Clementia Announces Plan to Submit a New Drug Application for Palovarotene for the Treatment of FOP Based on Positive Phase 2 Results

Updated Regulatory Strategy Based on 12-Week Flare-up Data from Completed Phase 2 Program and Consultation with the FDA

NDA Submission Planned for the Second Half of 2019

MONTREAL, Oct. 23, 2018 – Clementia Pharmaceuticals Inc. (Nasdaq: CMTA), a clinical-stage biopharmaceutical company innovating treatments for people with ultra-rare bone disorders and other diseases, today announced that it plans to submit a New Drug Application (NDA) for palovarotene to the U.S. Food and Drug Administration (FDA) in the second half of 2019. Based on recent interactions between Clementia and the FDA, including a Type B meeting held earlier today as part of palovarotene's Breakthrough Therapy Designation, the FDA has agreed that available data would support filing of an NDA for palovarotene for the prevention of heterotopic ossification (HO) associated with flare up symptoms in patients with fibrodysplasia ossificans progressiva (FOP). The FDA based its assessment on the efficacy and safety data available from the completed Phase 2 clinical program, which showed a statistically significant reduction in mean new HO volume, or bone volume, associated with flare-ups of FOP at 12 weeks as compared to placebo or untreated flare-ups.

The data provided to the FDA included the results from Clementia's Phase 2 studies, which were recently presented at ASBMR 2018. In these studies, 92 palovarotene-treated flare-ups in 62 patients across three different dosing regimens were evaluated compared to 46 placebo or untreated flare-ups in 41 patients from the Company's natural history study. Patients treated with palovarotene at the time of a flare-up demonstrated a greater than 70 percent reduction in mean new HO volume at 12 weeks compared to the untreated group. Further, the reduction in mean new HO volume among those treated with the episodic 20/10 mg dosing regimen (20 mg for 4 weeks starting at the time of a flare-up followed by 10 mg for 8 weeks) was statistically significant ($p=0.02$). Palovarotene was generally well tolerated across all dosing regimens of the Phase 2 clinical program. There were dose-related increases in retinoid-associated adverse events (AEs) with most being mild or moderate in severity, and only one patient discontinued participation in the study because of an AE.

Following the completion of standard non-clinical, clinical pharmacology and CMC studies to be agreed upon with the FDA, Clementia plans to submit its NDA in the second half of 2019 to seek approval of the palovarotene 20/10 mg episodic dosing regimen.

"The identification of a path to an NDA submission in the second half of 2019 is a significant milestone for Clementia and for patients with this ultra-rare and devastating genetic bone disease," said Clarissa Desjardins, Ph.D., founder and chief executive officer of Clementia. "We are thankful for the collaboration with the FDA's Division of Bone, Reproductive and Urologic Products, potentially bringing the first approved treatment option to individuals affected by FOP. We are also grateful to the patients and their families, as well as the investigators and clinical sites, without whom none of this work would have been possible."

Clementia's ongoing Phase 3 MOVE Trial will continue as planned, evaluating a dosing regimen of palovarotene which includes a chronic 5 mg daily dose in addition to the episodic 20/10 mg dosing regimen at the time of a flare-up. The Company believes that, if successful, the data from the MOVE Trial may provide the basis for a supplemental NDA for an additional treatment regimen option for patients with FOP.

Clementia also plans to engage international regulatory authorities in 2019 to discuss the registration strategy for palovarotene and anticipates providing an update at a later stage.

About Palovarotene
Palovarotene is an RARγ agonist being developed as a treatment for patients with ultra-rare and debilitating bone diseases, including fibrodysplasia ossificans progressiva (FOP) and multiple osteochondromas (MO), as well as other diseases. Palovarotene was in-licensed from Roche Pharmaceuticals, where it was previously evaluated in more than 800 subjects, including 450 patients treated for up to two years. Palovarotene has received Orphan Drug status for FOP and MO from the U.S. Food and Drug Administration (FDA) and the European Medicines Agency (EMA). In addition, palovarotene has been granted Fast Track and Breakthrough Therapy designations for FOP from the FDA.

About Fibrodysplasia Ossificans Progressiva (FOP)
FOP is a rare, severely disabling disorder characterized by heterotopic ossification (HO), or bone that forms outside the normal skeleton, in muscles, tendons or soft tissue. In FOP, HO progressively restricts movement by locking joints, leading to a cumulative loss of function, progressive disability, and increased risk of early death. FOP is caused by a mutation in the ACVR1 gene, resulting in excess signaling in the bone morphogenetic pathway, a key pathway controlling bone growth and development, by way of both ligand-dependent and independent mechanisms. There are currently no approved treatments for FOP.

About Clementia Pharmaceuticals Inc.
Clementia is a clinical-stage company innovating treatments for people with ultra-rare bone disorders and other diseases with high medical need. The Company is preparing for a 2019 NDA submission to the FDA to seek approval of its lead product candidate, palovarotene, a novel RARγ agonist, for the treatment of fibrodysplasia ossificans progressiva (FOP). The ongoing Phase 3 MOVE Trial is evaluating an additional dosing regimen of palovarotene which includes a chronic 5 mg daily dose in addition to the episodic 20/10 mg dosing regimen at the time of a flare-up. Palovarotene is also in a Phase 2 trial, the MO-Ped Trial, for the treatment of multiple osteochondromas (MO, also known as multiple hereditary exostoses, or MHE). In addition, Clementia has commenced a Phase 1 trial for an eye drop formulation of palovarotene for the potential treatment of dry eye disease and is also investigating other conditions that may benefit from RARγ therapy. For more information, please visit www.clementiapharma.com and connect with us on Twitter @ClementiaPharma.

Cautionary Note Regarding Forward-Looking Statements
This press release may include "forward-looking statements" within the meaning of the applicable securities laws, including with respect to the proposed timing of submission of the NDA for palovarotene. Each forward-looking statement contained in this press release is subject to known and unknown risks and uncertainties and other unknown factors that could cause actual results to differ materially from historical results and those expressed or implied by such statement. In addition to statements which explicitly describe such risks and uncertainties, readers are urged to consider statements labeled with the terms "believes," "belief," "expects," "intends," "anticipates," "will," or "plans" to be uncertain and forward-looking. Applicable risks and uncertainties include, among others, the Company's ability to successfully complete in a timely manner the studies required to be completed in order to submit the NDA, the Company's ability to generate revenue and become profitable; the risks related to its heavy

reliance on palovarotene, its only current product candidate; the risks associated with the development of palovarotene and any future product candidate, including the demonstration of efficacy and safety; its dependence on licensed intellectual property, including the ability to source and maintain licenses from third-party owners; as well as the risks identified under the heading "Risk Factors" in the Company's Annual Report on Form 20-F filed with the Securities and Exchange Commission ("SEC"), as well as the other information its file with the SEC or on SEDAR. Clementia cautions investors not to rely on the forward-looking statements contained in this press release when making an investment decision in its securities. Investors are encouraged to read the Company's filings with the SEC or on SEDAR, available at www.sec.gov or www.sedar.com, for a discussion of these and other risks and uncertainties. The forward-looking statements in this press release speak only as of the date of this press release, and the Company undertakes no obligation to update or revise any of these statements, whether as a result of new information, future events or otherwise, except as required by law.

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