Repare Therapeutics Provides Business Update and Reports First Quarter 2023 Financial Results

May 9, 2023

Reported initial camonsertib data from ongoing Phase 1/2 TRESR and Phase 1b/2 ATTACC clinical trials in a plenary oral presentation at 2023 AACR Annual Meeting

Camonsertib combination therapy showed 48% CBR across tumor types regardless of choice of PARP inhibitor or platinum resistance, with a favorable safety and tolerability profile

Company on track to report initial RP-6306 monotherapy data from Phase 1 MYTHIC clinical trial in June 2023

Announced the appointment of Susan Molineaux, Ph.D., and departure of Jerel Davis, Ph.D. on our Board of Directors, as well as the addition of Daniel Belanger to the senior leadership team

CAMBRIDGE, Mass. & MONTREAL--(BUSINESS WIRE)--May 9, 2023-- 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, 2023.

“We continue to execute clinically and across our pipeline programs, including presenting initial clinical data of camonsertib in combination with various PARP inhibitors from the ongoing TRESR and ATTACC trials at this year’s AACR conference,” said Lloyd M. Segal, President and Chief Executive Officer of Repare. “These findings are consistent with our preclinical data demonstrating that low, intermittent dosing of the camonsertib and PARP inhibitor combinations appears well tolerated and exhibited antitumor activity, most notably in advanced ovarian cancer. In addition, we are on track to present initial Phase 1 monotherapy data for RP-6306 in June.”

First Quarter 2023 Review and Operational Updates:

- **Announced initial clinical data from the Phase 1/2 TRESR and ATTACC trials evaluating camonsertib (RP-3500/RG6526, partnered with Roche), a potent and selective oral small molecule inhibitor of ATR (Ataxia-Telangiectasia and Rad3-related protein kinase), in combination with three poly (ADP-ribose) polymerase (PARP) inhibitors in a Clinical Trials Plenary Session at the 2023 American Association for Cancer Research (AACR) Annual Meeting.**
    - Camonsertib combinations appear to be well tolerated. Dose limiting toxicity in 68 patients treated with the proposed combination doses were related to myelotoxicity (Grade 3+ anemia 3%, thrombocytopenia 6%, neutropenia 7%, and febrile neutropenia 3%). No prophylactic growth factors were required when administering the PARP inhibitors at evaluated doses.
    - Camonsertib combination resulted in durable clinical benefit across tumor types and genomic alterations, regardless of choice of PARP inhibitor and presence of platinum resistance. Overall clinical benefit rate (CBR) for all patients was 48%. Patients with platinum-resistant tumors had an overall response rate (ORR) of 12% and CBR of 49% and benefited similarly to non-platinum-resistant tumors (ORR 13%, CBR 46%).
    - Compelling results were observed particularly in patients with advanced ovarian cancer (n = 19). In these patients, overall response was 32%, CBR was 58% and median progression-free survival was approximately 7 months with treatment greater than 16 weeks and ongoing in 9 patients.
    - Early circulating tumor DNA molecular responses in 66% (31/47) of evaluable patients confirm antitumor activity of low dose, intermittent PARP inhibitor + ATR inhibitor therapy. The molecular response rate (MRR) was significantly higher in patients with clinical benefit (83%) compared to those without (48%; p=0.015) and significantly higher than camonsertib monotherapy that was also administered at higher doses (43% or 27/63; p=0.02). Molecular responses were also observed in patients with prior PARP inhibitor exposure (57%) and platinum resistance (64%).
    - Repare is conducting dose optimization and efficacy assessments in tumor specific expansions in the ATTACC study in collaboration with Roche to support future clinical development plans for camonsertib combinations with PARP inhibitors.

- **Evaluating RP-6306, a first-in-class, oral PKMYT1 inhibitor as a monotherapy and in combinations in multiple early clinical studies.**
    - Repare presented two poster presentations for RP-6306 at the 2023 AACR Annual Meeting regarding the co-mutation landscape in CCNE1 amplifications and the tumor heterogeneity of copy number in ovarian and uterine cancers. Additionally, several collaborators presented preclinical findings on the potential benefits of combining a Wee1 inhibitor with RP-6306 and the effect of RP-6306 in triple negative breast cancer.
    - Repare expects to report initial Phase 1 monotherapy clinical data for RP-6306 for the treatment of molecularly selected advanced solid tumors (MYTHIC) in June 2023. The Company expects to report initial Phase 1 combination therapy clinical data for RP-6306 for the treatment of molecularly selected advanced solid tumors in the fourth quarter of the year.

- Repare is working with clinical investigators to initiate clinical testing, as part of an investigator-sponsored trial (IST), of a fourth RP-6306 combination with carboplatin, with first patient dosing expected this year.
- Repare is collaborating with the Canadian Cancer Trials Group for a basket Phase 2 IST to evaluate RP-6306 in patients with selected, advanced cancers receiving standard agents that is expected to begin this year. A sub-study under the master clinical trial protocol will evaluate RP-6306 in combination with gemcitabine in patients with CD4/6i-resistant ER+/HER2- metastatic breast cancer.

- **Advancing preclinical programs into clinical development.**
  - Repare initiated IND-enabling studies in the first half of this year for a small molecule, now designated RP-1664, against an undisclosed target with potential to enter the clinic in late 2023 or early 2024.
  - Repare is also pursuing development of an inhibitor of polymerase theta (Polθ) that is expected to enter the clinic in 2024.

- **In April 2023, Repare announced the appointment of Susan Molineaux, Ph.D., to its Board of Directors, effective as of the date of the Company's upcoming annual meeting of shareholders in June 2023. Concurrent with Dr. Molineaux's appointment as of the date of the annual meeting, Jerel Davis, Ph.D., Managing Director at Versant Ventures and a founding member of Repare's Board of Directors, will step down from the Board. Additionally, Repare has expanded the senior leadership team with the appointment of Daniel Belanger as EVP Human Resources in May 2023.**

First Quarter 2023 Financial Results:

- **Cash and cash equivalents and marketable securities:** Cash and cash equivalents and marketable securities as of March 31, 2023 were $314.1 million.
- **Revenue from collaboration agreements:** Revenue from collaboration agreements were $5.7 million and $0.4 million for the three months ended March 31, 2023 and 2022, respectively. The year-over-year increase in revenue was due to revenue recognized from our collaboration and license agreement with Roche.
- **Research and development expenses, net of tax credits (Net R&D):** Net R&D expenses were $31.8 million and $26.5 million for the three months ended March 31, 2023 and 2022, respectively. The year-over-year increase in Net R&D expenses was primarily due to higher personnel related costs from headcount in support of our development activities, and direct external costs related to the advancement of preclinical programs into IND-enabling studies.
- **General and administrative (G&A) expenses:** G&A expenses were $8.6 million and $8.8 million for three months ended March 31, 2023 and 2022, respectively. The year-over-year decrease in G&A was primarily due to lower professional fees associated with our collaboration and license agreement with Roche and lower D&O insurance premiums, offset by higher personnel related costs.
- **Net loss:** Net loss was $34.9 million, or $0.83 per share, in the three months ended March 31, 2023, and $34.8 million, or $0.83 per share in the three months ended March 31, 2022.

About Repare Therapeutics' SNIPRx® Platform

Repare's SNIPRx® platform is a genome-wide CRISPR-based screening approach that utilizes proprietary isogenic cell lines to identify novel and known synthetic lethal gene pairs and the corresponding patients who are most likely to benefit from the Company's therapies based on the genetic profile of their tumors. Repare's platform enables the development of precision therapeutics in patients whose tumors contain one or more genomic alterations identified by SNIPRx® screening, in order to selectively target those tumors in patients most likely to achieve clinical benefit from resulting product candidates.

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 RP-6306, a PKMYT1 inhibitor currently in Phase 1 clinical development; camonsertib (also known as RP-3500 or RG6526), a potential leading ATR inhibitor currently in Phase 1/2 clinical development and partnered with Roche; a preclinical Polθ inhibitor program; as well as several undisclosed preclinical programs, including RP-1664. For more information, please visit reparerx.com.

SNIPRx® 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 safety, efficacy and clinical progress of the Company's clinical programs, including RP-6306 and camonsertib; the clinical and preclinical development of the Company's pipeline and its research and development programs, including the anticipated timing, anticipated patient enrollment, trial outcomes or associated costs of its clinical trials of RP-6306 and camonsertib and ongoing preclinical studies and future clinical trials of the Company's RP-1664 and Polθ inhibitor programs; the Company's continued development of camonsertib in partnership with Roche; the status of clinical trials (including, without limitation, expectations regarding the data that is being presented, the expected timing of data releases and development, as well as the completion of clinical trials) and development timelines for the Company's product candidates; ; and the expected benefits of the Company's collaborations and partnerships. 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 evolving impact of macroeconomic events, including the COVID-19 pandemic, rising inflation, the U.S. Federal Reserve raising interest rates, recent disruptions in access to bank deposits or lending commitments due to bank failures and the Russia-Ukraine war, 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; 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 Annual Report on Form 10-K for the year ended December 31, 2022 filed with the Securities and Exchange Commission ("SEC") and the Québec Autorité des Marchés Financiers ("AMF") on February 28, 2023, and its other documents subsequently filed with or furnished to the SEC and AMF. 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, 2023	As of December 31, 2022
ASSETS		
CURRENT ASSETS:		
Cash and cash equivalents	$ 121,461	$ 159,521
Marketable securities	192,663	184,420
Research and development tax credits receivable	1,659	1,280
Collaboration revenue receivable	3,996	1,525
Other receivables	1,358	1,518
Prepaid expenses	4,389	5,715
Total current assets	325,526	353,979
Property and equipment, net	5,396	4,228
Operating lease right-of-use assets	4,976	5,371
Other assets	408	497

TOTAL ASSETS	$ 336,306	$ 364,075
LIABILITIES AND SHAREHOLDERS' EQUITY		
CURRENT LIABILITIES:		
Accounts payable	$ 3,620	$ 461
Accrued expenses and other current liabilities	17,442	21,645
Operating lease liability, current portion	2,257	2,171
Deferred revenue, current portion	52,760	53,102
Income tax payable	4,856	1,240
Total current liabilities	80,935	78,619
Operating lease liability, net of current portion	2,780	3,257
Deferred revenue, net of current portion	1,347	2,682
TOTAL LIABILITIES	85,062	84,558
SHAREHOLDERS' EQUITY		
Preferred shares, no par value per share; unlimited shares authorized as of March 31, 2023 and December 31, 2022, respectively; 0 shares issued and outstanding as of March 31, 2023, and December 31, 2022, respectively	—	—
Common shares, no par value per share; unlimited shares authorized as of March 31, 2023 and December 31, 2022; 42,079,896 and 42,036,193 shares issued and outstanding as of March 31, 2023 and December 31, 2022, respectively	482,677	482,032
Additional paid-in capital	43,056	37,226
Accumulated other comprehensive loss	(235)	(428)
Accumulated deficit	(274,254)	(239,313)
Total shareholders' equity	251,244	279,517
TOTAL LIABILITIES AND SHAREHOLDERS' EQUITY	$ 336,306	$ 364,075

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,

	2023	2022
Revenue:		
Collaboration agreements	$ 5,678	$ 408
Operating expenses:		
Research and development, net of tax credits	31,830	26,458
General and administrative	8,529	8,779
Total operating expenses	40,359	35,237
Loss from operations	(34,681)	(34,829)
Other income (expense), net		
Realized and unrealized loss on foreign exchange	(56)	(17)
Interest income	3,427	129
Other expense	(15)	(8)
Total other income, net	3,356	104
Loss before income taxes	(31,325)	(34,725)
Income tax expense	(3,616)	(32)
Net loss	$ (34,941)	$ (34,757)
Other comprehensive gain:		
Unrealized gain on available-for-sale marketable securities	$ 193	$ —
Total other comprehensive gain	193	—
Comprehensive loss	$ (34,748)	$ (34,757)
Net loss per share attributable to common shareholders - basic and diluted	$ (0.83)	$ (0.83)
Weighted-average common shares outstanding - basic and diluted	42,040,674	41,861,613

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Repare Contact:
Robin Garner
Executive Director and Head of Investor Relations
Repare Therapeutics Inc.
investor@reparerx.com

Investors:

Matthew DeYoung
Argot Partners
repare@argotpartners.com

Media:
David Rosen
Argot Partners
david.rosen@argotpartners.com
212-600-1902

Source: Repare Therapeutics Inc.