A New Path to Medicine

BIO CEO & Investor Conference

Sanjay S. Shukla, M.D., M.S., President & CEO

February 11, 2020

Forward-Looking Statements

The following slides and any accompanying oral presentation contain forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995 and other federal securities laws. The use of words such as "may," "might," "will," "should," "expect," "plan," "anticipate," "believe," "estimate," "project," "intend," "future," "potential," "opportunity," or "continue," and other similar expressions are intended to identify forward-looking statements. For example, all statements regarding: the potential therapeutic benefits of proteins derived from tRNA synthetase genes and our product candidates, including ATYR1923, and development programs; the ability to successfully advance our product candidates and undertake certain development activities (such as the initiation of clinical trials, clinical trials enrollment, the conduct of clinical trials and announcement of top-line results) and accomplish certain development goals, and the timing of such events; our ability to receive regulatory approvals for, and commercialize, our product candidates; our ability to identify and discover additional product candidates; potential activities and payments under collaboration agreements; and the ability of our intellectual property portfolio to provide protection are forward-looking statements. All forward-looking statements are based on estimates and assumptions by our management that, although we believe to be reasonable, are inherently uncertain. All forward-looking statements are subject to risks and uncertainties that may cause actual results to differ materially from those that we expected. These risks, uncertainties and other factors are more fully described in our filings with the U.S. Securities and Exchange Commission, including our Annual Report on Form 10-K, our Quarterly Reports on Form 10-Q, and in our other filings. The forward-looking statements in this presentation speak only as of the date of this presentation and neither we nor any other person assume responsibility for the accuracy and completeness of any forward-looking statement. We undertake no obligation to publicly update or revise any forward-looking statement, whether as a result of new information, future events or otherwise, except as required by law.

We own various U.S. federal trademark applications and unregistered trademarks, including our company name. All other trademarks or trade names referred to in this presentation are the property of their respective owners. Solely for convenience, the trademarks and trade names in this presentation are referred to without the symbols ® and ™, but such references should not be construed as any indicator that their respective owners will not assert, to the fullest extent under applicable law, their rights thereto.

aTyr Company Overview

aTyr: A New Path to Medicine

- Mission: develop a new class of medicine based on proprietary biology
- ATYR1923: Potential first-in-class immunomodulator for interstitial lung diseases (ILD) currently enrolling proof-of-concept trial in pulmonary sarcoidosis
  - Recent license agreement with Kyorin for the development and commercialization of ATYR1923 for ILDs in Japan
- Discovery pipeline focused on NRP2(1) antibodies for cancer and inflammation and new tRNA synthetase(2) candidates for immunology
- Cash, cash equivalents and investments at $38.1m as of 9/30/19
  - Does not include $8m upfront from Kyorin or $18m raised in equity offering
- Top investors include Federated, Fidelity, Dr. Paul Schimmel

(1) NRP2 = Neuropilin 2
(2) In collaboration with CSL Behring

tRNA Synthetases May Have Novel Functions Extracellularly

INTRACELLULAR

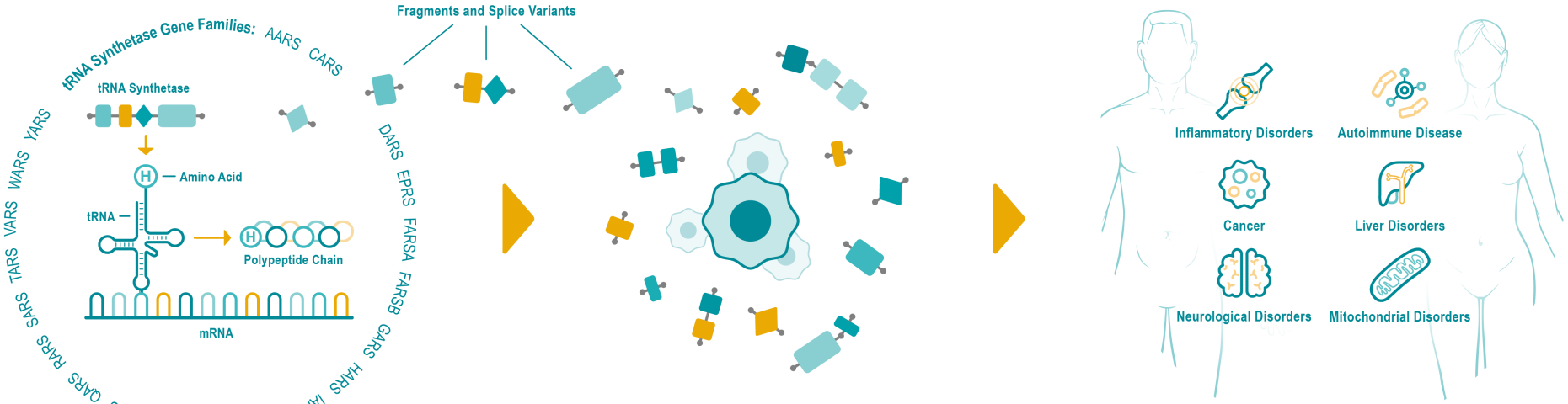
Catalyze Protein Synthesis

EXTRACELLULAR

Secreted in Circulation and Tissue to Regulate Diverse Pathways

PHYSIOLOGICAL

Pathway Disruption Associated with Disease



aTyr IP covers protein compositions from all 20 tRNA synthetase gene families and certain associated signaling pathways

aTyr

aTyr Development Pipeline

PROGRAM	DISEASES	DISCOVERY	PRECLINCAL	PHASE 1	PHASE 2	PHASE 3	PARTNERS
ATYR1923	Pulmonary Sarcoidosis						**Kyorin** ILD in Japan
	Chronic Hypersensitivity Pneumonitis (CHP)						
	Connective Tissue Disease ILD (CTD-ILD)						
tRNA synthetase candidates	Immunology						**CSL Behring** 4 candidates
NRP2 antibodies	Cancer; Inflammation						**Academic collaborations**

ATYR1923

First-in-class Immunomodulator for ILD

ATYR1923: Potential First-in-Class Immunomodulator for ILD

- Binds selectively to NRP2, a novel cell surface receptor upregulated in inflamed lung tissue
- Downregulates inflammatory and pro-fibrotic cytokines and chemokines *in vitro* and *in vivo*
- Demonstrated anti-inflammatory and anti-fibrotic effects in multiple ILD animal models
- Generally well tolerated in healthy volunteers with PK supporting once-monthly IV dosing
- Currently enrolling first-in-patient trial in pulmonary sarcoidosis; expect to announce results Q3 2020(1)
- Future development planned in other ILDs, e.g. CTD-ILD or CHP

(1) Dependent on patient enrollment

Persistent Immune Insult is Central to ILD Pathology

Pulmonary Sarcoidosis
- Non-caseating granulomas (CD4+ T cell driven)
- SOC: steroids - cytotoxic agents - TNF inhibitors

Chronic Hypersensitivity Pneumonitis (CHP)
- Exaggerated immune response to environmental antigen
- No effective SOC

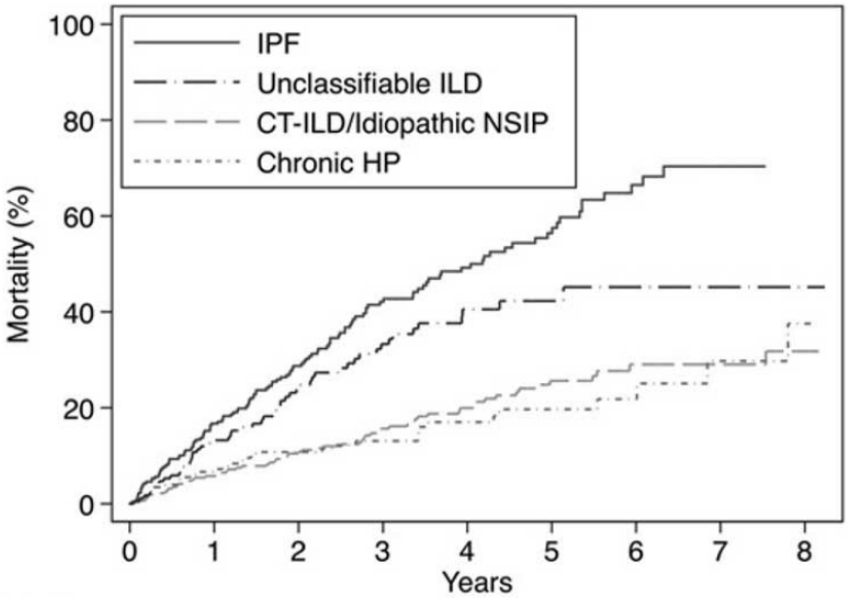
Connective Tissue Disease – ILD (CTD-ILD)
- >50% of Scleroderma patients and 10% RAs
- SOC: Immunosuppressants (2019 nintedanib label expansion for SSc-ILD)

Idiopathic Pulmonary Fibrosis (IPF)
- Irreversible fibrotic disease
- SOC: Nintedanib or pirfenidone (>$2.3b combined 2018 net revenue)

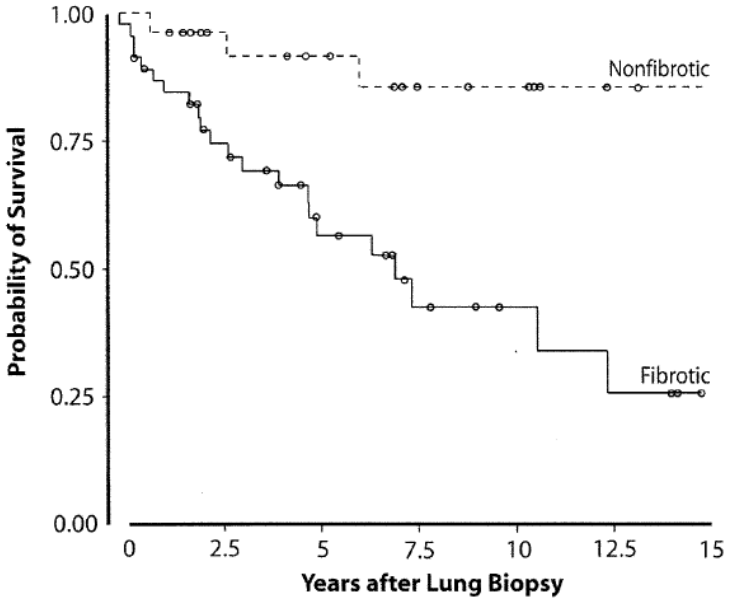
Slide adapted from Dr. Steven Nathan, Medical Director, Advanced Lung Disease and Transplant Program at Inova Fairfax Hospital, Falls Church, VA

Fibrosing ILDs Share Poor Clinical Outcomes

High Mortality Burden Beyond IPF



Fibrosis Associated with Mortality in CHP



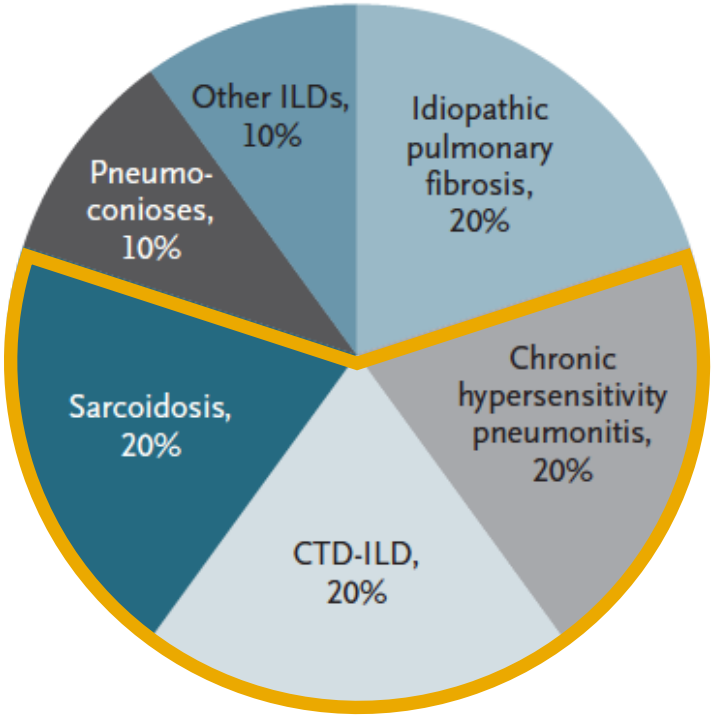
Intervening early to avoid progression to fibrosis may improve outcomes

Ryerson, CHEST 2014; Vourlekis, Am J Med 2004

aTyr

Initial Target: Pulmonary Sarcoidosis is a Major Form of ILD

Relative Distribution of Specific ILDs in the USA[1] – All ILDs Eligible for Orphan Drug Designation



$2-3b Global Opportunity[2]

Pulmonary Sarcoidosis

50% require systemic therapy

30% with chronic progressive disease despite currently available treatment

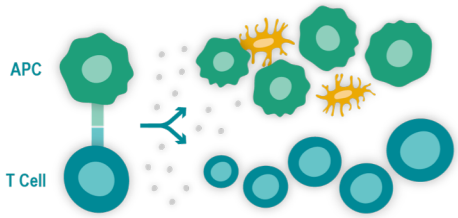
(1) Lederer, Martinez. NEJM 2018
(2) aTyr estimates for inflammatory ILD: Pulmonary Sarcoidosis, CHP, CTD-ILD; excludes IPF

ATYR1923 Mechanism of Action in ILD

Disease Trigger

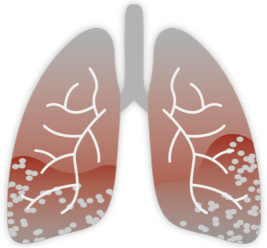
Organic; inorganic; infectious; autoimmune

Aberrant Immune Response



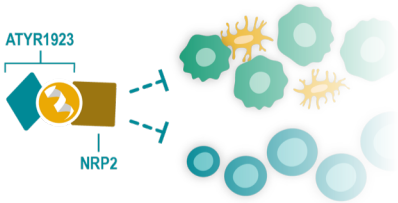
T-cell activation; pro-inflammatory cytokines/chemokines triggering fibrotic pathways; NRP2 upregulation on immune cells

Lung Inflammation & Fibrosis



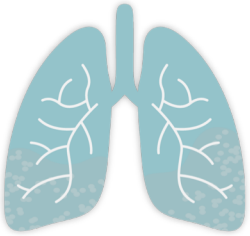
Persistent, unresolved inflammation in the lung can lead to fibrosis; patients experience chronic cough, dyspnea, mortality

ATYR1923 Dampens Immune Responses



ATYR1923 binds to NRP2 and downregulates cytokine and chemokine production and T-cell activation

Stabilized Lung



Reduced inflammation and fibrotic deposition; symptom relief, stabilized lung function*

*aTyr hypothesis

Demonstrated Effect in Multiple ILD Models*

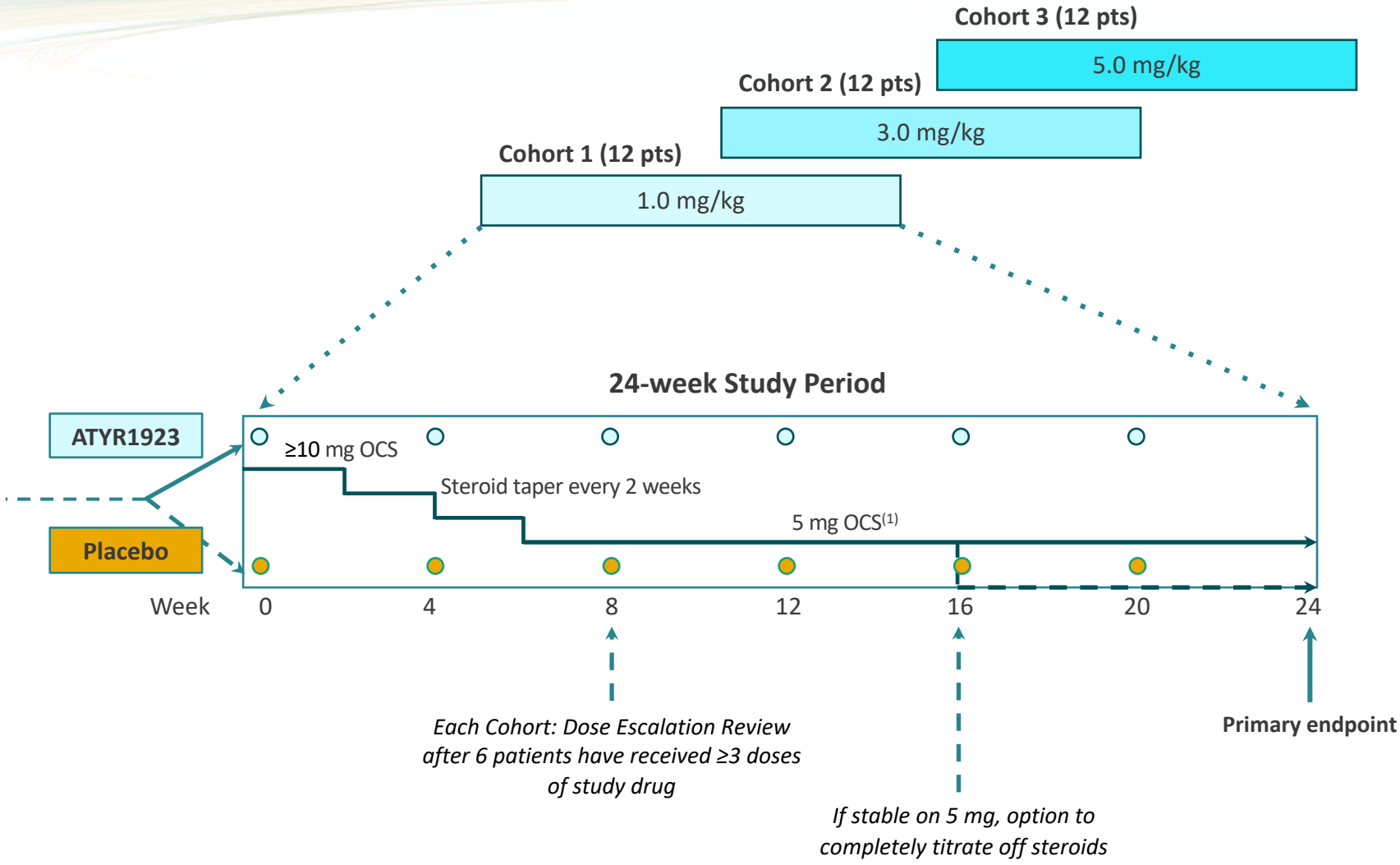
Bleomycin-Induced Lung Fibrosis	↓inflammation / fibrosis
SKG mice [joint (100%), lung (20%)] (RA-ILD)	↓ immune infiltrates
Sclerodermatous GvHD (lung and skin) (SSc-ILD)	↓fibrosis
S. Rectivirgula (CHP)	↓ tissue cytokines/chemokines and enzymes
P. acnes challenge (Sarcoidosis)	↓ pro-fibrotic proteins

*Data available at https://www.atyrpharma.com/our-science/publications/

ATYR1923 Phase 1b/2a Study in Pulmonary Sarcoidosis

Design	• Randomized (2:1), double-blind, placebo-controlled, multiple ascending dose
Population	• 36 histologically confirmed pulmonary sarcoidosis patients • ≥10 mg stable oral corticosteroid treatment • Symptomatic/active disease at baseline
Endpoints	• Primary ◦ Safety and tolerability of multiple ascending IV ATYR1923 doses • Secondary ◦ Steroid-sparing effect ◦ Immunogenicity ◦ Pharmacokinetics (PK) ◦ Exploratory efficacy measures: FDG-PET/CT imaging; Lung function (FVC); Serum biomarkers (ACE, sIL-2R); Health-related quality of life scales

Phase 1b/2a Study Schema



(1) subtherapeutic dose

ATYR1923 Program Snapshot

Status	• Phase 1 in 36 healthy volunteers completed in 2018 • Patient enrollment ongoing in Phase 1b/2a in 17 leading pulmonary sarcoidosis centers • Positive interim safety data reported December 2019
Timelines	• Expect to announce results in Q3 2020(1)
Possible Future Development	• Registrational trial in pulmonary sarcoidosis • Initiate P2 studies in other types of interstitial lung disease (e.g. CTD-ILD; CHP)

(1) Dependent on patient enrollment

ATYR1923 Japan Collaboration

Kyorin Overview

- Founded: 1923
- Focus: Respiratory, ENT, Urology
- 1600 employees: incl. 350 in R&D; 750 sales reps covering top respiratory centers in Japan
- Sales: ~$1b USD
- Market cap: $1.1b USD (4569:JP TSE)

Key Terms

- Scope: ATYR1923; Japan; ILD
- Upfront payment: $8m
- Development, regulatory and commercial milestones: $167m
- Tiered sales royalties into double digits
- Kyorin to fund all development and commercial activities in Japan

NRP2 Antibodies

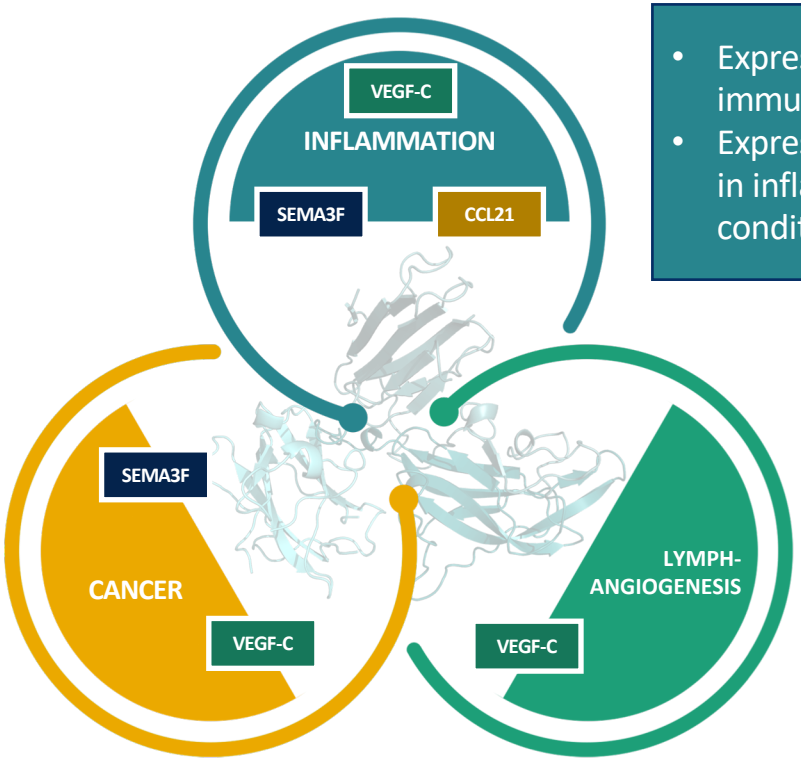
Regulating Diverse Disease Pathways

NRP2: A Novel Therapeutic Target

- NRP2 is a potentially novel target for cancer and inflammatory disorders
- Acts as a co-receptor for VEGF-C, class 3 Semaphorins and CCL21
- NRP2 expression is upregulated on tumors and immune cells during inflammation
- NRP2 expression is linked to worse outcomes in cancer
- aTyr has developed antibodies to selectively target different NRP2 epitopes for diverse therapeutic applications

NRP2 is a Compelling Target for Cancer and Inflammation

- Overexpressed in a variety of cancers
- Tumor expression linked to worse outcomes

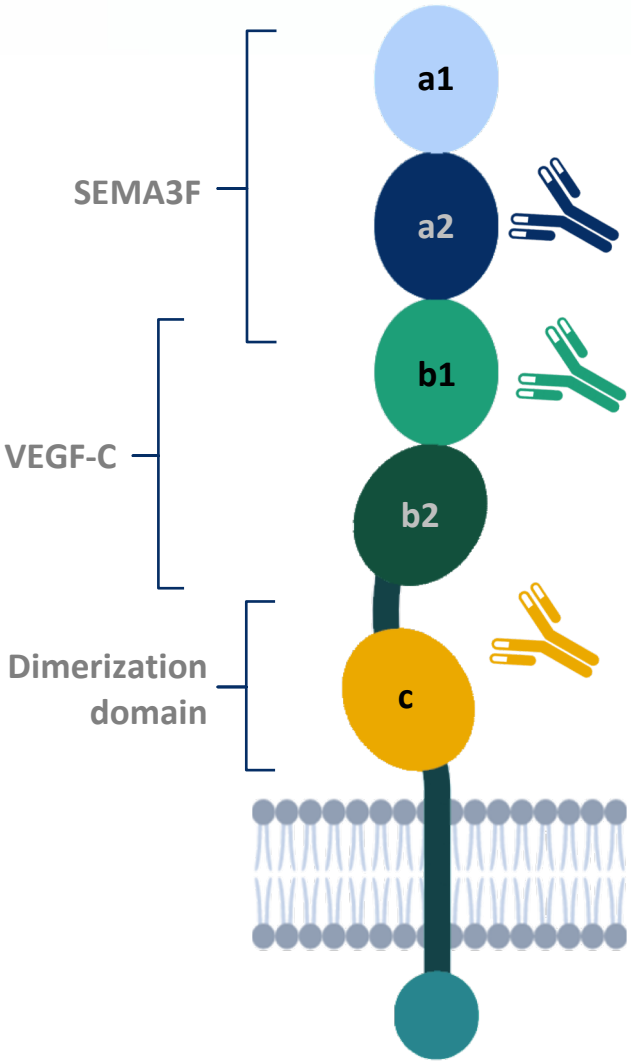


- Expressed on multiple immune cell types
- Expression upregulated in inflammatory conditions

- Lymphatic development and function impaired in NRP2 knockout

SEMA3F and VEGF-C are the most well characterized ligands for NRP2

aTyr Human NRP2 Blocking Antibodies



aTyr lead antibodies

SEMA3F Blocker
α-NRP2-14

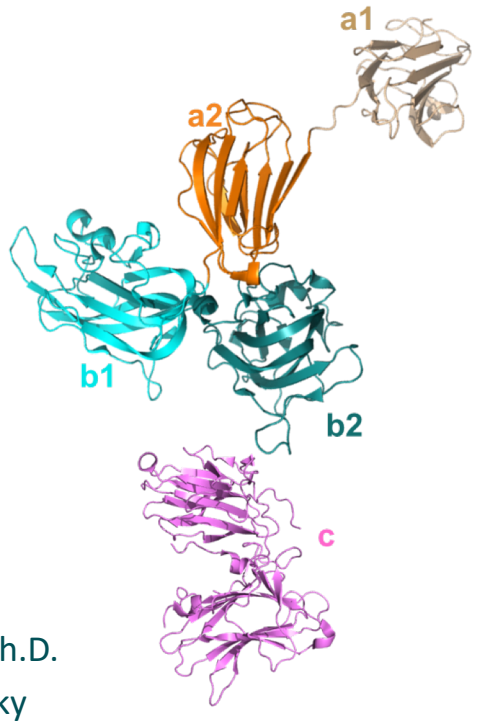
VEGF-C Blocker
α-NRP2-10

CCL21 Blocker(s)
Under evaluation

Leading World Researchers in NRP2

Diane Bielenberg, Ph.D.
Boston Children's Hospital
Harvard Medical School

David Briscoe, MB CHB.
Harvard Medical School



Kaustubh Datta, Ph.D.
University of Nebraska Medical Center

Arthur Mercurio, Ph.D.
University of Massachusetts
Medical School

Michael Muders, M.D., Ph.D.
Oncology, University of Bonn Medical Center

Craig Vander Kooi, Ph.D.
University of Kentucky

Robert M. Gemmill, Ph.D.
Medical University of South Carolina

tRNA Synthetases

A Potential New Class of Medicine

CSL Behring Collaboration to Identify New IND Candidates

Goal	• Identify new IND candidates from up to four tRNA synthetases from aTyr's pipeline (non-HARS derived)
Terms	• CSL to fund all R&D costs • aTyr eligible for up to $17m in option fees if CSL Behring advances all four programs ($4.25m per synthetase program) • CSL has an option to negotiate licenses for worldwide rights to each IND candidate that emerges from the collaboration
About CSL	• Leading global biotherapeutics company specializing in immunology, hematology and other rare and serious medical conditions • Employs >25,000 people globally, and delivers therapies to >60 countries
Status	• aTyr received first phase of funding totaling $630k, and of that recognized $278k of collaboration revenue through Q3 2019

aTyr

Value Drivers

Translating Novel Biology into First-in-Class Therapeutics

- ✓ Platform of proprietary new biology
- ✓ ATYR1923 in clinic for interstitial lung disease
  - Novel MOA for ILD
  - Demonstrated effect in multiple ILD animal models
  - Phase 1b/2a clinical study in pulmonary sarcoidosis enrolling in US
  - Positive interim safety data reported December 2019
  - Kyorin collaboration for ILD in Japan with upfront and potential milestone payments totaling $175m
- ✓ Supported by top tier investors
- ✓ Cash, cash equivalents, and investment at $38.1m as of 9/30/2019
  - Does not include $8m upfront from Kyorin or $18m raised in equity offering

Thank You