PRESS RELEASE

Stockholm, Sweden, 17 October 2023

Sobi announces positive Phase 2 NOBLE results of pegcetacoplan in post-transplant recurrence of primary IC-MPGN and C3G

- *After 12 weeks, 50% of patients treated with pegcetacoplan showed a reduction in C3c staining by two or more orders of magnitude of intensity from baseline.*

- *Pegcetacoplan patients showed improvements across key clinical measures, including kidney function and proteinuria.*

- *No approved treatment for IC-MPGN and C3G, rare diseases that often lead to kidney failure.*

Sobi® and Apellis Pharmaceuticals, Inc. today announced that positive results from the Phase 2 NOBLE study investigating pegcetacoplan for the treatment of post-transplant recurrence of primary immune complex membranoproliferative glomerulonephritis (IC-MPGN) and C3 glomerulopathy (C3G) will be presented at the American Society of Nephrology (ASN) Kidney Week Annual Meeting.

The results showed the potential for a treatment effect in both IC-MPGN and C3G patients treated with pegcetacoplan. At Week 12, of the 10 patients (IC-MPGN: n=2; C3G: n=8) treated with pegcetacoplan:

- Eight (80%) patients showed a reduction in C3c staining (reflective of damage-causing deposits) by one or more orders of magnitude of intensity from baseline.
- Five (50%) patients showed a reduction in C3c staining by two or more orders of magnitude of intensity from baseline.
- Four (40%) patients showed zero staining intensity, indicating that C3 deposits were cleared.

Excessive deposition of C3 breakdown products in the kidney lead to inflammation and damage of the kidney, often causing kidney failure.

“In as early as 12 weeks, these positive results show that pegcetacoplan has the capacity to clear the deposits that are causing kidney damage and may block future damage from occurring,” said Andrew Bomback, M.D., presenting author and co-director of the Centre for glomerular diseases at Columbia University Irving Medical Centre. “People living with post-transplant C3G and IC-MPGN have high rates of disease recurrence, creating a significant burden on patients both physically and emotionally. These data are very promising, especially given there are no approved treatments currently available.”

Additionally, in 12 weeks, pegcetacoplan showed a mean reduction of proteinuria (39.2% change from baseline), which is a key marker of disease progression[1], in a subgroup of patients with high baseline levels (≥1g per day). Other biomarkers also improved, including an increase in mean serum C3, reduction in mean serum C5b-9 and stabilisation of kidney function, as measured by estimated

glomerular filtration rate (eGFR). There were no discontinuations due to treatment-emergent adverse events.

"These findings further bolster our conviction in pegcetacoplan's potential to address both the primary and post-transplant variants of these rare, severe and life-threatening conditions," said Lydia Abad-Franch MD, Head of R&D and Medical Affairs, and Chief Medical Officer at Sobi. "We remain committed to advancing pegcetacoplan and bringing it to more people in need, guided by our unwavering commitment to transform the lives of people with rare diseases."

The Phase 3 VALIANT study investigating pegcetacoplan in adolescent and adult patients with native and post-transplant recurrence of IC-MPGN and C3G is ongoing, with top-line results expected in 2024. Data from the Phase 2 DISCOVERY study investigating pegcetacoplan in patients with C3G was recently published in KI Reports. Development for pegcetacoplan for IC-MPGN and C3G is being led by Apellis.

Presentation Details at ASN Kidney Week 2023

Efficacy of 12-week pegcetacoplan in kidney transplant recipients with recurrent C3 glomerulopathy (C3G) or immune complex membranoproliferative glomerulonephritis (IC-MPGN),
Poster #SA-PO923 – November 4, 2023 from 10:00 a.m. – 12:00 p.m. ET

About the Phase 2 NOBLE Study

The Phase 2 NOBLE study (NCT04572854) is a multicentre, open-label, randomised, controlled study designed to evaluate the efficacy and safety of pegcetacoplan in 13 adults who have post-transplant recurrence of C3G or primary IC-MPGN. Study participants were randomised in a 3:1 ratio to receive pegcetacoplan or maintain standard of care for 12 weeks, and patients will receive pegcetacoplan from week 13 to week 52. The primary endpoint of the study is the proportion of patients with a reduction in C3c staining on renal biopsy after 12 weeks of treatment with pegcetacoplan. Secondary endpoints include an evaluation of safety, the proportion of patients with a reduction in C3c staining on renal biopsy after 52 weeks of treatment, and the proportion of patients achieving at least a 50% reduction in proteinuria.

About the VALIANT Study

The VALIANT Phase 3 study (NCT05067127) is a randomised, placebo-controlled, double-blinded, multicentre study designed to evaluate pegcetacoplan efficacy and safety in approximately 90 patients who are 12 years of age and older with primary IC-MPGN or C3G. It is the only study to include both native kidney patients and patients who have recurrent disease after receiving a kidney transplant. Study participants will be randomised to receive 1080 mg of pegcetacoplan or placebo twice weekly for 26 weeks. Following this 26-week randomised controlled period, patients will proceed to a 26-week open-label phase in which all patients receive pegcetacoplan. The primary endpoint of the study is the log transformed ratio of urine protein-to-creatinine ratio (uPCR) at week 26 compared to baseline. uPCR is an important indicator of kidney function.

About Immune-Complex Membranoproliferative Glomerulonephritis (IC-MPGN) and C3 Glomerulopathy (C3G)

IC-MPGN and C3G are rare, debilitating kidney diseases that are estimated to affect 5,000 people in the United States and up to 8,000 in Europe.[2] Symptoms include blood in the urine, dark foamy urine due to the presence of protein, swelling, and high blood pressure.[3] Approximately 50% of people living with IC-MPGN and C3G ultimately suffer from kidney failure within five to 10 years of diagnosis.[4] There are no treatments available that target the underlying complement-mediated mechanism of these diseases and prevent loss of kidney function before or after renal transplant. Although IC-MPGN is considered a distinct disease from C3G, the underlying cause and progression of the two diseases are remarkably similar and include overactivation of the complement cascade, with excessive accumulation of C3 breakdown products in the kidney, causing inflammation and damage to the organ.[5-9]

About Pegcetacoplan in Rare Diseases

Pegcetacoplan is a targeted C3 therapy designed to regulate excessive activation of the complement cascade, a part of the body's immune system, which can lead to the onset and progression of many serious diseases. Pegcetacoplan is under investigation for several rare diseases across haematology and nephrology. Pegcetacoplan is approved for the treatment of

paroxysmal nocturnal haemoglobinuria (PNH) as EMPAVELI®/ASPAVELI in the United States, European Union, and other countries globally.

About the Sobi and Apellis Collaboration
Sobi and Apellis have global co-development rights for systemic pegcetacoplan. Sobi has exclusive ex-U.S. commercialisation rights for systemic pegcetacoplan, and Apellis has exclusive U.S. commercialisation rights for systemic pegcetacoplan and worldwide commercial rights for ophthalmological pegcetacoplan, including for geographic atrophy.

Sobi®
Sobi is a specialised international biopharmaceutical company transforming the lives of people with rare and debilitating diseases. Providing reliable access to innovative medicines in the areas of haematology, immunology and specialty care, Sobi has approximately 1,800 employees across Europe, North America, the Middle East, Asia and Australia. In 2022, revenue amounted to SEK 18.8 billion. Sobi's share (STO:SOBI) is listed on Nasdaq Stockholm. More about Sobi www.sobi.com, LinkedIn and YouTube.

Contacts
For details on how to contact the Sobi Investor Relations Team, please click here. For Sobi Media contacts, click here.

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