Press release

Cantargia AB
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Cantargia selects CAN10 as development project in systemic sclerosis and myocarditis

Cantargia AB today announced that its research platform CANxx has progressed with the start of a drug development project, CAN10. The project originates from inhibition of three inflammatory cytokine systems (IL-1, IL-33 and IL-36) by antibody targeting of interleukin 1 receptor accessory protein (IL1RAP). The project will focus on unmet medical need in systemic sclerosis and myocarditis. The goal is to initiate clinical development late 2021.

Cantargia develops antibody-based pharmaceuticals against the interleukin 1 receptor accessory protein (IL1RAP). The lead project CAN04 is in phase IIa clinical development for the treatment of pancreatic cancer and non-small cell lung cancer. With the CANxx platform, the goal is to build on the platform of novel IL1RAP-binding antibodies. IL1RAP is a unique target as it mediates signaling from three different disease-associated cytokine systems (IL-1, IL-33 and IL-36) involved in autoimmunity and inflammatory diseases.

As part of the advancing CANxx platform, the company has generated a library of more than 100 well-characterized anti-IL1RAP antibodies. The development project CAN10 includes a proprietary group of antibodies that bind strongly to IL1RAP (affinity below 1 nM) and shows a potent inhibition of the three inflammatory cytokines IL-1, IL-33 and IL-36 (low nM range). With CAN10, Cantargia now advances two of these fully humanized lead antibodies to the next step of development. Here, generation of stable production systems will be constructed, before selecting one of these antibodies as clinical candidate and focusing development to one antibody. Taking two antibodies with similar biological properties into initial development offers an opportunity compared to the original strategy; it allows further studies of production and biophysical properties in parallel with other development activities, thus decreasing the project risk without jeopardizing timelines.

To focus the development process of CAN10, independent experts have performed an analysis of its potential to treat approximately 150 autoimmune and inflammatory diseases. The analysis included interviews with key opinion leaders and covered a number of aspects, such as scientific rationale for the blockade of the three inflammatory cytokines, medical need, development opportunities and competition. Based on this analysis, the initial focus for development will be the two life-threatening diseases systemic sclerosis and myocarditis.

In parallel to the activities above, Cantargia has generated data showing strong and unique anti-inflammatory effects in mouse disease models of inflammatory disease. The data show much stronger anti-inflammatory activity than antibodies blocking only IL-1β. CAN10 will now advance with further studies on preclinical efficacy and safety, with the goal to initiate the phase I clinical trial late 2021.

"The development of our IL1RAP platform is a big step forward for the company. We look forward to advancing the CAN10 project for the treatment of life-threatening diseases with few therapeutic options", said Göran Forsberg, CEO at Cantargia.

As previously announced, Cantargia has an ongoing partnership with Panorama Research Inc in Sunnyvale, CA, USA, which will continue independently of CAN10 within the CANxx project.

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This is information that Cantargia AB is obliged to make public pursuant to the EU Market Abuse Regulation. The information was submitted for publication, through the agency of the contact person set out above, at 10.30 CET on 5 December 2019.

About Cantargia
Cantargia AB (publ), reg. no. 556791-6019, is a biotechnology company that develops antibody-based treatments for life-threatening diseases. The basis for this is the protein IL1RAP that is involved in a number of diseases and

where Cantargia has established a platform. The main project, the antibody CAN04 is being studied in the clinical phase I/IIa CANFOUR with a primary focus on non-small cell lung cancer and pancreatic cancer. The study is focused on combination therapies, but also includes a monotherapy arm. Positive interim data from the combination therapies were presented in December 2019. Cantargia's platform project, CANxx, is in the research phase and is aiming to develop a IL1RAP binding antibody optimized for the treatment of autoimmune and inflammatory diseases.

Cantargia is listed on Nasdaq Stockholm (ticker: CANTA). More information about Cantargia is available at http://www.cantargia.com.

About Systemic sclerosis
Systemic Sclerosis (also known as Scleroderma) is a chronic, autoimmune connective tissue disorder characterized by inflammation and fibrosis of the skin and internal organs (e.g., lungs, kidneys, heart, and gastrointestinal tract)[1].

Systemic Sclerosis is a complex, heterogenous disease that can present with a wide variety of clinical manifestations ranging from minor to life-threatening. The estimated annual incidence of systemic sclerosis is about 4.5 per 100 000 in North America and 1.8 per 100 000 in Europe[2,3,4]. The leading cause of death in patients with systemic sclerosis is interstitial lung disease and the unmet need is in particularly high in these patients.

1. Nat Rev Dis Primers. 2015 Apr 23;1:15002
2. Best Pract Res Clin Rheumatol. 2018 Apr;32(2):223-240
3. Clin Epidemiol. 2019 Apr 18;11:257-2
4. Ann Rheum Dis. 2014 Oct;73(10):1788-92

About myocarditis
Myocarditis is characterized by inflammation of muscular tissues of the heart (myocardium) that arise from different etiologies, including genetic and infectious mechanisms that are not well characterized1,2. Regardless of its etiology, myocarditis is characterized by initial acute inflammation that can progress to subacute and chronic stages resulting in tissue remodeling, fibrosis, and loss of myocardium architecture and contractile function[1,2].

The estimated incidence of myocarditis is approximately 22 per 100,000[3] (1.70M) and the disease accounts for approximately 0.6 per 100,000 deaths[4] (46.4K) annually worldwide. The unmet need is high for subsets of patients with fulminant myocarditis (acute disease) and dilated cardiomyopathy (chronic disease), where mortality rates are very high in certain immune subtypes. For these patients, the only definitive therapy is currently heart transplantation.

1. Adv Exp Med Biol. 2017;1003:187-221
2. Circ Res. 2019 May 24;124(11):1568-1583
3. J Am Coll Cardiol. 2016 Nov 29;68(21):2348-2364
4. Lancet. 2018;392:1736-88