted by law deemed to be an "at the market offering" as defined in Rule 415 of the Securities Act. Citizens JMP will use commercially reasonable efforts to sell the common stock from time to time, based upon instructions from the Company (including any price, time or size limits or other customary parameters or conditions the Company may impose). The Company will pay Citizens JMP a commission equal to 3.0% in the aggregate of the gross sales proceeds of any common stock sold through Citizens JMP under the 2024 ATM Agreement.

Termination of 2021 ATM Agreement

On July 2, 2021, the Company entered into an equity distribution agreement (the "2021 ATM Agreement") with JMP Securities LLC ("JMP") and RBC Capital Markets, LLC ("RBCCM") under which the Company may offer and sell, from time to time at its sole discretion, shares of its common stock having an aggregate offering price of up to \$75.0 million through JMP and RBCCM as its sales agents. The issuance and sale, if any, of common stock by the Company under the 2021 ATM Agreement will be made pursuant to the registration statement on Form S-3 (File No. 333-257661) which was declared effective on July 12, 2021, the accompanying prospectus, and the related prospectus supplement dated July 12, 2021. JMP and RBCCM may sell the common stock by any method permitted by law deemed to be an "at the market offering" as defined in Rule 415 of the Securities Act. JMP and RBCCM will use commercially reasonable efforts to sell the common stock from time to time, based upon instructions from the Company (including any price, time or size limits or other customary parameters or conditions the Company may impose). The Company will pay JMP and RBCCM a commission equal to 3.0% in the aggregate of the gross sales proceeds of any common stock sold through JMP and RBCCM under the 2021 ATM Agreement. As of June 30, 2024, no shares have been issued or sold under the 2021 ATM Agreement. The 2021 ATM Agreement terminated on July 11, 2024.

Share Repurchase Program

On December 20, 2021, the Company initiated a share repurchase program (the "Share Repurchase Program") pursuant to which the Company was able to repurchase up to \$50 million of shares of its common stock through December 31, 2023. On December 31, 2023, the Share Repurchase Program ended, and the Company had repurchased 1,575,692 shares of its common stock for approximately \$ 11.0 million.

B. Summary of Significant Accounting Policies

Use of Estimates

The preparation of financial statements in conformity with U.S. GAAP requires the Company to make estimates and assumptions that affect the amounts reported in the unaudited condensed consolidated financial statements and accompanying notes. Actual results could differ from those estimates.

On an ongoing basis, the Company evaluates its estimates and assumptions, including those related to revenue recognition, the useful lives of property and equipment, the recoverability of long-lived assets, the incremental borrowing rate for leases, and assumptions used for purposes of determining stock-based compensation, income taxes, the fair value of investments and the fair value of the warrant liabilities, contingent value right ("CVR") liabilities and discount and rebate liabilities, among others. The Company bases its estimates on historical experience and on various other assumptions that it believes to be reasonable, the results of which form the basis for making judgments about the carrying value of assets and liabilities.

Investments

The Company maintains investment securities that are classified as available-for-sale securities for which the Company has elected the fair value option under Accounting Standards Codification ("ASC") 825, *Financial Instruments* ("ASC 825"). As such, these securities are carried at fair value with unrealized gains and losses included in fair value adjustment related to investments on the unaudited condensed consolidated statements of operations. The securities primarily consist of U.S. Treasury securities and are included in securities at fair value in the unaudited condensed consolidated balance sheets. As of June 30, 2024, and December 31, 2023, the Company held securities with an aggregate fair value of \$ 10.0 million and \$24.7 million, respectively, that contained aggregate unrealized (losses) gains of approximately (\$0.1) million and \$0.6 million, respectively. Applying fair value accounting to these debt securities more accurately represents the Company's investment strategy due to the fact that excess cash is currently being invested for the purpose of funding future operations. Interest income is recognized as earned using an effective yield method giving effect to the amortization of premium and accretion of discount and is based on the economic life of the securities. Interest income is included in interest and other income, net in the unaudited condensed consolidated statements of operations.

Variable Interest Entities

The primary beneficiary of a variable interest entity ("VIE") is required to consolidate the assets and liabilities of the VIE. When the Company obtains a variable interest in another entity, it assesses at the inception of the relationship and upon occurrence of certain significant events whether the entity is a VIE, and if so, whether the Company is the primary beneficiary of the VIE based on its power to direct the activities of the VIE that most significantly impact the VIE's economic performance and the Company's obligation to absorb losses or the rights to receive benefits from the VIE that could potentially be significant to the VIE.

To assess whether the Company has the power to direct the activities of the VIE that most significantly impact the VIE's economic performance, the Company considers all the facts and circumstances, including the Company's role in establishing the VIE and the Company's ongoing rights and responsibilities. The assessment includes identifying the activities that most significantly impact the VIE's economic performance and identifying which party, if any, has the power to direct those activities. In general, the parties that make the most significant decisions affecting the VIE (management and representation on the Board of Directors) are deemed to have the power to direct the activities of a VIE.

To assess whether the Company has the obligation to absorb losses of the VIE or the rights to receive benefits from the VIE that could potentially be significant to the VIE, the Company considers all of its economic interests that are deemed to be variable interests in the VIE.

This assessment requires judgement in determining whether these interests, in the aggregate, are considered potentially significant to the VIE. As of June 30, 2024, and December 31, 2023, the Company identified Acer to be the Company's sole interest in a VIE (Note L). As Zevra is the final decision maker for all of Acer's research, development, and commercialization of drug candidates that it is producing, the Company directs the activities of Acer that most significantly impact its performance. Therefore, the Company is the primary beneficiary of this VIE for accounting purposes.

Revenue Recognition

The Company recognizes revenue in accordance with the provisions of ASC 606, *Revenue from Contracts with Customers* ("ASC 606") and, as a result, follows the five-step model when recognizing revenue: 1) identifying a contract; 2) identifying the performance obligations; 3) determining the transaction price; 4) allocating the price to performance obligations; and 5) recognizing revenue when the performance obligations have been fulfilled.

Licensing Agreements

The terms of the Company's licensing agreements typically include one or more of the following: (i) upfront fees; (ii) milestone payments related to the achievement of development, regulatory, or commercial goals; and (iii) royalties on net sales of licensed products. Each of these payments may result in licensing revenues.

As part of the accounting for these agreements, the Company must develop estimates and assumptions that require judgment to determine the underlying stand-alone selling price for each performance obligation which determines how the transaction price is allocated among the performance obligations. Generally, the estimation of the stand-alone selling price may include such estimates as independent evidence of market price, forecasted revenues or costs, development timelines, discount rates, and probability of regulatory success. The Company evaluates each performance obligation to determine if they can be satisfied at a point in time or over time, and it measures the services delivered to the licensee which are periodically reviewed based on the progress of the related program. The effect of any change made to an estimated input component and, therefore revenue or expense recognized, would be recorded as a change in estimate. In addition, variable consideration (e.g., milestone payments) must be evaluated to determine if it is constrained and, therefore, excluded from the transaction price.

Up-front Fees: If a license to the Company's intellectual property is determined to be distinct from the other performance obligations identified in the arrangement, the Company recognizes revenues from the transaction price allocated to the license when the license is transferred to the licensee and the licensee is able to use and benefit from the license. For licenses that are bundled with other promises, the Company utilizes judgment to assess the nature of the combined performance obligation to determine whether the combined performance obligation is satisfied over time or at a point in time.

Milestone Payments: At the inception of each arrangement that includes milestone payments (variable consideration), the Company evaluates whether the milestones are considered probable of being reached and estimates the amount to be included in the transaction price using the most likely amount method. If it is probable that a significant revenue reversal would not occur, the associated milestone value is included in the transaction price. Milestone payments that are not within the Company's or the licensee's control, such as non-operational developmental and regulatory approvals, are generally not considered probable of being achieved until those approvals are received. At the end of each reporting period, the Company re-evaluates the probability of achievement of milestones that are within its or the licensee's control, such as operational developmental milestones and any related constraint, and if necessary, adjusts its estimate of the overall transaction price. Any such adjustments are recorded on a cumulative catch-up basis, which would affect collaboration revenues and earnings in the period of adjustment. Revisions to the Company's estimate of the transaction price may also result in negative licensing revenues and earnings in the period of adjustment.

Product Revenues, Net

Net revenues from product sales is recognized at the transaction price when the customer obtains control of the Company's products, which occurs at a point in time, typically upon receipt of the product by the customer.

The Company's net revenues represent total revenues adjusted for discounts and allowances, including estimated cash discounts, chargebacks, rebates, returns, copay assistance, data fees and wholesaler fees for services. These adjustments represent variable consideration under ASC 606 and are recorded as a reduction of revenue. These adjustments are established by management as its best estimate based on available information and will be adjusted to reflect known changes in the factors that impact such allowances. Adjustments for variable consideration are determined based on the contractual terms with customers, historical trends, communications with customers and the levels of inventory remaining in the distribution channel, as well as expectations about the market for the product and anticipated introduction of competitive products.

Inventories

The value of inventory is recorded at its net realizable value. The Company determines the cost of its other inventories, which includes amounts related to materials and manufacturing overhead, on a first-in, first-out basis.

The Company may scale-up and make commercial quantities of its product candidates prior to the date it anticipates that such product will receive final regulatory approval. The scale-up and commercial production of pre-launch inventory involves the risk that such products may not be approved for marketing on a timely basis, or ever. This risk notwithstanding, the Company may scale-up and build pre-launch inventory of product that have not received final regulatory approval when the Company believes such action is appropriate in relation to the commercial value of the product launch opportunity. Inventory manufactured prior to regulatory approval is recorded as research and development expense until regulatory approval for the product is obtained. Inventory used in clinical trials is also expensed as research and development expense, when selected for such use. Inventory that can be used in either the production of clinical or commercial products is expensed as research and development costs when identified for use in a clinical manufacturing campaign. The cost of finished goods inventory that is shipped to a customer to support the Company's patient assistance programs is expensed when those shipments take place. As of June 30, 2024, and December 31, 2023, the Company did not have pre-launch inventory that qualified for capitalization.

The Company performs an assessment of the recoverability of capitalized inventory during each reporting period and writes down any excess and obsolete inventory to its net realizable value in the period in which the impairment is first identified. Such impairment charges, should they occur, are recorded as a component of cost of product revenue in the statements of operations and comprehensive loss. The determination of whether inventory costs will be realizable requires the use of estimates by management. If actual market conditions are less favorable than projected by management, additional write-downs of inventory may be required. Additionally, the Company's product is subject to strict quality control and monitoring that it performs throughout the manufacturing process. In the event that certain batches or units of product do not meet quality specifications, the Company will record a charge to cost of product revenue, to write-down any unmarketable inventory to its estimated net realizable value. For the three and six months ended June 30, 2024, the Company recognized a charge of approximately \$ 3.2 million related to write-downs for unmarketable inventory. No such write downs were recognized for the three and six months ended June 30, 2023.

Foreign currency

Assets and liabilities are translated into the reporting currency using the exchange rates in effect on the unaudited condensed consolidated balance sheet dates. Equity accounts are translated at historical rates, except for the change in retained earnings during the year, which is the result of the income statement translation process. Revenue and expense accounts are translated using the weighted average exchange rate during the period. The cumulative translation adjustments associated with the net assets of foreign subsidiaries are recorded in accumulated other comprehensive income/loss in the accompanying unaudited condensed consolidated statements of stockholders' equity.

Debt Issuance Costs

Debt issuance costs incurred in connection with financing arrangements are recorded as a reduction of the related debt on the balance sheet and amortized over the life of the respective financing arrangement using the effective interest method.

Warrants

The Company accounts for warrants as either equity-classified or liability-classified instruments based on an assessment of the warrant's specific terms and applicable authoritative guidance in FASB ASC Topic 480, *Distinguishing Liabilities from Equity* ("ASC 480") and FASB ASC Topic 815, *Derivatives and Hedging* ("ASC 815"). The assessment considers whether the warrants are freestanding financial instruments pursuant to ASC 480, meet the definition of a liability pursuant to ASC 480, and whether the warrants meet all of the requirements for equity classification under ASC 815, including whether the warrants are indexed to our own stock and whether the warrant holders could potentially require "net cash settlement" in a circumstance outside of the company's control, among other conditions for equity classification. This assessment, which requires the use of professional judgment, is conducted at the time of warrant issuance and as of each subsequent quarterly period end date while the warrants are outstanding.

For warrants that meet all criteria for equity classification, the warrants are required to be recorded as a component of additional paid-in capital, on the unaudited condensed consolidated statement of stockholders' deficit at the time of issuance. For warrants that do not meet all the criteria for equity classification, the warrants are required to be recorded at their initial fair value on the date of issuance, and on each balance sheet date thereafter. Changes in the estimated fair value of the warrants are recognized as a non-cash gain or loss in other expense, net, on the unaudited condensed consolidated statement of operations. The fair value of the warrants was estimated using the Black-Scholes option pricing model.

New Accounting Pronouncements Not Yet Adopted

Segment Reporting (Topic 280): Improvements to Reportable Segment Disclosures

In November 2023, the FASB issued Accounting Standards Update ("ASU") No. 2023-07, *Segment Reporting (Topic 280): Improvements to Reportable Segment Disclosures*, which modifies the disclosure and presentation requirements of reportable segments. The amendments in the update require the disclosure of significant segment expenses that are regularly provided to the chief operating decision maker ("CODM") and included within each reported measure of segment profit and loss. The amendments also require disclosure of all other segment items by reportable segment and a description of its composition. Additionally, the amendments require disclosure of the title and position of the CODM and an explanation of how the CODM uses the reported measure(s) of segment profit or loss in assessing segment performance and deciding how to allocate resources. This update is effective for annual periods beginning after December 15, 2023, and interim periods within fiscal years beginning after December 15, 2024. Early adoption is permitted. The Company is currently evaluating the impact that this guidance will have on the presentation of its consolidated financial statements and accompanying notes.

Income Taxes (Topic 740): Improvements to Income Tax Disclosures

In December 2023, the FASB issued ASU No. 2023- 09, *Income Taxes (Topic 740): Improvements to Income Tax Disclosures*, which expands disclosures in an entity's income tax rate reconciliation table and disclosures regarding cash taxes paid both in the U.S. and foreign jurisdictions. The update will be effective for annual periods beginning after December 15, 2024. The Company is currently evaluating the impact that this guidance will have on the presentation of its consolidated financial statements and accompanying notes.

C. Inventories

	June 30, 2024	December 31, 2023
Raw materials	\$ 4,822	\$ 2,938
Work in progress	3,321	1,884
Finished goods	2,055	5,019
Total inventory	<u>\$ 10,198</u>	<u>\$ 9,841</u>

D. Debt Obligations

Long-term debt consisted of the following (in thousands):

	June 30, 2024	December 31, 2023
Notes payable	\$ 60,484	\$ —
Secured promissory note	—	5,000
Unamortized original issue (discount) premium	(938)	148
Less: debt issuance costs	(1,218)	(82)
	<u>\$ 58,328</u>	<u>\$ 5,066</u>

Secured Promissory Note

In connection with the Merger (Note L), on August 30, 2023, the Company and Nantahala, entered into a secured promissory note payable by Zevra to Nantahala in the original principal amount of \$5.0 million (the "Nantahala Note"). The Nantahala Note initially bore interest at 9.0% per annum, payable quarterly in arrears in cash. The interest rate increased to 12.0% per annum effective March 1, 2024, as the Nantahala Note remained unpaid six months from its issue date. The additional 3.0% interest would have been paid in shares of Zevra's common stock based on the volume weighted average trading price ("VWAP") of Zevra's common stock during the twenty consecutive trading days ending on the date before such interest payment date. Beginning on the first interest payment date following the second anniversary of the Nantahala Note, and on each interest payment date thereafter, Zevra was required to make \$0.6 million amortization payments on the Nantahala Note until it was paid in full. All principal and unpaid interest on the Nantahala Note would have been due on August 30, 2026, the third anniversary of the Nantahala Note. Zevra was entitled to prepay the Nantahala Note at any time without penalty. The Nantahala Note was secured by Zevra's interest in (i) the loan assets under the Loan Purchase Agreement described in Note L; (ii) the note assets under the Note Purchase Agreement described in Note L; (iii) the Bridge Loan described in Note L; and (iv) the proceeds therefrom. The Company used the proceeds from the Nantahala Note, along with \$12.0 million in cash and 98,683 shares of Zevra's common stock, to acquire Acer's term loans, as more fully described in Note L. In April 2024, the Nantahala Note was repaid in full and terminated. At the time of repayment, Nantahala elected to receive a cash payment in lieu of shares of Zevra's common stock in exchange for the additional 3.0% interest accrued for the period from March 1, 2024, through April 5, 2024.

Line of Credit

On January 26, 2023, the Company and Wells Fargo, as lender, entered into a revolving margin account agreement. The Company's investments were used as collateral for the loan and the amount the Company was able to borrow was limited to 80-90% of its outstanding investment balance held with Wells Fargo. The margin account bore interest at the Prime rate minus 225 basis-points. In April 2024, the Company repaid the outstanding balance under the margin account with Wells Fargo, and upon such repayment, the margin capabilities were removed from the account. As of December 31, 2023, \$37.7 million was outstanding under the margin account.

Term Loans

On April 5, 2024 (the "Term Loans Closing Date"), the Company entered into a credit agreement (the "Credit Agreement") with HCR Stafford Fund II, L.P., HCR Potomac Fund II, L.P., and Perceptive Credit Holdings IV, LP (collectively, the "Lenders"), and Alter Domus (US) LLC, as administrative agent (the "Administrative Agent").

Under the terms of the Credit Agreement, the Lenders provided a senior secured loan facility to the Company in the aggregate principal amount of \$ 100.0 million, which is divided into three tranches as follows: (i) \$60.0 million which was funded in full on the Term Loans Closing Date; (ii) \$20.0 million which is available to the Company in up to two drawings, each in an amount not to exceed \$10.0 million, at the Company's option until 18 months following the Term Loans Closing Date; and (iii) \$20.0 million which is available to the Company upon approval by the FDA of the NDA for arimoclomol for the treatment of NPC, at the Company's option until December 31, 2024 (collectively, the "Term Loans").

The principal amount of the Term Loans outstanding (the "Outstanding Principal Amount") will bear interest at a rate equal to 3-Month Term SOFR *plus* 7.00% per annum. If the net product sales for the calendar year ending December 31, 2025, exceed \$100.0 million, the Outstanding Principal Amount will bear interest at 3-Month Term SOFR *plus* 6.00% per annum. If the net product sales for the calendar year ending December 31, 2025, do not exceed \$100.0 million, then for any subsequent period of four consecutive fiscal quarters ending on or after March 31, 2026, in which net product sales exceed \$125.0 million, the Outstanding Principal Amount will bear interest at 3-Month Term SOFR *plus* 6.50% per annum. In all cases, the 3-Month Term SOFR rate will be subject to a floor of 4.00% per annum. Interest will be payable quarterly in arrears on the last day of each calendar quarter. The Company has the option to pay up to 25% of the interest in-kind beginning on the Term Loans Closing Date, through and including June 30, 2026. The Company has recognized approximately \$0.4 million of interest-in-kind as of June 30, 2024, which is included in long-term debt in the unaudited condensed consolidated balance sheet. The Term Loans will mature on the fifth anniversary of the Term Loans Closing Date. In connection with the Credit Agreement, the Company incurred approximately \$2.2 million of costs, which primarily consisted of underwriting, legal and other professional fees, and are included as a reduction to the carrying amount of the related debt liability and are deferred and amortized over the remaining life of the financing using the effective interest method.

The Credit Agreement contains customary affirmative and negative covenants by the Company, which among other things, will require the Company to provide certain financial reports to the Lenders within 60 days after the end of each of the first three fiscal quarters of each fiscal year and 105 days after the end of each fiscal year, meet certain minimum net product sales amounts, and limit the ability of the Company to incur or guarantee additional indebtedness, engage in certain transactions, and effect a consolidation or merger without consent. In addition, as long as the line of credit remains active, the Company must maintain a minimum cash balance of \$20.0 million to ensure the Company can meet its immediate capital needs. The obligations of the Company under the Credit Agreement may be accelerated upon customary events of default, including non-payment of principal, interest, fees and other amounts, covenant defaults, insolvency, material judgments, or inaccuracy of representations and warranties. The Term Loans are secured by a first priority perfected lien on, and security interest in, substantially all current and future assets of the Company. The proceeds of the Term Loans were used to refinance certain existing indebtedness of the Company and its subsidiaries. The Company will use the remaining proceeds to pay fees and expenses related to the debt financing and fund the development and commercialization of OLPRUVA, and arimoclomol, if approved.

Future minimum principal payments under the Term Loans as of June 30, 2024, were as follows (in thousands):

Year Ending December 31,	
2024 (excluding the six months ending June 30, 2024)	\$ —
2025	—
2026	—
2027	—
2028	—
Thereafter	60,442
Total minimum payments	60,442
Less: unamortized debt discount, debt issuance costs and paid in kind interest	(2,114)
Secured promissory note, net	\$ 58,328

E. Revenue, net

Licensing Agreements

AZSTARYS License Agreement

The Company entered into a Collaboration and License Agreement (the "AZSTARYS License Agreement") with Commave Therapeutics SA ("Commave"), an affiliate of Gurnet Point Capital. Under the AZSTARYS License Agreement, as amended, the Company granted to Commave an exclusive, worldwide license to develop, manufacture and commercialize the Company's product candidates containing SDX and d-MPH, including AZSTARYS, or any other product candidates containing SDX and developed to treat ADHD or any other central nervous system disorder. Corium Inc. was tasked by Commave to lead all commercialization activities for AZSTARYS under the AZSTARYS License Agreement. Pursuant to the AZSTARYS License Agreement, Commave agreed to pay milestone payments up to an aggregate of \$590.0 million upon the occurrence of specified regulatory milestones related to AZSTARYS, additional fixed payments upon the achievement of specified U.S. sales milestones, and quarterly, tiered royalty payments based on a range of percentages of net sales (as defined in the AZSTARYS License Agreement). Commave is obligated to make such royalty payments on a product-by-product basis until expiration of the royalty term for the applicable product.

The AZSTARYS License Agreement is within the scope of ASC 606, as the transaction represents a contract with a customer where the participants function in a customer / vendor relationship and are not exposed equally to the risks and rewards of the activities contemplated under the AZSTARYS License Agreement.

The Company concluded that these regulatory milestones, sales milestones and royalty payments each contain a significant uncertainty associated with a future event. As such, these milestone and royalty payments are constrained at contract inception and are not included in the transaction price as the Company could not conclude that it is probable a significant reversal in the amount of cumulative revenue recognized will not occur surrounding these milestone payments. At the end of each reporting period, the Company updates its assessment of whether the milestone and royalty payments are constrained by considering both the likelihood and magnitude of the potential revenue reversal. For the three and six months ended June 30, 2024, the Company recognized \$1.3 million and \$2.5 million of revenue under the AZSTARYS License Agreement, respectively, primarily related to royalties. For the three and six months ended June 30, 2023, the Company recognized revenue under the AZSTARYS License Agreement of \$ 5.7 million and \$6.3 million, respectively, which includes recognition of a \$5.0 million net sales milestone that was met in June 2023. There was no deferred revenue related to this agreement as of June 30, 2024, or December 31, 2023.

In accordance with the terms of the Company's Termination Agreement with Aquestive Therapeutics ("Aquestive") dated March 20, 2012, Aquestive has the right to receive an amount equal to 10% of any royalty or milestone payments made to the Company related to AZSTARYS or KP1077 under the AZSTARYS License Agreement.

Relief Exclusive License Agreement

As a condition to entering into the Merger Agreement, Acer and Relief Therapeutics SA ("Relief") entered into an exclusive license agreement on August 30, 2023 (the "Relief License Agreement"). Pursuant to the Relief License Agreement, Relief will hold exclusive development and commercialization rights for OLPRUVA in the European Union, Liechtenstein, San Marino, Vatican City, Norway, Iceland, Principality of Monaco, Andorra, Gibraltar, Switzerland, United Kingdom, Albania, Bosnia, Kosovo, Montenegro, Serbia and North Macedonia ("Geographical Europe"). The Company will have the right to receive a royalty of up to 10% of the net sales of OLPRUVA in Geographical Europe. For the three and six months ended June 30, 2024, the Company did not recognize any revenue under the Relief License Agreement. There was no deferred revenue related to this agreement as of June 30, 2024, and December 31, 2023. For further discussion of the Relief License Agreement, see Note L.

Product Revenues, Net

Arimoclomol Expanded Access Program

Net revenue includes revenue from the sale of arimoclomol for the treatment of NPC under the remunerated expanded access compassionate use program in France ("French nATU"). An expanded access compassionate use program is a program giving specific patients access to a drug, which is not yet approved for commercial sale. Only drugs targeting serious or rare indications and for which there is currently no appropriate treatment are considered for expanded access compassionate use programs. Further, to be considered for the expanded access compassionate use program, the drug must have proven efficacy and safety and must either be undergoing price negotiations or seeking marketing approval.

In accordance with ASC 606, the Company recognizes revenue when fulfilling its performance obligation under the Arimoclomol Expanded Access Program ("Arimoclomol EAP") by transferring control of promised goods or services to its customer, in an amount that reflects the consideration that the Company expects to receive in exchange for those goods or services. In determining when the customer obtains control of the product, the Company considers certain indicators, including whether the Company has a present right to payment from the customer, whether title and/or significant risks and rewards of ownership have transferred to the customer and whether the customer acceptance has been received. Revenue is recognized net of sales deductions, including discounts, rebates, applicable distributor fees, and revenue-based taxes.

The French Health Authorities and the manufacturer have agreed to a price for sales during the French nATU, but the final transaction price depends on the terms and conditions in the contracts with the French Health Authorities and is subject to price negotiations with the French Health Authorities following market approval. Any excess in the price charged the manufacturer compared to the price agreed with the health authorities once the drug product is approved in France must be repaid. The repayment is considered in the clawback liability (rebate). An estimate of net revenue and clawback liability are recognized using the 'expected value' method. Accounting for net revenue and clawback liability requires determination of the most appropriate method for the expected final transaction price. This estimate also requires assumptions with respect to inputs into the method, including current pricing of comparable marketed products within the rare disease area in France. Management has considered the expected final sales price as well as the price of similar drug products. The Company is operating within a rare disease therapeutic area where there is unmet treatment need and hence a limited number of comparable commercialized drugs products. The limited available relevant market information for directly comparable commercialized drugs within rare disease increases the uncertainty in management's estimate.

For the three and six months ended June 30, 2024, the Company recognized revenue related to the Arimoclomol EAP in France of \$3.1 million and \$5.4 million, respectively, which is net of a clawback liability of \$1.7 million and \$3.0 million, respectively, and other gross to net adjustments. For the three and six months ended June 30, 2023, the Company recognized revenue related to the Arimoclomol EAP in France of \$2.8 million and \$4.5 million, respectively, which is net of a clawback liability of \$1.6 million and \$3.4 million, respectively, and other gross to net adjustments. The total estimated reserve liability as of June 30, 2024, and December 31, 2023, was \$14.9 million and \$12.2 million, respectively. As of June 30, 2024, and December 31, 2023, this estimated reserve liability is recorded as discount and rebate liabilities in the unaudited condensed consolidated balance sheets and is separated into current and long-term based upon the timing of the expected payment to the French regulators.

OLPRUVA Product Sales

On December 27, 2022, the FDA approved OLPRUVA (sodium phenylbutyrate), a prescription medicine used along with certain therapy, including changes in diet, for the long-term management of adults and children with UCDs weighing 44 pounds (20 kg) or greater and with a body surface area of 1.2m² or greater. On November 17, 2023, the Company acquired OLPRUVA in connection with the Merger (Note L). To commercialize OLPRUVA for oral suspension in the U.S. the Company has built marketing, sales, medical affairs, distribution, managerial and other non-technical capabilities or making arrangements with third parties to perform these services. The Company's current distributor for sales of OLPRUVA is a single specialty pharmacy provider. However, the Company intends to establish additional distributors such as other retail pharmacies and certain medical centers or hospitals. In addition to distribution agreements, the Company enters into arrangements with health care providers and payors that provide for government mandated and/or privately negotiated rebates with respect to the purchase of its products.

For the three and six months ended June 30, 2024, sales of OLPRUVA were de minimis.

Accounts and Other Receivables

Accounts and other receivables consist of receivables from product sales, receivables under the AZSTARYS License Agreement and Arimoclomol EAP, as well as income tax receivables and other receivables due to the Company. Receivables under the AZSTARYS License Agreement are recorded for amounts due to the Company related to reimbursable third-party costs and royalties on product sales. Receivables under the Arimoclomol EAP are recorded for product sales under the French nATU. These receivables are evaluated to determine if any reserve or allowance should be established at each reporting date. As of June 30, 2024, the Company had receivables related to the Arimoclomol EAP of \$5.2 million, AZSTARYS License Agreement of \$1.5 million, and other receivables of \$2.3 million. As of December 31, 2023, the Company had receivables related to the Arimoclomol EAP of \$4.7 million, AZSTARYS License Agreement of \$11.4 million, and other receivables of \$1.3 million. As of June 30, 2024, and December 31, 2023, no reserve or allowance for doubtful accounts had been established.

F. Commitments and Contingencies

From time to time, the Company is involved in various legal proceedings arising in the normal course of business. For some matters, a liability is not probable, or the amount cannot be reasonably estimated and, therefore, an accrual has not been made. However, for such matters when it is probable that the Company has incurred a liability and can reasonably estimate the amount, the Company accrues and discloses such estimates.

Stockholder Litigation Related to the Merger

On October 12, 2023, Brodsky & Smith, purporting to act as counsel for Jerry Beavee, who was asserted to be a stockholder of Acer, filed a complaint entitled *Jerry Beavee v. Acer Therapeutics Inc., et al.*, No. 1:23-cv-08995 in the United States District Court for the Southern District of New York (the "Action") alleging that defendants violated Section 14(a) and 20(a) of the Securities Exchange Act of 1934 by filing the Preliminary Merger Registration Statement which allegedly omitted certain information that such counsel asserts is material to Acer's required disclosure. On October 30, 2023, Acer filed with the SEC a Schedule DEF14A that contained additional information regarding the Merger, which mooted the disclosure claims alleged in the Action. On December 8, 2023, Jerry Beavee filed with the court a notice of dismissal of the Action without prejudice.

On October 20, 2023, Long Law, LLC and Acocelli Law, PLLC, purporting to act as counsel for Kevin Turner, who was asserted to be a stockholder of Acer, filed a complaint entitled *Kevin Turner v. Acer Therapeutics Inc., et al.*, No. 1:23-cv-01185 in the United States District Court for the District of Delaware alleging that defendants violated Section 14(a) and 20(a) of the Securities Exchange Act of 1934 as well as SEC Rule 14a-9 by filing the Definitive Proxy Statement which allegedly omitted certain information that such counsel asserts is material to Acer's required disclosure. The complaint prays that, if asserted omissions are not adequately corrected, then Turner will seek to enjoin Acer from holding a stockholder meeting to approve the Merger and, if the Merger closes, will seek to rescind it and seek an award of damages.

On October 20, 2023, Long Law, LLC, purporting to act as counsel for Matthew Jones, who was asserted to be a stockholder of Acer, filed a complaint entitled *Matthew Jones v. Acer Therapeutics Inc., et al.*, No. 1:23-cv-01186 in the United States District Court for the District of Delaware alleging that defendants violated Section 14(a) and 20(a) of the Securities Exchange Act of 1934 as well as SEC Rule 14a-9 by filing the Definitive Proxy Statement which allegedly omitted certain information that such counsel asserts is material to the Acer's required disclosure. The complaint prays that, if asserted omissions are not adequately corrected, then Jones will seek to enjoin Acer from holding a stockholder meeting to approve the Merger and, if the Merger closes, will seek to rescind it and seek an award of damages.

On June 19, 2024, the Company entered into a confidential fee agreement (the "Fee Agreement") to settle the above stockholder litigation matters related to the Merger. The parties to the Fee Agreement were entitled to an aggregate payment of approximately \$0.3 million, which was paid during the second quarter of 2024.

As of June 30, 2024, and December 31, 2023, no accruals were made related to commitments and contingencies.

G. Stock and Warrants

Authorized, Issued, and Outstanding Common Shares

As of June 30, 2024, and December 31, 2023, the Company had authorized shares of common stock of 250,000,000 shares. Of the authorized shares, 43,567,156 and 43,110,360 shares of common stock were issued as of June 30, 2024, and December 31, 2023, respectively, and 41,991,464 and 41,534,668 shares of common stock were outstanding as of June 30, 2024, and December 31, 2023, respectively.

As of June 30, 2024 and December 31, 2023, the Company had reserved authorized shares of common stock for future issuance as follows:

	June 30, 2024	December 31, 2023
Outstanding awards under equity incentive plans	9,849,495	8,023,142
Outstanding common stock warrants	5,483,537	5,603,729
Possible future issuances under equity incentive plans	3,824,574	1,728,885
Possible future issuances under employee stock purchase plans	1,242,425	1,340,172
Total common shares reserved for future issuance	20,400,031	16,695,928

Common Stock Activity

The following table summarizes common stock activity for the six months ended June 30, 2024:

	Shares of Common Stock
Balance as of January 1, 2024	41,534,668
Common stock issued as compensation to third parties	9,000
Common stock issued as a result of stock options exercised	306,826
Balance as of March 31, 2024	41,850,494
Common stock issued as a result of the Employee Stock Purchase Plan	97,747
Common stock issued as compensation to third parties	41,820
Common stock issued as a result of stock options exercised	1,403
Balance as of June 30, 2024	41,991,464

Authorized, Issued, and Outstanding Preferred Stock

As of June 30, 2024, and December 31, 2023, the Company had 10,000,000 shares of authorized preferred stock, none of which were designated, issued, or outstanding.

Warrants to Purchase Common Stock

The Company has issued warrants to purchase common stock to various third parties, of which 5,483,537 remain outstanding as of June 30, 2024, and are immediately exercisable. These warrants qualify as participating securities under ASC Topic 260, *Earnings per Share*, and are treated as such in the net loss per share calculation (Note J). The Company may be required to redeem these warrants for a cash amount equal to the Black-Scholes value of the portion of the warrants to be redeemed (the "Put Option").

In connection with the Merger (Note L), in November 2023, the Company directly issued to certain investors an aggregate of 1,382,489 shares of its common stock, par value \$0.0001 per share, and accompanying warrants to purchase up to 1,382,489 shares of its common stock (the "2023 Warrants") at a combined offering price of \$4.34 per share of common stock and the Warrants and an aggregate of 917,934 shares of its common stock in exchange for the cancellation of a warrant to purchase 2,920,306 shares of common stock of Acer. The Warrants are immediately exercisable and expire on November 22, 2028. The Company used the net proceeds of approximately \$6.0 million from the offering for general corporate purposes. These warrants are separately exercisable by the warrant holders. While the warrants are outstanding (but unexercised), the warrant holders will participate in any dividend or other distribution of the Company's assets to its common stockholders by way of return of capital or otherwise. As of June 30, 2024, and December 31, 2023, none of the warrants have been exercised. The warrants have been evaluated to determine the appropriate accounting and classification pursuant to ASC 480 and ASC 815. Generally, freestanding warrants should be classified as (i) liabilities if the warrant terms allow settlement of the warrant exercise in cash and (ii) equity if the warrant terms only allow settlement in shares of common stock.

The Company determined that its outstanding warrants and the Put Option should be recorded as a liability and stated at fair value at each reporting period. Changes to the fair value of the warrant liability are recorded through the unaudited condensed consolidated statements of operations as a fair value adjustment related to warrant and CVR liability. As of June 30, 2024, and December 31, 2023, the fair value of the liability associated with these warrants and the Put Option was approximately \$7.9 million and \$16.1 million, respectively. The fair value adjustment related to these warrants and the Put Option was approximately \$3.6 million and \$8.2 million of income for the three and six months ended June 30, 2024, respectively. The fair value adjustment related to these warrants and the Put Option was approximately \$2.1 million and \$0.6 million of income for the three and six months ended June 30, 2023, respectively.

H. Stock-Based Compensation

The Company maintains a stock-based compensation plan (the "Incentive Stock Plan") that governs stock awards made to employees and directors prior to completion of the IPO.

In November 2014, the Board of Directors of the Company ("the Board"), and in April 2015, the Company's stockholders, approved the Company's 2014 Equity Incentive Plan (the "2014 Plan"), which became effective in April 2015. The 2014 Plan provides for the grant of stock options, other forms of equity compensation, and performance cash awards. In June 2021, the Company's stockholders approved an Amended and Restated 2014 Equity Incentive Plan (the "A&R 2014 Plan"), following its adoption by the Board in April 2021, which among other things added 4,900,000 shares to the maximum number of shares of common stock to be issued under the plan and extended the annual automatic increases (discussed further below) until January 1, 2031 and eliminated individual grant limits that applied under the 2014 Plan to awards that were intended to comply with the exemption for "performance-based compensation" under Code Section 162(m). The maximum number of shares of common stock that may be issued under the A&R 2014 Plan is 9,932,883 as of June 30, 2024. The number of shares of common stock reserved for issuance under the A&R 2014 Plan will automatically increase on January 1 of each year, until and including January 1, 2031, by 4% of the total number of shares of the Company's capital stock outstanding on December 31 of the preceding calendar year, or a lesser number of shares determined by the Board. Pursuant to the terms of the 2014 Plan, on January 1, 2024, the common stock reserved for issuance under the 2014 Plan automatically increased by 1,661,386 shares.

During the three and six months ended June 30, 2024, 16,875, and 733,750 stock options were exercised, respectively. During the three and six months ended June 30, 2023, no stock options were exercised.

In June 2021, the Company's stockholders approved an Employee Stock Purchase Plan (the "ESPP"), following its adoption by the Board in April 2021. The maximum number of shares of common stock that may be issued under the ESPP is 1,500,000. The first offering period under the ESPP began on October 1, 2021, and the first purchase date occurred on May 31, 2022. As of June 30, 2024, 257,575 shares have been issued under the ESPP.

In January 2023, the Board approved the 2023 Employment Inducement Award Plan (the "2023 Plan"). The maximum number of shares of common stock that were initially available for issuance under the 2023 Plan was 1,500,000. In February 2024, the Board approved an amendment to the 2023 Plan to increase the aggregate number of shares of common stock available for issuance under the 2023 Plan from 1,500,000 to 4,500,000 shares.

In May 2023, the Board approved the Ninth Amended and Restated Non-Employee Director Compensation Policy (the "Non-Employee Director Compensation Policy"). The equity compensation made pursuant to the Non-Employee Director Compensation Policy will be granted under the A&R 2014 Plan.

Stock-based compensation expense recorded under the Incentive Stock Plan, A&R 2014 Plan, ESPP and 2023 Plan is included in the following line items in the accompanying unaudited condensed consolidated statements of operations (in thousands):

	Three months ended June 30,		Six months ended June 30,	
	2024	2023	2024	2023
Research and development	\$ 1,028	\$ 731	\$ 1,881	\$ 1,303
Selling, general and administrative	1,604	372	2,870	1,793
Total stock-based compensation expense	<u>\$ 2,632</u>	<u>\$ 1,103</u>	<u>\$ 4,751</u>	<u>\$ 3,096</u>

There was no stock-based compensation expense related to performance-based awards recognized during the six months ended June 30, 2024, and 2023.

I. Fair Value of Financial Instruments

The accounting standard for fair value measurements provides a framework for measuring fair value and requires disclosures regarding fair value measurements. Fair value is defined as the price that would be received to sell an asset or paid to transfer a liability in an orderly transaction between market participants at the measurement date, based on the Company's principal or, in absence of a principal, most advantageous market for the specific asset or liability.

The Company uses a three-tier fair value hierarchy to classify and disclose all assets and liabilities measured at fair value on a recurring basis, as well as assets and liabilities measured at fair value on a non-recurring basis, in periods subsequent to their initial measurement. The hierarchy requires the Company to use observable inputs when available, and to minimize the use of unobservable inputs, when determining fair value. The three tiers are defined as follows:

- Level 1—Observable inputs that reflect quoted market prices (unadjusted) for identical assets or liabilities in active markets;
- Level 2—Observable inputs other than quoted prices in active markets that are observable either directly or indirectly in the marketplace for identical or similar assets and liabilities; and
- Level 3—Unobservable inputs that are supported by little or no market data, which require the Company to develop its own assumptions.

The carrying amounts of certain financial instruments, including cash and cash equivalents, investments, and accounts payable and accrued expenses, approximate their respective fair values due to the short-term nature of such instruments.

Assets and Liabilities Measured at Fair Value on a Recurring Basis

The Company evaluates its financial assets and liabilities subject to fair value measurements on a recurring basis to determine the appropriate level in which to classify them for each reporting period. This determination requires significant judgments to be made. The following table summarizes the conclusions reached regarding fair value measurements as of June 30, 2024, and December 31, 2023 (in thousands):

	Balance as of June 30, 2024	Quoted Prices in Active Markets for Identical Assets (Level 1)	Significant Other Observable Inputs (Level 2)	Significant Unobservable Inputs (Level 3)
CVR liability (Note L)	6,100	\$ —	\$ —	\$ 6,100
Warrant liabilities	7,856	—	—	7,856
Total liabilities	<u>\$ 13,956</u>	<u>\$ —</u>	<u>\$ —</u>	<u>\$ 13,956</u>
Securities:				
U.S. Treasury securities	9,998	9,998	—	—
Total assets	<u>\$ 9,998</u>	<u>\$ 9,998</u>	<u>\$ —</u>	<u>\$ —</u>

	Balance as of December 31, 2023	Quoted Prices in Active Markets for Identical Assets (Level 1)	Significant Other Observable Inputs (Level 2)	Significant Unobservable Inputs (Level 3)
CVR liability (Note L)	\$ 7,262	\$ —	\$ —	\$ 7,262
Warrant liabilities	16,100	—	—	16,100
Total liabilities	<u>\$ 23,362</u>	<u>\$ —</u>	<u>\$ —</u>	<u>\$ 23,362</u>
Securities:				
U.S. Treasury securities	\$ 24,688	\$ 24,688	\$ —	\$ —
Total assets	<u>\$ 24,688</u>	<u>\$ 24,688</u>	<u>\$ —</u>	<u>\$ —</u>

The common stock warrant liabilities were recorded at fair value using the Black-Scholes option pricing model. The following assumptions were used in determining the fair value of the warrant liabilities valued using the Black-Scholes option pricing model as of June 30, 2024, and December 31, 2023:

	June 30, 2024	December 31, 2023
Risk-free interest rate	4.27% - 4.77%	3.76% - 4.12%
Volatility	57.09% - 86.67%	62.01% - 92.42%
Dividend yield	0%	0%
Expected term (years)	1.5 - 4.4	2.0 - 4.9
Weighted average fair value	\$ 1.43	\$ 2.94

The following table is a reconciliation for the common stock warrant liabilities measured at fair value using Level 3 unobservable inputs (in thousands):

Balance as of December 31, 2023	\$	16,100
Change in fair value measurement		(8,244)
Balance as of June 30, 2024	\$	7,856

For the six months ended June 30, 2024, and 2023, the changes in fair value of the warrant liabilities primarily resulted from the volatility of the Company's common stock and the change in the risk-free interest rates.

J. Net Loss Per Share

For all periods presented herein, the Company did not use the two-class method to compute net loss per share of common stock, even though it had issued securities, other than common stock, that contractually entitled the holders to participate in dividends and earnings, because these holders are not obligated to participate in a loss. The two-class method requires earnings for the period to be allocated between common stock and participating securities based upon their respective rights to receive distributed and undistributed earnings.

Under the two-class method, for periods with net income, basic net income per share of common stock is computed by dividing the undistributed net income by the weighted average number of shares of common stock outstanding during the period. Undistributed net income is computed by subtracting from net income the portion of current period earnings that participating securities would have been entitled to receive pursuant to their dividend rights had all of the period's earnings been distributed and subtracting the actual or deemed dividends declared. No such adjustment to earnings is made during periods with a net loss as the holders of the participating securities have no obligation to fund losses. Diluted net income per share of common stock is computed under the two-class method by using the weighted average number of shares of common stock outstanding plus the potential dilutive effects of stock options, warrants and other outstanding convertible securities. In addition to analyzing under the two-class method, the Company analyzes the potential dilutive effect of stock options and warrants, under the treasury-stock method and other outstanding convertible securities under the if-converted method when calculating diluted income (loss) per share of common stock, in which it is assumed that the stock options, warrants and other outstanding convertible securities convert into common stock at the beginning of the period or date of issuance, if the stock option, warrant or other outstanding convertible security was issued during the period. The Company reports the more dilutive of the approaches (two-class or treasury-stock/if-converted) as its diluted net income (loss) of common stock during the period.

As noted above, for all periods presented herein, the Company did not utilize the two-class approach as the Company was in a net loss position and the holders of the participating securities have no obligation to fund losses. The Company did analyze diluted net loss per share of common stock under the treasury-stock/if-converted method and noted that all outstanding stock options and warrants were anti-dilutive for the periods presented. For all periods presented, basic net loss per share of common stock was the same as diluted net loss per share of common stock.

The following securities, presented on a common stock equivalent basis, have been excluded from the calculation of weighted average number of shares of common stock outstanding because their effect is anti-dilutive:

	Three months ended June 30,		Six months ended June 30,	
	2024	2023	2024	2023
Awards under equity incentive plans	9,849,495	7,027,739	9,849,495	7,027,739
Common stock warrants	5,483,537	4,252,490	5,483,537	4,252,490
Total securities excluded from the calculation of weighted average number of shares of common stock outstanding	15,333,032	11,280,229	15,333,032	11,280,229

A reconciliation from net loss to basic and diluted net loss per share of common stock for the six months ended June 30, 2024, and 2023, is as follows (in thousands):

	Three months ended June 30,		Six months ended June 30,	
	2024	2023	2024	2023
Basic and diluted net loss per share of common stock:				
Net loss, basic and diluted	\$ (19,925)	\$ (2,573)	\$ (36,547)	\$ (15,790)
Weighted average number of shares of common stock outstanding, basic and diluted	41,899	33,898	41,840	34,181
Basic and diluted net loss per share of common stock	\$ (0.48)	\$ (0.08)	\$ (0.87)	\$ (0.46)

K. Leases

The Company has operating and finance leases for office space, laboratory facilities and various laboratory equipment, furniture and office equipment and leasehold improvements. The Company determines if an arrangement is a lease at contract inception. Lease assets and lease liabilities are recognized based on the present value of lease payments over the lease term at the commencement date. The Company does not separate lease and non-lease components. Leases with a term of twelve (12) months or less at commencement are not recorded on the unaudited condensed consolidated balance sheets. Lease expense for these arrangements is recognized on a straight-line basis over the lease term. The Company's leases have remaining lease terms of less than 1 year to approximately 3 years, some of which include options to extend the leases for up to 5 years, and some which include options to terminate the leases within 1 year.

Effective June 1, 2021, the Company agreed to sublease office space in Florida, comprised of one of the two contiguous suites, under a non-cancelable operating lease, which expires in February 2026.

The components of lease expense were as follows (in thousands):

Lease Cost	Three months ended June 30,		Six months ended June 30,	
	2024	2023	2024	2023
Finance lease cost:				
Amortization of right-of-use assets	\$ 12	\$ 32	\$ 24	\$ 64
Interest on lease liabilities	—	—	—	—
Total finance lease cost	12	32	24	64
Operating lease cost	118	114	211	227
Short-term lease cost	59	55	118	110
Variable lease cost	—	13	13	26
Less: sublease income	(39)	(39)	(78)	(78)
Total lease costs	\$ 150	\$ 175	\$ 288	\$ 349

Supplemental cash flow information related to leases was as follows (in thousands):

	Six months ended June 30,	
	2024	2023
Cash paid for amounts included in the measurement of lease liabilities:		
Financing cash flows from finance leases	\$ —	\$ 3
Operating cash flows from operating leases	263	284
Operating cash flows from short-term leases	118	110
Operating cash flows from variable lease costs	13	26
Right-of-use assets obtained in exchange for lease liabilities:		
Finance leases	\$ —	\$ —
Operating leases	419	—

Supplemental balance sheet information related to leases was as follows (in thousands, except weighted average remaining lease term and weighted average discount rate):

	June 30, 2024	December 31, 2023
Finance Leases		
Property and equipment, at cost	\$ 1,031	\$ 1,031
less: accumulated depreciation and amortization	(906)	(882)
Property and equipment, net	\$ 125	\$ 149
Other current liabilities	\$ —	\$ —
Other long-term liabilities	—	—
Total finance lease liabilities	\$ —	\$ —
Operating Leases		
Operating lease right-of-use assets	\$ 911	\$ 790
Total operating lease right-of-use assets	\$ 911	\$ 790
Current portion of operating lease liabilities	\$ 596	\$ 543
Operating lease liabilities, less current portion	544	456
Total operating lease liabilities	\$ 1,140	\$ 999
Weighted Average Remaining Lease Term		
Operating leases (in years)	3	2
Weighted Average Discount Rate		
Finance leases	0.0%	14.3%
Operating leases	8.5%	7.6%

Maturities of lease liabilities were as follows (in thousands):

Year Ending December 31,	Finance Leases	Operating Leases
2024 (excluding the six months ended June 30, 2024)	\$ —	\$ 332
2025	—	532
2026	—	141
2027	—	130
2028	—	154
Thereafter	—	39
Total lease payments	—	1,328
Less: future interest expense	—	(188)
Lease liabilities	\$ —	\$ 1,140

L. Merger

On August 30, 2023, in connection with the Merger Agreement with Acer, the following transactions occurred prior to Closing:

- *Bridge Loan* - Zevra and Acer entered into a bridge loan agreement (the "Bridge Loan Agreement"), providing for Zevra to make loans to Acer up to an aggregate principal amount of \$16.5 million. The Bridge Loan was provided to Acer to support its termination agreement with Relief Therapeutics Holding SA and to provide Acer with working capital, including for payments of accounts payable to support the commercial launch of OLPRUVA and the development of celiprolol pending the Merger's closure. On October 31, 2023, the Company and Acer entered into an amendment to the Bridge Loan Agreement, which increased the aggregate principal amount available under the loan from \$16.5 million to \$18.0 million.
- *Purchase of Acer's Term Loans* - Zevra purchased certain indebtedness of Acer held by Nantahala. Under the loan purchase with Nantahala, Zevra purchased (i) an original senior secured term loan facility made available to Acer in an aggregate amount of \$ 6.5 million and funded on March 14, 2022, and (ii) an additional senior secured term loan made to Acer in an aggregate amount of \$ 7.0 million in a single borrowing which funded on January 31, 2023 for (1) \$12.0 million in cash; (2) 98,683 shares of Zevra Common Stock; and (3) a secured Promissory Note payable by Zevra to Nantahala in the original principal amount \$5.0 million. These were recorded as receivables from Acer and were treated as a settlement of a preexisting relationship in connection with the closing of the transaction and recorded as a component of purchase consideration.
- *Purchase of Acer's Convertible Notes ("Marathon Convertible Notes")* - Under the Note Purchase Agreement with Nantahala, Zevra purchased the Marathon Convertible Notes that Nantahala had acquired on June 16, 2023. Zevra acquired the Marathon Convertible Notes in exchange for the issuance of 2,171,038 shares of Zevra Common Stock at \$ 5.0667 per share for a total purchase price of \$ 11.0 million.
- *Amendment to IP License Agreement and IP Termination Agreement* - As a condition to entering into the Merger Agreement, Acer and Relief entered into the Exclusive License Agreement and the Termination Agreement terminating the collaboration and license agreement, dated March 19, 2021, by and between Acer and Relief. Pursuant to the Exclusive License Agreement, Relief holds exclusive development and commercialization rights for OLPRUVA in Geographical Europe. Acer has the right to receive a royalty of up to 10.0% of the net sales of OLPRUVA in Geographical Europe. In accordance with the terms of the Termination Agreement, Relief received an upfront payment from Acer of \$10.0 million (which payment was funded with the Bridge Loan described above) with an additional payment of \$ 1.5 million due on the first-year anniversary of the \$10.0 million payment. Acer also agreed to pay a 10.0% royalty on net sales of OLPRUVA worldwide, excluding Geographical Europe, and 20.0% of any value received by Acer from certain third parties relating to OLPRUVA licensing or divestment rights, all of the foregoing which are capped at \$45.0 million, for total payments to Relief of up to \$ 56.5 million.

In connection with the closing of the Merger on November 17, 2023, each share of common stock of Acer was converted into the right to receive (i) 0.1210 fully paid and non-assessable shares of common stock of Zevra, par value \$ 0.0001 per share, and (ii) one non-transferable CVR to be issued by Zevra, which will represent the right to receive one or more contingent payments up to an additional \$ 76 million upon the achievement, if any, of certain commercial and regulatory milestones for Acer's OLPRUVA and celiprolol products 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).

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The assets acquired and liabilities assumed were recorded based on their acquisition date fair values. Consideration for the Merger was \$ 72.6 million and consists of (i) approximately 2.96 million shares of Zevra common stock valued at \$ 12.8 million, (ii) the Bridge Loan advances of \$ 17.8 million, (iii) \$12.0 million in cash paid to Nantahala; (iv) 2.27 million shares of Zevra Common Stock issued to Nantahala valued at \$ 11.5 million based on the VWAP of shares of Zevra Common Stock during the 20 consecutive trading days ending on the trading date prior to August 30, 2023; (v) a secured promissory note payable by Zevra to Nantahala in the original principal amount of \$5.0 million, as disclosed in Note C, (vi) \$8.5 million in the estimated fair value of contingent consideration related to the CVRs, (vii) approximately 0.9 million shares of Zevra Common Stock issued to a former holder of Acer warrants valued at \$4.0 million based on Zevra's common stock price on the Effective Date and (viii) \$ 1.0 million in notes payable paid by the Company on Acer's behalf. In addition, effective as of immediately prior to the Effective Time, all of the outstanding and unexercised Acer stock options were automatically cancelled and ceased to exist without any cash or other consideration being paid or provided in respect thereof. The following purchase price allocation reflects the preliminary estimates of and assumptions related to the fair values of assets acquired and liabilities assumed:

Assets		
Cash	\$	575
Prepaid expenses		278
Other current assets		11
Inventory		9,376
Property, plant, and equipment		35
Other noncurrent assets		209
Approved product - OLPRUVA		68,000
IPR&D - celiprolol		2,000
Goodwill acquired		4,701
		<u>85,185</u>
Liabilities		
Accounts payable and accrued expenses	\$	10,881
Deferred collaboration funding		1,500
Operating lease liabilities		175
		<u>12,556</u>
Fair Value of Net Assets Acquired	\$	<u>72,629</u>

The preliminary fair values assigned to the tangible and intangible assets acquired and liabilities assumed were determined using an income approach based on management's estimates and assumptions, as well as other information compiled by management, including third-party valuations that utilize customary valuation procedures and techniques. These preliminary fair values are subject to change within the one-year measurement period. The estimated fair values were developed by discounting future net cash flows to their present value at market-based rates of return. The goodwill acquired represents the excess of the purchase price and related costs over the value assigned to the net tangible and identifiable intangible assets of the business acquired. The useful lives of the intangible assets for amortization purposes were determined by considering the period of expected cash flows used to measure the fair values of the intangible assets adjusted as appropriate for entity-specific factors including legal, regulatory, contractual, competitive, economic and other factors that may limit the useful life. The marketed product asset is amortized on a straight-line basis over its estimated useful life. As of June 30, 2024, the in-process research and development ("IPR&D") project had not been completed or abandoned and, therefore, the IPR&D intangible asset is not currently subject to amortization.

The results of operations and changes in stockholders' equity for Acer were included in the Company's consolidated financial statements beginning November 18, 2023.

The following pro forma combined results of operations present the acquisition as if it had occurred on January 1, 2023. The pro forma combined results of operations do not necessarily represent the Company's consolidated results of operations had the acquisition occurred on the date assumed, nor are these results necessarily indicative of the Company's future consolidated results of operations. The Company expects to realize certain benefits from integrating Acer into the Company and to incur certain one-time costs. The pro forma combined results of operations do not reflect these benefits or costs.

	Three Months Ended June 30, 2023	Six Months Ended June 30, 2023
Pro forma revenue	\$ 8,470	\$ 11,646
Pro forma net loss	(8,211)	(28,863)

Cancellation of Acer Warrant

On November 22, 2023, the Company sold an aggregate of 917,934 shares of its common stock to a healthcare focused investment fund (the "Investor") to cancel a warrant held by the Investor to purchase 2,920,306 shares of common stock of Acer. The shares of common stock were offered and sold to the Investor in a registered direct offering without an underwriter or placement agent.

Contingent Consideration

Contingent consideration liabilities relate to our liabilities arising in connection with the CVRs issued as a result of the Merger. The contingent consideration is classified as Level 3 in the fair value hierarchy. The fair value is measured based on a Monte Carlo simulation or a scenario-based method, depending on the earn-out achievement objectives, utilizing projections about future performance. Significant inputs include volatility and projected financial information, including projections representative of a market participant's view of the expected cash payments associated with the agreed upon regulatory milestones based on probabilities of technical success, timing of the potential milestone events for the compounds, and estimated discount rates.

The following table provides a reconciliation of the beginning and ending balances related to the contingent consideration liabilities for the CVRs (dollars in thousands):

Balance at December 31, 2023	\$ 7,262
Change in fair value recognized in earnings	(1,162)
Balance at June 30, 2024	<u>\$ 6,100</u>

For the six months ended June 30, 2024, the Company recorded a \$1.2 million gain on the change in fair value of contingent consideration, primarily due to changes in market data and revenue projections.

M. Goodwill & Intangible Assets

The Company's goodwill balance was \$4.7 million as of June 30, 2024, and December 31, 2023.

As of June 30, 2024, and December 31, 2023, non-amortizable intangible assets include IPR&D of \$ 2.0 million

As of June 30, 2024, and December 31, 2023, the Company had a definite-lived intangible asset, net related to the acquisition of OLPRUVA as a result of the Merger of \$64.2 million and \$67.2 million, respectively. This is amortized on a straight-line basis over its estimated economic life of eleven years and is reviewed periodically for impairment. Amortization expense is recorded as intangible asset amortization in the unaudited condensed consolidated statements of operations and was \$1.5 million and \$3.1 million for the three and six months ended June 30, 2024. No amortization expense related to definite-lived intangible assets was recognized for the three and six months ended June 30, 2023.

For intangible assets subject to amortization, estimated amortization expense for the five fiscal years subsequent to June 30, 2024, is expected to be \$6.2 million per year.

N. Subsequent Events

The Company evaluated events and transactions occurring subsequent to June 30, 2024, through August 13, 2024, the date the accompanying unaudited condensed consolidated financial statements were issued. During this period, other than the August 2024 Offering and the related entry into the Underwriting Agreement, the entry into the Equity Distribution Agreement, the Genetic Metabolic Diseases Advisory Committee meeting, and receipt of the first round of arimoclomol labeling comments from the FDA, all of which are disclosed in Note A, there were no subsequent events that required recognition in the accompanying unaudited condensed consolidated financial statements, nor were there any additional non-recognized subsequent events that required disclosure.

ITEM 2. MANAGEMENT'S DISCUSSION AND ANALYSIS OF FINANCIAL CONDITION AND RESULTS OF OPERATIONS

You should read the following discussion and analysis of our financial condition and results of operations together with our unaudited condensed consolidated financial statements and related notes thereto included elsewhere in this Quarterly Report on Form 10-Q. Some of the information contained in this discussion and analysis or set forth elsewhere in this Quarterly Report on Form 10-Q, including information with respect to our plans and strategy for our business and related financing, includes forward-looking statements that involve risks and uncertainties. As a result of many factors, including those factors set forth in Part II, Item 1A. "Risk Factors" of this Quarterly Report on Form 10-Q and Part I, Item 1A. "Risk Factors" of our Annual Report on Form 10-K for the fiscal year ended December 31, 2023, filed with the SEC on April 1, 2024, our actual results could differ materially from the results described in or implied by the forward-looking statements contained in the following discussion and analysis.

Overview

We are a rare disease company combining science, data and patient need to create transformational therapies for diseases with limited or no treatment options. Our mission is to bring life changing therapeutics to people living with rare diseases. With unique, data-driven development and commercialization strategies, we are overcoming complex drug development challenges to make new therapies available to the rare disease community. We have a diverse portfolio of products and product candidates, which includes preclinical developmental programs, clinical stage pipeline and commercial stage assets. Our team has specialized expertise and a track record of success in advancing promising therapies that face complex clinical and regulatory challenges with an approach that balances science and data with patient need.

Following the U.S. approval of AZSTARYS® (further described below) in March 2021, we undertook a strategic process to evaluate how to leverage and potentially augment the Company's existing capabilities while also considering where to invest in our pipeline to generate long-term shareholder value. With a track record of drug development success leading to approvals for products which had either difficult pathways to approval or where approvals were won following a complete response letter ("CRL") from the U.S. Food and Drug Administration ("FDA"), the Company determined to focus its expertise on rare disease indications, as well as seeking value-creating opportunities by building and directly commercializing product candidates in lieu of an out-licensing model. We are executing on this balanced approach by building a culture that is patient-focused and driven by our commitment to developing and making available therapies which address the myriad unmet needs within the rare disease community.

As part of our commitment to serving the rare disease community, in February 2023, we changed our name to Zevra Therapeutics, Inc. Our name, Zevra, is the Greek word for zebra, which is the internationally recognized symbol for rare disease. This name reflects our intense focus and dedication to developing transformational, patient-focused therapies for rare diseases with limited or no treatment options available, or treatment areas with significant unmet needs.

In May 2022, we purchased all of the assets and operations of Orphazyme A/S related to arimocloamol, settled all of Orphazyme's actual outstanding liabilities to its creditors with a cash payment of \$12.8 million, and agreed to assume an estimated reserve clawback liability of \$5.2 million related to revenue generated from Orphazyme's Expanded Access Program in France (the "Arimocloamol EAP").

On November 17, 2023, Zevra completed the acquisition of Acer Therapeutics, Inc. ("Acer"). Pursuant to the Merger Agreement, Acer continues as a wholly-owned subsidiary of Zevra (the "Merger"). The Merger included the acquisition of OLPRUVA® (sodium phenylbutyrate) for oral suspension, which was approved by the FDA on December 27, 2022, for the treatment of certain urea cycle disorders ("UCDs"). Acer also had a pipeline of investigational product candidates, including celirolol for the treatment of Vascular Ehlers-Danlos syndrome ("VEDS") in patients with a confirmed type III collagen (COL3A1) mutation. At the effective time of the Merger (the "Effective Time"), each share of common stock of Acer, par value \$0.0001 per share, issued and outstanding immediately prior to the Effective Time (excluding cancelled shares and any shares held by holders who have exercised their appraisal rights) were converted into the right to receive (i) 0.1210 fully paid and non-assessable shares of common stock of Zevra, par value \$0.0001 per share, and (ii) one non-transferable contingent value right ("CVR") issued by Zevra, which represents the right to receive one or more contingent payments up to an additional \$76.0 million upon the achievement, if any, of certain commercial and regulatory milestones for Acer's OLPRUVA and celirolol products 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).

In order to accomplish our mission, we are seeking to further expand our pipeline through both internal development and through our business development activities to collaborate, partner, and potentially acquire additional assets. We intend to target assets that will allow us to leverage the expertise and infrastructure that we have built in order to mitigate risk and enhance our probability of success. In addition, we may consider external opportunities within neurology and neurodegenerative diseases, psychiatric disorders, and other rare diseases, along with adjacent or related therapeutic categories. If we are successful, expanding our pipeline could be accretive to our value proposition and has the potential to create incremental long-term value for stockholders.

Our recurring operating losses and negative cash flows from operations raise substantial doubt about our ability to continue as a going concern. The Company's ability to continue operating as a going concern is contingent upon its ability to generate revenue from approved products or obtain product candidate regulatory approvals, which would generate revenue, milestones, and cash flow sufficient to support ongoing operations and the satisfaction of financial covenants. We are early in our commercialization effort for OLPRUVA and do not yet have a substantial basis to project future earnings, and our other sources of revenue are not sufficient to sustain our present activities on their own. Accordingly, our ability to continue as a going concern may require us to obtain additional financing to fund our operations. The perception of our inability to continue as a going concern may make it more difficult for us to obtain financing for the continuation of our operations and could result in the loss of confidence by investors, suppliers and employees. Adequate additional financing may not be available to us on acceptable terms, or at all. To the extent that we raise additional capital through the sale of equity or debt, the terms of these securities may restrict our ability to operate. If we raise additional funds through collaborations, strategic alliances or marketing, distribution or licensing arrangements with third parties, we may be required to relinquish valuable rights. If we are unable to raise capital when needed or on attractive terms, we could be forced to delay, reduce or altogether cease our research and development programs or future commercialization efforts.

Our Product Candidates and Approved Products

We have built a diverse portfolio of products and product candidates through a combination of internal development and strategic investments through acquisition. For example, we have employed our proprietary Ligand Activated Technology ("LAT") platform to develop approved products (e.g., AZSTARYS), and clinical development candidate (KP1077IH and KP1077N). Through our business development efforts, we have added a commercial product (OLPRUVA), and clinical development candidates (arimoclomol, celiprolol). We furthermore have a variety of product candidates and compounds that are early-stage, pre-clinical and clinical-stage designed to address a variety of rare diseases and other indications.

Currently active commercial products and development assets are summarized in the table below:

Active Zevra Commercial and Development Assets

Parent Drug	Indication	Product / Candidate	Development Status	Next Milestone(s)
Sodium phenylbutyrate	Urea Cycle Disorders (UCD)	OLPRUVA	FDA Approved	Tracking Commercial Progress
Arimoclomol	Niemann Pick disease type C (NPC)	Arimoclomol	Pending FDA Review	PDUFA target date September 21, 2024
Celiprolol	Vascular Ehlers Danlos Syndrome (VEDS)	Celiprolol	Clinical - Phase 3	Phase 3 ongoing
Serdexmethylphenidate	Idiopathic Hypersomnia (IH)	KP1077IH	Clinical - Phase 2	Evaluation of potential Phase 3 Trial
Serdexmethylphenidate	Narcolepsy	KP1077N	Clinical - Phase 1/2	Evaluation of next steps based on potential Phase 3 Trial for KP1077IH
Serdexmethylphenidate and dexamethylphenidate	Attention Deficit and Hyperactivity Disorder (ADHD)	AZSTARYS	FDA Approved and Partnered	Collecting royalties and milestones

These anticipated milestones are based on information currently available to us. Our current plans and expectations are subject to a number of uncertainties, risks and other important factors that could materially impact our plans, including risks which are not solely within our control. See Part I, Item 1A. "Risk Factors" of our Annual Report on Form 10-K for the fiscal year ended December 31, 2023, filed with the SEC on April 1, 2024, as updated by Part II, Item 1A. "Risk Factors" of this Quarterly Report on Form 10-Q.

OLPRUVA

OLPRUVA (sodium phenylbutyrate) for oral suspension is approved in the U.S. as adjunctive therapy to standard of care, which includes dietary management, for the chronic management of UCDs involving deficiencies of carbamylphosphate synthetase (CPS), ornithine transcarbamylase (OTC), or argininosuccinic acid synthetase (AS). OLPRUVA is a proprietary and novel formulation of sodium phenylbutyrate powder, packaged in pre-measured single-dose envelopes, that has shown bioequivalence to existing sodium phenylbutyrate powder but with a pH-sensitive polymer coating that is designed to minimize dissolution of the coating for up to five minutes after preparation.

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. 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. Approximately 1 in 100,000 people have UCD, and there are an estimated 800 patients who are actively treated with nitrogen scavenging therapy in the U.S. While there are therapies currently approved for the treatment of UCDs - specifically RAVICTI®, marketed by Amgen, Inc. (formerly Horizon Therapeutics) and PHEBURANE®, marketed by Medunik USA - there remain unmet needs for this community of patients. OLPRUVA offers benefits over other UCD treatments by eliminating issues with palatability, offering improved portability with its single-dose envelopes, and it comes in a dosage that is personalized to the patient based on weight.

To commercialize OLPRUVA for oral suspension in the U.S. we have built marketing, sales, medical affairs, distribution, managerial and other non-technical capabilities or making arrangements with third parties to perform these services. During the quarter ended December 31, 2023, we began generating revenue from the sale of OLPRUVA in the U.S. For additional information regarding the Merger, see Note L of our unaudited condensed consolidated financial statements included elsewhere in this Quarterly Report on Form 10-Q. Zevra has a partnership with Relief Therapeutics who has rights to commercialize OLPRUVA in various European Union (EU) countries, if approved. In addition, Zevra pays royalties of 10% of U.S. net sales plus milestones to Relief Therapeutics.

During the first half of 2024, we initiated the commercial launch of OLPRUVA in the U.S. We have focused our initial efforts on the approximately 40 metabolic treatment centers of excellence across the United States which treat the majority of UCD patients to build awareness with physicians regarding the benefits of OLPRUVA. In the months since launch, our team has been able to engage with more than 90% of our customers. We have seen meaningful growth in reimbursement coverage, which was approximately 55% of U.S. covered lives at the time of acquisition, to now approximately 75%.

OLPRUVA summary:

- **OLPRUVA is available in the U.S. for the treatment of UCD.** OLPRUVA is an adjunctive therapy for long-term management of adults and children weighing 20kg or greater with UCD from deficiencies of CPS, OTC, or AS.
- **OLPRUVA is differentiated from currently available forms of phenylbutyrate.** OLPRUVA is formulated to improve palatability while providing patients with a portable and discrete pre-measured dose.
- **Zevra has assembled a team to support OLPRUVA and additional future commercial products.** We have established an efficient commercial team which is designed to fully service the patients and prescribers within the rare disease indications we are pursuing.

Arimoclomol

Arimoclomol is our product candidate being developed for the treatment of Niemann-Pick disease type C ("NPC"), an ultra-rare neurodegenerative lysosomal storage disorder ("LSD"). Arimoclomol is an orally delivered, first in-class investigational product candidate which has been granted orphan drug designation, Fast-Track designation, Breakthrough Therapy designation and rare pediatric disease designation for the treatment of NPC by the FDA, and orphan medicinal product designation for the treatment of NPC by the European Commission. The arimoclomol New Drug Application ("NDA") was submitted to the FDA on December 21, 2023, and is currently undergoing review by the FDA. On August 2, 2024, the FDA convened a meeting with the recently formed Genetic Metabolic Diseases Advisory Committee to review the NDA for arimoclomol at which the GeMDAC voted favorably that the data supports that arimoclomol is effective in the treatment of patients with NPC. On August 9, 2024, the Company received the first round of arimoclomol labeling comments from the FDA and is evaluating the feedback. The FDA has assigned a Prescription Drug User Fee Act (PDUFA) date of September 21, 2024. We believe that, if approved by the FDA, arimoclomol will be eligible to receive a Rare Pediatric Disease Priority Review Voucher ("PRV"), which is transferrable.

As an LSD, NPC is characterized by an inability of the body to transport cholesterol and lipids inside of cells. Symptoms of NPC include a progressive impairment of mobility, cognition, speech, and swallowing, often culminating in premature death. The incidence of NPC is estimated to be one in 100,000 to 130,000 live births. We estimate that there are approximately 1,800 individuals with NPC in the U.S. and Europe, of these, approximately 300 have been diagnosed in the U.S. However, diagnostic challenges may affect the number of potential patients, and we believe that the availability of treatment options could increase awareness of the disease and assist in more accurately identifying patients. Effective therapies to treat NPC are desperately needed, and for this reason, arimoclomol is currently being made available to NPC patients in the United States, France, Germany, and other EU member states under various expanded access programs ("EAPs").

On September 16, 2020, the previous sponsor of the arimoclomol program, Orphazyme, submitted an NDA to the FDA, seeking approval for arimoclomol to treat NPC. In June 2021, the FDA issued a complete response letter ("CRL"), meaning it determined that it could not approve the NDA in its present form.

Zevra acquired the assets of Orphazyme A/S ("Orphazyme") in May 2022, and took over the responsibility for arimoclomol, including the preparation and resubmission of the NDA designed to respond to the FDA's specific deficiencies identified in the CRL and feedback in subsequent meetings between the FDA and Orphazyme. Since that time, we have worked diligently to characterize the meaningful evidence of safety and efficacy of arimoclomol for its intended use and the substantial data generated since the CRL, including the recently completed four-year open-label safety trial, an interim analysis of which was presented at the 19th WorldSymposium™ in February 2023. Upon fulfilling the randomized double-blinded portion of the Phase 2/3 clinical trial, both placebo- and arimoclomol-treated patients were given the option to continue into the four-year (48 month) open-label-extension ("OLE"), phase of the study with arimoclomol treatment provided in addition to their current standard of care. We believe that the results from this analysis, based on up to four years of continuous treatment, suggest that arimoclomol may reduce the long-term progression of NPC.

In preparation of the arimoclomol NDA resubmission, we completed a meeting with the FDA in August 2023, receiving feedback that was used to finalize the NDA submission. The updated NDA package for arimoclomol was resubmitted to the FDA in December 2023. Zevra believes it has addressed the issues previously raised by the FDA in the 2021 CRL. Zevra has conducted additional studies to support the potential mechanism of action of arimoclomol. Additionally, new data was included in the resubmission as supportive evidence from multiple non-clinical studies, natural history comparisons, real-world data generated from the ongoing early access programs in the U.S. and the European Union, as well as data from the four-year open-label extension of the Phase 2/3 clinical trial (NCT02612129).

In January 2024, the FDA acknowledged receipt of the resubmission and, under PDUFA, deemed the arimoclomol NDA resubmission to be a Class II complete response which has a six-month review period from the date of resubmission. On March 4, 2024, we announced that the FDA had extended the review period for the NDA for arimoclomol and set a new PDUFA date of September 21, 2024. The FDA also presented the resubmission for discussion at an advisory committee meeting in August 2024. On August 2, 2024, the FDA convened a meeting with the recently formed Genetic Metabolic Diseases Advisory Committee to review the NDA for arimoclomol at which the GeMDAC voted favorably that the data supports that arimoclomol is effective in the treatment of patients with NPC.

In April 2024, the Company presented new long-term, real-world data from the expanded access program (EAP: NCT04316637) for the treatment of NPC at the *Society for Inherited Metabolic Disorders*. The study demonstrated that adults treated with arimoclomol, including those with and without miglustat use, generally had a stable disease course with a clinically meaningful slowing of disease progression over two years of treatment and follow-up and the safety profile was consistent with that observed in the Phase 2/3 study where no new safety adverse events were identified.

Zevra holds the global rights for arimoclomol. We are evaluating the possibility of seeking regulatory approval and commercialization outside of the US.

Arimoclomol summary:

- **Currently, no approved treatments for NPC in the U.S.** There are no currently approved products in the U.S. to treat the underlying disease of NPC and we believe, if approved, arimoclomol could be considered a foundational therapy for patients in the U.S.
- **Designed to address disease progression.** Arimoclomol is designed to address the symptoms of NPC by slowing the progression of the disease itself, rather than serving as a symptomatic treatment only. The Phase 2/3 trial data for arimoclomol in NPC demonstrated reduced disease progression, and long-term data from the 4-year OLE of the Phase 2/3 trial suggest improved outcomes vs. historical controls.
- **Ease of flexible administration as an oral treatment.** Arimoclomol is administered as an oral capsule that can be swallowed whole, opened and contents mixed with foods or liquids, or delivered through a feeding tube.
- **Extensive clinical experience with favorable safety data.** No significant safety findings have been reported with more than 600 patients treated in various clinical trials and through our EAPs.
- **Advantageous regulatory designations.** Arimoclomol has been granted orphan drug designation, Fast Track designation, and Breakthrough Therapy designation for the treatment of NPC. If approved for the treatment of NPC, we believe arimoclomol will be eligible to receive a PRV.

Celiprolol

The Merger with Acer included the acquisition of celiprolol. We are advancing celiprolol as an investigational product candidate for the treatment of VEDS in patients with a confirmed type III collagen (*COL3A1*) mutation. Celiprolol is a selective adrenergic modulator ("SAM") and, if we receive the first approval in the U.S. for celiprolol, we believe it would be deemed a new chemical entity ("NCE") in the U.S. Celiprolol is currently approved in the EU for the treatment of hypertension and angina.

Ehlers-Danlos Syndrome is an inherited disorder caused by mutations in the genes responsible for the structure, production, or processing of collagen, an important component of the connective tissues in the human body, or proteins that interact with collagen. VEDS 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. Gastrointestinal and uterine fragility or rupture also commonly occur in VEDS patients. Spontaneous arterial rupture has a peak incidence in the third or fourth decade of life in VEDS patients but may occur earlier and is the most common cause of sudden death in VEDS patients. Arterial rupture or dissection events occur in about 25% of patients before the age of 20 but increase to roughly 90% of patients by the age of 40. The median survival age of VEDS patients in the U.S. is 51 years, with arterial rupture being the most common cause of sudden death. Pregnancy-related complications also occur in women with VEDS and include arterial dissection or rupture, uterine rupture, hemorrhage, premature rupture of membranes, lacerations, and complications during and after surgery. The incidence of VEDS is estimated to be one in 50,000 to 200,000 people. There are approximately 7,500 patients in the U.S.

Currently, there are no approved therapies anywhere in the world for VEDS. However, celiprolol, prescribed off label, has become the standard of care therapy for VEDS in some European countries. Medical intervention for VEDS focuses on surgery, symptomatic treatment, genetic counseling, and prophylactic measures, such as avoiding intense physical activity, scuba diving, and violent sports. Arterial, digestive, or uterine complications in VEDS patients typically require immediate hospitalization, observation in an intensive care unit, and sometimes surgery. Pregnant women with VEDS are considered to be at risk and receive special care. While VEDS patients are encouraged to take steps to minimize the chances of an arterial rupture or dissection, there are no pharmacologic options to reduce the likelihood of such an event, and accordingly current treatments for VEDS focus on the repair of arterial ruptures or dissection. Therefore, patients must adopt a "watch and wait" approach following any confirmed diagnosis. Unfortunately, many of these arterial events have high mortality associated with them, and thus, a pharmacologic intervention that reduces the rate of events would be clinically meaningful.

Celiprolol has not been approved for any indication in the U.S. In the past, an NDA for celiprolol for the treatment for hypertension was submitted to the FDA by Rorer (subsequently acquired by Aventis Pharma SA (Aventis)) in June 1987, but was subsequently withdrawn prior to completion of the FDA review and therefore never approved. We have obtained the exclusive right in North and South America from Aventis to reference the celiprolol data included in the marketing authorization application dossier filed with and approved by the UK Medicines and Healthcare Products Regulatory Agency ("MHRA"). In addition, our wholly-owned subsidiary, Acer has licensed exclusive worldwide rights to the data from the Phase 3 clinical trial known as the BBEST trial which was sponsored by L'Assistance Publique Hôpitaux de Paris ("AP-HP").

Celiprolol received orphan drug designation from the FDA for the treatment of VEDS in 2015. In October 2018, a new celiprolol NDA was submitted to the FDA by Acer based on data obtained from the BBEST trial and was subsequently accepted by the FDA in October 2018 with priority review status. Following FDA review, Acer received a CRL from the FDA stating that it will be necessary to conduct an adequate and well-controlled trial to determine whether celiprolol reduces the risk of clinical events in patients with VEDS. Subsequently, Acer appealed the FDA decision, and while the FDA denied the appeal, it described possible paths forward toward approval. In a May 2021 Type B meeting with the FDA, Acer discussed the conduct of an U.S.-based prospective, randomized, double-blind, placebo-controlled, decentralized clinical trial in patients with *COL3A1* positive VEDS, and sought the FDA's opinion on various proposed design features of the study.

Based on FDA's feedback during the Type B meeting, we adopted a decentralized (virtual) event-based clinical trial design and use of an independent centralized adjudication committee with a primary endpoint based on clinical events associated with disease outcome. In April 2022, the FDA granted celiprolol Breakthrough Therapy designation ("BTD") in the U.S. for the treatment of patients with *COL3A1*-positive VEDS.

In July 2022, Acer initiated enrollment in a Phase 3 long-term event-driven clinical trial designed based on the discussions from the May 2021 Type B meeting with the FDA, also known as the DiSCOVER trial. The DiSCOVER trial intends to enroll 150 VEDS patients, with 100 patients receiving celiprolol and 50 patients receiving placebo. Recruitment in the Phase-3 trial was restarted earlier this year after a brief hiatus and the trial is actively enrolling patients. We believe that celiprolol could address significant unmet needs as there are currently no approved treatments for VEDS in the U.S.

Celiprolol summary:

- **Currently, no approved treatments for VEDS in the U.S.** There are currently no approved treatments of VEDS in the U.S. and we believe that celiprolol, if approved, could be a significant innovation in the treatment of VEDS in the U.S. where current treatment options are focused primarily on surgical intervention.
- **Unique pharmacological profile.** Mechanism of action in VEDS patients is thought to be through vascular dilatation and smooth muscle relaxation, the effect of which is to reduce the mechanical stress on collagen fibers in the arterial wall, and thereby potentially less incidence of vascular ruptures.
- **Evidence of efficacy in the EU and extensive clinical experience from multiple trials.** Celiprolol has become the primary treatment for VEDS patients in several European countries. BBEST Clinical Trial data showed 76% reduction in risk of arterial events observed in *COL3A1*+ subpopulation, with additional data from a long-term observational study in France.
- **Regulatory designations.** Celiprolol for VEDS would be considered an NCE in the U.S. and has been granted Orphan Drug designation and Breakthrough Therapy designation.
- **Solid patent protection through 2038.** Celiprolol is generally protected by U.S. patents that will expire, after utilizing all appropriate patent term adjustments but excluding possible term extensions, in 2038.

KP1077

KP1077 is being developed for the treatment of IH and narcolepsy. IH is a rare neurological sleep disorder affecting approximately 37,000 patients in the United States. The cardinal feature of IH is excessive daytime sleepiness ("EDS"), characterized by daytime lapses into sleep, or an irrepressible need to sleep that persists even with adequate or prolonged nighttime sleep. Additionally, those with IH have extreme difficulty waking, otherwise known as "sleep inertia," suffer from severe and debilitating brain fog, and may fall asleep unintentionally or at inappropriate times, also known as narcolepsy. These symptoms often further lead to reported memory problems, difficulty maintaining focus, and depression.

There is currently only one approved product for the treatment of IH, XYWAV®, developed by Jazz Pharmaceuticals. A second product, WAKIX®, developed by Harmony Biosciences and originally approved for the treatment of EDS or cataplexy in adult patients with narcolepsy, but in October 2023, Harmony announced that the difference in outcome for EDS when comparing WAKIX and placebo in its Phase 3 trial with IH patients did not reach statistical significance. Prescribers also utilize narcolepsy medications and various stimulant products "off-label" to treat IH symptoms, with methylphenidate, a stimulant which has been classified by the DEA as a Schedule II controlled substance, being one of the most commonly used stimulants for treating IH. While each of these medications can help to address certain IH symptoms, there are also potential shortcomings, including dosing inconvenience, serious adverse events, such as elevated blood pressure and heart rate, and significant drug-to-drug interactions ("DDIs"), including with medications used to manage contraception and depression. In addition, patients have indicated that the effectiveness of their current medication was poor.

Narcolepsy is a rare, chronic, debilitating neurologic disorder of sleep-wake state instability that impacts up to 200,000 Americans and is primarily characterized by EDS and cataplexy (sudden loss of muscle tone while a person is awake) along with other manifestations of rapid eye movement ("REM"), sleep dysregulation, which intrude into wakefulness. In most patients, narcolepsy is caused by the loss of hypocretin, a neuropeptide in the brain that supports sleep-wake state stability. Typical symptom onset occurs in adolescence or young adulthood, but it can take up to a decade to be properly diagnosed. Although there are several approved medications for narcolepsy, we believe a treatment option based on serdexmethylphenidate ("SDX"), our proprietary prodrug of d-methylphenidate ("d-MPH") which has previously been classified as a Schedule IV controlled substance, with superior exposure/duration characteristics and low abuse potential may be beneficial.

We reported top-line data from a Phase 1 proof-of-concept study of SDX in the fourth quarter of 2021 and final data for the Phase 1 proof-of-concept study of SDX in the first quarter of 2022. The proof-of-concept study was a dose-escalation study to evaluate the pharmacokinetics, pharmacodynamic stimulant effects, and safety of single oral doses of SDX in subjects with a history of high-dose stimulant use. In the trial, 240 mg and 360 mg doses of SDX were observed to be well-tolerated and produced d-MPH exposure that appeared to increase proportionally with dose. Mean d-MPH plasma concentrations showed a gradual increase after SDX administration, reaching a broad peak from eight to twelve hours post-dose, followed by a shallow decline thereafter. Increased wakefulness, alertness, hypervigilance, and insomnia effects were reported by study participants, which we believe suggests that SDX produced targeted pharmacodynamic effects that have the potential to benefit patients with IH and other sleep disorders. In November 2022, we announced that the FDA has granted the orphan drug designation to SDX for the treatment of IH.

In January 2022, we announced that we had selected KP1077 for the treatment of IH and narcolepsy as our lead clinical development candidate. KP1077 utilizes SDX, our prodrug of d-MPH, as its API. During the first quarter of 2022, we initiated a Phase 1 clinical trial comparing the cardiovascular safety of SDX to immediate-release and long-acting formulations of RITALIN®, a commonly prescribed central nervous system ("CNS") stimulant. In September 2022, we announced topline data from our exploratory Phase 1 clinical trial, which showed the potential for higher dose formulations of SDX to be safe and well tolerated while avoiding the potential for greater cardiovascular safety risk compared to immediate-release and long-acting formulations of Ritalin.

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Based on the data, in December 2022, we announced the initiation of a double-blind, placebo-controlled, randomized-withdrawal, dose-optimizing, multi-center Phase 2 clinical trial evaluating the efficacy and safety of KP1077 for the treatment of IH. The trial concluded in March 2024 and provided meaningful information of the optimal dose and dosing regimen to inform Phase 3 trial design.

We enrolled 48 adult patients with IH in more than 30 centers in the United States. Part 1 of the trial consisted of a five-week open-label titration phase during which patients were optimized to one of four doses of SDX (80, 160, 240, or 320 mg/day). Part 2 of the trial entailed a two-week randomized, double-blind, withdrawal phase, during which two-thirds of the trial participants will continue to receive their optimized dose while the remaining one-third will receive placebo. Participants were further assigned into two evenly divided cohorts. The first cohort received a single daily dose just before bedtime, and the second cohort received half the daily dose shortly after awakening and half the daily dose prior to bedtime.

Clinically meaningful improvements were observed across all studied endpoints. The trial was not powered for statistical significance, and this was not the primary endpoint. The exploratory endpoints of sleep inertia and brain fog performed in-line with expectations and were stable when compared across a variety of other endpoints. Symptom improvements in patients receiving KP1077 were similar after both once-per-day, and twice-per-day dosing.

In the Phase 2 trial, KP1077 was observed to be well-tolerated at all dose levels and both dosing regimens, with adverse events that are typical for stimulants and mostly mild in severity. These results are consistent with data from the Phase 1 trial with SDX that indicated no greater cardiovascular safety risk despite higher overall exposure levels when compared to both immediate and long-acting methylphenidate products currently used off-label for the treatment of IH. The trial concluded in March 2024 and provided meaningful information of the optimal dose and dosing regimen to inform Phase 3 trial design. On June 3, 2024, we announced final results from the Phase 2 Clinical Trial of KP1077 for IH. The proof-of-concept study was designed to demonstrate safety and tolerability and was not powered to demonstrate statistical significance. However, the trial included several important secondary and exploratory endpoints, such as the change in Epworth Sleepiness Scale ("ESS") total score, the IH Severity Scale ("IHSS"), the Sleep Inertia Visual Analog Scale ("SIVAS"), and a new scale to assess the symptoms and severity of brain fog. These data gathered from the secondary endpoints will help inform the study design for a potential Phase 3 clinical trial of KP1077. We submitted a briefing book to the FDA for an end-of-Phase 2 meeting to be held by the end of Q3 2024. Additionally, we expect to meet with the FDA to discuss the design of a pivotal Phase 3 trial to study KP1077 in IH at the end of the third quarter.

Key Takeaways from Phase 2 Clinical Trial of KP 1077 for Idiopathic Hypersomnia include:

- KP1077 was well tolerated at all dose levels evaluated in the trial, including the highest dose of 320 mg daily, regardless of the dosing regimen: once daily (QD) or twice daily (BID).
 - o Adverse events (AEs) were similar to other methylphenidate products
 - o Most common AEs included insomnia, headache, anxiety, decreased appetite, and nausea
 - o Most AEs occurred during the titration period, were mild, and did not lead to early discontinuation
- KP1077 produced clinically meaningful improvements in EDS as assessed by change from baseline in the ESS during both the 5-week open-label (OL) titration period which was maintained during the 2-week double-blind withdrawal period for both dosing regimens.
 - o Mean total ESS scores decreased by approximately 9 points after 5 weeks of OL treatment.
- At the end of 7 weeks of treatment, patients administered KP1077 showed clinically meaningful benefits in change from baseline for the ESS, IHSS, SIVAS, and Brain Fog Scale (BFS):
 - o Mean total ESS score decreased by 9.4 (QD) and 8.8 (BID)
 - o Mean total IHSS score decreased by 16.1 (QD) and 12.3 (BID)
 - o Mean SIVAS score decreased by 25.9 (QD) and 17.2 (BID)
 - o Mean total BFS symptom score decreased by 23.8 (QD) and 22.3 (BID)
- The study successfully fulfilled the objectives of providing key information for the design of a pivotal efficacy trial, and the results of the secondary efficacy endpoints were supportive of initiating a Phase 3 trial of KP1077.

In the second quarter of 2023, we initiated a Phase 1 clinical trial in healthy volunteers to assess proposed dosing regimen for the narcolepsy indication. This study was completed in September 2023. By leveraging the data from the IH program, Zevra is evaluating the potential to initiate a Phase 3 trial in narcolepsy.

KP1077 is subject to the terms and conditions of the AZSTARYS License Agreement (as defined below) but is not currently licensed to Commave thereunder.

KP1077 Summary:

- **No drug-to-drug interactions.** We have not observed drug-to-drug interactions in clinical drug-drug interaction studies.
- **Potential for reduced abuse potential as a Schedule IV controlled substance.** All other methylphenidate-based products have been designated as Schedule II controlled substances, which indicates stricter control over the prescribing and use of such products. KP1077 is based on SDX, which has been designated a Schedule IV controlled substance.
- **No currently approved generic equivalent product.** KP1077 contains SDX, our proprietary prodrug of d-methylphenidate, also known as the new chemical name, serdexmethylphenidate, by the U.S. Adopted Names Council of the American Medical Association ("USAN"), which means that there may be no generic equivalent product for KP1077 in most states, making drug-equivalent substitution potentially difficult at the pharmacy.
- **Orphan drug designation.** Because small size of the IH patient population, the FDA has granted KP1077 orphan drug designation for the treatment of IH. We believe KP1077 may potentially be eligible for fast-track and breakthrough therapy designation, which may provide various regulatory benefits for the development program.

AZSTARYS (Partnered product)

AZSTARYS contains d-MPH and our prodrug of dexamethylphenidate, SDX. On March 2, 2021, the FDA approved AZSTARYS as a once-daily treatment for attention deficit hyperactivity disorder (ADHD), in patients age six years and older. AZSTARYS is currently being marketed in the U.S. under our September 2019 collaboration and license agreement, or the AZSTARYS License Agreement, with Commave Therapeutics SA (formerly known as Boston Pharmaceutical S.A.) ("Commave"), an affiliate of Gurnet Point Capital, L.P. Under the AZSTARYS License Agreement, we granted to Commave an exclusive, worldwide license, to develop, manufacture, and commercialize AZSTARYS and any of our product candidates containing SDX and used to treat ADHD or any other CNS disease. In July 2020, we entered into the Corium Consulting Agreement (the "Corium Consulting Agreement"), under which Corium and Commave, respectively, engaged us to guide the product development and regulatory activities for certain current and potential future products in their portfolio, as well as continue supporting preparation for the potential commercial launch of AZSTARYS.

Commave has tasked Corium, Inc. ("Corium"), another affiliate of Gurnet Point Capital, L.P., to lead all commercialization activities for AZSTARYS in the U.S. Corium commercially launched AZSTARYS in the U.S. during the third quarter of 2021. In December 2021, Commave entered into a sublicense of commercialization rights for AZSTARYS in greater China to Shanghai Ark Biopharmaceutical Ltd.

Pursuant to the AZSTARYS License Agreement, Commave agreed to pay up to \$63.0 million in milestone payments upon the occurrence of specified regulatory milestones related to AZSTARYS, including FDA approval and specified conditions with respect to the final approval label. In addition, Corium agreed to make additional payments upon the achievement of specified U.S. sales milestones of up to \$420 million in the aggregate. Further, Commave will pay us quarterly, tiered royalty payments based on a percentage of net sales on a product-by-product basis. Corium also agreed to be responsible for and reimburse us for all of development, commercialization and regulatory expenses for any products or product candidates containing SDX, subject to certain limitations as set forth in the AZSTARYS License Agreement, including consultation fees to be paid to us for services provided to Corium in performing such activities.

In April 2021, we entered into the AZSTARYS Amendment. Pursuant to the AZSTARYS Amendment, we and Commave agreed to modify the compensation terms of the AZSTARYS License Agreement. Commave paid us \$10.0 million in connection with the execution of the AZSTARYS Amendment following the FDA approval of AZSTARYS in the United States. Corium also paid us \$10.0 million following the SDX scheduling determination by the DEA, which occurred on May 7, 2021. In addition, the AZSTARYS Amendment increased the total remaining future regulatory and sales milestone payments related to AZSTARYS up to an aggregate of \$590.0 million. The AZSTARYS License Agreement will continue on a product-by-product basis (i) until expiration of the royalty term for the applicable product candidate in the United States and (ii) perpetually for all other countries.

In May 2021, we announced that SDX, our proprietary prodrug of d-MPH and the primary active pharmaceutical ingredient ("API") in AZSTARYS, was classified as a Schedule IV controlled substance by the DEA. AZSTARYS is classified as a Schedule II controlled substance as its formulation includes a 70:30 mixture of SDX (Schedule IV) and d-MPH (Schedule II), respectively.

During the first half of 2023, annual net sales of AZSTARYS surpassed \$25 million, triggering the first annual net sales milestone payment of \$5.0 million under the AZSTARYS License Agreement, which was earned and recognized as revenue in the second quarter of 2023, and received after quarter-end. During the second half of 2023, annual net sales of AZSTARYS surpassed \$50 million, triggering the second net sales milestone payment of \$10.0 million under the AZSTARYS License Agreement, which was earned and recognized in the fourth quarter of 2023.

Other Third-Party Agreements

Aquestive Termination Agreement

Under our March 2012 termination agreement with Aquestive Therapeutics ("Aquestive") has the right to receive a royalty amount equal to 10% of any value generated by AZSTARYS and any product candidates containing SDX. In connection with the AZSTARYS License Agreement, we paid Aquestive a royalty equal to 10% of the quarterly royalty payments and of the regulatory and net sales milestones earned in 2020, 2021, and 2023.

Distributor Agreement

Our current single distributor for sales of our approved product, OLPRUVA, is a specialty pharmacy provider, however, the Company intends to establish additional distributors such as other retail pharmacies and certain medical centers or hospitals. In addition to distribution agreements, we enter into arrangements with health care providers and payors that provide for government mandated and/or privately negotiated rebates with respect to the purchase of our products.

Relief Exclusive License Agreement

As a condition to entering into the Merger Agreement, Acer and Relief Therapeutics SA ("Relief") entered into an exclusive license agreement on August 30, 2023 (the "Relief License Agreement"). Pursuant to the Relief License Agreement, Relief will hold exclusive development and commercialization rights for OLPRUVA in the EU, Liechtenstein, San Marino, Vatican City, Norway, Iceland, Principality of Monaco, Andorra, Gibraltar, Switzerland, United Kingdom, Albania, Bosnia, Kosovo, Montenegro, Serbia and North Macedonia ("Geographical Europe"). The Company will have the right to receive a royalty of up to 10% of the net sales of OLPRUVA in Geographical Europe.

Results of Operations

Comparison of the three months ended June 30, 2024, and 2023(in thousands):

	Three months ended June 30,		Period-to- Period Change
	2024	2023	
Revenue, net	\$ 4,449	\$ 8,470	\$ (4,021)
Cost of product revenue (excluding \$1,546 in intangible asset amortization for the three months ended June 30, 2024, shown separately below)	3,573	677	2,896
Intangible asset amortization	1,546	—	1,546
Operating expenses:			
Research and development	10,521	7,433	3,088
Selling, general and administrative	12,604	6,612	5,992
Total operating expenses	23,125	14,045	9,080
Loss from operations	(23,795)	(6,252)	(17,543)
Other income (expense):			
Interest expense	(2,110)	(197)	(1,913)
Fair value adjustment related to warrant and CVR liability	5,779	2,118	3,661
Fair value adjustment related to investments	1	131	(130)
Interest and other income (expense), net	270	1,553	(1,283)
Total other income	3,940	3,605	335
Loss before income taxes	(19,855)	(2,647)	(17,208)
Income tax benefit	(70)	74	(144)
Net loss	\$ (19,925)	\$ (2,573)	\$ (17,352)

Net Loss

Net loss for the three months ended June 30, 2024, was \$19.9 million, compared to net loss of \$2.6 million for the three months ended June 30, 2023, an increase in net loss of \$17.3 million. The change was primarily attributable to an increase in loss from operations of \$17.5 million, partially offset by an increase in other income of \$0.3 million.

Revenue

Revenue for the three months ended June 30, 2024, was \$4.4 million, compared to revenue of \$8.5 million for the three months ended June 30, 2023, a decrease of \$4.0 million. The decrease was primarily due to a decrease in net sales milestone revenue under the AZSTARYS License Agreement of \$5.0 million, partially offset by an increase in royalties and other reimbursements under the AZATARYS License Agreement of \$0.6 million and an increase in French EAP reimbursements of \$0.4 million. OLPRUVA revenue was de minimis for the period.

Cost of product revenue

Cost of product revenue for the three months ended June 30, 2024, increased by approximately \$2.9 million compared to the cost of product revenue for the three months ended June 30, 2023, primarily due to recognition of \$3.2 million of inventory obsolescence.

Intangible asset amortization

Intangible asset amortization for the three months ended June 30, 2024, was due to \$1.5 million in amortization expense related to definite lived intangible assets acquired in the Merger.

Research and Development

Research and development expenses increased by \$3.1 million, from \$7.4 million for the three months ended June 30, 2023, to \$10.5 million for the three months ended June 30, 2024. This increase was primarily driven by an increase in spending for the ongoing Phase 2 clinical study in KP1077 and an increase in personnel-related costs, partially offset by a decrease in third-party costs related to arimoclomol.

Selling, General and Administrative

Selling, general and administrative expenses increased by \$6.0 million, from \$6.6 million for the three months ended June 30, 2023, to \$12.6 million for the three months ended June 30, 2024. The period-over-period increase was primarily related to an increase in personnel costs due to an increase in headcount and an increase in other expenses as we built our commercial organization.

Other Income

Other income increased by \$0.3 million, from \$3.6 for the three months ended June 30, 2023, to \$3.9 million for the three months ended June 30, 2024. This increase was primarily attributable to an increase in the fair value adjustment related to warrant and CVR liability of \$3.7 million, partially offset by an increase in interest expense of \$1.9 million and a decrease in interest and other income of \$1.3 million.

Comparison of the six months ended June 30, 2024, and 2023 (in thousands):

	Six months ended June 30,		Period-to-Period Change
	2024	2023	
Revenue, net	\$ 7,874	\$ 11,646	\$ (3,772)
Cost of product revenue (excluding \$1,546 and \$3,074 in intangible asset amortization for the three and six months ended June 30, 2024, respectively shown separately below)	3,748	802	2,946
Intangible asset amortization	3,074	—	3,074
Operating expenses:			
Research and development	22,798	16,088	6,710
Selling, general and administrative	22,535	13,839	8,696
Total operating expenses	45,333	29,927	15,406
Loss from operations	(44,281)	(19,083)	(25,198)
Other (expense) income:			
Interest expense	(2,845)	(379)	(2,466)
Fair value adjustment related to warrant and CVR liability	9,406	575	8,831
Fair value adjustment related to investments	(26)	327	(353)
Interest and other income (expense), net	1,199	2,593	(1,394)
Total other income	7,734	3,116	4,618
Loss before income taxes	(36,547)	(15,967)	(20,580)
Income tax benefit	—	177	(177)
Net loss	\$ (36,547)	\$ (15,790)	\$ (20,757)

Net Loss

Net loss for the six months ended June 30, 2024, was \$36.5 million, compared to net loss of \$15.8 million for the six months ended June 30, 2023, an increase in net loss of \$20.8 million. The change was primarily attributable to an increase in loss from operations of \$25.2 million, partially offset by an increase in other income of \$4.6 million.

Revenue

Revenue for the six months ended June 30, 2024, was \$7.9 million, compared to revenue of \$11.6 million for the six months ended June 30, 2023, a decrease of \$3.7 million. This decrease was primarily due to a decrease in net sales milestones under the AZSTARYS License Agreement of \$5.0 million, partially offset by an increase in royalties and other reimbursements under the AZSTARYS License Agreement of \$1.2 million and an increase in French EAP reimbursements of \$0.3 million. OLPRUVA revenue was de minimis for the six months ended June 30, 2024.

Cost of product revenue

Cost of product revenue for the six months ended June 30, 2024 increased by approximately \$2.9 million compared to cost of product revenue for the six months ended June 30, 2023 primarily due to recognition of \$3.2 million of inventory obsolescence.

Intangible asset amortization

Intangible asset amortization for the six months ended June 30, 2024, was due to \$3.1 million in amortization expense related to definite lived intangible assets acquired in the Merger.

Research and Development

Research and development expenses increased by \$6.7 million, from \$16.1 million for the six months ended June 30, 2023, to \$22.8 million for the six months ended June 30, 2024. This increase was primarily driven by an increase in spending for the ongoing Phase 2 clinical study in KP1077 and an increase in personnel-related costs, partially offset by a decrease in third-party costs related to arimoclomol, and a decrease in expenses incurred during the six months ended June 30, 2023, to terminate the collaboration and license agreement with KVK Tech, Inc.

Selling, General and Administrative

Selling, general and administrative expenses increased by \$8.7 million, from \$13.8 million for the six months ended June 30, 2023, to \$22.5 million for the six months ended June 30, 2024. The period-over-period increase was primarily related to an increase in personnel costs and fees associated with our commercial and business development activities.

Other Income

Other income increased by \$4.6 million, from \$3.1 million of expense for the six months ended June 30, 2023, to \$7.7 million for the six months ended June 30, 2024. This increase was primarily attributable to a change in the fair value adjustment related to warrant and CVR liability of \$8.8 million, offset by an increase in interest expense of \$2.5 million, a decrease in interest and other income of \$1.4 million and a decrease in fair value adjustment related to investments of \$0.3 million.

Liquidity and Capital Resources

Sources of Liquidity

Through June 30, 2024, we have funded our research and development and operating activities primarily through the issuance of debt and equity and from revenue received under the Arimoclomol EAP, AZSTARYS License Agreement, OLPRUVA product sales and consulting arrangements. As of June 30, 2024, we had cash, cash equivalents and investments of \$49.3 million.

To date, we have generated revenue from the Arimoclomol EAP, AZSTARYS License Agreement, reimbursement of out-of-pocket third-party costs, the performance of consulting services, and sales of OLPRUVA.

We have had recurring negative net operating cash flows and we anticipate that we may continue to incur minimal positive net cash flows from operations or negative net cash flows from operations for at least the next several years. We expect that our sources of revenue will be through payments arising from our license agreement with Commave, the Arimoclomol EAP, sales of OLPRUVA and other potential consulting arrangements and any other future arrangements related to one of our product candidates.

We filed a registration statement on Form S-3 covering the sale of the shares of our common stock up to \$350.0 million, \$75.0 million of which was allocated to the sales of the shares of common stock issuable under the Equity Distribution Agreement (described below). The Form S-3 was declared effective on July 12, 2021. As of June 30, 2024, no shares had been issued or sold under the Equity Distribution Agreement and this registration statement under S-3 expired on July 12, 2024.

On June 4, 2024, we filed a registration statement on Form S-3 (File No. 333-279941) (the "June 2024 Registration Statement") under which we may sell securities, including as may be issuable upon conversion, redemption, repurchase, exchange or exercise of securities, in one or more offerings up to a total aggregate offering price of \$350.0 million, \$75.0 million of which was allocated to the sales of the shares of common stock issuable under the 2024 ATM Agreement (as described further below). The registration statement was declared effective on June 13, 2024.

On August 8, 2024, we entered into an underwriting agreement (the "Underwriting Agreement") with Cantor Fitzgerald & Co. and William Blair & Company, L.L.C., as representatives of the several underwriters named therein (collectively, the "Underwriters"), in connection with the offering, issuance and sale by us of 9,230,770 shares of our common stock at a public offering price of \$6.50 per share, pursuant to the June 2024 Registration Statement and a related prospectus supplement dated August 8, 2024 filed with the SEC (the "August 2024 Offering"). Under the terms of the Underwriting Agreement, we also granted the Underwriters an option exercisable for 30 days to purchase up to an additional 1,384,615 shares of our common stock at the public offering price, less underwriting discounts and commissions, which the Underwriters exercised in full on August 9, 2024. The August 2024 Offering closed on August 12, 2024. Total shares issued were 10,615,385. Net proceeds from the offering were approximately \$64.5 million, after deducting underwriting discounts and commissions and estimated offering expenses payable by us. We intend to use the net proceeds of the offering to support the pre-commercial launch activities for arimoclomol, continued commercial support for OLPRUVA and the continued development of celiprolol and KP1077 through potential NDA filings and other general corporate purposes.

We have incurred operating losses since our inception and, as of June 30, 2024, had an accumulated deficit of \$436.3 million. Our recurring operating losses and negative cash flows from operations raise substantial doubt about our ability to continue as a going concern. The Company's ability to continue operating as a going concern is contingent upon its ability to generate revenue from approved products or obtain product candidate regulatory approvals, which would generate revenue, milestones, and cash flow sufficient to support ongoing operations and the satisfaction of financial covenants. We are early in our commercialization effort for OLPRUVA and do not yet have a substantial basis to project future earnings, and our other sources of revenue are not sufficient to sustain our present activities on their own. Accordingly, our ability to continue as a going concern may require us to obtain additional financing to fund our operations. The perception of our inability to continue as a going concern may make it more difficult for us to obtain financing for the continuation of our operations and could result in the loss of confidence by investors, suppliers and employees. Adequate additional financing may not be available to us on acceptable terms, or at all. To the extent that we raise additional capital through the sale of equity or debt, the terms of these securities may restrict our ability to operate. If we raise additional funds through collaborations, strategic alliances or marketing, distribution or licensing arrangements with third parties, we may be required to relinquish valuable rights. If we are unable to raise capital when needed or on attractive terms, we could be forced to delay, reduce or altogether cease our research and development programs or future commercialization efforts.

Equity Distribution Agreements

On July 12, 2024, we entered into an equity distribution agreement ("2024 ATM Agreement") with Citizens JMP Securities LLC ("Citizens JMP") under which we may offer and sell, from time to time in our sole discretion, shares of our common stock having an aggregate offering price of up to \$75.0 million through Citizens JMP as our sales agent. The issuance and sale, if any, of common stock by us under the 2024 ATM Agreement will be made pursuant to a registration statement on Form S-3. Citizens JMP may sell the common stock by any method permitted by law deemed to be an "at the market offering" as defined in Rule 415 of the Securities Act. Citizens JMP will use commercially reasonable efforts to sell the common stock from time to time, based upon instructions from us (including any price, time or size limits or other customary parameters or conditions we may impose). We will pay Citizens JMP a commission equal to 3.0% in the aggregate of the gross sales proceeds of any common stock sold through Citizens JMP under the 2024 ATM Agreement. We filed the June 2024 Registration Statement which was declared effective on June 13, the accompanying prospectus, and the related prospectus supplement dated July 12, 2024, covering the sale of the shares of our common stock up to \$350.0 million, \$75.0 million of which was allocated to the sale of the shares of common stock issuable under the 2024 ATM Agreement.

On July 2, 2021, we entered into an equity distribution agreement ("2021 ATM Agreement") with JMP Securities LLC ("JMP") and RBC Capital Markets, LLC ("RBCCM"), under which we had the ability to offer and sell shares of our common stock having an aggregate offering price of up to \$75.0 million through JMP and RBCCM as our sales agents. The 2021 ATM Agreement expired on July 12, 2024.

Merger Transactions and Documents

On August 30, 2023, in connection with the Merger Agreement with Acer, the following transactions occurred prior to Closing:

- **Bridge Loan** – Zevra and Acer entered into a bridge loan agreement (the "Bridge Loan Agreement"), providing for Zevra to make loans (collectively, the "Bridge Loan") to Acer up to an aggregate principal amount of \$16.5 million. The Bridge Loan was provided to Acer to support its termination agreement with Relief and to provide Acer with working capital, including for payments of accounts payable to support the commercial launch of OLPRUVA and the development of celiprolol pending the Merger's closure. On October 31, 2023, the Company and Acer entered into an amendment to the Bridge Loan Agreement, which increased the aggregate principal amount available under the loan from \$16.5 million to \$18.0 million.
- **Purchase of Acer's Term Loans** – Zevra purchased certain indebtedness of Acer held by Nantahala Capital Management, LLC ("Nantahala"). Under the loan purchase with Nantahala, certain of its affiliates and certain other parties (collectively with Nantahala, "Nantahala Holders") Zevra purchased (i) an original senior secured term loan facility made available to Acer in an aggregate amount of \$6.5 million and funded on March 14, 2022, and (ii) an additional senior secured term loan made to Acer in an aggregate amount of \$7.0 million in a single borrowing which funded on January 31, 2023 for (1) \$12.0 million in cash; (2) 98,683 shares of Zevra Common Stock; and (3) a secured Promissory Note payable by Zevra to Nantahala in the original principal amount \$5.0 million (the "Nantahala Note"). These were recorded as receivables from Acer and were treated as a settlement of a preexisting relationship in connection with the closing of the transaction and recorded as a component of purchase consideration. In April 2024, the Nantahala Note was repaid in full and terminated.
- **Purchase of Acer's Convertible Notes** ("Marathon Convertible Notes") – Under the Note Purchase Agreement with the Nantahala Holders, Zevra purchased the Marathon Convertible Notes that Nantahala had acquired on June 16, 2023. Zevra acquired the Marathon Convertible Notes in exchange for the issuance of 2,171,038 shares of Zevra Common Stock at \$5.0667 per share for a total purchase price of \$11.0 million.
- **Amendment to IP License Agreement and IP Termination Agreement:** As a condition to entering into the Merger Agreement, Acer and Relief entered into the Exclusive License Agreement and the Termination Agreement terminating the collaboration and license agreement, dated March 19, 2021, by and between Acer and Relief. Pursuant to the Exclusive License Agreement, Relief holds exclusive development and commercialization rights for OLPRUVA in Geographical Europe. Acer has the right to receive a royalty of up to 10% of the net sales of OLPRUVA in Geographical Europe. In accordance with the terms of the Termination Agreement, Relief received an upfront payment from Acer of \$10.0 million (which payment was funded with the Bridge Loan described above) with an additional payment of \$1.5 million due on the first-year anniversary of the \$10.0 million payment. Acer has also agreed to pay a 10% royalty on net sales of OLPRUVA worldwide, excluding Geographical Europe, and 20.0% of any value received by Acer from certain third parties relating to OLPRUVA licensing or divestment rights, all of the foregoing which are capped at \$45.0 million, for total payments to Relief of up to \$56.5 million.

In connection with the closing of the Merger on November 17, 2023, each share of common stock of Acer was converted into the right to receive (i) 0.1210 fully paid and non-assessable shares of common stock of Zevra, par value \$0.0001 per share, and (ii) one non-transferable CVR to be issued by Zevra, which will represent the right to receive one or more contingent payments up to an additional \$76 million upon the achievement, if any, of certain commercial and regulatory milestones for Acer's OLPRUVA and celiprolol products 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).

Registration Rights Agreement

Zevra and Nantahala concurrently entered into a registration rights agreement (the "Registration Rights Agreement"), pursuant to which Zevra agreed to file a resale registration statement with respect to the resale of the Zevra common stock issuable under the Loan and Note Purchase Agreements and the Nantahala Note. On February 5, 2024, Zevra filed a registration statement on Form S-3 (File No. 333-276856) registering an aggregate of 2,269,721 shares of Zevra's common stock that were issued pursuant to the Loan and Note Purchase Agreements. On April 5, 2024, we filed an amendment to the registration statement on Form S-3 (File No. 333-250945) covering the issuance of the shares of our common stock issuable upon the exercise of warrants issued in connection with the Merger and remaining unexercised as of the date of the amendment, which was declared effective on April 8, 2024.

Cancellation of Acer Warrant

On November 22, 2023, we sold an aggregate of 1,382,489 shares of our common stock and accompanying warrants to purchase up to 1,382,489 shares of our common stock at a price of \$4.34 per share to a healthcare focused investment fund (the "Investor") for gross proceeds of approximately \$6.0 million and an aggregate of 917,934 shares of our common stock to cancel a warrant held by the Investor to purchase 2,920,306 shares of common stock of Acer. The shares of common stock and warrants were offered and sold to the Investor in a registered direct offering without an underwriter or placement agent.

Line of Credit

On January 26, 2023, we and Wells Fargo, as lender, entered into a margin account agreement. Our investments were used as collateral for the loan and the amount we were able to borrow was limited to 80-90% of our outstanding investment balance held with Wells Fargo. The margin account bore interest at the Prime Rate minus 225 basis-points. This facility was paid off on April 5, 2024, and the margin capability was removed from the account.

Term Loans

On April 5, 2024 (the "Term Loans Closing Date"), we entered into a credit agreement (the "Credit Agreement") with HCR Stafford Fund II, L.P., HCR Potomac Fund II, L.P., and Perceptive Credit Holdings IV, LP (collectively, the "Lenders"), and Alter Domus (US) LLC, as administrative agent (the "Administrative Agent").

Under the terms of the Credit Agreement, the Lenders provided a senior secured loan facility to us in the aggregate principal amount of \$100.0 million, which is divided into three tranches as follows: (i) \$60.0 million which was funded in full on the Term Loans Closing Date; (ii) \$20.0 million which is available to us in up to two drawings, each in an amount not to exceed \$10.0 million, at the Company's option until 18 months following the Term Loans Closing Date; and (iii) \$20.0 million which is available to us upon approval by the FDA of the NDA for arimoclomol for the treatment of NPC, at our option until December 31, 2024 (collectively, the "Term Loans").

The principal amount of the Term Loans outstanding (the "Outstanding Principal Amount") will bear interest at a rate equal to 3-Month Term SOFR *plus* 7.00% per annum. If the net product sales for the calendar year ending December 31, 2025, exceed \$100.0 million, the Outstanding Principal Amount will bear interest at 3-Month Term SOFR *plus* 6.00% per annum. If the net product sales for the calendar year ending December 31, 2025, do not exceed \$100.0 million, then for any subsequent period of four consecutive fiscal quarters ending on or after March 31, 2026, in which net product sales exceed \$125.0 million, the Outstanding Principal Amount will bear interest at 3-Month Term SOFR *plus* 6.50% per annum. In all cases, the 3-Month Term SOFR rate will be subject to a floor of 4.00% per annum. Interest will be payable quarterly in arrears on the last day of each calendar quarter. We have the option to pay up to 25% of the interest in-kind beginning on the Term Loans Closing Date, through and including June 30, 2026. The Term Loans will mature on the fifth anniversary of the Term Loans Closing Date. In connection with the Credit Agreement, we incurred approximately \$2.2 million of costs, which primarily consisted of underwriting, legal and other professional fees, and are included as a reduction to the carrying amount of the related debt liability and are deferred and amortized over the remaining life of the financing using the effective interest method.

The Credit Agreement contains customary affirmative and negative covenants by us, which among other things, will require us to provide certain financial reports to the Lenders, meet certain minimum net product sales amounts, and limit our ability to incur or guarantee additional indebtedness, engage in certain transactions, and effect a consolidation or merger without consent. In addition, as long as the line of credit remains active, we must maintain a minimum cash balance of \$20.0 million to ensure that we can meet our immediate capital needs. Our obligations under the Credit Agreement may be accelerated upon customary events of default, including non-payment of principal, interest, fees and other amounts, covenant defaults, insolvency, material judgments, or inaccuracy of representations and warranties. The Term Loans are secured by a first priority perfected lien on, and security interest in, substantially all of our current and future assets. The proceeds of the Term Loans were used to refinance certain of our previously existing indebtedness. We will use the remaining proceeds to pay fees and expenses related to the debt financing and fund the development and commercialization of OLPRUVA and arimoclomol.

Cash Flows

The following table summarizes our cash flows for the six months ended June 30, 2024, and 2023 (in thousands):

	Six months ended June 30,	
	2024	2023
Net cash used in operating activities	\$ (35,274)	\$ (12,749)
Net cash provided by investing activities	14,664	16,481
Net cash provided by (used in) financing activities	16,547	(2,799)
Effect of exchange rates on cash and cash equivalents	274	(203)
Net (decrease) increase in cash and cash equivalents	<u>\$ (3,789)</u>	<u>\$ 730</u>

Operating Activities

For the six months ended June 30, 2024, net cash used in operating activities of \$35.3 million consisted of a net loss of \$36.5 million and \$1.6 million in changes in working capital, partially offset by \$2.8 million in adjustments for non-cash items. Net loss was primarily attributable to our spending on research and development programs and operating costs; partially offset by revenue received under the AZSTARYS License Agreement, and the Arimoclomol EAP. The changes in working capital consisted of \$8.6 million related to a change in accounts payable and accrued expenses, \$3.6 million change in inventories, \$0.3 million related to a change in operating lease liabilities, and a decrease of \$0.9 million in prepaids and other assets, partially offset by \$8.4 million related to a change in accounts and other receivables, \$2.7 million related to a change in discount and rebate liabilities, \$0.3 million related to a change in operating lease right-of-use assets, and \$0.3 million related to a change in other liabilities. The adjustments for non-cash items primarily consisted of stock-based compensation expense of \$4.8 million, an inventory obsolescence charge of \$3.2 million, interest expense of \$0.7 million, and \$3.5 million related to depreciation, amortization and other items, partially offset by a change in the fair value of warrant and CVR liability of \$9.4 million.

For the six months ended June 30, 2023, net cash used in operating activities of \$12.7 million consisted of a net loss of \$15.8 million, partially offset by \$2.4 million in adjustments for non-cash items and \$0.7 million in changes in working capital. Net loss was primarily attributable to our spending on research and development programs and operating costs, partially offset by revenue received under the AZSTARYS License Agreement, Arimoclomol EAP and the Corium Consulting Agreement. The changes in working capital consisted of \$3.1 million related to a change in discount and rebate liabilities, \$2.9 million related to a change in accounts payable and accrued expenses, \$0.2 million related to a change in operating lease right-of-use assets, \$0.1 million related to a change in inventories and \$0.6 million related to a change in other liabilities, partially offset by \$5.7 million related to a change in accounts and other receivables, \$0.2 million related to a change in operating lease liabilities and \$0.3 million related to a change in prepaid expenses and other assets. The adjustments for non-cash items primarily consisted of stock-based compensation expense of \$1.7 million, non-cash severance expense of \$1.4 million, and \$0.2 million related to depreciation, amortization and other items, partially offset by \$0.5 million change in fair value adjustment related to derivative and warrant liability, a change in the fair value adjustment related to investments of \$0.3 million and a gain on foreign currency exchange rates of \$0.1 million.

Investing Activities

For the six months ended June 30, 2024, net cash provided by investing activities was \$14.7 million, which was primarily attributable to maturities of investments.

For the six months ended June 30, 2023, net cash provided by investing activities was \$16.5 million, which was primarily attributable to maturities of investments of \$34.0 million, partially offset by purchases of investments of \$17.5 million.

Financing Activities

For the six months ended June 30, 2024, net cash provided by financing activities was \$16.5 million, which was primarily attributable to proceeds from the issuance of debt of \$59.0 million, partially offset by repayments of debt of \$42.7 million.

For the six months ended June 30, 2023, net cash used in financing activities was \$2.8 million, which was primarily attributable to repayment of debt of \$13.0 million, payments to repurchase shares as part of the Share Repurchase Program of \$3.4 million, and payments of principal on insurance financing arrangements of \$0.6 million, partially offset by proceeds from the issuance of debt of \$12.8 million, proceeds from insurance financing programs of \$1.2 million and proceeds from the Employee Stock Purchase Plan of \$0.1 million.

Future Funding Requirements

While under applicable accounting principles factors exist that raise substantial doubt about our ability to continue as a going concern, based on our current operating forecast and the net proceeds from the August 2024 Offering (as described above), we believe that our existing cash, cash equivalents and investments will be sufficient to fund our operations into the first quarter of 2027, subject to continuing compliance with our debt covenants. This estimate includes the ongoing reimbursements from the French expanded access program for arimoclomol, completion of the arimoclomol NDA resubmission, commercial activities to support the launch of arimoclomol, if approved, and completion of the KP1077 development program for IH up to NDA submission. This estimate does not include revenue from arimoclomol after potential FDA approval, or the potential sale of the priority review voucher of arimoclomol, which would be received at that time, as well, or the costs of a Phase 3 trial for KP1077 in narcolepsy. Certain of the milestones are associated with regulatory matters that are outside our control. In addition, we maintain the majority of our cash and cash equivalents in accounts with major U.S. and multi-national financial institutions, and our deposits at these institutions exceed insured limits. Market conditions can impact the viability of these institutions. In the event of a failure of any of the financial institutions where we maintain our cash and cash equivalents, there can be no assurance that we would be able to access uninsured funds in a timely manner or at all. Any inability to access or delay in accessing these funds could adversely affect our business and financial position.

Potential near-term sources of additional funding include:

- any royalties or net sales milestone payments generated under the AZSTARYS License Agreement;
- any product sales under the Arimoclomol EAP;
- any product sales of OLPRUVA;
- any product sales of arimoclomol, if approved; and
- any consulting services revenue generated under other potential consulting agreements;

We cannot guarantee that we will be able to generate sufficient proceeds from any of these potential sources to fund our operating expenses. We anticipate that our expenses will fluctuate substantially as we:

- continue our ongoing preclinical studies, clinical trials and our product development activities for our pipeline of product candidates;
- seek regulatory approvals for any product candidates that successfully complete clinical trials;
- continue research and preclinical development and initiate clinical trials of our product candidates;
- seek to discover and develop additional product candidates either internally or in partnership with other pharmaceutical companies;
- adapt our regulatory compliance efforts to incorporate requirements applicable to marketed products;
- maintain, expand and protect our intellectual property portfolio; and
- incur additional legal, accounting and other expenses in operating as a public company.

To date, we have generated revenue from the AZSTARYS License Agreement, reimbursements of out-of-pocket third-party costs, the performance of consulting services, OLPRUVA product sales, and product sales under the Arimoclomol EAP. We expect that, for the foreseeable future, our only sources of revenues will be through payments arising from the AZSTARYS License Agreement, product sales of OLPRUVA, through potential consulting arrangements and any other future arrangements related to one of our product candidates and product sales under the Arimoclomol EAP. While we have entered into the AZSTARYS License Agreement to develop, manufacture and commercialize AZSTARYS, we cannot guarantee that this, or any strategy we adopt in the future, will be successful. For instance, we received milestone payments under the AZSTARYS License Agreement, but we cannot guarantee that we will earn any additional milestone or royalty payments under this agreement in the future. We also cannot guarantee that we will continue to generate revenue under the Arimoclomol EAP or successfully commercialize OLPRUVA. We also expect to continue to incur additional costs associated with operating as a public company.

We have based our estimates of our cash needs and cash runway on assumptions that may prove to be wrong, and we may use our available capital resources sooner than we currently expect and we cannot guarantee that we will be able to generate sufficient proceeds from the AZSTARYS License Agreement, product reimbursements under the Arimoclomol EAP, product sales of OLPRUVA, potential consulting arrangements or other funding transactions to fund our operating expenses. To meet any additional cash requirements, we may seek to sell additional equity or convertible securities that may result in dilution to our stockholders, issue additional debt or seek other third-party funding, including potential strategic transactions, such as licensing or collaboration arrangements. Because of the numerous risks and uncertainties associated with the development and commercialization of product candidates and products, we are unable to estimate the amounts of increased capital outlays and operating expenditures necessary to complete the commercialization and development of our partnered product or product candidates, should they obtain regulatory approval.

Critical Accounting Estimates

This management's discussion and analysis of our financial condition and results of operations is based on our unaudited condensed consolidated financial statements, which we have prepared in accordance with accounting principles generally accepted in the United States. The preparation of our unaudited condensed consolidated financial statements requires us to make estimates that affect the reported amounts of assets and liabilities and the disclosure of contingent assets and liabilities at the date of our unaudited condensed consolidated financial statements, as well as the reported revenues and expenses during the reported periods. We evaluate these estimates on an ongoing basis. We base our estimates on historical experience and on various other factors that we believe are reasonable under the circumstances. Actual results may differ from these estimates under different assumptions or conditions.

Our critical accounting policies have not changed materially from those described in *Part II, Item 7 – Management's Discussion and Analysis of Financial Condition and Results of Operations* of our Annual Report on Form 10-K for the fiscal year ended December 31, 2023, filed with the SEC on April 1, 2024.

ITEM 3. QUANTITATIVE AND QUALITATIVE DISCLOSURES ABOUT MARKET RISK

Not applicable.

ITEM 4. CONTROLS AND PROCEDURES

Limitations on Effectiveness of Controls and Procedures

In designing and evaluating our disclosure controls and procedures, management recognizes that any controls and procedures, no matter how well designed and operated, can provide only reasonable assurance of achieving the desired control objectives. In addition, the design of disclosure controls and procedures must reflect the fact that there are resource constraints and that management is required to apply judgment in evaluating the benefits of possible controls and procedures relative to their costs.

Evaluation of Disclosure Controls and Procedures

Our management, with the participation of our chief executive officer and our chief financial officer, evaluated the effectiveness of our disclosure controls and procedures as of June 30, 2024. Based on the evaluation of our disclosure controls and procedures as of June 30, 2024, our chief executive officer and our chief financial officer concluded that, as of such date, our disclosure controls and procedures were effective at the reasonable assurance level.

Material Weakness

As previously disclosed in our Annual Report on Form 10-K for the year ended December 31, 2023, and our subsequent Quarterly Report on Form 10-Q for the three months ended March 31, 2024, management identified a material weakness in our internal control over financial reporting related to the accounting for warrants to purchase the Company's common stock. The material weakness in our internal control resulted in the restatement of the Company's consolidated financial statements as of and for the year ended December 31, 2022, and the condensed consolidated financial statements for the interim periods ended March 31, 2022, June 30, 2022, September 30, 2022, March 31, 2023, June 30, 2023, and September 30, 2023, included in our Annual Report on Form 10-K. The material weakness, which related to internal controls over the application of specific technical accounting guidance, was identified during the performance of our year-end control procedures.

Remediation of Material Weakness

In 2024, we implemented certain remedial measures including, but not limited to, a review of all existing accounting for warrants to purchase the Company's common stock to confirm compliance with GAAP prior to filing the consolidated financial statements as of and for the year ended December 31, 2023, with our Annual Report on Form 10-K.

In addition, we have developed enhanced control procedures designed to ensure proper accounting for our warrant related accounts and balances, which included adding technical resources to perform and oversee technical accounting. During the quarter ended June 30, 2024, we successfully completed the testing necessary, and the applicable controls have been in place for a sufficient period of time, to conclude that the material weakness has been remediated and that these controls are operating effectively.

Changes in Internal Control Over Financial Reporting

Except as disclosed above, there was no change in our internal control over financial reporting (as defined in Rules 13a-15(f) and 15d-15(f) under the Exchange Act) identified in connection with the evaluation required by Rules 13a-15(d) and 15d-15(d) of the Exchange Act that occurred during our fiscal quarter ended June 30, 2024, that materially affected, or is reasonably likely to materially affect, our internal control over financial reporting.

**PART II
OTHER INFORMATION**

ITEM 1. LEGAL PROCEEDINGS

From time to time, we may be involved in routine legal proceedings, as well as demands, claims and threatened litigation, which arise in the normal course of our business. We believe there is no litigation pending that would reasonably be expected to, individually or in the aggregate, have a material adverse effect on our results of operations or financial condition.

ITEM 1A. RISK FACTORS

In addition to the other information set forth in this Quarterly Report on Form 10-Q, you should carefully consider all the risk factors and uncertainties described in Part I, Item 1A. "Risk Factors" of our Annual Report on Form 10-K for the fiscal year ended December 31, 2023, filed with the SEC on April 1, 2024, before investing in our common stock. Other than as described below, there have been no material changes to the risk factors described in that report. If any of those risks materialize, our business, financial condition and results of operations could be seriously harmed. This Quarterly Report on Form 10-Q also contains forward-looking statements that involve risks and uncertainties. Our actual results could differ materially from those anticipated in the forward-looking statements because of the risk factors in our Annual Report on Form 10-K and below, and the other factors described in in this Quarterly Report on Form 10-Q.

If we experience additional material weaknesses or otherwise fail to maintain effective internal control over financial reporting and disclosure controls and procedures, we may not be able to accurately report our financial results or report them in a timely manner, which may adversely affect investor confidence in us and, as a result, the value of our common stock.

Our management is required to report annually on the effectiveness of our internal control over financial reporting pursuant to Section 404 of the Sarbanes-Oxley Act, or Section 404. The rules governing the standards that must be met for our management to assess our internal control over financial reporting are complex and require significant documentation, testing and possible remediation.

A material weakness is a deficiency, or a combination of deficiencies, in internal control over financial reporting, such that there is a reasonable possibility that a material misstatement of the Company's annual or interim financial statements will not be prevented or detected on a timely basis. We previously identified and disclosed a material weakness in our internal control over financial reporting in our Annual Report on Form 10-K for the year ended December 31, 2023, and our Quarterly Report on Form 10-Q for the three months ended March 31, 2024. This material weakness has since been remediated.

Our failure to certify the effectiveness of our internal control over financial reporting or our disclosure controls and procedures, or the identification of the material weakness, could subject us to regulatory scrutiny and a loss of public confidence, which could have a material adverse effect on our business and our stock price. In the future, we may identify additional material weaknesses or significant deficiencies, and we may not be able to remediate them in time to meet the deadline imposed by the Sarbanes-Oxley Act for compliance with the requirements of Section 404. In addition, we may encounter problems or delays in completing the implementation of any requested improvements and, if and when such a report is required, receiving a favorable attestation report from our independent registered public accounting firm. If we do not maintain adequate financial and management personnel, processes and controls, we may not be able to manage our business effectively or accurately report our financial performance on a timely basis, which could cause a decline in our common stock price and adversely affect our results of operations and financial condition.

ITEM 2. UNREGISTERED SALES OF EQUITY SECURITIES AND USE OF PROCEEDS

Recent Sales of Unregistered Securities

None.

Purchases of Equity Securities By the Issuer and Affiliated Purchasers

None.

ITEM 3. DEFAULTS UPON SENIOR SECURITIES

Not applicable.

ITEM 4. MINE SAFETY DISCLOSURES

Not applicable.

ITEM 5. OTHER INFORMATION

(a) Disclosure in lieu of reporting on a Current Report on Form 8-K.

None.

(b) Material changes to the procedures by which security holders may recommend nominees to the board of directors.

None.

(c) Insider Trading Arrangements and Policies.

During the three months ended June 30, 2024, no director or officer of the Company adopted or terminated a "Rule 10b5-1 trading arrangement" or "non-Rule 10b5-1 trading arrangement," as each term is defined in Item 408(a) of Regulation S-K.

ITEM 6. EXHIBITS

The following is a list of exhibits filed as part of this Form 10-Q (the SEC file number for all items incorporated by reference herein from reports on Forms 10-K, 10-Q, and 8-K is 001-36913):

Exhibit No.	Description
3.1	Amended and Restated Certificate of Incorporation of Zevra Therapeutics, Inc. (incorporated herein by reference to the Registrant's Current Report on Form 8-K as filed with the SEC on April 21, 2015).
3.1.1	Certificate of Amendment of Amended and Restated Certificate of Incorporation of the Registrant, effective as of December 23, 2020 (incorporated herein by reference to Registrant's Current Report on Form 8-K as filed with the SEC on December 23, 2020).
3.1.2	Certificate of Amendment of Amended and Restated Certificate of Incorporation of Zevra Therapeutics, Inc. (incorporated herein by reference to the Registrant's Current Report on Form 8-K as filed with the SEC on February 24, 2023).
3.2	Amended and Restated Bylaws, as currently in effect, of Zevra Therapeutics, Inc. (incorporated herein by reference to the Registrant's Current Report on Form 8-K as filed with the SEC on February 28, 2024).
4.1	Specimen stock certificate evidencing shares of Common Stock (incorporated herein by reference to the Registrant's Annual Report on Form 10-K as filed with the SEC on March 12, 2021).
10.1*	Employment Agreement by and between the Registrant and Rahsaan Thompson, dated as of June 21, 2024.
31.1*	Certification of the Principal Executive Officer pursuant to Rule 13a-14(a) or Rule 15d-14(a) of the Securities Exchange Act of 1934, as amended.
31.2*	Certification of the Principal Financial Officer pursuant to Rule 13a-14(a) or Rule 15d-14(a) of the Securities Exchange Act of 1934, as amended.
32.1**	Certification of the Principal Executive Officer pursuant to Rule 13a-14(b) of the Securities Exchange Act of 1934, as amended, and 18. U.S.C. Section 1350, as adopted pursuant to Section 906 of the Sarbanes-Oxley Act of 2002.
32.2**	Certification of the Principal Financial Officer pursuant to Rule 13a-14(b) of the Securities Exchange Act of 1934, as amended, and 18. U.S.C. Section 1350, as adopted pursuant to Section 906 of the Sarbanes-Oxley Act of 2002.
101.INS*	Inline XBRL Instance Document
101.SCH*	Inline XBRL Taxonomy Extension Schema Document
101.CAL*	Inline XBRL Taxonomy Extension Calculation Linkbase Document
101.DEF*	Inline XBRL Taxonomy Extension Definition Linkbase Document
101.LAB*	Inline XBRL Taxonomy Extension Label Linkbase Document
101.PRE*	Inline XBRL Taxonomy Extension Presentation Linkbase Document
104**	Cover page Interactive Data File (embedded within the Inline XBRL and combined in Exhibit 101)

* Filed herewith

** Furnished herewith

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 thereunto duly authorized.

Zevra Therapeutics, Inc.

Date: August 13, 2024

By: /s/ Neil F. McFarlane
Neil F. McFarlane
President and Chief Executive Officer
(Principal Executive Officer)

Date: August 13, 2024

By: /s/ R. LaDuane Clifton
R. LaDuane Clifton, MBA, CPA
Chief Financial Officer and Treasurer
(Principal Financial Officer)

ZEVRA THERAPEUTICS, INC.

EMPLOYMENT AGREEMENT

RAHSAAN THOMPSON

EFFECTIVE AS OF JUNE 20, 2024

ZEVRA THERAPEUTICS, INC.
EMPLOYMENT AGREEMENT

This Employment Agreement ("Agreement") is made and entered into on June 20, 2024, by and between Zevra Therapeutics, Inc., a Delaware corporation (the "Company"), including its affiliates, parent, subsidiaries, successors, and assigns and Rahsaan Thompson ("Executive") (each being a "Party" hereto and together constituting the "Parties").

WHEREAS, THE COMPANY DESIRES TO EMPLOY THE EXECUTIVE AS ITS CHIEF LEGAL OFFICER, SECRETARY AND COMPLIANCE OFFICER, UNDER THE TERMS AND CONDITIONS SET FORTH BELOW.

NOW, THEREFORE, IN CONSIDERATION OF THE MUTUAL PROMISES AND COVENANTS CONTAINED HEREIN, AND FOR OTHER GOOD AND VALUABLE CONSIDERATION, THE RECEIPT, ADEQUACY AND SUFFICIENCY OF WHICH IS HEREBY ACKNOWLEDGED, THE PARTIES HERETO AGREE AS FOLLOWS:

1. Employment.

A. Employment. Company hereby desires to employ Executive and Executive hereby accepts such employment with Company as its Chief Legal Officer, Secretary and Compliance Officer, or in such other capacities as Company shall reasonably determine from time to time, upon the terms and conditions set forth in this Agreement.

A. Effective Date and Term. Company's employment of Executive under this Agreement shall commence effective as of June 20, 2024, (the "Effective Date"), and continue until the Date of Termination (defined in Section 4(A)) (hereinafter such period of time from the commencement until termination of employment shall be referred to as the "Employment Term"). Company and Executive acknowledge that Executive's employment is and shall continue to be at-will, as defined under applicable law, and that Executive's employment with Company may be terminated by either Party at any time for any or no reason (subject to the notice requirements of Section 4). This "at-will" nature of Executive's employment shall remain unchanged during Executive's tenure as an employee and may not be changed, except in an express writing signed by Executive and a duly authorized officer of Company.

B. Duties of Executive. During the Employment Term, all of the following shall apply: Executive shall carry out, perform and comply with such reasonable and lawful orders, directions, and written rules and policies (including those rules and policies memorialized in meeting minutes) as are assigned or set by Company's Chief Executive Officer ("CEO") from time to time. Executive shall report to, receive directions from and be reviewed by the CEO. Executive's duties shall include the duties and responsibilities commonly associated with and that are appropriate for an individual of Executive's or a similar position of a company similar to Company. Subject to the limitations of Section 4(E)(3)(iv), the CEO retains the right to modify Executive's job title and responsibilities pursuant to the legitimate business needs of Company.

C. Duty of Loyalty. During the Employment Term, Executive shall not, without the prior written consent of the CEO, accept other employment or render or perform other services for compensation, or engage in outside business activities (including serving on outside boards or committees). Executive shall devote Executive's full business time and attention and Executive's best efforts to the faithful performance of Executive's duties as an officer and employee of Company. However, Executive's expenditure of reasonable amounts of time (i) devoted to serving on the boards of directors set forth on Exhibit A hereto; (ii) for teaching and/or personal business; and (iii) on behalf of charitable or professional organizations in a capacity other than a board member, shall not be deemed a breach of this Agreement, provided such activities do not interfere with the performance of Executive's duties and responsibilities hereunder, including the limitations in Sections 6 through 9, and Executive has provided prior written notice to CEO and CEO has provided prior written approval of such activities, as determined in CEO's reasonable sole discretion, which approval will not be unreasonably withheld. Nothing in this Agreement shall preclude Executive's expenditure of reasonable amounts of time on managing Executive's personal, financial and legal affairs; provided such activities do not materially interfere with the performance of Executive's duties and responsibilities hereunder, including the limitations of Sections 6 through 9, as determined in CEO's good faith sole discretion.

D. Place of Performance. Executive's initial place of employment during the Employment Term will be in Oakland, California. Based upon specified business milestones expected to occur in the third quarter of 2024, Executive will be expected to relocate residency from Oakland, California to the Boston, Massachusetts Metro Area no later than January 31, 2025 (the "Relocation"). The Company agrees to cover reasonable and customary expenses associated with the Relocation, as outlined in Exhibit A to the offer letter between the Parties dated as of June 19, 2024 (the "Relocation Exhibit"), subject to specified repayment requirements if the Executive's Date of Termination (as defined below in Section 4), is prior to the first anniversary of the date Relocation is completed, provided that such date shall not be later than January 31, 2026. All payments from Company in respect of the payments and benefits set forth in the Relocation Exhibit shall be paid no later than March 15, 2025. Notwithstanding the foregoing, Executive understands and agrees that Executive's presence may be required at other Company worksites, or Executive may be required to travel for business, in each case, in accordance with Executive's duties and responsibilities under this Agreement, as business needs require or may change over time and as reasonably requested by the CEO.

2. Compensation and Benefits. In consideration of the services to be rendered by Executive pursuant to this Agreement, as well as Executive's covenants set forth in this Agreement, Company shall pay to Executive the following compensation, which shall be the entire and exclusive compensation for all of Executive's services rendered and other obligations taken on Company's behalf:

A. Annual Base Salary. During the Employment Term, Company shall pay to Executive an annualized base salary of \$450,000 (the "Base Salary"). For calendar years in which Executive is employed for less than the full year, the Base Salary shall be prorated and accrue on a per diem basis for only those days on which Executive was employed during the Employment Term. The Base Salary will be paid by Company in equal installments according to Company's customary payroll practices, but in any event not less frequently than monthly, and shall be subject to all mandatory and voluntary payroll deductions. Executive's Base Salary shall be reviewed periodically by the Company's Board of Directors ("Board of Directors") or the Compensation Committee of the Board of Directors (the "Compensation Committee") if so designated and may be appropriately increased from time to time in the sole discretion of Board of Directors or the Compensation Committee, as applicable.

B. Incentive Compensation. During the Employment Term, Executive shall be entitled to participate in all short-term and long-term incentive programs established by Company, at such levels as the Board of Directors or Compensation Committee determines. Executive's annual short-term cash incentive opportunity target shall be no less than 40% of the Base Salary (as such percentage may be increased from time to time, the "Target Annual Bonus"). The actual amount of such annual incentive compensation (the "Annual Bonus") shall be determined in accordance with the applicable plans based on 1) the determination of the Company as to whether the Executive has achieved relevant performance goals and 2) Company performance objectives established in advance by the Board of Directors or the Compensation Committee, taking into account input from the CEO. Such actual annual short term incentive compensation amount may be more or less than the target amount and no incentive is guaranteed. The payment of any Annual Bonus pursuant to the incentive program shall be subject to Executive's continued employment with Company through the date of payment, except as otherwise provided in Section 4. For the 2024 calendar year, the Annual Bonus shall be pro-rated for Executive's partial year of employment.

C. Equity Compensation. Upon the terms and conditions set forth in this subsection, in special consideration for the Non-Competition Covenant (as defined in Section 8 below), and subject to the approval of the Board of Directors or Compensation Committee, on or after the Effective Date, Company shall grant Executive 200,000 Restricted Stock Units ("RSUs") pursuant to and in accordance with the terms and conditions of Company's 2023 Employment Inducement Award Plan (the "Plan") and Company's form of restricted stock unit grant agreement. Each RSU represents a contingent right to receive one share of common stock based upon the RSU's vesting provisions. The RSUs will vest over a three (3) year period, with one-third vesting on the first anniversary of the Effective Date and the remainder vesting in two (2) equal annual installments thereafter until such time that all such shares are fully vested, subject to Executive's continuous employment to Company through each such vesting date; provided, that the RSUs, and each other outstanding equity award granted to Executive, shall accelerate so as to be fully vested and immediately exercisable immediately prior to any Change in Control (as defined in the Plan) of the Company, subject to Executive's continuous employment to Company through such Change in Control.

D. Retirement, Welfare and Other Benefit Plans and Programs. During the Employment Term, Executive shall be entitled to participate in the employee retirement and welfare benefit plans and programs made available to Company's other senior level executives as a group, as such retirement and welfare plans may be in effect from time to time and subject to the eligibility requirements of such plans, including but not limited to, life, health and disability plans, and a 401(k) retirement plan and similar or other plans. During the Employment Term, Executive shall be eligible for Paid Time off and holidays in accordance with the Company's policies. Nothing in this Agreement or otherwise shall prevent Company from amending or terminating after the Effective Date any retirement, welfare or other employee benefit plans, programs, policies or perquisites from time to time as Company deems appropriate, and Executive's participation in any such plan, program, policy and perquisite shall be subject to the terms, provisions, rules and regulations thereof.

E. Reimbursement of Expenses. During the Employment Term, Company shall reimburse Executive for all reasonable and necessary business expenses that Executive incurs while performing Executive's duties under this Agreement in accordance with Company's general policies of expense reimbursement in effect from time to time.

F. Relocation Bonus. To assist with the Relocation as defined in Section 1(D) herein, Company will provide Executive with a one-time cash payment of \$25,000, less deductions and withholdings required by applicable law (the "Relocation Bonus"), if the Relocation has been completed by January 31, 2025, subject to specified repayment requirements if the Executive's Date of Termination (as defined below in Section 4), is prior to the first anniversary of the date Relocation is completed. The Relocation Bonus shall be paid within thirty (30) days of completion of the Relocation, but no later than March 2, 2025.

3. Company Policies and Procedures. Executive agrees to observe and comply with the reasonable and lawful policies and procedures of Company as adopted by the Board of Directors in writing or reflected in the formal minutes of the Board of Directors or committee thereof, respecting performance of Executive's duties and to carry out and to perform the reasonable and lawful orders and directions stated by Company to Executive, from time to time, either orally or in writing. Executive agrees that Executive will be subject to any compensation clawback, recoupment and anti-hedging policies that may be applicable to Executive as an executive of Company, as in effect from time to time and as approved by the Board of Directors or a duly authorized committee thereof.

4. TERMINATION.

A. Notice of Termination and Date of Termination. Each Party must give written notice to the other of the intent to terminate this Agreement and Executive's employment hereunder ("Notice of Termination"). The Notice of Termination must specify a date of termination of employment, which shall incorporate any period of notice required by this Section 4 ("Date of Termination"). Executive may terminate Executive's employment at any time by giving the Company Notice of Termination at least 30 days prior to the Date of Termination designated by Executive. Company may terminate Executive's employment at any time by giving Executive a Notice of Termination at least 30 days prior to the Date of Termination designated by the Company.

B. Executive's Death or Total Disability. Executive's employment under this Agreement shall terminate upon the date of Executive's death. Additionally, if, during the Employment Term, Executive suffers a Total Disability (as defined in Section 4(E)(3)(iii)), then Company may terminate Executive's employment under this Agreement by giving Executive a Notice of Termination specifying the Date of Termination. Upon such termination due to death or Total Disability, Company shall pay to Executive or Executive's estate (i) any Base Salary that has fully accrued but not been paid as of the Date of Termination, as well as any vested and accrued employment benefits subject to the terms of any applicable employment benefit arrangements and applicable law ("Accrued Benefits") and (ii) an amount in cash equal to the Pro-Rated Target Bonus (as defined below), which amount shall be calculated and paid in the same manner as set forth below in Section 4(E)(1)(b). All other rights and benefits of Executive and Executive's dependents hereunder shall terminate upon such termination, except for any right to the continuation of benefits otherwise provided by law.

C. Company with Cause. Company may terminate with Cause (as defined in Section 4(E)(3)(i)) Executive's employment hereunder at any time. In order to terminate Executive's employment hereunder with Cause, Company must give Notice of Termination to Executive specifying the Cause and the Date of Termination (as defined in Section 4(A)). Upon termination with Cause, Company shall pay to Executive all Accrued Benefits. All other rights and benefits of Executive hereunder shall terminate upon such termination, except for any right to the continuation of benefits otherwise provided by law.

D. By Executive without Good Reason or by Mutual Agreement Executive may terminate Executive's employment without Good Reason (as defined in Section 4(E)(3)(iv)) at any time by giving Company Notice of Termination at least 30 days prior to the Date of Termination designated by Executive. In addition, this Agreement may be terminated at any time by written mutual agreement of the Parties with or without notice. Upon termination of Executive's employment by Executive without Good Reason or termination by mutual agreement of the parties, Company shall pay to Executive all Accrued Benefits. All other rights and benefits of Executive hereunder shall terminate upon such termination, except for any right to the continuation of benefits otherwise provided by law.

E. Without Cause by Company or For Good Reason by Executive Company may terminate Executive's employment at any time without Cause by giving Executive a Notice of Termination at least one day prior to the Date of Termination, and Executive may terminate Executive's employment for Good Reason by giving Company a Notice of Termination in accordance with Section 4(E)(3)(iv) below. Upon termination of Executive's employment without Cause by Company or for Good Reason by Executive, Company will pay Executive (i) all Accrued Benefits and (ii) the severance compensation payable set forth below in this Section 4(E). All other rights and benefits of Executive hereunder shall terminate upon such termination, except for any right to the continuation of benefits otherwise provided by law.

(1) In the event that Company terminates Executive's employment without Cause or Executive terminates his or her employment for Good Reason, and contingent upon the Executive's execution of a release of claims in the form attached hereto as Exhibit B, then Company shall pay to Executive as severance compensation, the following amounts:

(a) An amount in cash equal to 1.0 times Executive's Base Salary (at the rate payable at the time of such termination) for a period of twelve (12) months following the Date of Termination. Such severance compensation shall be paid by Company in the form of salary continuation in equal installments according to Company's customary payroll practices, with the first payment made on the first regularly scheduled pay day immediately following the Date of Termination, but in any event payments shall be made not less frequently than monthly; provided, however, that (a) Company shall pay such severance in a lump sum on the first regularly scheduled pay day immediately following the Date of Termination if such termination of employment occurs upon or within one year following a Sale, and the Sale constitutes a "change in control event" as defined under Section 409A of the Internal Revenue Code of 1986, as amended (the "Code"), to the extent required to comply with Section 409A of the Code; and (b) notwithstanding the preceding clause (a), if the Sale is not a "change in control event" as defined under Section 409A of the Code and penalty taxes may result under Section 409A if such severance compensation is paid in a lump sum, then the severance compensation will be paid in equal installments according to Company's customary payroll practices, with the first payment made on the first regularly scheduled pay day immediately following the effective date of termination, but in any event payments shall be made not less frequently than monthly.

(b) To the extent Executive is entitled to participate in an annual cash incentive compensation program established by the Company for the year in which the Date of Termination occurs based upon the determination of the Company in its sole discretion that the Executive has achieved relevant performance goals and Company performance objectives, as established in advance by the Board of Directors or the Compensation Committee, have occurred, Executive shall receive an amount in cash equal to (i) the Target Annual Bonus for the year in which the Date of Termination occurs (measured at the target level, identified "goal" target or other similar target, without taking into account any incentive override for above goal performance, or any project-specific or other non-standard incentives) times (ii) a fraction, (A) the numerator of which is the number of days in which Executive was employed by Company during the year in which the Date of Termination occurs, including the Date of Termination, and (B) the denominator of which is 365 (the "Pro-Rated Target Bonus"), which shall be paid within 60 days following the Date of Termination.

(c) During the twelve (12) month period following the Date of Termination, if Executive timely elects continued coverage under Section 4980B of the Code ("COBRA"), Company will reimburse Executive for the monthly COBRA premiums necessary to continue health coverage under the health plans of Company paid by Executive for Executive, and, if applicable, Executive's spouse and dependents, less the amount that Executive would be required to contribute for health coverage if Executive were an active employee of Company; provided that such reimbursements shall not continue beyond the first to occur of (x) the date on which Executive fails to pay the COBRA premiums and (y) the date on which Executive is eligible for substantially similar coverage from a subsequent employer (such period from the Date of Termination through the earlier of (x)-(y), the "COBRA Payment Period"). These reimbursements will commence on the first regularly scheduled pay day immediately following the Date of Termination and will be paid on the first regularly scheduled pay day of each month, provided that Executive demonstrates proof of payment of the applicable premiums prior to the applicable reimbursement payment date. Notwithstanding the foregoing, if at any time Company determines that its payment of COBRA premiums on Executive's behalf would result in a violation of applicable law or the incurrence of an excise tax (including, but not limited to, the 2010 Patient Protection and Affordable Care Act, as amended by the 2010 Health Care and Education Reconciliation Act), then in lieu of paying COBRA premiums pursuant to this section, Company shall pay Executive on the last day of each remaining month of the COBRA Payment Period, a fully taxable cash payment equal to the COBRA premium for such month, subject to applicable tax withholding, for the remainder of the COBRA Payment Period, and Executive shall not be obligated to use any monthly, taxable cash payment to pay COBRA premiums. In the event that Executive is on COBRA continuation coverage and Company is required by law to subsidize all or any portion of the monthly COBRA premium that Executive is required to pay, Executive shall not be entitled to reimbursement of the monthly COBRA premium (except as required by law) or the taxable monthly payment of the COBRA premium amount to the extent of the amount of the required subsidy. Nothing in this Agreement shall deprive Executive of Executive's rights under COBRA or ERISA for benefits under plans and policies arising under his employment by Company.

(d) The vesting of each outstanding equity award granted to Executive will accelerate so that such awards will be fully vested as of the Date of Termination. If any equity awards vest based on the attainment of performance goals, the performance goals will be deemed to have been met as of the Date of Termination, unless such greater amount of vesting is provided for in the applicable award agreements.

(2) Payment of the severance compensation shall be subject to all mandatory and voluntary payroll deductions. In the event that Executive materially breaches any of his or her post-employment covenants or obligations set forth in this Agreement and fails to cure such breach within fifteen (15) calendar days following receipt from Company of notice to cure such breach, then the payment of severance compensation pursuant to this section shall terminate

immediately and permanently. During the period that Executive is paid the foregoing severance compensation, Executive shall not further accrue any other benefits under any benefit plans of which Executive was a participant while employed by Company, except as otherwise required by applicable federal or state law, by the express terms of this Agreement, or by the express terms of such benefit plans.

(3) For purposes of this Agreement:

(i) Executive's employment will be deemed to have been terminated by Company "with Cause" if the termination arises from or relates to a determination by the Board of Directors that (a) Executive performed an act or acts of willful and material malfeasance or misconduct with respect to the performance of Executive's duties and responsibilities as an employee and officer of Company or under this Agreement that resulted in material harm to Company that remains uncorrected for fifteen (15) days after receipt of written notice by Company to Executive; or (b) except as otherwise provided in Section 1(C), Executive's continued failure to devote his or her full business time and attention and his or her best efforts to the faithful performance of his or her material duties and responsibilities (other than a failure resulting from Executive becoming disabled) that remains uncorrected for fifteen (15) days after receipt of written notice by Company to Executive; or (c) Executive's material breach of any material provision of this Agreement that remains uncorrected for fifteen (15) days after receipt of written notice by Company to Executive; or (d) Executive committed an act of fraud, embezzlement, misappropriation, or personal dishonesty against Company; or (e) the conviction, or plea of *nolo contendere*, of Executive to a crime constituting a felony; or (f) Executive engaged in discrimination, harassment, or other behaviors that would be reasonably likely to bring the company negative publicity or embarrassment "resulting in or intending to result in personal gain to Executive at the expense of the Company."

(ii) Executive's employment shall be deemed to have been terminated by Company "without Cause" if such termination does not arise from or relate to any of acts or omissions constituting "Cause" as set forth in clauses (a) through (f) of the immediately preceding subsection, and such termination is not the result of Executive's death or Executive suffering a Total Disability.

(iii) Executive shall be deemed to have suffered a "Total Disability" if (a) Executive is granted long-term disability benefits under Company's long-term disability plan or (b) Executive becomes physically or mentally disabled so that Executive is unable to perform the essential functions of Executive's job, with or without reasonable accommodation in accordance with the Americans with Disabilities Act and its amendments, for a period of one hundred eighty (180) consecutive days.

(iv) Executive shall be deemed to have terminated his or her employment for "Good Reason" if Executive terminates his or her employment on account of the occurrence of one or more of the following without Executive's consent:

(a) A material diminution by Company of Executive's authority, duties or responsibilities the duration of which is greater than fifteen (15) days and which is not the result of Executive's acts or omissions;

(b) A material change in the geographic location at which Executive must perform services under this Agreement (which, for purposes of this Agreement, means the requirement by Company that Executive work at a location more than fifty (50) miles from the location at which Executive performs services immediately prior to the relocation) (provided that, for the avoidance of doubt, the relocation of Executive as contemplated by Section 1(D) shall not give rise to "Good Reason");

(c) A material diminution in Executive's Base Salary and/or Target Annual Bonus which is not the result of Executive's acts or omissions; or

(d) Any action or inaction that constitutes a material breach by Company of this Agreement, including the failure of Company to pay any amounts due under Section 2 or the failure of Company to obtain from its successors the express assumption and agreement required under Section 16(A).

Executive must provide Notice of Termination for Good Reason to Company within sixty (60) days after the event constituting Good Reason, which must state with reasonable specificity the applicable facts and circumstances underlying such finding of Good Reason. Company shall have a period of thirty (30) days in which it may correct the act or failure to act that constitutes the grounds for Good Reason as set forth in Executive's Notice of Termination. If Company does not correct the act or failure to act, then, in order for the termination to be considered a Good Reason termination, Executive must terminate his or her employment for Good Reason by giving Notice of Termination with a Date of Termination designated by Executive which is at least thirty (30) days after the date on which the Notice of Termination is given but not more than ninety (90) days after the end of the cure period.

(4) In the event Company terminates Executive's employment with Cause, Executive voluntarily terminates his or her employment with Company other than for Good Reason, or such employment is terminated by mutual agreement or as the result of Executive's death or Total Disability, Executive shall not be entitled to payment of any severance compensation under this Agreement.

F. Cooperation after Notice of Termination. Following any Notice of Termination by either Company or Executive, Executive, if requested by Company, shall reasonably cooperate with Company in all matters relating to the winding up of Executive's pending work on behalf of Company and the orderly transfer of any such pending work to other employees of Company as may be reasonably designated by Company following the Notice of Termination. Executive shall not receive any additional compensation during the Employment Term, other than Executive's Base Salary, for any services that Executive renders as provided in this Section 4(F), provided that, if Executive is not receiving any severance compensation pursuant to this Section 4, for each day that Executive performs services under this Section 4(F) after the Employment Term, Executive shall be reimbursed for his or her reasonable out-of-pocket expenses and Company shall pay Executive a per diem cash amount equal to 130% of Executive's Base Salary rate (expressed on a daily basis) on the Date of Termination.

G. Relocation Repayment Upon Termination. If the Executive's Date of Termination is prior to the earlier of the first anniversary of the completion of the Relocation, or January 31, 2026, Executive shall repay a pro-rated portion of the actual costs of the Relocation which shall include the sum of (x) the actual costs paid by the Company, either directly to Executive or to the Company's third party relocation provider, up to \$125,000; and (y) the Relocation Bonus (together, the "Relocation Payments"), in an amount equal to (i) the Relocation Payments times (ii) a fraction, (A) the numerator of which is the number of days remaining from the Date of Termination until the earlier of the first anniversary of the completion of the Relocation, or January 31, 2026, and (B) the denominator of which is 365, which shall be paid in cash by Executive to Company and/or withheld from any payments due to Executive as of the Date of Termination.

H. Surrender of Records and Property. Upon termination of employment or earlier upon the Company's request or demand, Executive shall promptly turn-over or deliver to Company at Company's expense all property of Company in Executive's possession, custody, or control, including without limitation thereto: records (paper and electronic), files (paper and electronic), documents (paper and electronic), electronic mail (e-mail) on Company accounts, letters, financial information, memorandum, notes, notebooks, contracts, project manuals, specifications, reports, data, tables, calculations, data, electronic information, and computer disks, in all cases whether or not such property constitutes Confidential Information (as defined below), and all copies thereof; all keys to motor vehicles, offices or other property of Company; and all computers, cellular phones and other property of Company. If any of the foregoing property of Company is electronically stored on a computer or other storage medium owned by Executive or a friend, family member or agent of Executive, such information shall be copied onto a computer disk to be delivered to Company together with a written statement of Executive that the information has been deleted from such person's computer or other storage medium.

5. Section 280G of the Code

A. The following terms shall have the meanings set forth below for purposes of this Section 5:

(1) "Accounting Firm" means a certified public accounting firm chosen by the Company.

(2) "After-Tax" means after taking into account all applicable Taxes and Excise Tax.

(3) "Excise Tax" means the excise tax imposed by Section 4999 of the Code, together with any interest or penalties imposed with respect to such excise tax.

(4) "Safe Harbor Amount" means one dollar less than three times Executive's "base amount," within the meaning of Section 280G(b)(3) of the Code.

(5) "Taxes" MEANS ALL FEDERAL, STATE, LOCAL AND FOREIGN INCOME, EXCISE, SOCIAL SECURITY AND OTHER TAXES, OTHER THAN THE EXCISE TAX, AND ANY ASSOCIATED INTEREST AND PENALTIES.

B. If any payment, individually or in the aggregate, due to Executive under this Agreement (a Payment) is subject to the Excise Tax, then such Payment shall be adjusted, if necessary, to equal the greater of (x) the Safe Harbor Amount or (y) the Payment, whichever results in such Executive's receipt, After-Tax, of the greatest amount of the Payment. The reduction of Executive's Payments pursuant to this Section 5.B., if applicable, shall be made by first reducing the acceleration of Executive's stock option vesting (if any), the acceleration of the vesting of Executive's other equity securities (if any), and then by reducing any cash payments owed to the Executive, in that order.

All determinations required to be made under this Section 5, including whether and in what manner any Payments are to be reduced pursuant to the second sentence of Section 5.B., and the assumptions to be utilized in arriving at such determinations, shall be made by the Accounting Firm, and shall be binding upon the Company and Executive, except to the extent the Internal Revenue Service or a court of competent jurisdiction makes an inconsistent final and binding determination. The Accounting Firm shall provide detailed supporting calculations both to the Company and Executive within fifteen (15) business days after receiving notice from Executive that there has been a Payment or such earlier time as may be requested by the Company. All fees and expenses of the Accounting Firm shall be borne solely by the Company.

6. Intellectual Property.

A. Work Product. During the Employment Term, Executive will be expected to perform duties which may lead to and include the discovery, creation, development, or expression of inventions, discoveries, developments, modifications, procedures, ideas, innovations, systems, programs, know-how, literary properties, chemical or biological data, computer software, improvements, processes, methods, formulas, systems, creative works and techniques (collectively, hereinafter "Work Product").

B. Assignment. Executive hereby assigns and transfers to Company, and agrees that Company shall be the sole owner of all Work Product conceived, developed or made by Executive (alone or with others), whether during working hours or at any other time, in whole or in part during Executive's employment with Company, whether at the request or upon the suggestion of Company or otherwise, which are useful in, or directly or indirectly related to Company's business or any contemplated business of Company or which relate to, or are conceived, developed, or made in the course of, Executive's employment or which are developed or made from, or by reason of knowledge gained from, such employment.

C. Work for Hire. Executive hereby agrees that all work or other material containing or reflecting any Work Product shall be deemed a work made for hire under the U.S. Copyright Act. To the extent any such Work Product is determined that it is not a work made for hire, Executive hereby assigns to Company all of Executive's right, title and interest, including all rights of copyright, patent, trade secret and other intellectual property rights, in, to and under the Work Product.

D. Continuing Obligations. Executive agrees to disclose promptly all Work Product conceived or made by Executive (alone or with others) to which Company is entitled to as provided herein, and agrees not to disclose such Work Product to others except as required by law or as is reasonably necessary or appropriate in connection with the performance of Executive's duties as an employee and officer of Company, without the express written consent of Company. Executive further agrees that during the Employment Term and at any time thereafter, Executive will, upon request by Company, provide all assistance reasonably required to protect, perfect and use the Work Product, including execution of proper assignments to Company of any and all such Work Product to which Company is entitled, execution of all papers and performance all other lawful acts which Company may deem necessary or advisable for the preparation, prosecution, procurement and maintenance of trademarks, copyrights and or patent applications, and execution of any and all proper documents as shall be required or necessary to vest title in Company to such Work Product. It is understood that all expenses in connection with such trademarks, copyrights or patents, and all applications related thereto, shall be borne by Company, however Company is under no obligation to protect such Work Product, except at its own discretion and to such extent as Company shall deem desirable. Executive shall not receive any additional compensation during the Employment Term, other than Executive's Base Salary, for any services that Executive renders as herein provided. For each day that Executive performs services under this Section 6(D) after the Employment Term, Executive shall be reimbursed for his or her reasonable out-of-pocket expenses and Company shall pay Executive a per diem cash amount equal to 130% of Executive's Base Salary rate (expressed on a daily basis) on the Date of Termination.

7. Confidential Information.

A. Confidential Information. The term "Confidential Information" means all information related to Company's business, which exists or is developed at any time while Executive is an employee, officer and/or director of Company, including without limitation: (i) strategic and development plans, financial information, equity investors, business plans, co-developer identities, business relationships, business records, project records, market reports, information relating to processes and techniques, technology, research, data, development, trade secrets, know-how, discoveries, ideas, concepts, specifications, diagrams, inventions, technical and statistical data, designs, drawings, models, flow charts, engineering, products, invention disclosures, patent applications, chemical and molecular structures, synthetic pathways, biological data, safety data, clinical data, developmental data, development route, manufacturing processes, synthetic techniques, analytical data, Work Product, and any and all other proprietary and sensitive information, disclosed or learned, whether oral, written, graphic or machine-readable, whether or not marked confidential or proprietary, whether or not patentable, whether or not copyrightable, including the manner and results in which any such Confidential Information may be combined with other information or synthesized or used by Company, which could prove beneficial in enabling a competitor to compete with Company; or (ii) information that satisfies the definition of a "trade secret" as that term is defined in the Uniform Trade Secrets Act or any applicable state trade secret law, each as amended from time to time; provided, however, that information that is in the public domain (other than as a result of a breach by Executive of this Section 7), was released in accordance and compliance with an approval from the Company for the release, or lawfully obtained from a third party who is not known by Executive (after Executive's reasonable inquiry) to be bound by any confidentiality obligations owed to the Company with regard to the information is not Confidential Information. The Executive understands that the above list is not exhaustive, and that Confidential Information also includes other information that is marked or otherwise identified as confidential or proprietary, or that would otherwise appear to a reasonable person to be confidential or proprietary in the context and circumstances in which the information is known or used.

B. Acknowledgements. Executive acknowledges and agrees that: (1) Executive's position with Company is one of high trust and confidence, (2) the Confidential Information constitutes a valuable, special and unique asset which Company uses to obtain a competitive advantage over its competitors, (3) Executive's protection of such Confidential Information against unauthorized use or disclosure is critically important to Company in maintaining its competitive advantage, (4) all Confidential Information is the property of Company, and (5) Executive shall acquire no right, title or interest in, to or under any such Confidential Information.

C. Nondisclosure. Executive promises that, unless legally compelled to do so, Executive will never (before, during or after the Employment Term): (1) disclose any Confidential Information to any person other than (i) an officer or director of Company; or (ii) any other person who is bound by nondisclosure restrictive covenants to Company and to whom disclosure of such Confidential Information is reasonably necessary or appropriate in connection with performance by Executive of Executive's duties as an employee and officer of Company; or (2) use any Confidential Information except to the extent it is reasonably necessary or appropriate in connection with performance by Executive of Executive's duties as an employee and officer of Company. Executive promises to take all reasonable precautions to prevent the inadvertent or accidental disclosure or misuse of any Confidential Information. In the event Executive receives a request to disclose all or any part of the Confidential Information under the terms of a subpoena or order issued by a court or governmental body, Executive promises, to the extent permissible by law, to (a) notify Company immediately of the existence, terms and circumstances surrounding such request, (b) consult with Company on the advisability of taking legally available steps to resist or narrow such request, (c) if disclosure is required, furnish only such portion of the Confidential Information as Executive is legally compelled to disclose; and (e) exercise Executive's best efforts to obtain an order or other reliable assurance that confidential treatment will be accorded to the disclosed Confidential Information.

D. Executive acknowledges that Company has provided Executive notice of immunity rights under the Defend Trade Secrets Act, which states: (1) An individual shall not be held criminally or civilly liable under any Federal or State trade secret law for the disclosure of a trade secret that (A) is made (i) in confidence to a Federal, State, or local government official, either directly or indirectly, or to an attorney, and (ii) solely for the purpose of reporting or investigating a suspected violation of law, or (B) is made in a complaint or other document filed in a lawsuit or other proceeding, if such filing is made under seal; and (2) an individual who files a lawsuit for retaliation by an employer for reporting a suspected violation of law may disclose a trade secret to the attorney of the individual and use the trade secret information in the court proceeding, if the individual (A) files any document containing the trade secret under seal, and (B) does not disclose a trade secret, except pursuant to court order.

8. Noncompetition.

A. Restricted Period. As used in this Agreement, the term "Restricted Period" means throughout the Employment Term and, to the extent Executive has moved from California to Massachusetts or another state approved by the CEO, continuing until the end of the twelve (12) month period following the date on which Executive's employment with Company is terminated for any reason (whether voluntary or involuntary); provided, however, if Executive's employment is terminated without Cause and Executive resides in Massachusetts and/or Massachusetts law is deemed to apply to this Section 8, the Restricted Period shall extend only through the Employment Term.

B. Prohibition on Competition. Executive hereby covenants and agrees that, until the expiration of the Restricted Period, Executive will not serve as an officer, director, employee, independent contractor, consultant or agent of, or have any ownership interest in, any business entity which engages in any activities that are materially similar to or competitive with Company's pharmaceutical product development and Commercialization (as defined below) activities in the fields of (i) Niemann-Pick disease type C ("NPC"); (ii) idiopathic hypersomnia ("IH"), narcolepsy or other sleep disorders; (iii) Vascular Ehler-Danlos Syndrome ("VEDS"); and/or (iv) such other indications, products or product candidates which Company is actively and demonstrably developing and/or Commercializing at the time Executive's employment is terminated; provided, however, the restriction on Executive providing services as an employee, independent contractor, consultant or agent shall apply only to the extent Executive is providing (a) operational, executive, strategic, consulting, marketing, business or legal services, (b) Executive is providing services similar to the services Executive provided to Company during Executive's employment with Company, or (c) Executive could use Confidential Information in the scope of providing such services ("Non-Competition Covenant"). If a court of competent jurisdiction finds this non-competition provision invalid or unenforceable due to unreasonableness in time, geographic scope, or scope of Company's business, then Executive agrees that such court shall interpret and enforce this provision to the maximum extent that such court deems reasonable. For purposes of this Agreement, "Commercialize" or "Commercialization" means the sales and marketing phase with regard to a specific drug candidate in a specific country or region following the regulatory approval of said drug candidate in the applicable country or region.

C. Exceptions. Executive's ownership of less than 5% of the stock of a company that is competitive with the activities of Company as described in Section 8(B) and listed on a national securities exchange shall not be deemed to violate the prohibitions of Section 8(B). Also, Executive shall not be considered to have violated Section 8(B) with respect to the purchasing entity if there is a Sale and Executive becomes an employee, officer, director or shareholder of the purchasing entity. The term "Sale" means the sale of more than 50% of the equity of Company, a merger of Company with an entity the equity of which after the merger the stockholders of Company immediately prior to such merger own less than 50%, or the sale of all or substantially all of the assets of Company, in any case to a person or entity not affiliated with Company. Neither a recapitalization nor change of form of Company shall be considered a Sale. Additionally, a "Sale" shall not be deemed to have occurred as a result of a lender exercising any of its remedies in connection with the occurrence or continuation of an event of default under that certain Credit Agreement dated as of April 5, 2024, by and between Company, Alter Domus (US) LLC, and the lenders as defined therein, or any other indebtedness of the Company which may be added from time to time in the future.

9. Non-solicitation of Employees. Until the expiration of the Restricted Period, Executive shall not, directly or indirectly, either on Executive's own account or for any other person or entity: (a) solicit, induce, advise, or otherwise convince any then-employee of the Company to leave their employment with the Company; (b) interfere with Company's employment of any then-employee of the Company; (c) offer employment to any then-employee of Company; (d) interfere with Company's engagement with, or offer employment to, any then-consultant of Company; or (e) induce or attempt to induce any such employee or consultant to breach their employment agreement or relationship or consulting agreement or relationship with Company; provided, however, that Executive shall not be in breach of this provision if any such employee or consultant, without inducement or solicitation by Executive, applies for employment at Executive's subsequent employer in response to a general advertisement soliciting employment.

10. Reasonableness Of Restrictions; Remedies. Executive has carefully read and considered the restrictive covenants set forth in Sections 6 – 9 hereof, and understands Executive's obligations thereunder, the limitations such obligations will impose upon Executive after termination of Executive's employment with Company, and that the Restricted Period extends for 12 months after the termination of Executive's employment. Executive has had full opportunity to review with Executive's personal attorney this Agreement, including Sections 6 – 9, before executing the Agreement. Executive agrees that, as a result of Executive's position with Company, the length of the Restricted Period and each restriction set forth in Sections 6, 7, 8 and 9 herein are (1) fair and reasonable, (2) reasonably required for the protection of the legitimate business interests and goodwill established by Company, and (3) not overly broad or unduly burdensome to Executive. Executive acknowledges that Executive's compliance with Executive's obligations and restrictive covenants set forth in this Agreement is necessary to protect the business and goodwill of Company. Executive agrees that Executive's breach of Executive's obligations and/or restrictive covenants under this Agreement may irreparably and continually damage Company, for which money damages may not be adequate. Consequently, Executive agrees that in the event that Executive breaches or threatens to breach any of the covenants or agreements contained herein, Company shall be entitled to: (a) seek injunctive relief to prevent or halt Executive from breaching this Agreement; and (b) money damages as determined appropriate by a court of competent jurisdiction. Executive hereby agrees that injunctive relief may be granted by a court of competent jurisdiction without the necessity of Company to post bond, or if required to post bond, Executive agrees that the lowest amount permitted shall be adequate. Nothing in this Agreement shall be construed to prohibit Company from pursuing any other remedy available or from seeking to enforce any restrictive covenants to a lesser extent than set forth herein. The Parties agree that all remedies shall be cumulative.

11. No Prior Restrictions. Executive hereby represents and warrants to Company that the execution, delivery, and performance by Executive of Executive's duties under this Agreement do not violate any provision of any agreement or restrictive covenant which Executive has with any former employer or any other entity. Executive further agrees to honor and inform Company of any and all post-employment obligations Executive has to any former employer or any other entity with which Executive has or had a business relationship.

12. Notices. Any notice or communication required or permitted to be given hereunder may be delivered by hand, deposited with an overnight courier, sent by confirmed email, confirmed facsimile, or mailed by registered or certified mail, return receipt requested, postage prepaid, in the case of Company, addressed to Company's principal office marked attention to Company's president, and in the case of Executive, addressed to Executive's personal address as appearing in Company's payroll records, and in each case to such other mail address, e-mail address, or facsimile number as may hereafter be furnished in writing by either Party to the other Party. Such notice will be deemed to have been given as of the date it is hand delivered, emailed, faxed or three days after deposit in the U.S. mail.

13. PROTECTED RIGHTS. NOTHING IN THIS AGREEMENT IS INTENDED TO OR SHALL RESTRICT EXECUTIVE FROM (A) COMMUNICATING OR COOPERATING WITH OR OTHERWISE PARTICIPATING IN ANY INVESTIGATION OR PROCEEDING CONDUCTED BY THE SECURITIES AND EXCHANGE COMMISSION, THE FINANCIAL INDUSTRY REGULATORY AUTHORITY, THE EQUAL EMPLOYMENT OPPORTUNITY COMMISSION, THE NATIONAL LABOR RELATIONS BOARD, THE OCCUPATIONAL SAFETY AND HEALTH ADMINISTRATION, THE COMMODITY FUTURES TRADING COMMISSION, THE DEPARTMENT OF JUSTICE OR ANY OTHER FEDERAL, STATE OR LOCAL GOVERNMENT AGENCY OR COMMISSION, INCLUDING PROVIDING DOCUMENTS OR OTHER INFORMATION, WITHOUT NOTICE TO COMPANY; OR (B) EXERCISING ANY RIGHTS EXECUTIVE MAY HAVE UNDER SECTION 7 OF THE U.S. NATIONAL LABOR RELATIONS ACT, SUCH AS THE RIGHT TO ENGAGE IN CONCERTED ACTIVITY, INCLUDING COLLECTIVE ACTION OR DISCUSSION CONCERNING WAGES OR WORKING CONDITIONS.

14. Likeness. Executive hereby grants to Company a license to use, without further compensation or approval from Executive, Executive's name, image, portrait, voice, likeness and all other rights of publicity, or any derivative or modification thereto that Company may create, in any and all mediums, now known or hereafter developed, provided that such use is in relation to Company's business and consistent with professional business standards, and does not disparage or denigrate Executive. Provided, however, if written notice is provided to Company by Executive following termination of Executive's employment requesting that Company cease using Executive's likeness, Company has 30 days to cease using Executive's likeness in the manner set forth in the notice.

15. Section 409A.

A. This Agreement is intended to comply with Section 409A of the Code and its corresponding regulations, or to satisfy an exemption, and payments may only be made under this Agreement upon an event and in a manner permitted by Section 409A of the Code, to the extent applicable. Notwithstanding anything in this Agreement to the contrary, if required by Section 409A of the Code, if Executive is considered a "specified employee" for purposes of Section 409A and if payment of any amounts under this Agreement is required to be delayed for a period of six months after separation from service pursuant to Section 409A of the Code, payment of such amounts shall be delayed as required by Section 409A of the Code, and the accumulated amounts shall be paid in a lump sum payment within 10 days after the end of the six month period. If Executive dies during the postponement period prior to the payment of benefits, the amounts withheld on account of Section 409A of the Code shall be paid to the personal representative of Executive's estate within 60 days after the day of Executive's death. The Parties agree that this Section 14 shall not be construed in a manner so as to accelerate any payments due under this Agreement.

B. All payments to be made upon a termination of employment under this Agreement may only be made upon a "separation from service" under Section 409A of the Code. For purposes of Section 409A of the Code, each payment hereunder shall be treated as a separate payment and the right to a series of installment payments under this Agreement shall be treated as a right to a series of separate payments. In no event may Executive, directly or indirectly, designate the calendar year of a payment.

C. To the extent that any reimbursements under this Agreement are subject to Section 409A, (i) any such reimbursements payable to Executive shall be paid to Executive no later than December 31 of the year following the year in which the expense was incurred, (ii) Executive shall submit Executive's reimbursement request promptly following the date the expense is incurred, (iii) the amount of expenses reimbursed in one year shall not affect the amount eligible for reimbursement in any subsequent year, other than medical expenses referred to in Section 105(b) of the Code, and (iv) Executive's right to reimbursement under this Agreement will not be subject to liquidation or exchange for another benefit.

16. INDEMNIFICATION; LIABILITY INSURANCE. Company shall indemnify and hold Executive harmless to the fullest extent permitted by the laws of Company's state of organization or incorporation in effect at the time against and in respect of any and all actions, suits, proceedings, claims, demands, judgments, costs, expenses (including advancement of reasonable attorney's fees), losses, and damages resulting from Executive's performance of Executive's duties and obligations with Company. Executive will be entitled to be covered, both during and, while potential liability exists, by any insurance policies Company may elect to maintain generally for the benefit of officers and directors of Company against all costs, charges and expenses incurred in connection with any action, suit or proceeding to which Executive may be made a party by reason of being an officer or director of Company in the same amount and to the same extent as Company covers its other officers and directors. These obligations shall survive the termination of Executive's employment with Company.

17. General Provisions.

A. Successors and Assigns. This Agreement is personal to the Executive and shall not be assigned by the Executive. Any purported assignment by the Executive shall be null and void from the initial date of the purported assignment. The Company may assign this Agreement to any successor or assign (whether direct or indirect, by purchase, merger, consolidation, or otherwise) to all or substantially all of the business or assets of the Company. This Agreement shall inure to the benefit of the Company and permitted successors and assigns.

B. Survival of Certain Terms. The terms, conditions and covenants set forth in this Agreement which specifically relate to periods, activities or obligations upon or subsequent to the termination of Executive's employment, including, without limitation, the restrictive covenants contained in Sections 6 – 9, shall survive the termination of this Agreement and Company's employment of Executive hereunder, and the Parties shall remain bound by such terms, conditions and covenants.

C. Governing Law; Jurisdiction. This Agreement shall be governed by and construed and enforced in accordance with the procedural and substantive laws of the State of Florida (or, prior to the relocation of Executive's residence from California, the State of California), without regard to its conflicts of laws provisions. The litigation of any disputes arising out of this Agreement shall take place in the appropriate federal or state court located in Osceola County (or, prior to the relocation of Executive's residence from California, in the State of California). The Parties, to the extent they can legally do so, hereby consent to service of process, and to be sued in the State of Florida and in the State of California and consent to the exclusive jurisdiction of the courts of the State of Florida and the United States District Court for the Middle District of Florida (or, prior to the relocation of Executive's residence from California, in the State of California), as well as to the jurisdiction of all courts to which an appeal may be taken from such courts, for the purpose of any suit, action or other proceeding arising out of any of their obligations hereunder or with respect to the transactions contemplated hereby, and expressly waive any and all objections they may have to venue in such courts. Notwithstanding the foregoing, should Executive refuse to comply with an order or judgment of such court, then Company may enforce this Agreement and the order or judgment of such court in any jurisdiction it deems appropriate.

D. Severability, Reform. If any provision of this Agreement is determined to be void, invalid or unenforceable, the remainder shall be unaffected and shall be enforceable as if the void, invalid or unenforceable part was not a provision of the Agreement.

E. Entire Agreement. This Agreement and its attached exhibits, which by this reference are hereby incorporated into and made a part of this Agreement as if set forth herein verbatim, contain the entire understanding of the parties to this Agreement and supersede and replace all former agreements or understandings, oral or written, between Company and Executive, including any offer letter sent to Executive, regarding the subject matter hereof.

F. Modification and Waiver. This Agreement may not be amended except by a written instrument signed by both Parties which specifically refers to the particular provision or provisions being amended. No provision of this Agreement may be waived except in a written instrument that specifically refers to the particular provision or provisions being waived and is signed by the Party against whom the waiver is being asserted. No waiver by any Party of any right, power or privilege hereunder shall constitute a waiver of any other right, power or privilege hereunder, and no waiver by any party of any breach of a provision hereunder shall constitute a waiver of any other breach of that or any other provision of this Agreement.

G. Taxes; Withholding. All compensation and benefits payable to Executive under this Agreement shall be subject to all income and other employment tax withholding and reporting required by federal, state or local law with respect to compensation, benefits and reimbursable expenses paid by a corporation to an employee. Executive shall be responsible for all taxes applicable to amounts payable under this Agreement.

H. Assistance in Litigation. Executive shall reasonably cooperate with Company in the defense or prosecution of any claims or actions now in existence or that may be brought in the future against or on behalf of Company that relate to events or occurrences that transpired while Executive was employed by Company. Executive's cooperation in connection with such claims or actions shall include being available to meet with counsel to prepare for discovery or trial and to act as a witness on behalf of Company at mutually convenient times. Executive also shall cooperate fully with Company in connection with any investigation or review by any federal, state or local regulatory authority as any such investigation or review relates, to events or occurrences that transpired while Executive was employed by Company. Notwithstanding anything to the contrary in this Section 16(H), unless otherwise mutually agreed between Executive and Company in writing and, for each day that Executive performs services under this Section 16(H) after the final payment by Company of any and all severance compensation due to Executive under Section 4(E)(1), Executive shall be reimbursed for his reasonable out-of-pocket expenses and Company shall pay Executive a per diem cash amount equal to 130% of Executive's Base Salary rate (expressed on a daily basis) on the Date of Termination.

I. Beneficiaries; References. Executive shall be entitled to select (and change to the extent permitted under any applicable law) a beneficiary or beneficiaries to receive any compensation or benefit payable hereunder following Executive's death, and may change such election, in either case by giving Company written notice thereof. In the event of Executive's death or a judicial determination of Executive's incompetence, reference in this Agreement to Executive shall be deemed, where appropriate, to refer to Executive's beneficiary, estate or other legal representative. Any reference to any gender in this Agreement shall include, where appropriate, the other gender.

J. Voluntary Agreement. Each Party to this Agreement has read and fully understands the terms and provisions hereof, has had an opportunity to review this Agreement with legal counsel, has executed this Agreement based upon such party's own judgment and advice of counsel, and knowingly, voluntarily and without duress, agrees to all of the terms set forth in this Agreement. The Parties have participated jointly in the negotiation and drafting of this Agreement. If an ambiguity or question of intent or interpretation arises, this Agreement will be construed as if drafted jointly by the Parties and no presumption or burden of proof will arise favoring or disfavoring any party because of authorship of any provision of this Agreement. Except as expressly set forth in this Agreement, neither the Parties nor their affiliates, advisors and/or their attorneys have made any representation or warranty, express or implied, at law or in equity with respect of the subject matter contained herein. Without limiting the generality of the previous sentence, Company, its affiliates, advisors and/or attorneys have made no representation or warranty to Executive concerning the state or federal tax consequences to Executive regarding the transactions contemplated by this Agreement.

K. Effect of Headings. Headings to sections and paragraphs of this Agreement are for reference only, and do not form a part of this Agreement, or effect the interpretation of this Agreement.

L. Counterparts. This Agreement may be executed in counterparts, including by transmission of facsimile or PDF copies of signature pages, each of which shall for all purposes be deemed to be an original and all of which shall constitute an instrument. All signatures of the parties transmitted by facsimile or PDF shall be deemed to be their original signatures for all purposes.

[SIGNATURE PAGE FOLLOWS]

SIGNATURE PAGE
OF
EMPLOYMENT AGREEMENT

IN WITNESS WHEREOF, COMPANY HAS CAUSED THIS AGREEMENT TO BE DULY EXECUTED AND DELIVERED BY ITS DULY AUTHORIZED OFFICER, AND EXECUTIVE HAS DULY EXECUTED AND DELIVERED THIS AGREEMENT, EFFECTIVE AS OF THE DATE FIRST WRITTEN ON PAGE 1 OF THIS AGREEMENT.

ZEVRA THERAPEUTICS, INC.
("COMPANY"):

By _____
R. LaDuane Clifton, MBA, CPA
Chief Financial Officer, Secretary
and Treasurer

Rahsaan Thompson ("Executive"):

EXHIBIT A

LIST OF OUTSIDE BUSINESS ACTIVITIES

1. Board service for Oakland Museum of California (non-profit).
 2. Board service for The Just Society (non-profit).
-

EXHIBIT B

FORM OF RELEASE OF CLAIMS

Separation of Employment Agreement and General Release

THIS SEPARATION OF EMPLOYMENT AGREEMENT AND GENERAL RELEASE (the "Agreement") is made as of this ____ day of _____, _____, by and between Rahsaan Thompson ("Executive") and Zevra Therapeutics, Inc. (the "Company").

WHEREAS, Executive is employed by Company as Chief Legal Officer, Secretary and Compliance Officer;

WHEREAS, Executive and Company entered into an Employment Agreement, effective as of June 20, 2024, (the "Employment Agreement") which provides for certain benefits in the event that Executive's employment is terminated on account of a reason set forth in the Employment Agreement;

WHEREAS, Executive's employment with Company will terminate effective _____ (the "Termination Date"); and

WHEREAS, in connection with the termination of Executive's employment, the parties have agreed to a separation package and the resolution of any and all disputes between them.

NOW, THEREFORE, IT IS HEREBY AGREED by and between Executive and Company as follows:

1. Executive, for and in consideration of the commitments of Company as set forth in paragraph 6 of this Agreement, and intending to be legally bound, does hereby REMISE, RELEASE AND FOREVER DISCHARGE Company, its stockholders, its present and past affiliates, subsidiaries and parents, their respective officers, directors, investors, employees, and agents, and their respective predecessors, successors and assigns, heirs, executors, and administrators (collectively, "Releasees"), subject to the exceptions of Section 2 of the Agreement, from all causes of action, suits, debts, claims and demands whatsoever in law or in equity, which Executive ever had, now has, or hereafter may have, whether known or unknown, or which Executive's heirs, executors, or administrators may have, by reason of any matter, cause or thing whatsoever, from the beginning of time to the date of this Agreement, to the extent arising from or relating in any way to Executive's employment relationship with Company, the terms and conditions of that employment relationship, and/or the termination of that employment relationship, including, but not limited to, (i) any claims for monetary damages arising under the Age Discrimination in Employment Act ("ADEA"), the Older Workers Benefit Protection Act ("OWBPA"), Title VII of The Civil Rights Act of 1964, the Americans with Disabilities Act; (ii) any and all claims arising under the Family and Medical Leave Act of 1993, the Employee Retirement Income Security Act of 1974, as amended; (iii) any and all claims arising under any applicable state and local fair employment practice laws and wage and hour laws, including the Massachusetts Wage Act and the California Fair Employment and Housing Act; (iv) any other claims under any federal, state or local common law, statutory, or regulatory provision, now or hereafter recognized; and (v) any claims for attorneys' fees and costs.

2. The foregoing shall in no event apply to (i) enforcement by Executive of Executive's rights under this Agreement, (ii) Executive's rights as a stockholder in Company or any of its affiliates, (iii) Executive's rights to indemnifications under any separate contract or insurance policy, (iv) Executive's right to seek unemployment insurance benefits, (v) Executive's right to seek workers' compensation benefits, (vi) any rights Executive has to indemnification for service as an officer of Company, or (vii) any claims that, as a matter of applicable law, are not waivable. This Agreement is effective without regard to the legal nature of the claims raised and without regard to whether any such claims are based upon tort, equity, implied or express contract or discrimination of any sort.

Executive and Company agree that nothing in this Agreement prevents or prohibits Executive from (i) making any disclosure of relevant and necessary information or documents in connection with any charge, action, investigation, or proceeding relating to this Agreement, or as required by law or legal process; (ii) participating, cooperating, or testifying in any charge, action, investigation, or proceeding with, or providing information to, any self-regulatory organization, governmental agency or legislative body, and/or pursuant to the Sarbanes-Oxley Act, without notifying the Company; (iii) filing, testifying, participating in or otherwise assisting in a proceeding relating to an alleged violation of any federal, state or municipal law relating to fraud, or any rule or regulation of the Securities and Exchange Commission or any self-regulatory organization; (iv) challenging the knowing and voluntary nature of the release of ADEA claims pursuant to the OWBPA; or (v) or exercising in any rights Executive may have under Section 7 of the National Labor Relations Act. To the extent permitted by law, upon receipt of any subpoena, court order or other legal process compelling the disclosure of any such information or documents, Executive agrees to give prompt written notice to Company so as to permit Company to protect its interests in confidentiality to the fullest extent possible. To the fullest extent provided by law, Executive acknowledges and agrees, however, Executive is waiving any right to recover monetary damages and other relief in connection with any such charge, action, investigation or proceeding, but retains the right to recover any whistleblower awards. To the extent Executive receives any monetary relief in connection with any such charge, action, investigation or proceeding, Company will be entitled to an offset for the benefits made pursuant to this Agreement, to the fullest extent provided by law. Further, nothing in this Agreement is intended to or shall restrict Executive from discussing or disclosing information about unlawful acts in the workplace, such as harassment or discrimination or any other conduct that Executive has reason to believe is unlawful.

Executive and Company further agree that the Equal Employment Opportunity Commission (EEOC) and comparable state or local agencies have the authority to carry out their statutory duties by investigating charges, issuing determinations, and filing lawsuits in Federal or state court in their own name, or taking any action authorized by the EEOC or comparable state or local agencies. Executive retains the right to file and participate in any such action. Executive retains the right to communicate with the EEOC and comparable state or local agencies and such communication can be initiated by Executive or in response to the government and such right is not limited by any non-disparagement claims. Executive and Company agree that communication with employees plays a critical role in the EEOC's enforcement process because employees inform the agency of employer practices that might violate the law. For this reason, the right to communicate with the EEOC is a right that is protected by federal law and this Agreement does not prohibit or interfere with those rights. Notwithstanding the foregoing, Executive agrees to waive Executive's right to recover monetary damages or other relief in any charge, complaint or lawsuit filed by Executive or by anyone else on Executive's behalf.

3. Executive agrees to continue to comply with the covenants described in Sections 6 – 9 of the Employment Agreement. Further, Executive agrees:

A. Prohibition on Competition. Executive hereby covenants and agrees that, from the Termination Date through the twelve (12) month anniversary of the Termination Date (the "Restricted Period"), Executive will not serve as an officer, director, employee, independent contractor, consultant or agent of, or have any ownership interest in, any business entity which engages in any activities that are materially similar to or competitive with Company's pharmaceutical product development and Commercialization activities in the fields of (i) Niemann-Pick disease type C ("NPC"); (ii) idiopathic hypersomnia ("IH"), narcolepsy or other sleep disorders; (iii) Vascular Ehler-Danlos Syndrome ("VEDS"); and/or (iv) such other indications, approved products or product candidates which Company is actively and demonstrably developing and/or Commercializing at the time Executive's employment is terminated; provided, however, the restriction on Executive providing services as an employee, independent contractor, consultant or agent shall apply only to the extent Executive is providing (i) operational, executive, strategic, consulting, marketing, business or legal services, (ii) Executive is providing services similar to the services Executive provided to Company during Executive's employment with Company, or (ii) Executive could use Confidential Information in the scope of providing such services. If a court of competent jurisdiction finds this non-competition provision invalid or unenforceable due to unreasonableness in time, geographic scope, or scope of Company's business, then Executive agrees that such court shall interpret and enforce this provision to the maximum extent that such court deems reasonable. For purposes of this Agreement, "Commercializing" or "Commercialization" means the sales and marketing phase with regard to a specific drug candidate in a specific country or region following the regulatory approval of said drug candidate in the applicable country or region.

B. Exceptions. Executive's ownership of less than five percent (5%) of the stock of a company that is competitive with the activities of Company as described in Section 8(B) of the Employment Agreement and listed on a national securities exchange shall not be deemed to violate the prohibitions of Section 8(B) of the Employment Agreement.

C. Reasonableness of Restrictions; Remedies. Executive has carefully read and considered the restrictive covenants set forth in this Section and understands Executive's obligations thereunder, the limitations such obligations will impose upon Executive, and that the Restricted Period extends for twelve (12) months after the termination of Executive's employment. Executive has had full opportunity to review with Executive's personal attorney this Agreement, before executing the Agreement. Executive agrees that, as a result of Executive's position with Company, the length of the Restricted Period is (1) fair and reasonable, (2) reasonably required for the protection of the legitimate business interests and goodwill established by Company, and (3) not overly broad or unduly burdensome to Executive. Executive acknowledges that Executive's compliance with Executive's obligations and restrictive covenants set forth in this Agreement is necessary to protect the business and goodwill of Company. Executive agrees that Executive's breach of Executive's obligations and/or restrictive covenants under this Agreement or the Employment Agreement may irreparably and continually damage Company, for which money damages may not be adequate. Consequently, Executive agrees that in the event that Executive breaches or threatens to breach any of the covenants or agreements contained herein, Company shall be entitled to: (a) seek injunctive relief to prevent or halt Executive from breaching this Agreement; and (b) money damages as determined appropriate by a court of competent jurisdiction. Executive hereby agrees that injunctive relief may be granted by a court of competent jurisdiction without the necessity of Company to post bond, or if required to post bond, Executive agrees that the lowest amount permitted shall be adequate. Nothing in this Agreement shall be construed to prohibit Company from pursuing any other remedy available or from seeking to enforce any restrictive covenants to a lesser extent than set forth herein. The Parties agree that all remedies shall be cumulative.

4. Executive agrees that, subject to Section 2 herein, Executive will not disparage or subvert Company or the Releasees, or make any statement reflecting negatively on Company or the Releasees, including, but not limited to, any matters relating to the operation or management of Company, Executive's employment and the termination of Executive's employment, irrespective of the truthfulness or falsity of such statement.

5. In consideration for Executive's agreement as set forth herein, Company agrees to pay and provide Executive with the severance benefits described in Section 4(E)(1) of Executive's Employment Agreement. Executive agrees that Executive is not entitled to any payments, benefits, severance payments or other compensation beyond that expressly provided in Section 4(E)(1) of Executive's Employment Agreement and the Accrued Benefits (as defined in Section 4(B) of

the Employment Agreement).

6. Executive understands and agrees that the payments, benefits and agreements provided in this Agreement are being provided to Executive in consideration for Executive's acceptance and execution of, and in reliance upon Executive's representations in, this Agreement. Executive acknowledges that if Executive had not executed this Agreement containing a release of all claims against Company and the Releasees, Executive would only have been entitled to the payments provided in Company's standard severance pay plan for employees.

7. Executive acknowledges and agrees that Company previously has satisfied any and all obligations owed to Executive under any employment agreement or offer letter Executive has with Company or a Releasee and, further, that this Agreement supersedes any and all prior agreements or understandings, whether written or oral, between the parties, excluding only Executive's and Company's post-termination obligations under Executive's Employment Agreement, Executive's rights under any outstanding equity grants in accordance with the terms of the applicable grant agreements, any obligations relating to the securities of Company or any of its affiliates and Company's obligations under Section 4(E)(1) of Executive's Employment Agreement and to pay or provide the Accrued Benefits (as defined in Section 4(B) of the Employment Agreement), all of which shall remain in full force and effect to the extent not inconsistent with this Agreement, and further, that, except as set forth expressly herein, no promises or representations have been made to Executive in connection with the termination of Executive's Employment Agreement or the terms of this Agreement.

8. Except as may be necessary to obtain approval or authorization to fulfill Executive's or its obligations hereunder or as required by applicable law and subject to the exceptions of Section 2 of the Agreement, (a) Executive agrees not to disclose the terms of this Agreement to anyone, except Executive's spouse, attorney and, as necessary, tax/financial advisor, and (b) Company agrees that the terms of this Agreement will not be disclosed. It is expressly understood that any violation of the confidentiality obligation imposed hereunder constitutes a material breach of this Agreement.

9. Executive represents that Executive does not presently have in Executive's possession any records and business documents, whether on computer or hard copy, and other materials (including but not limited to computer disks and tapes, computer programs and software, office keys, correspondence, files, customer lists, technical information, customer information, pricing information, business strategies and plans, sales records and all copies thereof) (collectively, the "Corporate Records") provided by Company and/or its predecessors, parents, subsidiaries or affiliates or obtained as a result of Executive's employment with Company and/or its predecessors, parents, subsidiaries or affiliates, or created by Executive while employed by or rendering services to Company and/or its predecessors, parents, subsidiaries or affiliates. Executive acknowledges that all such Corporate Records are the property of Company. In addition, Executive shall promptly return in good condition any and all Company owned equipment or property, including, but not limited to, automobiles, personal data assistants, facsimile machines, copy machines, pagers, credit cards, cellular telephone equipment, business cards, laptops and computers. As of the Termination Date, Company will make arrangements to remove, terminate or transfer any and all business communication lines including network access, cellular phone, fax line and other business numbers.

10. Subject to the exceptions of Section 2 of the Agreement, Executive expressly waives all rights afforded by any statute which expressly limits the effect of a release with respect to unknown claims, including Section 1542 of the California Civil Code, which states:

"A GENERAL RELEASE DOES NOT EXTEND TO CLAIMS THAT THE CREDITOR OR RELEASING PARTY DOES NOT KNOW OR SUSPECT TO EXIST IN HIS OR HER FAVOR AT THE TIME OF EXECUTING THE RELEASE AND THAT, IF KNOWN BY HIM OR HER, WOULD HAVE MATERIALLY AFFECTED HIS OR HER SETTLEMENT WITH THE DEBTOR OR RELEASED PARTY."

Executive acknowledges the significance of this release of unknown claims and the waiver of statutory protection against a release of unknown claims which provides that a general release does not extend to claims which the creditor does not know or suspect to exist in Executive's favor at the time of executing the release, which if known by it must have materially affected its settlement with the debtor.

11. The parties agree and acknowledge that the agreements by Company described herein, and the settlement and termination of any asserted or unasserted claims against the Releasees, are not and shall not be construed to be an admission of any violation of any federal, state or local statute or regulation, or of any duty owed by any of the Releasees to Executive.

12. Executive agrees and recognizes that should Executive breach any of the obligations or covenants set forth in this Agreement, Company will have no further obligation to provide Executive with the consideration set forth herein, and will have the right to seek repayment of all consideration paid up to the time of any such breach. Further, Executive acknowledges in the event of a breach of this Agreement, Releasees may seek any and all appropriate relief for any such breach, including equitable relief and/or money damages.

13. This Agreement and the obligations of the parties hereunder shall be construed, interpreted and enforced in accordance with the laws of the State of Florida. This Agreement shall inure to the benefit of the Parties hereto.

14. Executive certifies and acknowledges as follows:

(a) That Executive has read the terms of this Agreement, and that Executive understands its terms and effects, including the fact that Executive has agreed to RELEASE AND FOREVER DISCHARGE Company and each of the Releasees from any legal action arising out of Executive's employment relationship with Company and the termination of that employment relationship;

(b) That Executive has signed this Agreement voluntarily and knowingly in exchange for the consideration described herein, which Executive acknowledges is adequate and satisfactory to Executive and which Executive acknowledges is in addition to any other benefits to which Executive is otherwise entitled;

(c) That Executive has been and is hereby advised in writing to consult with an attorney prior to signing this Agreement;

(d) That Executive does not waive rights or claims that may arise after the date this Agreement is executed;

(e) That Company has provided Executive with a period of **twenty-one (21)** or **forty-five (45)** days within which to consider this Agreement, and that Executive has signed on the date indicated below after concluding that this Separation of Employment Agreement and General Release is satisfactory to Executive; and

[Note: The applicable time period will depend on whether the termination is part of a reduction in force (45 days) or not (21 days). In addition, if the termination is in connection with a reduction in force, certain disclosures will need to be made to Executive to comply with the requirements of the ADEA if Executive is at least age 40.]

(f) Executive acknowledges that this Agreement may be revoked by Executive within seven (7) business days after execution, and it shall not become effective until the expiration of such seven (7) business day revocation period. In the event of a timely revocation by Executive, this Agreement will be deemed null and void and Company will have no obligations hereunder. Revocation may be achieved only by delivering a letter to **[NAME, TITLE, ADDRESS]**, clearly evidencing a decision to revoke within the seven (7) day revocation period.

[Signature Page Follows]

Intending to be legally bound hereby, Executive and Company executed the foregoing Separation of Employment Agreement and General Release this _____ day of _____, ____.

Rahsaan Thompson

Witness: _____

ZEVRA THERAPEUTICS, INC.

By: _____
Name:
Title:

Witness: _____

CERTIFICATION

I, Neil F. McFarlane, certify that:

1. I have reviewed this Quarterly Report on Form 10-Q of Zevra Therapeutics, Inc.;
2. Based on my knowledge, this report does not contain any untrue statement of a material fact or omit to state a material fact necessary to make the statements made, in light of the circumstances under which such statements were made, not misleading with respect to the period covered by this report;
3. Based on my knowledge, the financial statements, and other financial information included in this report, fairly present in all material respects the financial condition, results of operations and cash flows of the registrant as of, and for, the periods presented in this report;
4. The registrant's other certifying officer and I are responsible for establishing and maintaining disclosure controls and procedures (as defined in Exchange Act Rules 13a-15(e) and 15d-15(e)) and internal control over financial reporting (as defined in Exchange Act Rules 13a-15(f) and 15d-15(f)) for the registrant and have:
 - (a) Designed such disclosure controls and procedures, or caused such disclosure controls and procedures to be designed under our supervision, to ensure that material information relating to the registrant, including its consolidated subsidiaries, is made known to us by others within those entities, particularly during the period in which this report is being prepared;
 - (b) Designed such internal control over financial reporting, or caused such internal control over financial reporting to be designed under our supervision, to provide reasonable assurance regarding the reliability of financial reporting and the preparation of financial statements for external purposes in accordance with generally accepted accounting principles;
 - (c) Evaluated the effectiveness of the registrant's disclosure controls and procedures and presented in this report our conclusions about the effectiveness of the disclosure controls and procedures, as of the end of the period covered by this report based on such evaluation; and
 - (d) Disclosed in this report any change in the registrant's internal control over financial reporting that occurred during the registrant's most recent fiscal quarter (the registrant's fourth fiscal quarter in the case of an annual report) that has materially affected, or is reasonably likely to materially affect, the registrant's internal control over financial reporting; and
5. The registrant's other certifying officer and I have disclosed, based on our most recent evaluation of internal control over financial reporting, to the registrant's auditors and the audit committee of the registrant's board of directors (or persons performing the equivalent functions):
 - (a) All significant deficiencies and material weaknesses in the design or operation of internal control over financial reporting which are reasonably likely to adversely affect the registrant's ability to record, process, summarize and report financial information; and
 - (b) Any fraud, whether or not material, that involves management or other employees who have a significant role in the registrant's internal control over financial reporting.

August 13, 2024

/s/ Neil F. McFarlane

Name: Neil F. McFarlane

Title: President and Chief Executive Officer
(Principal Executive Officer)

CERTIFICATION

I, R. LaDuane Clifton, certify that:

1. I have reviewed this Quarterly Report on Form 10-Q of Zevra Therapeutics, Inc.;
2. Based on my knowledge, this report does not contain any untrue statement of a material fact or omit to state a material fact necessary to make the statements made, in light of the circumstances under which such statements were made, not misleading with respect to the period covered by this report;
3. Based on my knowledge, the financial statements, and other financial information included in this report, fairly present in all material respects the financial condition, results of operations and cash flows of the registrant as of, and for, the periods presented in this report;
4. The registrant's other certifying officer and I are responsible for establishing and maintaining disclosure controls and procedures (as defined in Exchange Act Rules 13a-15(e) and 15d-15(e)) and internal control over financial reporting (as defined in Exchange Act Rules 13a-15(f) and 15d-15(f)) for the registrant and have:
 - (a) Designed such disclosure controls and procedures, or caused such disclosure controls and procedures to be designed under our supervision, to ensure that material information relating to the registrant, including its consolidated subsidiaries, is made known to us by others within those entities, particularly during the period in which this report is being prepared;
 - (b) Designed such internal control over financial reporting, or caused such internal control over financial reporting to be designed under our supervision, to provide reasonable assurance regarding the reliability of financial reporting and the preparation of financial statements for external purposes in accordance with generally accepted accounting principles;
 - (c) Evaluated the effectiveness of the registrant's disclosure controls and procedures and presented in this report our conclusions about the effectiveness of the disclosure controls and procedures, as of the end of the period covered by this report based on such evaluation; and
 - (d) Disclosed in this report any change in the registrant's internal control over financial reporting that occurred during the registrant's most recent fiscal quarter (the registrant's fourth fiscal quarter in the case of an annual report) that has materially affected, or is reasonably likely to materially affect, the registrant's internal control over financial reporting; and
5. The registrant's other certifying officer and I have disclosed, based on our most recent evaluation of internal control over financial reporting, to the registrant's auditors and the audit committee of the registrant's board of directors (or persons performing the equivalent functions):
 - (a) All significant deficiencies and material weaknesses in the design or operation of internal control over financial reporting which are reasonably likely to adversely affect the registrant's ability to record, process, summarize and report financial information; and
 - (b) Any fraud, whether or not material, that involves management or other employees who have a significant role in the registrant's internal control over financial reporting.

August 13, 2024

/s/ R. LaDuane Clifton

Name: R. LaDuane Clifton, MBA, CPA

Title: Chief Financial Officer and Treasurer
(Principal Financial Officer)

**CERTIFICATION OF THE PRINCIPAL EXECUTIVE OFFICER
PURSUANT TO 18 U.S.C. SECTION 1350, AS ADOPTED PURSUANT TO
SECTION 906 OF THE SARBANES-OXLEY ACT OF 2002**

In connection with the Quarterly Report on Form 10-Q of Zevra Therapeutics, Inc., (the "Company") for the quarterly period ended June 30, 2024, as filed with the Securities and Exchange Commission on the date hereof (the "Report"), I, Neil F. McFarlane, Principal Executive Officer of the Company, certify, pursuant to 18 U.S.C. Section 1350, as adopted pursuant to Section 906 of the Sarbanes-Oxley Act of 2002, that, to my knowledge:

1. The Report fully complies with the requirements of Section 13(a) or 15(d) of the Securities Exchange Act of 1934; and
2. The information contained in the Report fairly presents, in all material respects, the financial condition and results of operations of the Company.

August 13, 2024

/s/ Neil F. McFarlane

Name: Neil F. McFarlane

Title: President and Chief Executive Officer
(Principal Executive Officer)

The foregoing certification is being furnished solely pursuant to 18 U.S.C. Section 1350, is not being "filed" by the Company as part of the Report or as a separate disclosure document and is not to be incorporated by reference into any filing of the Company under the Securities Act of 1933, as amended, or the Securities Exchange Act of 1934, as amended (whether made before or after the date of the Report), irrespective of any general incorporation language contained in such filing.

**CERTIFICATION OF THE PRINCIPAL FINANCIAL OFFICER
PURSUANT TO 18 U.S.C. SECTION 1350, AS ADOPTED PURSUANT TO
SECTION 906 OF THE SARBANES-OXLEY ACT OF 2002**

In connection with the Quarterly Report on Form 10-Q of Zevra Therapeutics, Inc., (the "Company") for the quarterly period ended June 30, 2024, as filed with the Securities and Exchange Commission on the date hereof (the "Report"), I, R. LaDuane Clifton, Principal Financial Officer of the Company, certify, pursuant to 18 U.S.C. Section 1350, as adopted pursuant to Section 906 of the Sarbanes-Oxley Act of 2002, that, to my knowledge:

1. The Report fully complies with the requirements of Section 13(a) or 15(d) of the Securities Exchange Act of 1934; and
2. The information contained in the Report fairly presents, in all material respects, the financial condition and results of operations of the Company.

August 13, 2024

/s/ R. LaDuane Clifton

Name: R. LaDuane Clifton, MBA, CPA

Title: Chief Financial Officer and Treasurer
(Principal Financial Officer)

The foregoing certification is being furnished solely pursuant to 18 U.S.C. Section 1350, is not being "filed" by the Company as part of the Report or as a separate disclosure document and is not to be incorporated by reference into any filing of the Company under the Securities Act of 1933, as amended, or the Securities Exchange Act of 1934, as amended (whether made before or after the date of the Report), irrespective of any general incorporation language contained in such filing.