PRESS RELEASE

Zambon Completes Enrollment in Phase 3 Clinical Development Program Evaluating Liposomal Cyclosporine A for Inhalation (L-CsA-i) in Patients with Bronchiolitis Obliterans Syndrome (BOS)

- Pivotal Phase 3 studies consist of BOSTON-1 and BOSTON-2, both designed to evaluate the safety and efficacy of L-CsA-i in patients with BOS following either single or double lung transplantation, respectively

- Approximately fifty percent of lung transplant patients develop BOS within five years post-transplant[1]

- If approved, L-CsA-i would be the first and only treatment indicated for BOS

Milan, Italy and **Boston, MA, May 1, 2023** —Zambon, a multinational pharmaceutical company focused on innovating cure and care to improve people's health and the quality of patients' lives, announced today that the Company has completed enrollment in its two pivotal Phase 3 studies. The trials are designed to evaluate the safety and efficacy of Liposomal Cyclosporine A for Inhalation (L-CsA-i) for the treatment of BOS in adults following single lung (BOSTON-1) or double lung (BOSTON-2) transplantation.

BOS is a rapidly progressive inflammatory rare disease that irreversibly destroys the airways of the lungs and usually leads to respiratory failure and death within two to four years after diagnosis. Unfortunately, approximately 50 percent of lung transplant patients develop BOS within five years post-transplant, the highest rejection rate of any transplanted organ. Currently, there is no approved treatment indicated for BOS.[1]

"The completion of enrollment in our Phase 3 BOSTON-1 and -2 studies represents a significant milestone for Zambon and the patient community we aim to serve," said Paola Castellani, chief medical officer and R&D head at Zambon. "On behalf of the Company, we would like to thank the many participants in our Phase 3 clinical program, as well as our study sites and investigators for their commitment. We look forward to reporting top-line results from the program during 2024.

L-CsA-i is a novel, inhaled therapy administered via the optimized investigational eFlow® Technology nebulizer system (PARI Pharma GmbH). The BOSTON-1 and -2 studies enrolled a total of 231 patients in Europe, Israel and the United States and had participation from 42 lung transplant centers globally.

"At Zambon, we are committed to advancing new therapeutic options that have the potential to make lives better for people affected by severe respiratory diseases such as BOS. The completion of enrollment in our pivotal trials brings us one step closer to our goal of bringing a much-needed therapy to the lung transplant recipients worldwide with

BOS," said Ilaria Villa, chief executive officer and interim chief operations officer of Zambon.

Zambon is also advancing one additional clinical study, BOSTON-3 – an extension study that is open to eligible participants who complete the BOSTON-1 and-2 studies.

L-CsA-i has received Orphan Drug Designation from the U.S. Food and Drug Administration (FDA) and European Medicines Agency for the treatment of BOS, reflecting the high unmet need of the disease. The Fast Track designation was also granted by FDA. If approved, L-CsA-i would be the first treatment indicated for BOS.

About Bronchiolitis Obliterans Syndrome (BOS)
Bronchiolitis Obliterans Syndrome (BOS), also known as obliterative bronchiolitis (OB), is caused by T-cell mediated inflammation that leads to blockage of bronchioles, the small and medium airways in the lungs, resulting in respiratory failure and death.[1] BOS most commonly affects people who have received lung transplant or allogeneic hematopoietic stem cell transplant (alloHSCT), although it is also associated with autoimmune disease and exposure to environmental contaminants.[2,3,4]

Approximately 50 percent of lung transplant patients develop BOS within five years post-transplant, the highest rejection rate of any transplanted organ.[5]

About Liposomal Cyclosporine A for Inhalation (L-CsA-i)
Liposomal Cyclosporine A for Inhalation (L-CsA-i) is a novel liposomal formulation of cyclosporine A developed for inhaled delivery to the lungs. Calcineurin inhibitors (CNIs), like cyclosporine A, are highly potent immunosuppressive drugs and a cornerstone of lung transplant medicine. L-CsA-i is administered via a drug-specific Investigational eFlow® Technology nebulizer system (PARI Pharma GmbH). The investigational drug-device combination is designed to deliver L-CsA-i to the site of disease in the lung.

About Zambon S.p.A.
Zambon Spa is a modern multinational pharmaceutical company established in 1906 in Vicenza, whose history is founded on the values of an Italian family committed to innovating cure & care to improve patients' lives. It employs 2,474 people worldwide, is present in 23 countries in Europe, America and Asia, and has production facilities in Italy, Switzerland, China and Brazil. Thanks to its innovative, quality products commercialized in 87 countries, Zambon Spa reported a revenue of 765 million euros in 2022. Alongside the three historical therapeutic areas - diseases of the respiratory system, urinary tract infections and pain treatment – Zambon is focused on developing treatments for neurodegenerative diseases such as Parkinson's or rare diseases such as cystic fibrosis, BOS, to which is linked the major 2019 acquisition of Breath Therapeutics, and NCFB. As part of Zambon's ambitious new path, an important role was also played by the agreement with Aquestive Therapeutics to market and distribute in Europe an innovative oral formulation of riluzole for patients suffering from

Amyotrophic Lateral Sclerosis.

For further information on Zambon please visit www.zambon.com

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[1] Chambers DC, et al. J Heart Lung Transplant. 2018;37(10):1169–1183

[2] Weigt, et al. Semin Respir Crit Care Med. 2013;34(3):336–351.

[3] Soubani, AO. Eur Respir J. 2007; 29: 1007-1019.

[4] Sheshadri, A, et al. Clin Ther. May 2022, https://doi.org/10.1016/j.clinthera.2022.03.011.

[5] Weigt, et al. Semin Respir Crit Care Med. 2013;34(3):336–351.