Avalyn Pharma Publishes Phase 1b ATLAS Results Demonstrating Stabilization of Lung Function at 24 and 48 Weeks with AP01 and Favorable Safety Profile

March 23, 2023 – SEATTLE -- Avalyn Pharma Inc., a clinical-stage biopharmaceutical company focused on the development of targeted therapies for life-threatening pulmonary diseases, today reported that results from the Phase 1b ATLAS study were published online in *Thorax*, an official journal of the British Thoracic Society. The study assessed safety and efficacy of AP01 (inhaled pirfenidone) in adults with idiopathic pulmonary fibrosis (IPF) through 72 weeks. Efficacy and safety results at weeks 24 and 48 are also reported.

"While oral pirfenidone has the potential to improve lung function by reducing fibrosis, its utility is limited by its poor tolerability profile," explained Lyn Baranowski, Avalyn's CEO. "Although the ATLAS study's primary endpoint was safety, secondary measures of efficacy showed a trend towards disease stabilization in participants with IPF who administered high-dose AP01. Based on this data, we believe AP01 has the potential to meet or exceed the efficacy of oral pirfenidone without the systemic toxicities that limit its adoption. We are eager to explore this hypothesis in a larger, controlled efficacy trial that we plan to begin later this year."

At baseline, mean forced vital capacity (FVC), a measure of lung capacity, was 2.5 L for ATLAS study participants in the 50 mg QD arm and 2.6 L in the 100 mg BID arm. Most participants in the 100 mg BID group experienced stabilization of FVC with a mean change of 0.6% at week 24 and -0.4% at week 48. These results suggest inhaled pirfenidone is successfully delivered to the target tissue with beneficial effect. Supporting these observations, FVC changes in the 100 mg BID group correlated with changes in quantitative lung fibrosis scores, as measured by high-resolution computed tomography (HRCT).

While oral pirfenidone has the potential to improve lung function by reducing fibrosis, its utility is limited by a poor tolerability profile and incidence of adverse events including elevated liver enzymes, diarrhea, vomiting and nausea.

"The tolerability profile we saw with AP01 in this study is really important because we see so many patients who may try to take the oral antifibrotics but eventually stop as the side-effects become increasingly difficult to bear as their disease progresses, and others who cannot tolerate them for even a short time," added Alex West, Interstitial Lung Disease Lead at Guy's and St Thomas' Hospital, London and an investigator in the ATLAS study. "AP01 has the potential to keep patients on effective therapy longer, which might translate into increased life expectancy and quality of life."

The doses of AP01 studied (50 mg QD or 100 mg BID) were well tolerated with no adverse effects on respiratory rate, spirometry, or oxygenation during or following administration. 48-week treatment-emergent adverse events (AEs) reported for at least 10% of patients in either dose group included cough, rash, dyspnea, nausea, IPF, fatigue, lower respiratory tract infection and upper respiratory tract

infection. Except for a Grade 3 parainfluenza virus infection, all other treatment-related events were mild or moderate. One treatment-related liver enzyme increase occurred but resolved after dose interruption.

About the ATLAS Study

The open-label ATLAS study enrolled 91 adults with IPF who were intolerant of or ineligible for oral pirfenidone or nintedanib. Participants were randomized to one of two treatment arms: 50 mg AP01 once daily (QD) or 100 mg twice daily (BID) delivered with the eFlow Nebulizer System (eFlow® nebulizer; PARI GmbH, Germany). The ATLAS study assessed the safety, tolerability and efficacy of two AP01 doses in patients with IPF. Of the 91 participants who enrolled, 77 (85%) completed 24 weeks of treatment.

Following review of week 24 data, the DSMB recommended all participants transition to the higher dose of AP01. Dosing continued in the ATLAS study through week 72. Five participants receiving low-dose AP01 transitioned to the higher dose before or during their week 48 visit; an additional 16 transitioned by week 72. All participants who reached week 72 were eligible to participate in an open-label extension study.

Of the 54 who reached week 72, 41 (76%) continued into the open-label extension. To date, 100 individuals with IPF and other forms of pulmonary fibrosis, including progressive pulmonary fibrosis, are enrolled in the open-label extension, eight of whom have been on study medication for over three years. Findings from the open-label extension study will be presented at upcoming medical meetings.

About AP01 (Pirfenidone Solution for Inhalation)

Pirfenidone is a small molecule shown to inhibit fibroblast differentiation and extracellular matrix (ECM) production. AP01 is an inhaled aerosol formulation of pirfenidone that is delivered using the eFlow® nebulizer, a high efficiency vibrating membrane nebulizer similar in design and operation to PARI's 510(k) cleared/FDA-approved eFlow-based nebulizers marketed with other products for other disease indications. This administration method allows a smaller pirfenidone dose to be delivered as a soft mist directly to the lung, maximizing pirfenidone's effect on diseased lung tissue while sparing tissue outside of the lungs from the debilitating toxicities associated with oral delivery. Avalyn is planning to advance AP01 (100 mg BID) into the next phase of the clinical program in 2023.

About Avalyn Pharma

Avalyn is a biopharmaceutical company developing targeted therapeutics for the treatment of rare respiratory diseases including IPF and other interstitial lung diseases (ILD). ILDs are characterized by scarring, a decline in lung function, reduced exercise capacity and quality of life, and are associated with increased mortality. Currently approved therapeutic options slow ILD progression but are associated with significant toxicities, which restrict their use and dosing. Avalyn is developing a pipeline of inhaled therapeutics designed to reduce systemic exposure and deliver medication to the site of disease. AP01, Avalyn's lead candidate, is an inhaled formulation of pirfenidone optimized for delivery via inhalation. In a recent clinical study of two doses assessed in 91 individuals with IPF, AP01 demonstrated the potential

to improve efficacy and safety over existing therapy. More information can be found at www.avalynpharma.com.

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