

Repare Therapeutics Provides Business and Clinical Update and Reports First Quarter 2024 Financial Results

May 7, 2024

Phase 1 MYTHIC clinical trial of lunresertib in combination with camonsertib demonstrated a significant reduction in Grade 3 anemia and continued trends of patient response and benefit; FDA agrees with RP2D

First patient dosed in Phase 1 MYTHIC clinical trial of lunresertib in combination with the WEE1 inhibitor, Debio 0123; first clinical trial inhibiting both PKMYT1 and WEE1

Initiating Phase 2 TRESR expansion in ~20 patients evaluating monotherapy camonsertib in NSCLC; initial data expected in 2025

First patient dosed in Phase 1 LIONS monotherapy trial for PLK4 inhibitor RP-1664

Announced the appointment of Steven H. Stein, M.D. to Repare's Board of Directors, effective in June 2024

\$237.0 million in cash and cash equivalents and marketable securities to advance clinical programs and portfolio to mid-2026

CAMBRIDGE, Mass. & MONTREAL--(BUSINESS WIRE)--May 7, 2024-- Repare Therapeutics Inc. ("Repare" or the "Company") (Nasdaq: RPTX), a leading clinical-stage precision oncology company, today reported financial results for the first quarter ended March 31, 2024.

"This was a quarter of clinical progress as we await key, near-term data on a rich set of distinctive clinical approaches for our four wholly-owned compounds in 2024," said Lloyd M. Segal, President and Chief Executive Officer of Repare. "We have agreement with the FDA regarding our recommended Phase 2 dose (RP2D) for our lunresertib plus camonsertib combination, with significantly improved tolerability at the RP2D with our updated dosing schedule. We are seeing continuing trends of patient response and benefit, and we are on track to report the updated dataset in the fourth quarter of 2024. Our objective is to determine the best opportunity for a registrational trial, to start in 2025. Additionally, we are initiating a small clinical trial to rapidly confirm a camonsertib monotherapy signal in non-small cell lung cancer (NSCLC) and expect that readout to be available in 2025. Our clinical portfolio also includes the LIONS trial of our RP-1664 PLK4 inhibitor, the PKMYT1 and WEE1 inhibitor combination in MYTHIC, and the upcoming clinical start of our Polθ inhibitor program, RP-3467, in the second half of 2024."

First Quarter 2024 and Recent Portfolio Highlights:

• Lunresertib (RP-6306)

- o On track for a potential registrational trial decision in gynecologic expansion cohorts in the fourth quarter of 2024 based on the Phase 1 expansion in MYTHIC trial evaluating lunresertib in combination with camonsertib in patients harboring CCNE1 amplification or FBXW7 or PPP2R1A deleterious alterations. Grade 3 anemia has been significantly reduced to 25% as of the March 2024 cut-off date in patients treated at the RP2D and updated dosing schedule, from 45% as previously presented at the September 2023 data cut-off date. The U.S. Food and Drug Administration (FDA) has agreed with the RP2D of lunresertib 80mg BID and camonsertib 80mg QD. Efficacy and tolerability assessment at RP2D is ongoing, and the Company expects to present data from the dose expansion cohorts in patients with ovarian and endometrial cancer in the fourth quarter of 2024.
- First patient was dosed in April 2024 in the Phase 1 MYTHIC clinical trial evaluating lunresertib in combination with Debio 0123, a highly selective, brain-penetrant, clinical WEE1 inhibitor, in advanced solid tumors harboring CCNE1 amplification or FBXW7 or PPP2R1a deleterious alterations. The primary endpoints are safety, tolerability and RP2D, as well as preliminary efficacy of the combination. Repare is expected to report initial data from this trial in 2025.
- o Initial data from the Phase 1 MINOTAUR trial evaluating lunresertib in combination with FOLFIRI for the treatment of advanced solid tumors demonstrated no significant incremental toxicities in the combination of lunresertib and FOLFIRI over FOLFIRI alone. In addition, Repare has observed favorable tolerability in colorectal and other gastrointestinal tumors, unlike some other agents combined with irinotecan. This data will be presented at the European Society of Medical Oncology (ESMO) Gastrointestinal (GI) Cancers Congress 2024, taking place in Munich, Germany on June 26-29.

Camonsertib (RP-3500)

- Regained global development and commercialization rights for camonsertib from Roche, effective May 7, 2024.
 Since inception of the Roche camonsertib collaboration, Repare has earned a cumulative total of \$182.6 million from Roche, including the upfront and milestone payments, in addition to certain additional reimbursements from Roche.
- Initiating Phase 2 TRESR expansion in approximately 20 patients with ATM-mutated (ATMm) NSCLC, supported by early, promising camonsertib monotherapy signal in patients with ATMm NSCLC from the ongoing Phase 1/2 TRESR trial. Repare is expected to report initial data in 2025.

 First patient dosed in the multicenter, open-label Phase 1 dose escalation trial (LIONS) of its polo-like kinase 4 (PLK4) inhibitor, RP-1664, in adult and adolescent patients with TRIM37-high and other biomarkers in February 2024.

• RP-3467

o Initiation of a Phase 1 dose finding trial of RP-3467, a potential best-in-class Polθ ATPase inhibitor, is expected in the second half of 2024.

· Other Highlights

- In March 2024, Bristol-Myers Squibb exercised its one remaining option to in-license an undruggable target for a combined total of five druggable targets and one undruggable target over the course of the collaboration.
- o In April 2024, Repare announced the appointment of Steven H. Stein, M.D., Chief Medical Officer of Incyte Corporation, to Repare's Board of Directors, effective as of June 17, 2024, the date of the Company's upcoming annual meeting of shareholders (the "Annual Meeting"). The Company also announced that Todd Foley has decided not to stand for re-election as a director of the Company following the end of his current term as a Class I director on the date of the Annual Meeting, after serving more than seven years on the Board.

Summary of Expected Milestones:

• H1 2024

o Initial Phase 1 MINOTAUR (lunresertib + FOLFIRI combination) data to be reported at ESMO GI in June 2024

• H2 2024

- Camonsertib monotherapy expansion to NSCLC in TRESR
- o Initiation of Phase 1 clinical trial of RP-3467
- Additional data from dose expansion cohorts for the MYTHIC lunresertib + camonsertib combination in ovarian and endometrial cancers by end of Q4 2024

2025

- o Lunresertib + Debio 0123 combination data
- o Camonsertib monotherapy data in NSCLC
- o Initiate first pivotal trial in an indication for lunresertib + camonsertib

First Quarter 2024 Financial Results:

- Cash, cash equivalents and marketable securities: Cash, cash equivalents and marketable securities as of March 31, 2024 were \$237.0 million, as compared to \$223.6 million as of December 31, 2023. The Company believes that its cash, cash equivalents, and marketable securities are sufficient to fund its current operational plans at least into mid-2026.
- Revenue from collaboration agreements: Revenue from collaboration agreements was \$52.4 million and \$5.7 million for the three months ended March 31, 2024 and 2023, respectively. The increase in revenue for the three-month period was primarily due to the \$40.0 million Roche milestone achievement in the first guarter of 2024.
- Research and development expenses, net of tax credits (Net R&D): Net R&D expenses were \$33.0 million and \$31.8 million for the three months ended March 31, 2024 and 2023, respectively. The increase in Net R&D for the three-month period was primarily due to higher direct external costs related to the progress of Repare's lunresertib clinical program, offset by lower direct external costs of its camonsertib clinical program.
- **General and administrative (G&A) expenses:** G&A expenses were \$8.6 million and \$8.5 million for the three months ended March 31, 2024 and 2023, respectively.
- Net income (loss): Net income was \$13.2 million, or \$0.30 per diluted share, for the three months ended March 31, 2024, and net loss was \$34.9 million, or \$0.83 per diluted share, for the three months ended March 31, 2023.

About Repare Therapeutics Inc.

Repare Therapeutics is a leading clinical-stage precision oncology company enabled by its proprietary synthetic lethality approach to the discovery and development of novel therapeutics. The Company utilizes its genome-wide, CRISPR-enabled SNIPRx® platform to systematically discover and develop highly targeted cancer therapies focused on genomic instability, including DNA damage repair. The Company's pipeline includes lunresertib (also known as RP-6306), a PKMYT1 inhibitor currently in Phase 1/2 clinical development; camonsertib (also known as RP-3500), a potential leading ATR inhibitor currently in Phase 1/2 clinical development; RP-1664, a Phase 1 PLK4 inhibitor; RP-3467, a preclinical Polθ ATPase inhibitor program; as well as additional, undisclosed preclinical programs. For more information, please visit www.reparerx.com and follow @Reparerx on X (formerly Twitter) and LinkedIn.

 $\ensuremath{\mathsf{SNIPRx}}\xspace$ is a registered trademark of Repare Therapeutics Inc.

Forward-Looking Statements

This press release contains "forward-looking statements" within the meaning of the Private Securities Litigation Reform Act of 1995 and securities laws in Canada. All statements in this press release other than statements of historical facts are "forward-looking statements. These statements may be identified by words such as "aims," "anticipates," "believes," "could," "estimates," "expects," "forecasts," "goal," "intends," "may," "plans," "possible," "potential," "seeks," "will" and variations of these words or similar expressions that are intended to identify forward-looking statements, although not all forward-looking statements contain these words. Forward-looking statements in this press release include, but are not limited to, statements

regarding: the design, objectives, initiation, timing, progress and results of current and future preclinical studies and clinical trials of the Company's product candidates, including the expansion of its Phase 1 MYTHIC trial evaluating lunresertib alone and in combination with camonsertib, its Phase 1 MINOTAUR trial evaluating lunresertib in combination with FOLFIRI, the Module of the Company's Phase 1/1b MYTHIC trial of, its Phase 1/1b trial of Debio 0123 and lunresertib in partnership with Debiopharm, the expansion of its Phase 2 TRESR trial of camonsertib in patients with ATMm, its Phase 1 LIONS trial of RP-1664, its Phase 1 trial of RP-3467; its planned expansion of development of lunresertib plus camonsertib combination; a potential registrational trial in 2025; the tolerability, efficacy and clinical progress of camonsertib, lunresertib, RP-1664 and RP-3467; the potential of RP-3467 as a best-in-class Pol0 ATPase inhibitor; the potential synergies of Debio 0123 in combination with lunresertib, lunresertib in combination with camonsertib and lunresertib in combination with FOLFIRI; the Company's anticipated cash runway; and the benefits and ability to discover further targets and clinical candidates from the Company's discovery platform. These forward-looking statements are based on the Company's expectations and assumptions as of the date of this press release. Each of these forward-looking statements involves risks and uncertainties that could cause the Company's clinical development programs, future results or performance to differ materially from those expressed or implied by the forward-looking statements. Many factors may cause differences between current expectations and actual results, including: the potential that success in preclinical testing and earlier clinical trials does not ensure that later clinical trials will generate the same results or otherwise provide adequate data to demonstrate the efficacy and safety of a product candidate; the impacts of macroeconomic conditions, including the conflict in Ukraine and the conflict in the Middle East, heightened inflation and uncertain credit and financial markets, on the Company's business, clinical trials and financial position; unexpected safety or efficacy data observed during preclinical studies or clinical trials; clinical trial site activation or enrollment rates that are lower than expected; the Company's ability to realize the benefits of its collaboration and license agreements; changes in expected or existing competition; changes in the regulatory environment; the uncertainties and timing of the regulatory approval process; and unexpected litigation or other disputes. Other factors that may cause the Company's actual results to differ from those expressed or implied in the forward-looking statements in this press release are identified in the section titled "Risk Factors" in the Company's Quarterly Report on Form 10-Q for the quarter ended March 31, 2024 filed with the Securities and Exchange Commission ("SEC") and the Québec Autorité des Marchés Financiers ("AMF") on May 7, 2024. The Company expressly disclaims any obligation to update any forward-looking statements contained herein, whether as a result of any new information, future events, changed circumstances or otherwise, except as otherwise required by law. For more information, please visit reparerx.com and follow Repare on Twitter at @RepareRx and on LinkedIn at https://www.linkedin.com/company/repare-therapeutics/,

Repare Therapeutics Inc.

Consolidated Balance Sheets

(Unaudited)

(Amounts in thousands of U.S. dollars, except share data)

	As of March 31,	As of December 31,
	2024	2023
ASSETS		
CURRENT ASSETS:		
Cash and cash equivalents	\$103,217	\$ 111,268
Marketable securities	133,784	112,359
Income tax receivable	10,829	10,813
Other current receivables	3,377	4,499
Prepaid expenses	3,463	4,749
Total current assets	254,670	243,688
Property and equipment, net	3,714	4,215
Operating lease right-of-use assets	2,763	3,326
Income tax receivable	1,630	2,276
Other assets	307	396

TOTAL ASSETS		\$263,084	\$ 253,901
LIABILITIES AND SHAREHOLDERS' EQUITY			
CURRENT LIABILITIES:			
Accounts payable		\$6,825	\$ 2,400
Accrued expenses and other current liabilities		20,454	24,057
Operating lease liability, current portion		2,218	2,400
Deferred revenue, current portion		1,073	10,222
Total current liabilities		30,570	39,079
Operating lease liability, net of current portion		561	1,010
Deferred revenue, net of current portion		_	1,730
TOTAL LIABILITIES		31,131	41,819
SHAREHOLDERS' EQUITY			
Preferred shares, no par value per share; unlimited shares authorized a respectively; 0 shares issued and outstanding as of March 31, 2024, and		_	_
Common shares, no par value per share; unlimited shares authorized as 42,445,406 and 42,176,041 shares issued and outstanding as of March		486,375	483,350
Additional paid-in capital		65,638	61,813
Accumulated other comprehensive (loss) income		(113)	28
Accumulated deficit		(319,947)	(333,109)
Total shareholders' equity		231,953	212,082
TOTAL LIABILITIES AND SHAREHOLDERS' EQUITY		\$263,084	\$ 253,901
Repare Therapeutics Inc.			
Consolidated Statements of Operations and Comprehensive Loss			
(Unaudited)			
(Amounts in thousands of U.S. dollars, except share and per share	data)		
	Three Months Ended March 31,		

Collaboration agreements

Revenue:

2024

2023

Operating expenses:

Research and development, net of tax credits	32,970		31,830	
General and administrative	8,618		8,529	
Total operating expenses	41,588		40,359	
Income (loss) from operations	10,816		(34,681)
Other income (expense), net				
Realized and unrealized gain (loss) on foreign exchange	31		(56)
Interest income	2,968		3,427	
Other expense	(24)	(15)
Total other income, net	2,975		3,356	
Income (loss) before income taxes	13,791		(31,325)
Income tax expense	(629)	(3,616)
Net income (loss)	\$13,162		\$ (34,941)
Net income (loss) Other comprehensive (loss) income:	\$13,162		\$ (34,941)
	\$13,162 \$(141		\$ (34,941 \$ 193)
Other comprehensive (loss) income:)
Other comprehensive (loss) income: Unrealized (loss) gain on available-for-sale marketable securities	\$ (141)	\$ 193)
Other comprehensive (loss) income: Unrealized (loss) gain on available-for-sale marketable securities Total other comprehensive (loss) income	\$ (141 (141 \$ 13,021)	\$ 193 193	
Other comprehensive (loss) income: Unrealized (loss) gain on available-for-sale marketable securities Total other comprehensive (loss) income Comprehensive income (loss)	\$ (141 (141 \$ 13,021)	\$ 193 193	
Other comprehensive (loss) income: Unrealized (loss) gain on available-for-sale marketable securities Total other comprehensive (loss) income Comprehensive income (loss) Net income (loss) per share attributable to common shareholders:	\$ (141 (141 \$ 13,021)	\$ 193 193 \$ (34,748)
Other comprehensive (loss) income: Unrealized (loss) gain on available-for-sale marketable securities Total other comprehensive (loss) income Comprehensive income (loss) Net income (loss) per share attributable to common shareholders: Basic	\$ (141 (141 \$ 13,021 \$ 0.31)	\$ 193 193 \$ (34,748 \$ (0.83)
Other comprehensive (loss) income: Unrealized (loss) gain on available-for-sale marketable securities Total other comprehensive (loss) income Comprehensive income (loss) Net income (loss) per share attributable to common shareholders: Basic Diluted	\$ (141 (141 \$ 13,021 \$ 0.31)	\$ 193 193 \$ (34,748 \$ (0.83)

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