

People living with rare lung diseases are at the center of everything we do

- Founded in 2016 and based in Munich, Germany and Menlo Park, California.
- Emerging biotech with 18+ employees and plans to grow operations.
- Series A financing in 2017 raised \$46 million.
- Pipeline focused on breakthrough inhalation therapies for respiratory orphan diseases.
- Lead product candidate, L-CsA-i, currently in Phase 3 clinical studies for the treatment of bronchiolitis obliterans syndrome (BOS) following lung transplantation.

Bronchiolitis Obliterans Syndrome (BOS)

- BOS is a rare, fatal lung disease with no approved therapies.¹
- Rapidly progressive inflammatory disease that irreversibly destroys the airways of the lung and usually leads to respiratory failure and death within 2 to 4 years after diagnosis.²
- BOS is most commonly seen in patients following lung transplantation (LTx) and allogeneic hematopoietic stem cell transplantation (alloHSCT).
- Historically known as “popcorn lung”, BOS was first seen in patients with environmental exposures and autoimmune disease and it is believed that BOS is often misdiagnosed.

BOS Prevalence Cases Currently Estimated for US, EU, JAPAN³

LTx	5,000 annual procedures	31,000 people living with	21,500 affected by BOS
alloHSCT	32,000 annual procedures	140,000 people living with	8,500 affected by BOS

Liposomal Cyclosporine A for Inhalation (L-CsA-i)

- L-CsA-i is a novel investigational liposomal formulation of cyclosporine A designed for inhaled delivery to the lungs via a customized nebulizer (PARI’s eFlow® Nebulizer).
 - Systemic cyclosporine is a highly potent immunosuppressive drug, cornerstone of lung transplant medicine.
 - L-CsA-i has ~1,100 patient-months of exposure.
- L-CsA-i has received orphan drug designation from FDA and EMA.
- Investigational drug-device combination is designed to increase effective dosing to small airways of the lung and minimize overall systemic drug exposure.
 - Drug nebulizer system is specifically designed for inhalation of L-CsA-i.
 - Fast, portable, with patient self-monitoring capability.

BOSTON Program

The BOSTON program is evaluating Liposomal Cyclosporine A for Inhalation (L-CsA-i), as a first potential therapy for the treatment of BOS.

- Two global Phase 3 studies were initiated in Q1, 2019.
- Additional studies include L-CsA-i for BOS following alloHSCT and for pediatric patients with BOS.

The BOSTON studies are designed to bring L-CsA-i to patients with BOS.

BOSTON-1

L-CsA-i for BOS following single lung transplant [Ph3] (Initiated Q1 2019)

BOSTON-2

L-CsA-i for BOS following double lung transplant [Ph3] (Initiated Q1 2019)

BOSTON-3

Extension trial of BOSTON-1 and BOSTON-2

BOSTON-4

L-CsA-i for BOS following Allogeneic Hematopoietic Stem Cell Transplantation

BOSTON-5

L-CsA-i for pediatric patients with BOS

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REFERENCES:

- An international ISHLT/ATS/ERS clinical practice guideline: diagnosis and management of bronchiolitis obliterans syndrome. Meyer KC, et al. Eur Respir J 2014; 44: 1479–1503 | DOI: 10.1183/09031936.00107514
- The International Thoracic Organ Transplant Registry of the International Society for Heart and Lung Transplantation: Thirty-fifth adult lung and heart-lung transplant report—2018; Focus theme: Multiorgan Transplantation. Chambers DC, et al. J Heart Lung Transplant; Volume 37, Issue 10, 1169 – 1183
- Company internal market research based on country-specific reports for lung transplants and stem cell transplantations

Forward-looking Statements

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