2019 R&D Day

May 22, 2019 - New York

Welcome and Agenda

Beth DelGiacco | VP, Investor Relations

Forward-Looking Statements

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Safe Harbor: Certain statements contained in this presentation, other than present and historical facts and conditions independently verifiable at the date hereof, may constitute forward-looking statements. Examples of such forward-looking statements include those regarding our investigational product candidates and preclinical studies and clinical trials, and the status, plans, timing of expected data readouts and related presentations and related results thereof, including the design of our trials and the availability of data from them, the timing and achievement of our product candidate development activities, future results of operations and financial positions, including potential milestones, business strategy, plans and our objectives for future operations. When used in this presentation, the words "anticipate," "believe," "can," "could," "estimate," "expect," "intend," "is designed to," "may," "might," "will," "plan," "potential," "predict," "objective," "should," or the negative of these and similar expressions identify forward-looking statements. Such statements, based as they are on the current analysis and expectations of management, inherently involve numerous risks and uncertainties, known and unknown, many of which are beyond the Company's control. Such risks include, but are not limited to: the impact of general economic conditions, general conditions in the biopharmaceutical industries, changes in the global and regional regulatory environments in the jurisdictions in which the Company does or plans to do business, market volatility, fluctuations in costs and changes to the competitive environment. Consequently, actual future results may differ materially from the anticipated results expressed in the forward-looking statements. In the case of forward-looking statements regarding investigational product candidates and continuing further development efforts, specific risks which could cause actual results to differ materially from the Company's current analysis and expectations include: failure to demonstrate the safety, tolerability and efficacy of our product candidates; final and quality controlled verification of data and the related analyses; the expense and uncertainty of obtaining regulatory approval, including from the U.S. Food and Drug Administration and European Medicines Agency; the possibility of having to conduct additional clinical trials; our ability to obtain and maintain intellectual property protection for our product candidates; and our reliance on third parties such as our licensors and collaboration partners regarding our suite of technologies and product candidates. Further, even if regulatory approval is obtained, biopharmaceutical products are generally subject to stringent on-going governmental regulation, challenges in gaining market acceptance and competition. These statements are also subject to a number of material risks and uncertainties that are described in the Company's filings with the U.S. Securities and Exchange Commission ("SEC"), including in argenx's most recent annual report on Form 20-F filed with the SEC as well as subsequent filings and reports filed by argenx with the SEC. The reader should not place undue reliance on any forward-looking statements included in this presentation. These statements speak only as of the date made and the Company is under no obligation and disavows any obligation to update or revise such statements as a result of any event, circumstances or otherwise, unless required by applicable legislation.

Agenda

8:30 AM:	**argenx Today: Building Leadership in Immunology** *Tim Van Hauwermeiren, CEO*
8:45 AM:	**ARGX-117: Renaissance in Complement - Therapeutic Potential of C2** *Karen Silence, Ph.D., Project Leader*
	Expert Insights from KOLs
10:00 AM:	**Q&A Session and Coffee Break**
10:20 AM:	**ARGX-118: Immunology Breakthrough in Airway Inflammation**
11:00 AM:	**Cusatuzumab Development Plan** *Wim Parys, M.D., CMO*
11:15 AM:	**argenx 2021: Global Commercial Vision** *Keith Woods, COO*
	Q&A Session and Closing Remarks

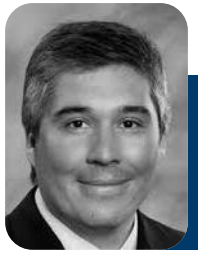
Today's Guest Speakers

Erik Hack, M.D., Ph.D.

Prof. Immunology
UMC Utrecht

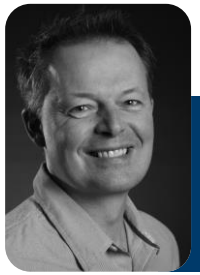
Ludo van der Pol, M.D., Ph.D.

Associate Prof. Neurology
UMC Utrecht



Rafael Villicana, M.D.

Medical Director, Kidney Transplantation
Loma Linda University



Bart Lambrecht, M.D., Ph.D.

Prof. Pulmonary Medicine, VIB
Ghent University

argenx Today:
Building Leadership in Immunology

Tim Van Hauwermeiren | Chief Executive Officer

argenx 2021:
Becoming an Integrated Immunology Global Biotech

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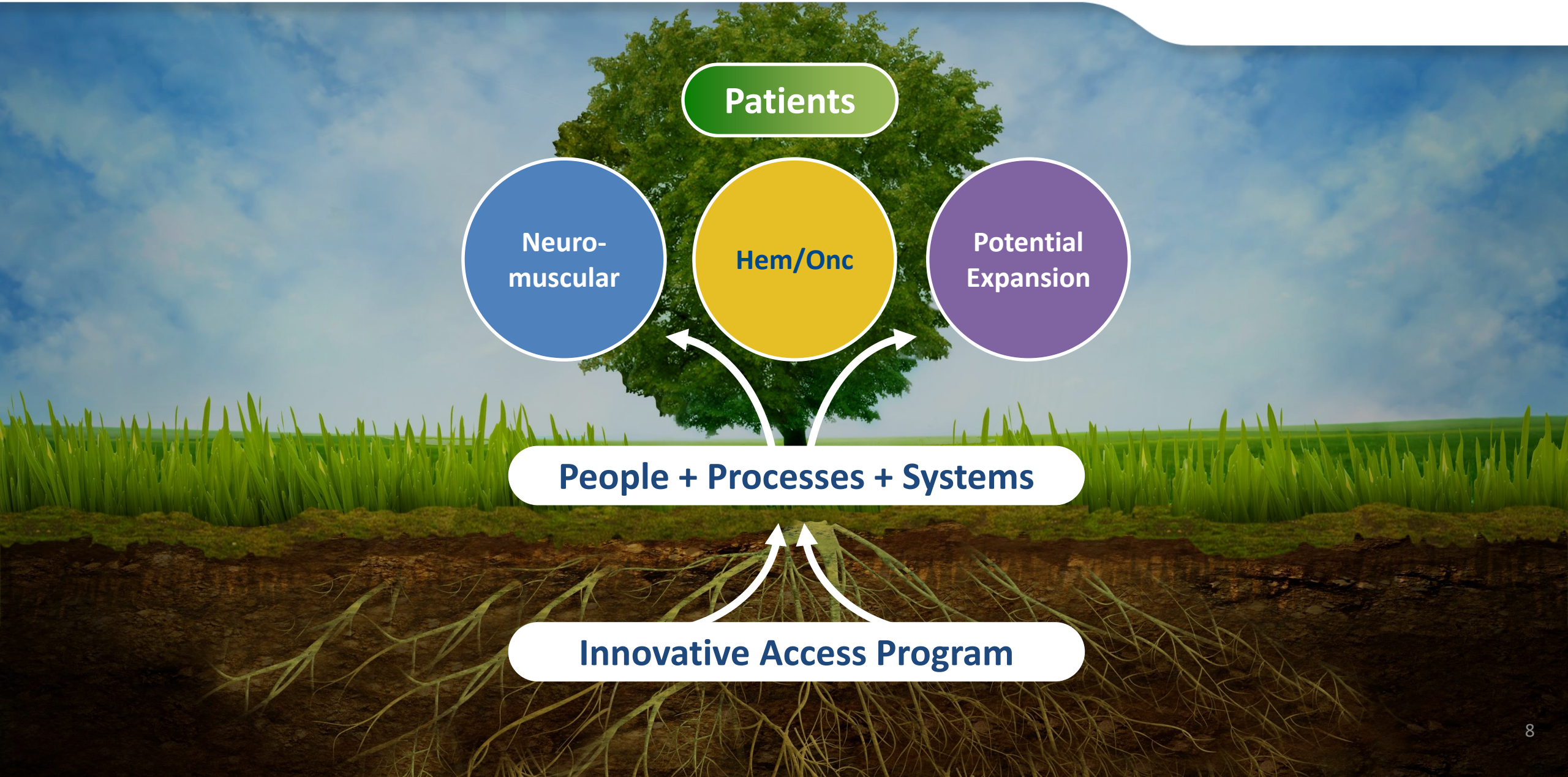
Becoming an Integrated Immunology Global Biotech



Appointment of Wim Parys, M.D. as Chief Medical Officer

- M.D. from KUL (Belgium)
- Janssen Research Foundation (BE): Reminyl® / Razadyne® for Alzheimer's disease
- Head of Development at Tibotec (BE): established Tibotec USA, HIV: Prezista®, Intelence® & Edurant®
- Head of Development of Janssen's Infectious Diseases and Vaccines: Incivo®, Olysio®/Sovriad®, TB: Sirturo®
- Head of R&D of Janssen's Global Public Health group: programs in HIV, TB, other mycobacterial infections, Dengue and malaria

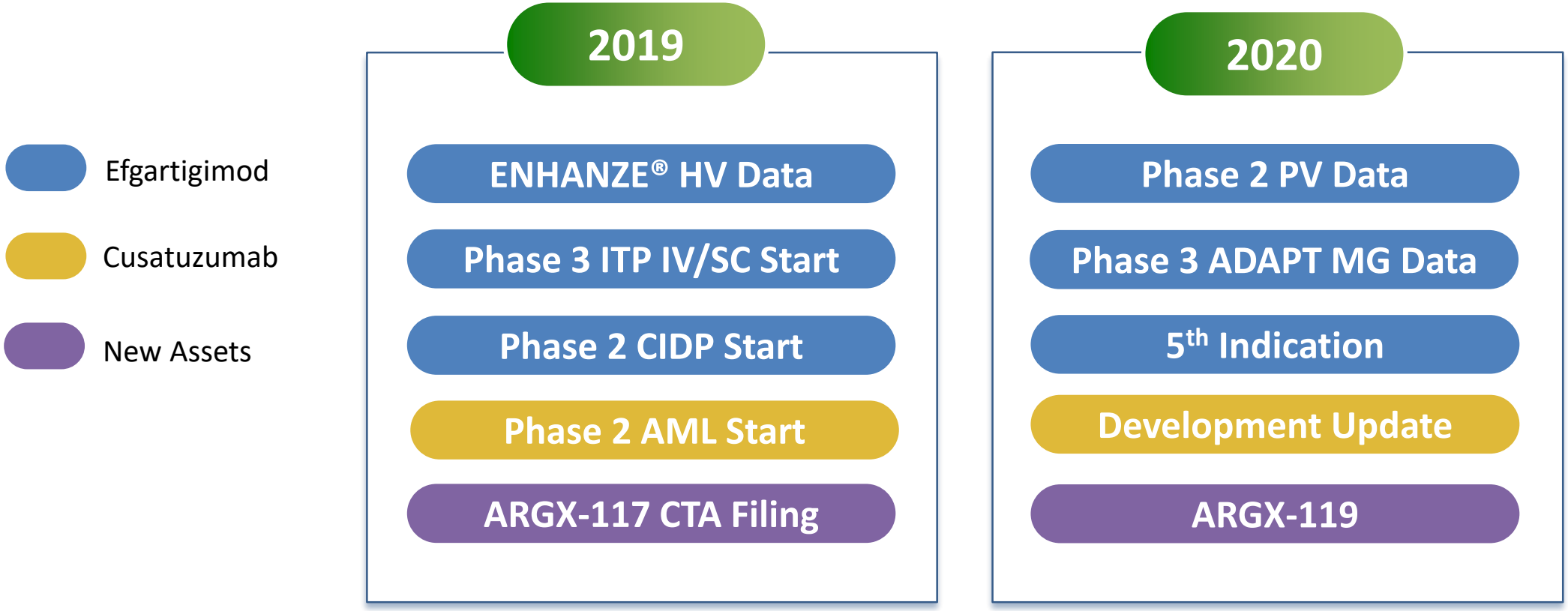
argenx Today: Building Leadership in Immunology

Late-stage immunology company	Two Phase 3 trials by end of 2019
Wholly-owned pipeline-in-a-product assets	Potential across multiple high-value indications
Proof-of-concept in two indications	Success in beachhead indications de-risks concept
Validating oncology collaborations	Maintained 50% of cusatuzumab commercial rights
Proven engine to grow pipeline	Innovative Access Program in action

Wholly-Owned Pipeline of Orphan Disease Candidates

Product Candidate	Target	Indication	Preclinical	Phase 1	Phase 2	Phase 3	BLA
ARGX-113 Efgartigimod	FcRn	Myasthenia Gravis (MG)					adapt myasthenia gravis study
		Immune Thrombocytopenia (ITP)					
		Pemphigus Vulgaris (PV)					
		Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)					
		ENHANZE® SC					
ARGX-110 Cusatuzumab	CD70	Acute Myeloid Leukemia (AML)				janssen	
ARGX-117	C2	Severe Autoimmunity IV/ENHANZE® SC					
ARGX-118	Galectin-10	Airway Inflammation					

Multiple Value-Creating Milestones Through 2020



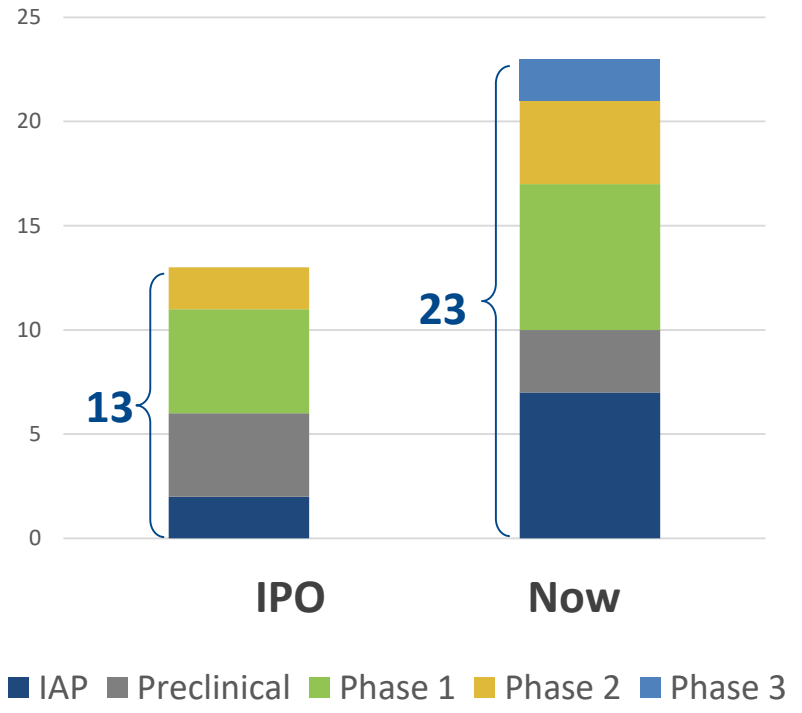
$1.1B in Cash; Funded Through 2021

Impressive Value Creation Since IPO

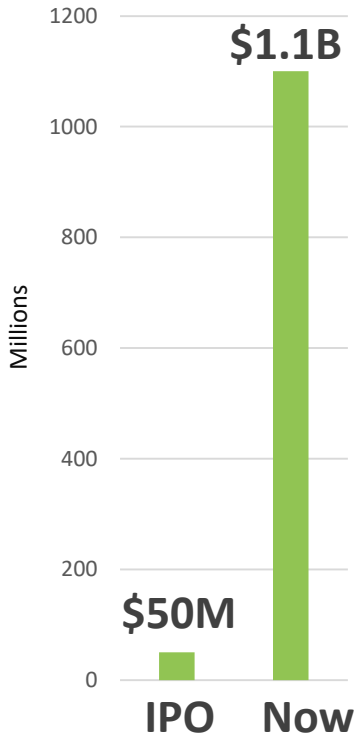
Accelerating & expanding development programs

Key Data

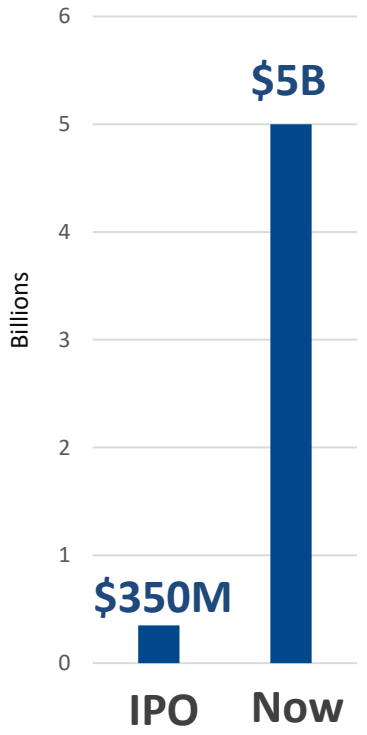
MG & ITP | Interim PV | AML



Well-capitalized to advance to the next level

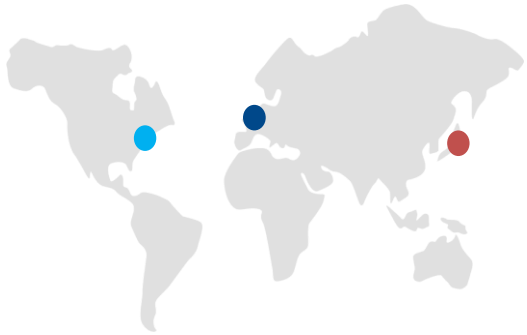


Cash



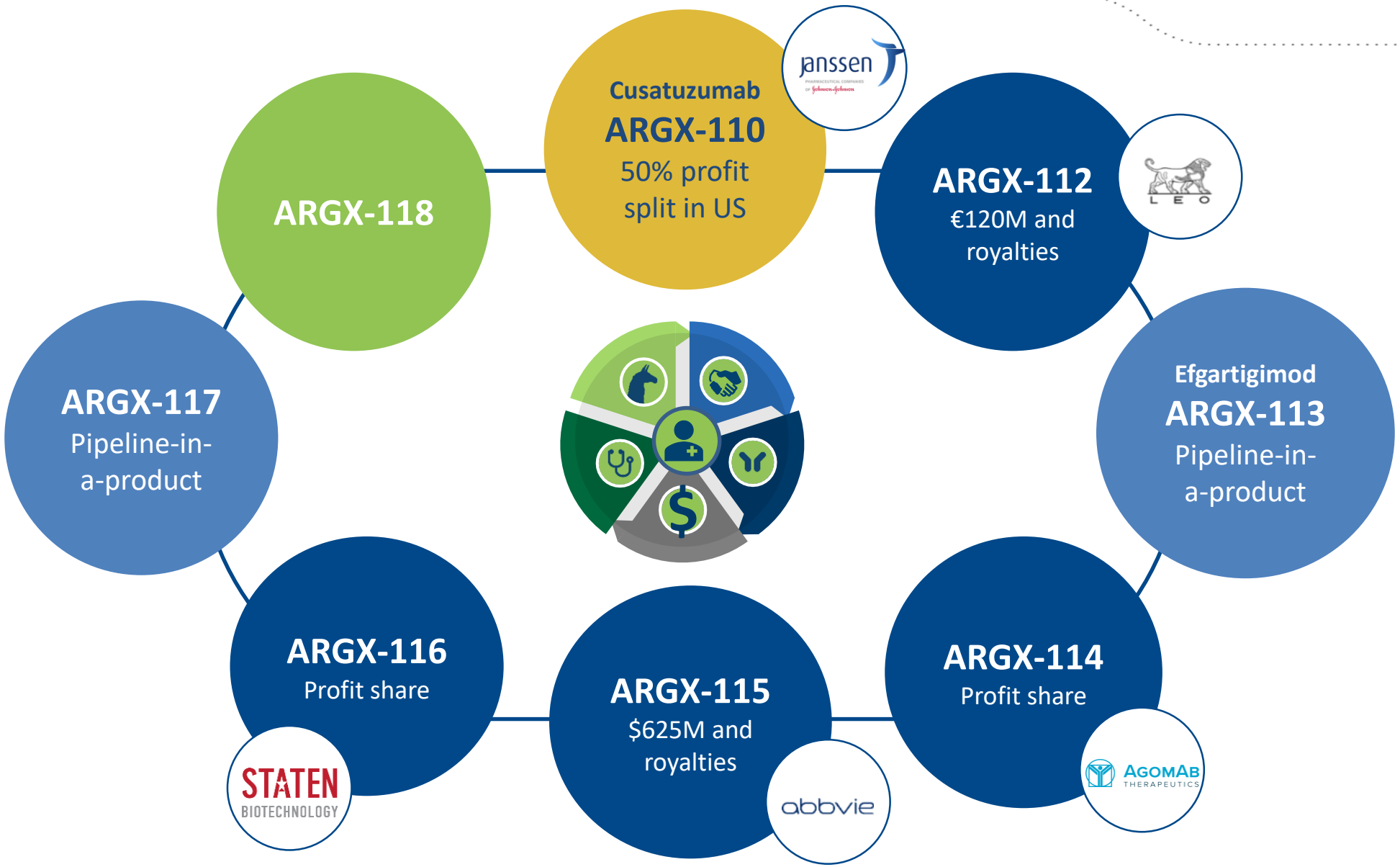
Market Cap

Global expansion



- **Ghent**
- **Boston** (2018)
- **Tokyo** (2019)

Serial Value Creation from Novel Targets



Innovative Access Program (IAP): How We Build Value

Accessing Novel Targets Through Collaboration

argenx

Antibody Expertise
SIMPLE Antibody™, NHance®, ABDEG™, POTELLIGENT®

Top Academic Institutions & Biotechs

Disease Biology Expertise
Texas, Bern, Utrecht, Louvain, Penn, Columbia, Torino, Institutes of Ludwig, de Duve, VIB, TAM

Co-creating first-in-class assets

WHOLLY-OWNED

ARGX-113
ARGX-110
(Co-developed Janssen)
ARGX-117
ARGX-118

PARTNERED

ARGX-115 abbvie
ARGX-112 LEO
ARGX-116 STATEN
ARGX-114 AGOMAB THERAPEUTICS

5-10 ongoing programs at any given time

Immunology Breakthroughs to Impact Patients' Lives

1

Novel Target Biology

- Academic collaboration
- First-in-class targets

2

Integrated Antibody Discovery Suite

- Meaningful differentiation
- Engineering excellence

5

Franchise Structure

- Neuromuscular and hem/onc
- Phase 3 studies ongoing in 2019

3

Rapid Pipeline Expansion

- Productive development engine
- One new pipeline asset per year

4

Maximum Value per Asset

- Pipeline-in-a-product strategy
- Driven by biological rationale

ARGX-117: Jumpstart Product Development

1
Novel Target Biology
Complement component C2
UMC Utrecht: Erik Hack

2
Integrated Antibody Discovery Suite
Sweeping antibody

3
Rapid Pipeline Expansion
Exercised option to bring ARGX-117 in house

Halozyme
Option exercised for C2 ENHANZE® SC technology

4
Maximum Value per Asset
Severe autoimmune diseases

5
Franchise Structure
CTA in 4Q 2019
Well-positioned within argenx franchises

ARGX-118: Build from Target Validation

1

Novel Target Biology

Galectin-10
VIB: Bart Lambrecht

2

Integrated Antibody Discovery Suite

Crystal-dissolving SIMPLE Antibody™

5

Preclinical Development

Lead optimization

3

Rapid Pipeline Expansion

Exercised option to bring ARGX-118 in house

4

Maximum Value per Asset

Range of immunology indications

IAP Enables Full Spectrum of Antibody Creation

Early Target Validation

Power of SIMPLE Antibody™ technology

→ Charcot-Leyden Crystal dissolving antibodies

Unravelling novel airway inflammation biology

→ Galectin-10 first novel airway inflammation target in decades

ARGX-118

Jumpstart Product Development

Power of NHance® technology and engineering know-how

→ Turn unique mouse V-regions into highly differentiated product candidate

Leveraging unique insights in complement disease biology

→ Pipeline-in-product opportunity

ARGX-117

ARGX-117: Renaissance in Complement Therapeutic Potential of C2

Erik Hack, M.D., Ph.D. | Prof. UMC Utrecht
Karen Silence, Ph.D. | Project Leader argenx

Professor Erik Hack, M.D., Ph.D., UMC Utrecht

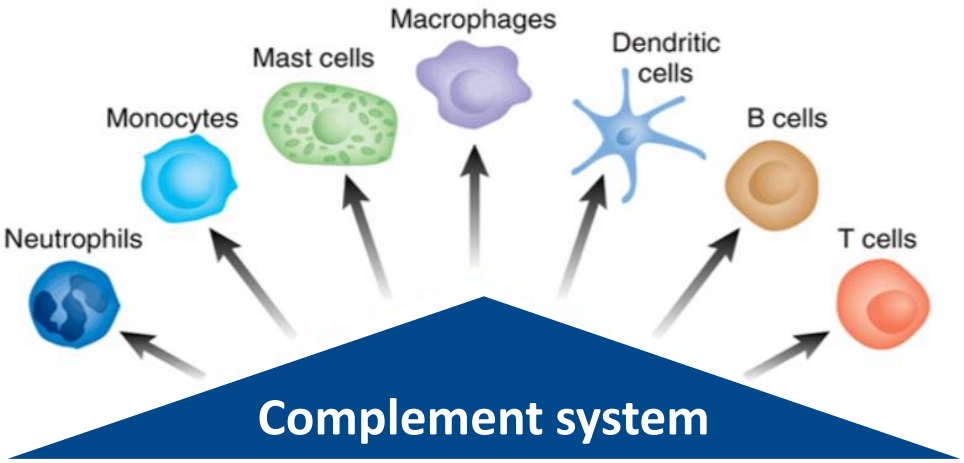
- Prof. Immunology at UMC Utrecht
- Sanquin, Head of Immunopathology
- Crucell, Head of Protein Biologics Group
- Research interests: role of inflammation and coagulation in experimental and human diseases
- Lead development of Cinryze® and Ruconest® (C1-inhibitor in angioedema)
- Co-authored over 500 peer-reviewed papers; more than 23,000 citations

Therapeutic Renaissance in Complement

- Growing number of immune-mediated and inflammatory diseases that involve complement
- ‘07 — Eculizumab approval in 2007 validated concept of therapeutic complement intervention
- 20+ — 20+ clinical-stage drug candidates targeting various stages of complement cascade
- ↔ — No one-size-fits-all; different complement-driven diseases require different points of intervention

Complement: Cornerstone of Powerful Innate Defense System

Defense & Surveillance



Innate	Disposal	Adaptive
• Opsonization • Lysis • Chemotaxis • Inflammation • Cell activation	• Clear immune complexes and apoptotic cells	• Augment Ab response • Promote T-cells • Eliminate self-reactive B-cells • Enhance immune memory

Clinical Complications

1 **Excessive activation**

↓

inflammation

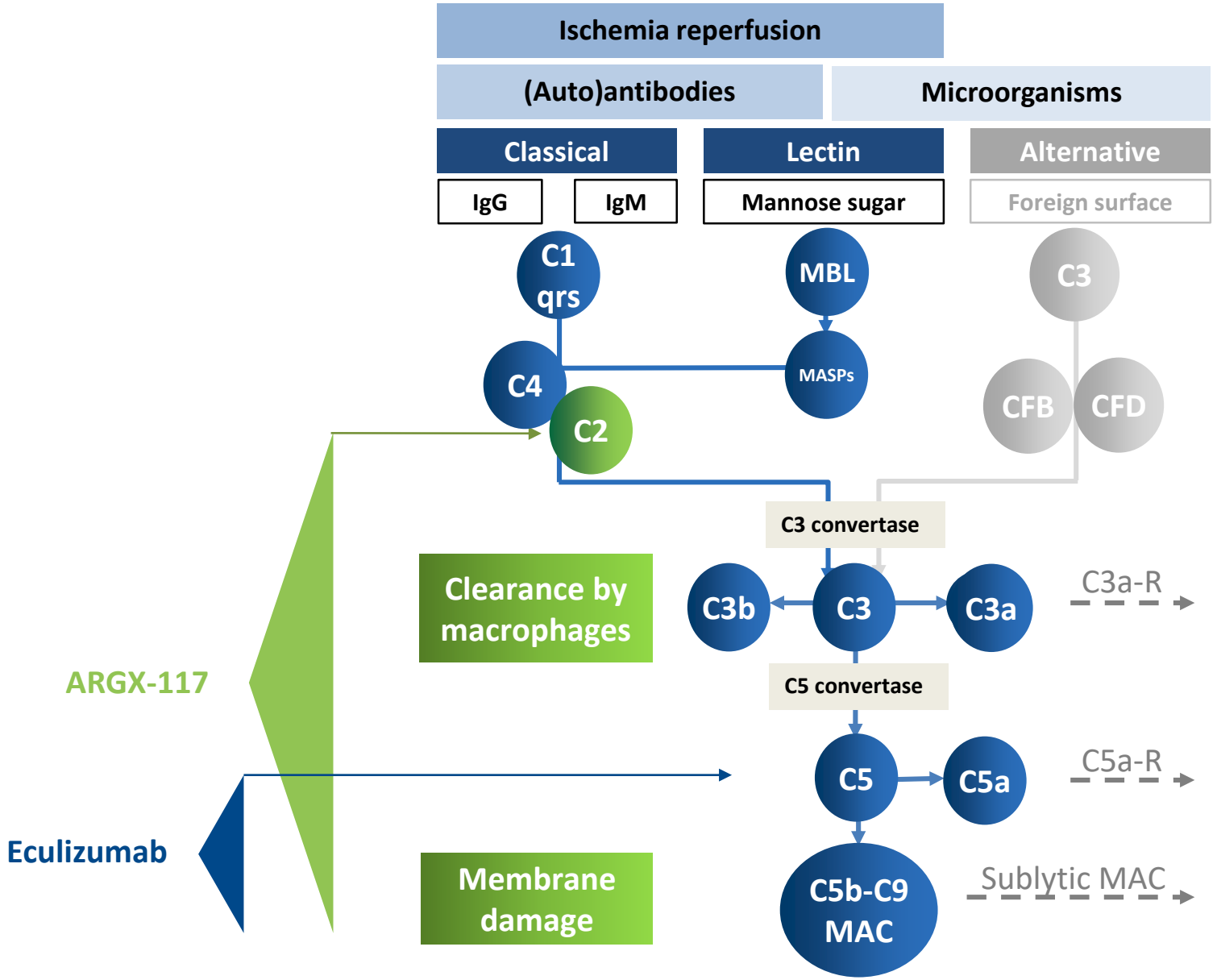
2 **Too little activation**

↓

risk for infections and lupus

Source: Katie Ris-Vicari

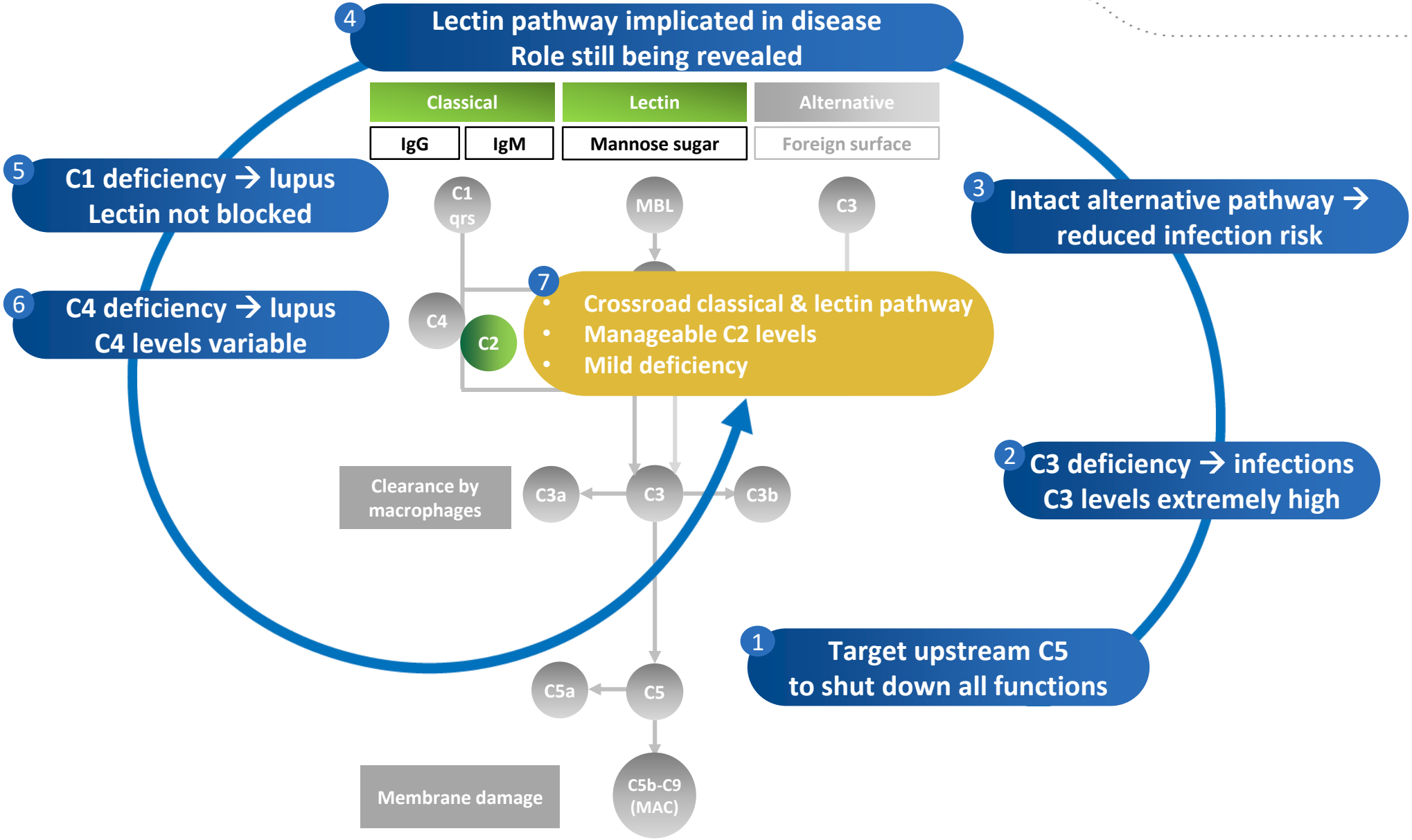
Targeting C2 Shuts Down Effector Functions Upstream



Inflammation

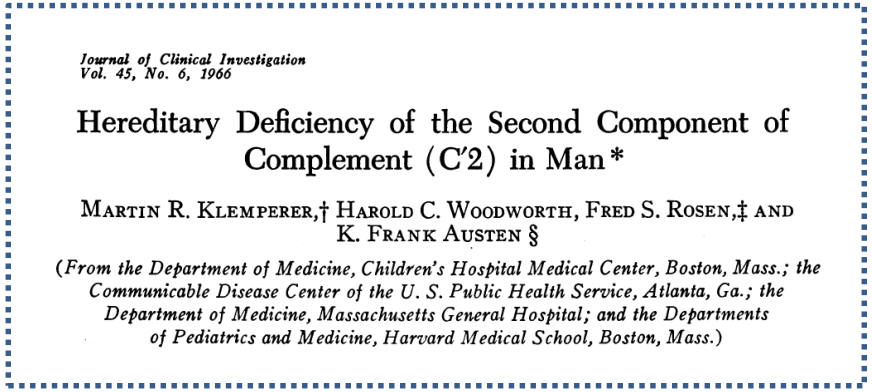
- Mast cell degranulation
- PMN chemotaxis
- PMN degranulation
- Expression endothelial adhesion molecules
- Increased endothelial permeability
- Tissue factor expression
- Vasodilatation
- Cytokine production
- Bronchial smooth cell contraction

Targeting C2 Preserves Key Complement Functionality



Potential Best-in-Class Tolerability Profile

C2 deficiency observed in healthy people



Journal of Clinical Investigation
Vol. 45, No. 6, 1966

Hereditary Deficiency of the Second Component of Complement (C'2) in Man*

MARTIN R. KLEMPERER,† HAROLD C. WOODWORTH, FRED S. ROSEN,‡ AND K. FRANK AUSTEN §

(From the Department of Medicine, Children's Hospital Medical Center, Boston, Mass.; the Communicable Disease Center of the U. S. Public Health Service, Atlanta, Ga.; the Department of Medicine, Massachusetts General Hospital; and the Departments of Pediatrics and Medicine, Harvard Medical School, Boston, Mass.)

C2 deficiency has mildest phenotype among classical pathway deficiencies:

- No symptoms in 50% of cases
- Recurrent infections, particularly in childhood
- SLE/SLE-like condition in 10-30%

Prevalence of homozygous C2 deficiency: 1 in 10-20,000 persons (Caucasians)

Longstanding acquired C2 deficiency in hereditary angioedema (HAE) not associated with increased infection risk

Innovative Access in Action

argenx

Antibody Expertise

Broteio Pharma | UMC Utrecht

Disease Biology Expertise

Antibody optimization: longer half-life and sweeping

Focus on the science: good science leads to good development

Open communication; fair economics; aligned goals

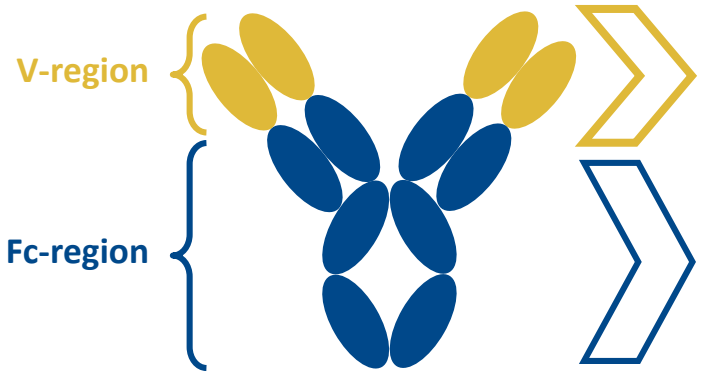
Next-generation complement therapeutics require **careful target selection**, patient population selection, **improved PK properties** (high plasma levels and rapid turnover of complement proteins) and **safety considerations**

Karen Silence, Ph.D., Project Leader

- **Ph.D. at Center for Molecular and Vascular Biology, University of Leuven, Belgium (Désiré Collen, M.D., Ph.D.)**
  - Preclinical development of staphylokinase – fibrinolytic drug
- **Postdoc at VIB (Belgium)**
  - Establishment of Ablynx NV
- **First employee at Ablynx (now Sanofi)**
  - Project leader caplacizumab - approved for acquired thrombotic thrombocytopenia purpura (aTTP)
  - Director pharmacology
- **One of the first employees at argenx**
  - Project leader cusatuzumab – partnered with Janssen Cilag
  - Project leader for ARGX-117

Innovative Access in Action

ARGX-117



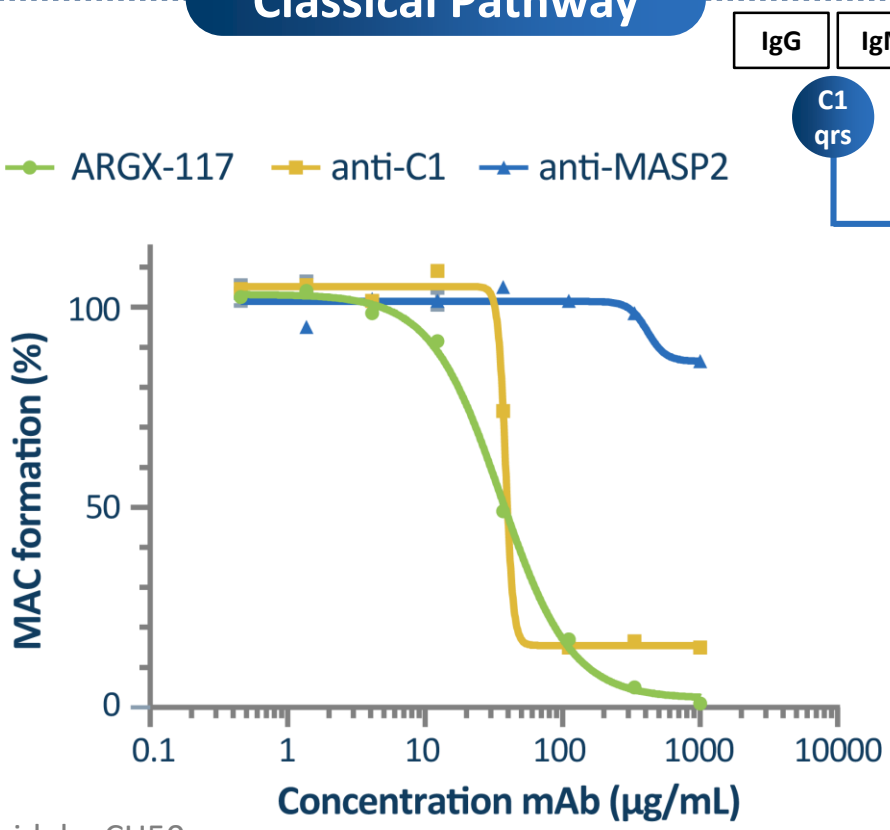
argenx | **Broteio Pharma | UMC Utrecht**

Antibody Expertise | Disease Biology Expertise

- BRO-2 antibody prototype optimized
- Translational data generated through UMC Utrecht collaboration
- True collaboration yielded unique end product

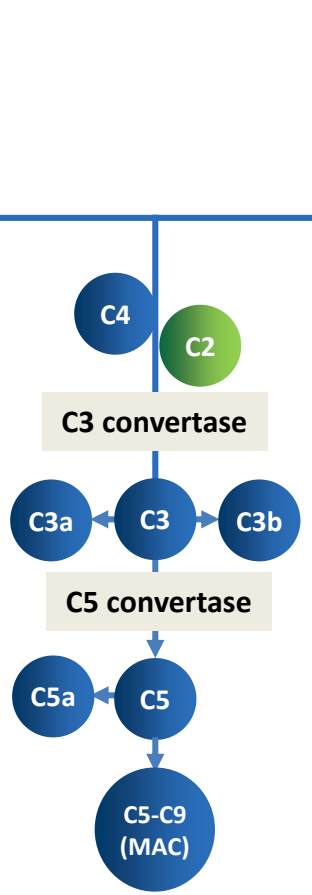
ARGX-117: Potently Blocks Classical and Lectin Pathways

Classical Pathway

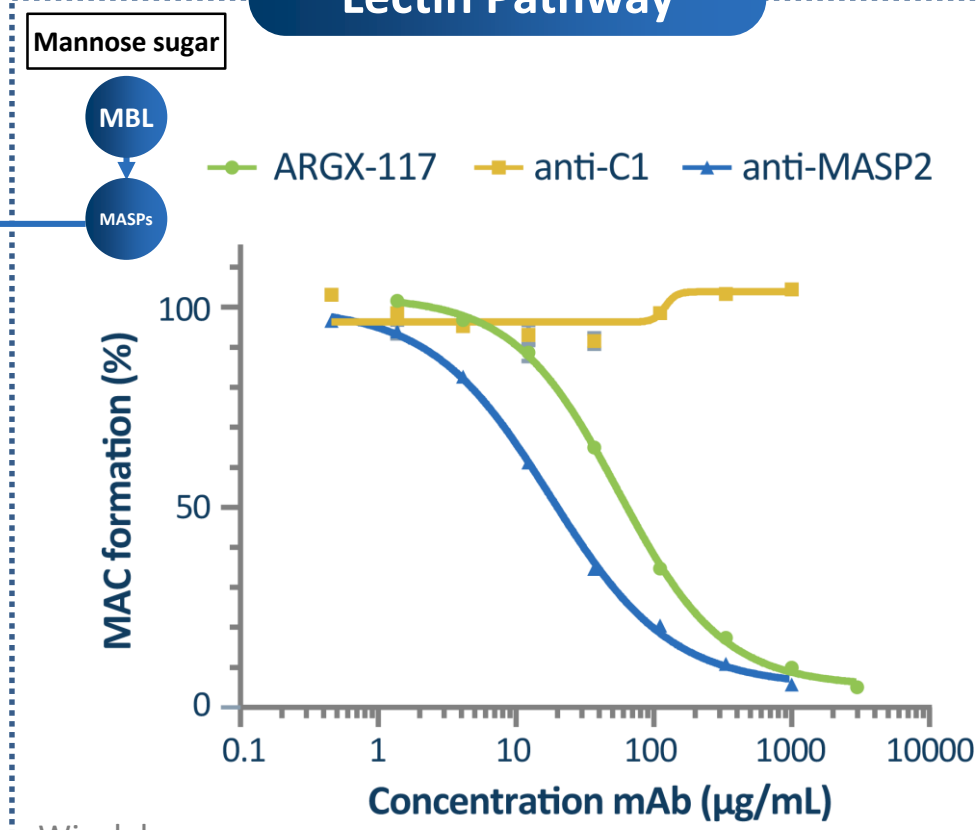


Quidel – CH50

EC50 (µg/mL)		
ARGX-117	anti-C1	anti-MASP-2
36	~39	>1000



Lectin Pathway



Wieslab

EC50 (µg/mL)		
ARGX-117	anti-C1	anti-MASP-2
55	>1000	18

ARGX-117: V and Fc Regions Act in Concert to Sweep C2

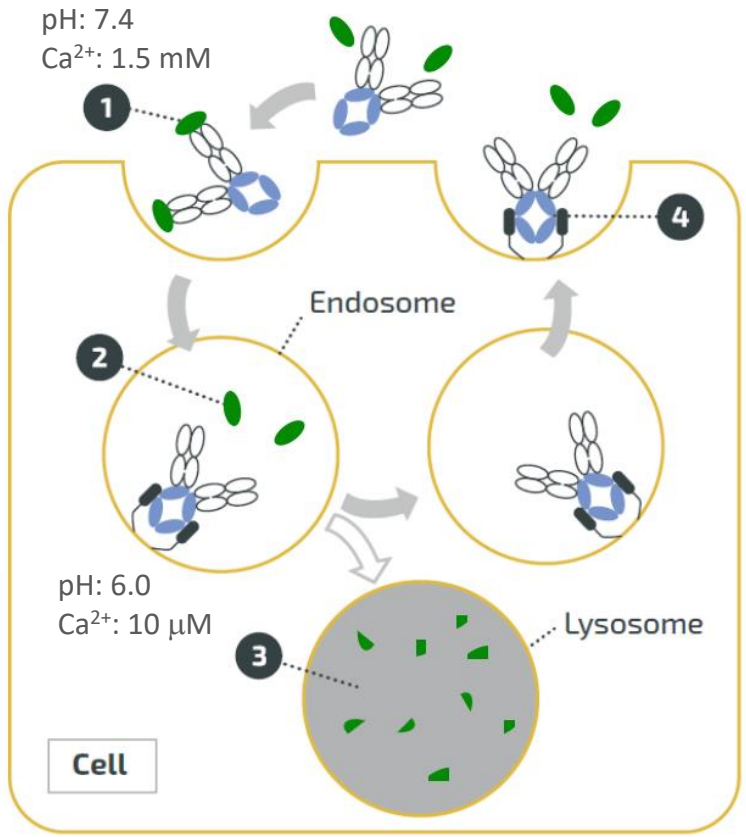
1 ARGX-117 binds C2 in circulation

2 ARGX-117/C2 complex taken up by pinocytosis entering endosome;

ARGX-117 releases C2 at lower pH in endosome

3 C2 is degraded in lysosome

4 ARGX-117 remains bound going back into circulation to bind to new C2 because of NHance® mutations

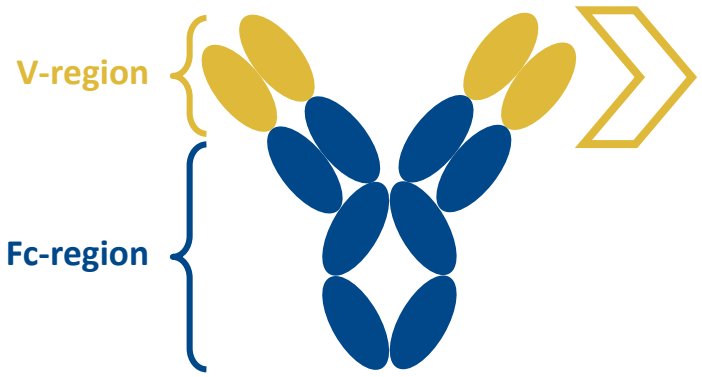


FcRn

ARGX-117

C2

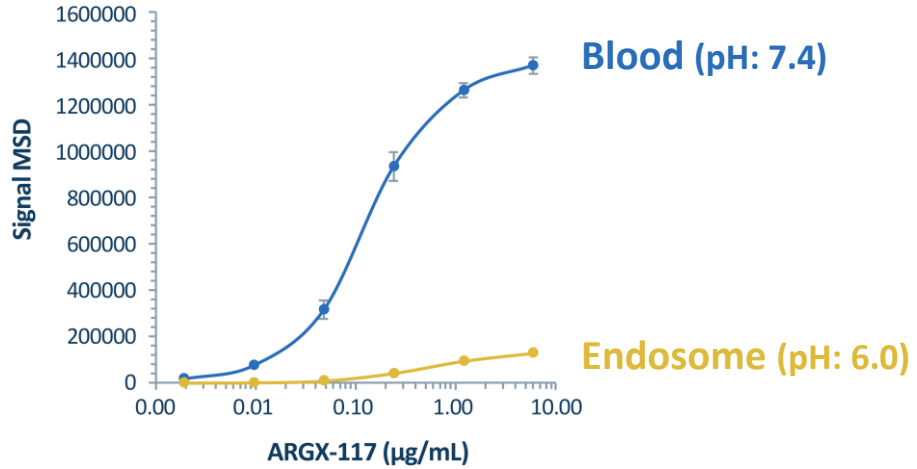
ARGX-117: Intrinsic pH and Ca^{2+} Dependent Target Binding



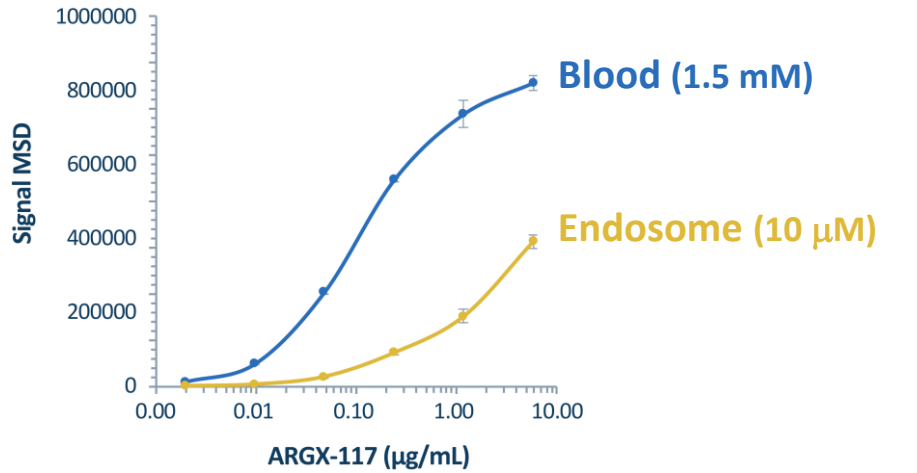
V-regions:

- 97% human identity
- Affinity ~3 nM

pH dependent C2 binding

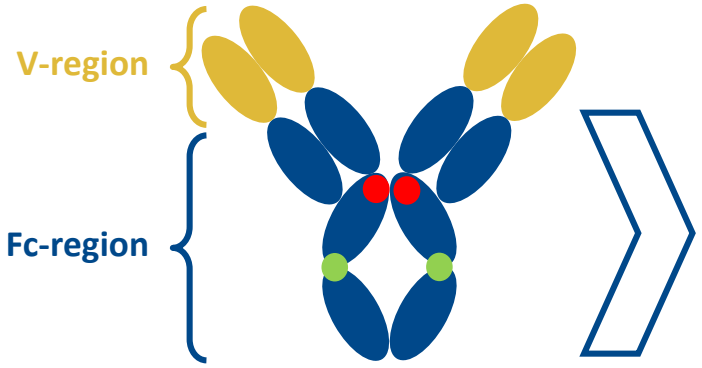


Calcium dependent C2 binding



Differential binding in blood and endosome

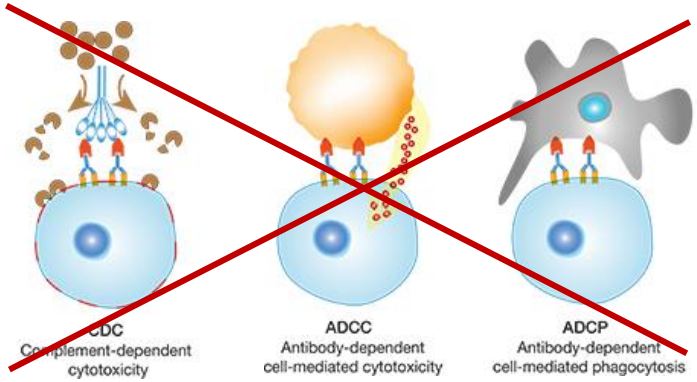
ARGX-117: Increase in Endosomal FcRn Binding and Knockout



Fc-region:

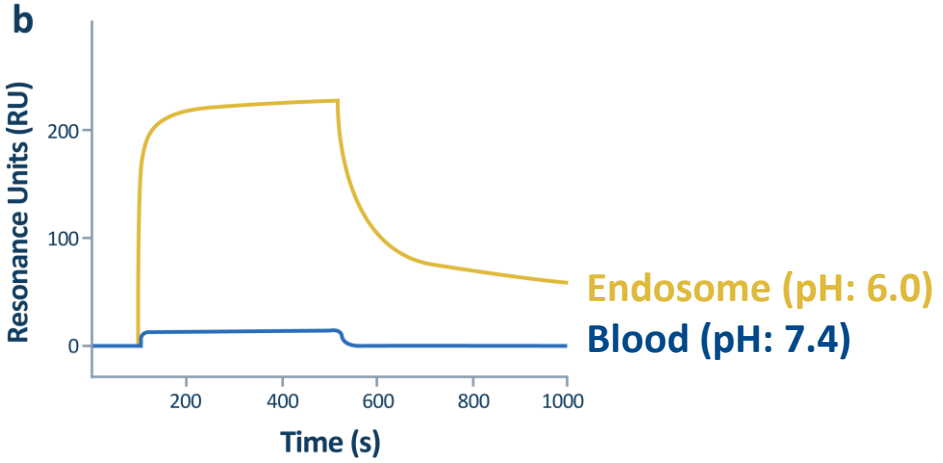
- Human IgG1
- LALA mutations knock out effector functions (●)
- NHance® modulates FcRn binding (●)

LALA mutations knock out effector functions



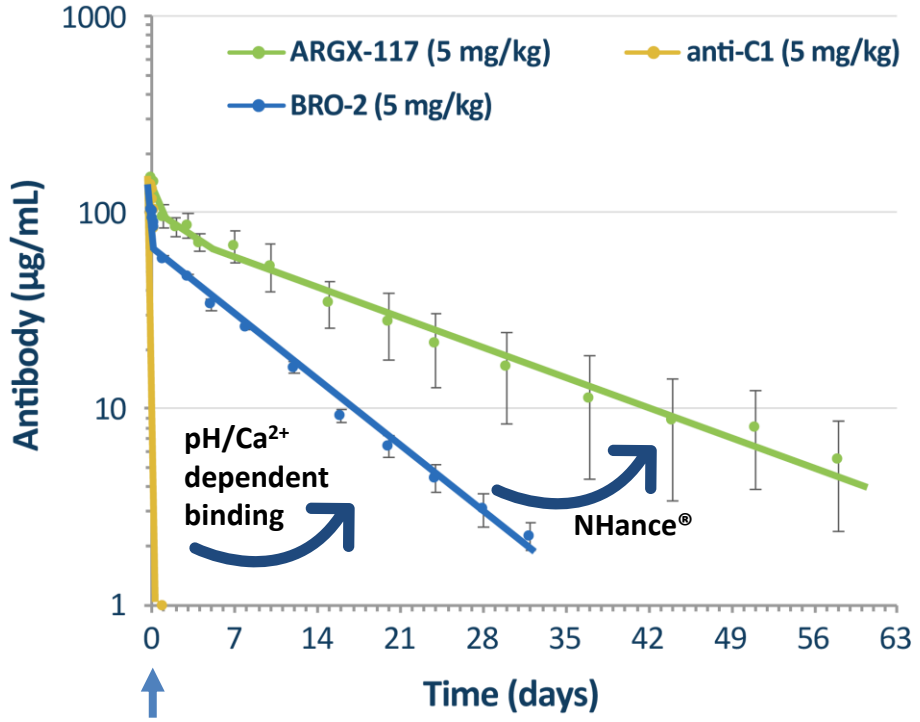
From Journal of Virology, 2001: 75: 12161–12168

NHance® modulates FcRn binding



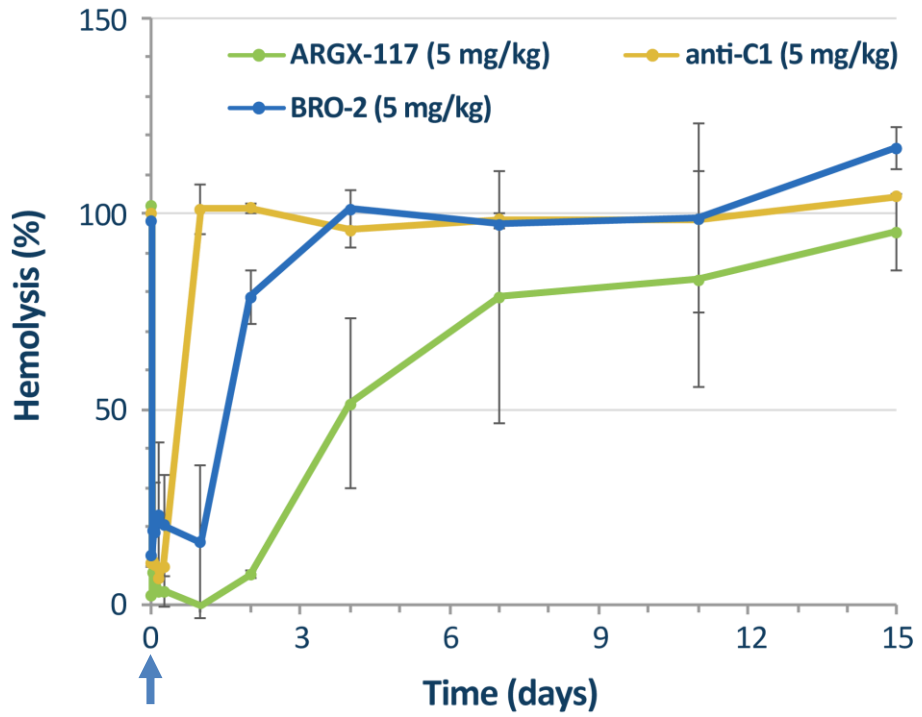
ARGX-117: Longer Half-life and Differentiated PD Effect

Pharmacokinetics



Prolonged half-life…

Pharmacodynamics

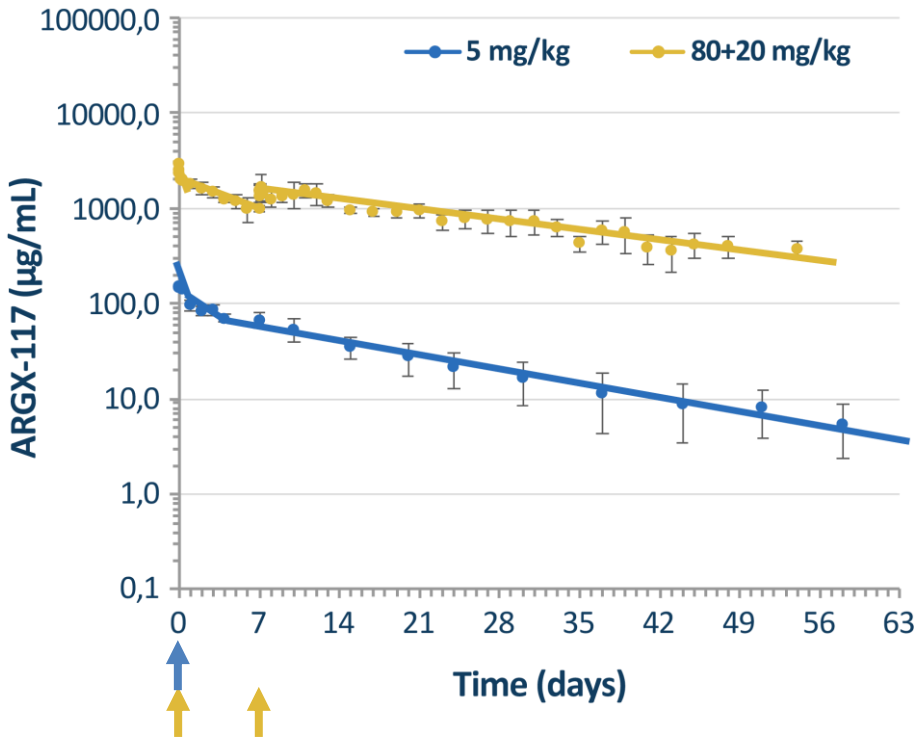


…results in extended PD profile

Cynomolgus monkey data

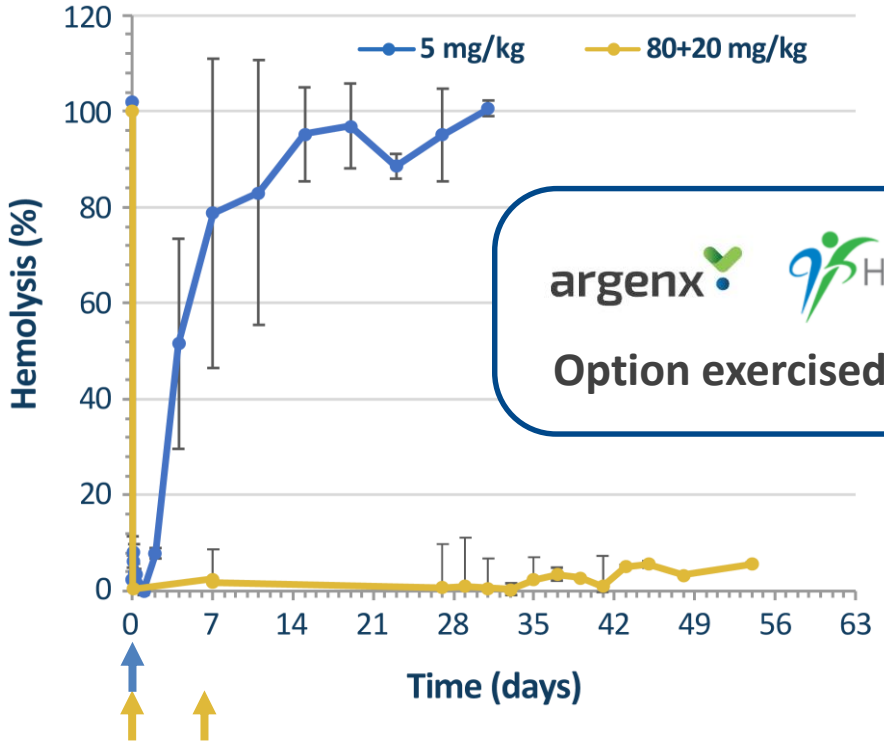
ARGX-117: Dosing Optionality

Pharmacokinetics



Half-life ARGX-117: 2-3 weeks

Pharmacodynamics



argenx Halozyme

Option exercised for C2

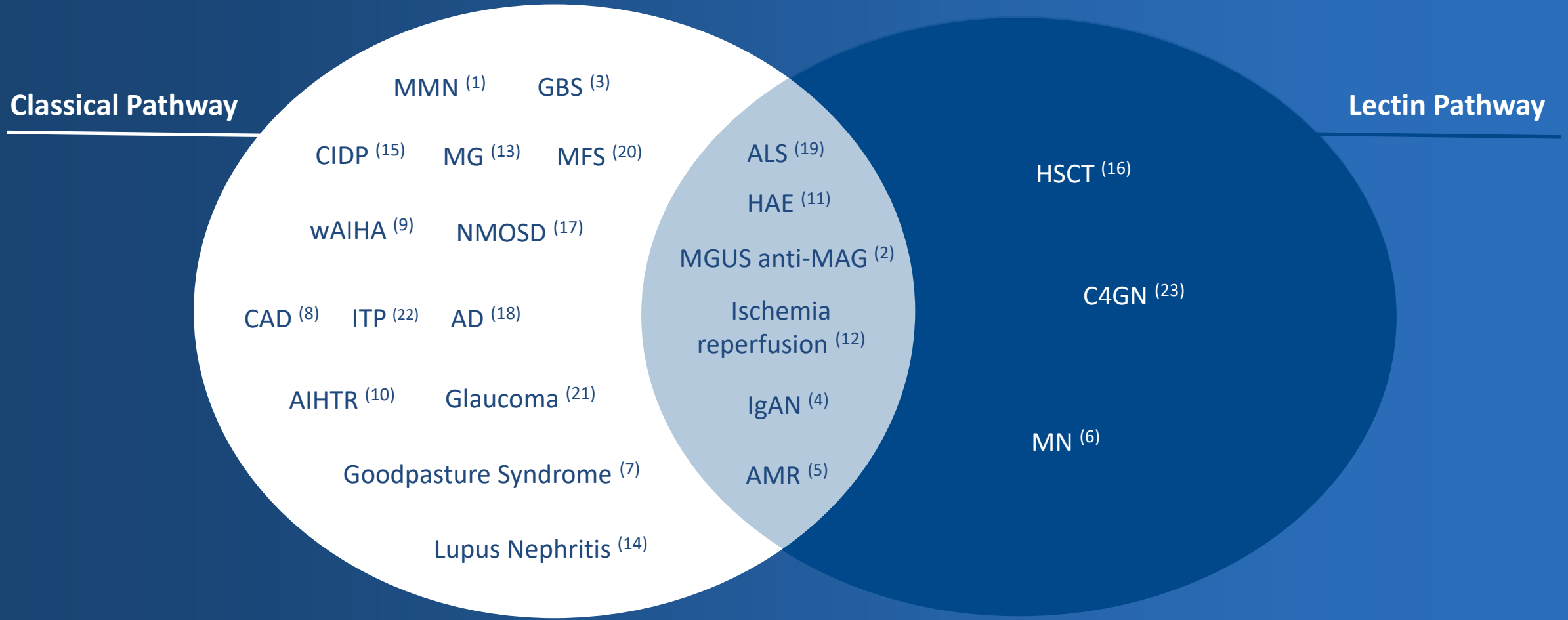
Blocking for 2 months after repeat dosing

Cynomolgus monkey data

C2 levels cynomolgus monkey = 4x human

Classical and Lectin Pathway Autoimmune Diseases

Complement Mediated Diseases



1. Vlam et al, 2015, Neurology 2. Stork et al, 2016, J of Neuroimmunol 3. Goodfellow et al, 2016, Nat Rev 4. Medjeral-Thomas et al, 2018, Kidney Int report 5. Montgomery et al, 2016, Am Soc of Transplant 6. Couser, 2017, Clin J Am Soc Nephrol 7. Greco et am, 2015, Autoimmun Rev 8. Jäger et al, 2018, Blood 9. Berentsen et al, 2015, Transfus Med Hemother 10. Sharp et al, 2014, Front in Immunol 11. Csuka et al, 2017, Mol Immunol 12. Gorsuch et al, 2012, Immunobiol 13. Howard, 2018, Ann NY Acad Sci 14. Bao et al, 2015, Kidney Dis 15. Querol et al, 2017, Scientific Reports 16. Swierzko et al, 2018, Front in ImmunolG 17. Yoshikura et al, 2017, J Neuroimmunol 18. Morgan, 2018, Semin Immunopathol 19. Kjaeldgaad et al., 2018, Mol Immunol 20. Lehmann et al., 2008, Brain 21. Williams e al., 2016, Mol. Neurodegeneration 22. Peerschke et al., 2016, Br J Haematol 23. Sethi et al., 2016, Am J of Kidney Diseases

ARGX-117: Pipeline-in-a-Product Opportunity

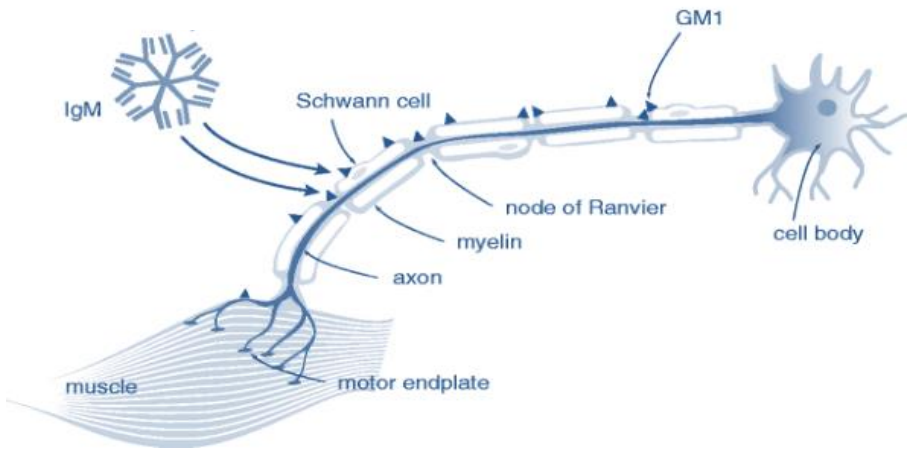
Classical and Lectin Pathway-Mediated Severe Autoimmune Diseases

Solid Biology Rationale

Feasible for Biotech
Orphan potential, economically viable, efficient clinical & regulatory pathway

- Neuromuscular
- Hem/Onc
- Kidney

Multifocal Motor Neuropathy (MMN)



- Monoclonal plasma cells produce **IgM** against **GM1**
- **Classical pathway** of complement activated
  - Demyelination/ disruption Schwann-cell-axolemma junctions
  - Displacement of ion channel clustering
  - Disturbing membrane integrity
- Results in loss of strength

Classical (IgM mediated) pathway activation

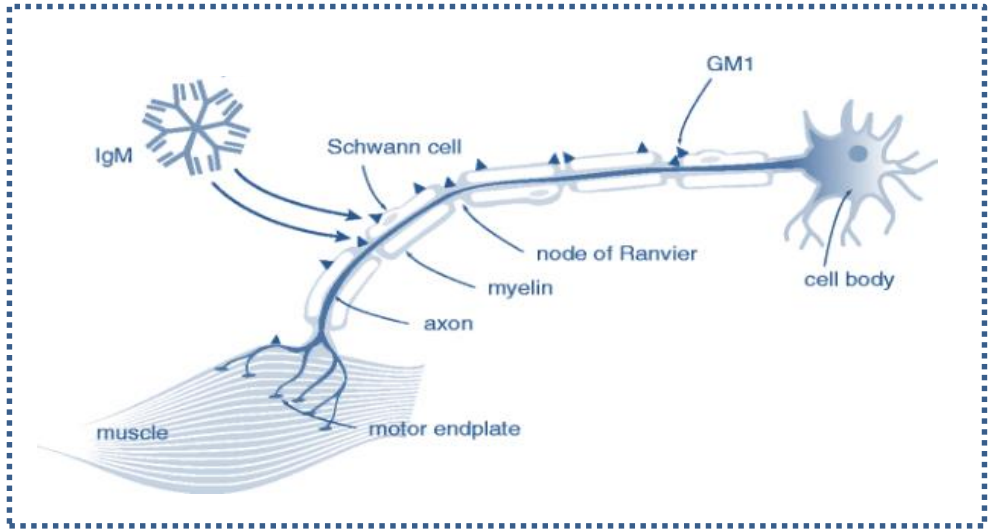
IVIg at high doses is effective

Eculizumab in MMN did not show benefit on top of IVIg

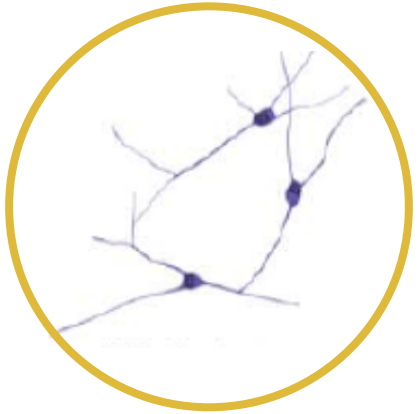
Hypothesis:
Other mechanisms must be in play which are not driven by MAC

J Clin Immunology, 2010: 30: 79–83
J Peripheral Nervous System, 2011: 16: 84-91

MMN: ARGX-117 Indication Selection



Motor neurons derived from pluripotent stem cells



Schwann cells

Complement activation using MMN patient-derived IgM

IgM

Serum

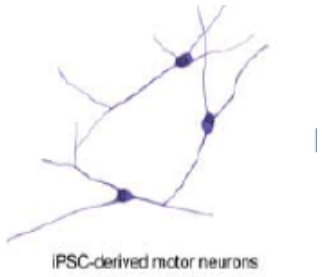
Complement active serum

Ann Neurol, 2016: 80: 71-88

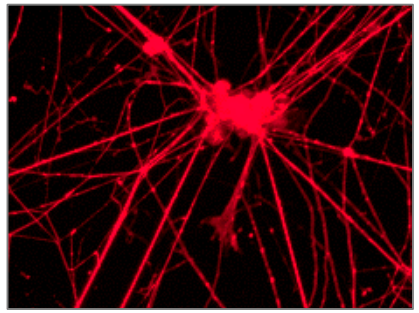
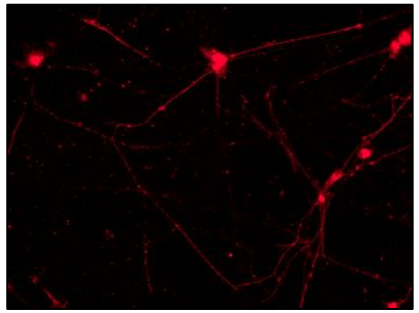
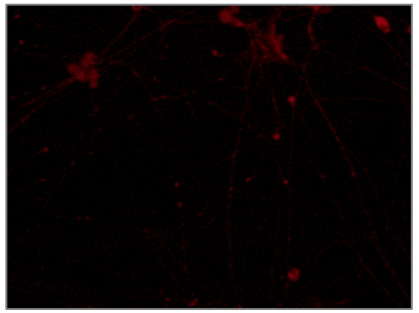
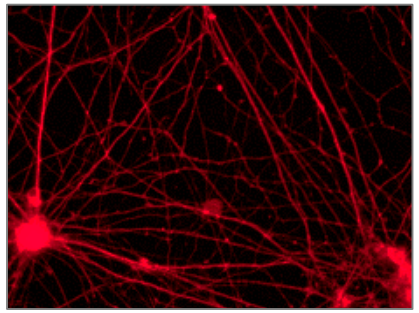
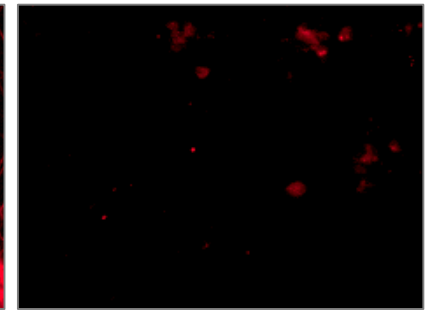
Biology Rationale:
C2 Critical for Complement Activation Induced by IgM in MMN

Serum MMN Patient

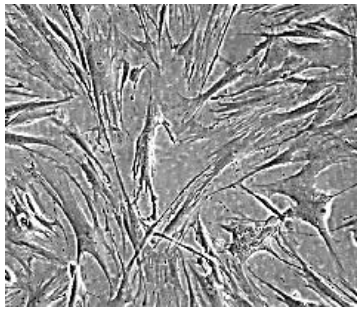
Motor neurons



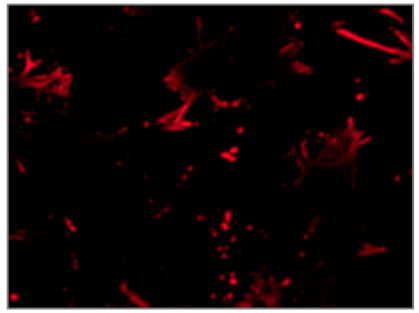
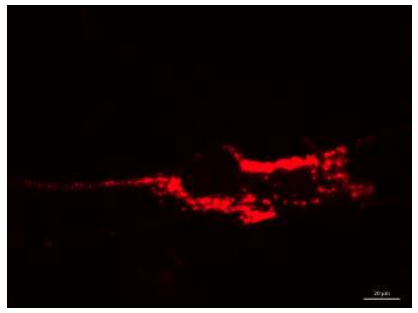
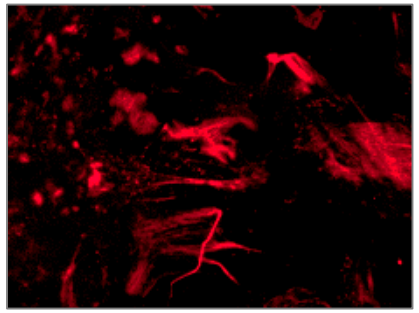
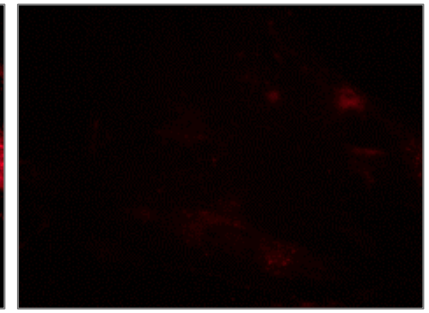
C3 deposition

Isotype control	IVIg	C2 depleted serum	C2 depleted serum + 100% C2	ARGX-117
				

Schwann cells



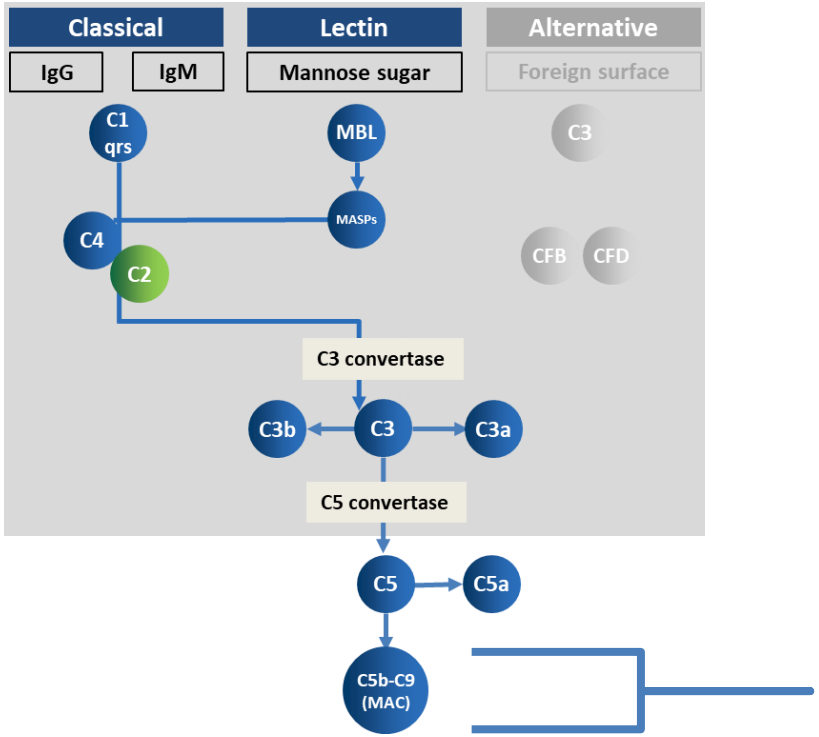
C3 deposition

C3 activation confirmed MOA in MMN - ARGX-117 blocks C3 deposition

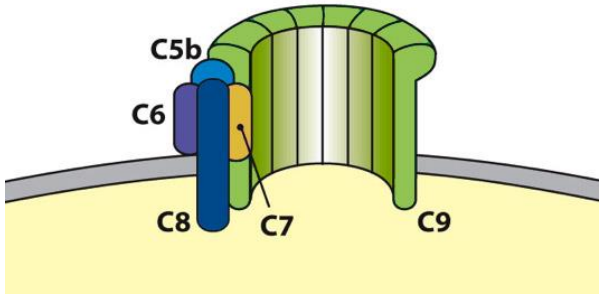
C3 Activation Confirmed MOA of IgM in MMN

No MAC formation due to high CD59 expression - complement activation upstream of C5

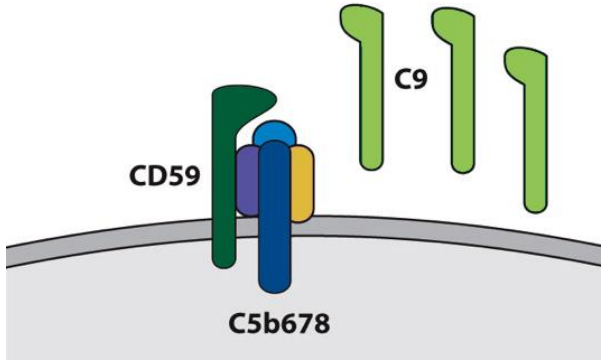


High CD59 expression inhibits MAC formation

C5-C9 activation: assembly of MAC complex



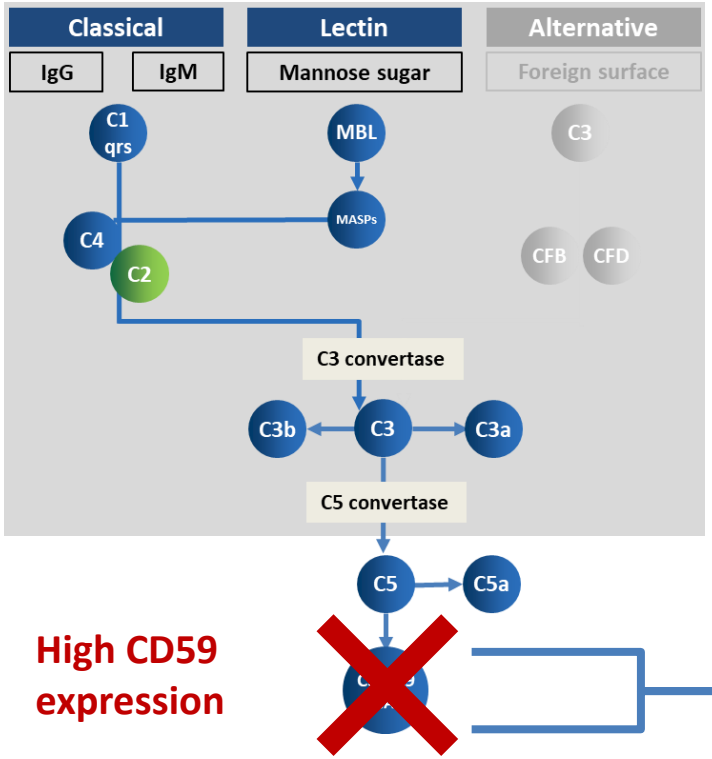
CD59 prevents recruitment of C9 – no MAC formation



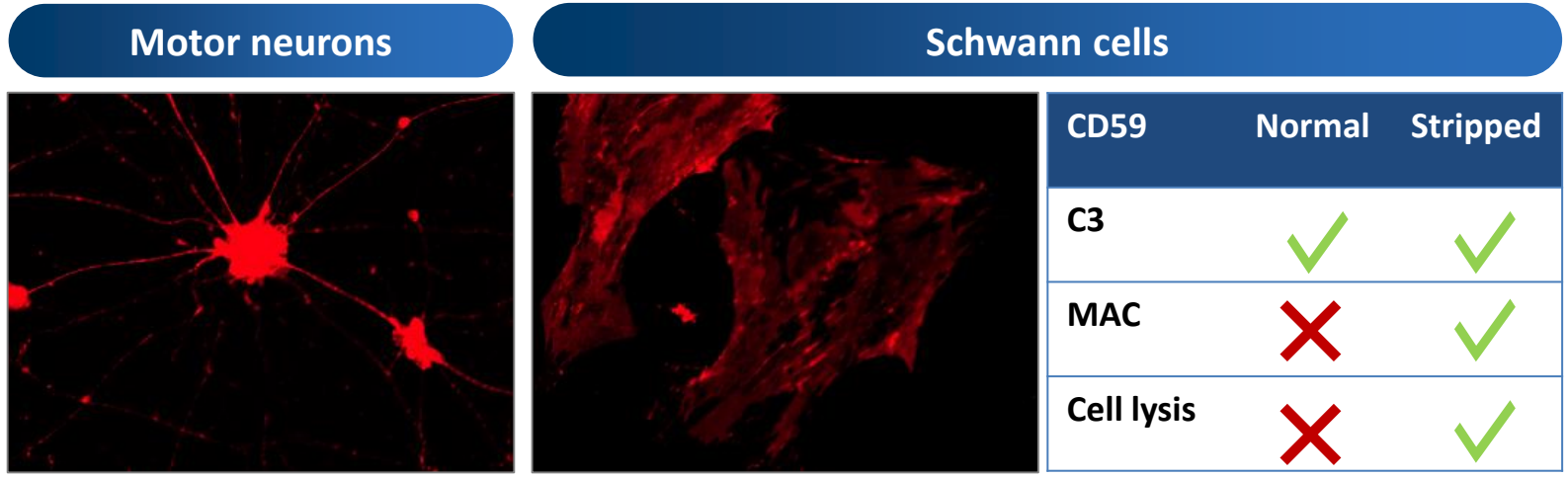
Hypothesis: pathology of MMN is upstream of C5

C3 Activation Confirmed MOA of IgM in MMN

High expression of CD59 protects motor neurons and Schwann cells against complement lysis



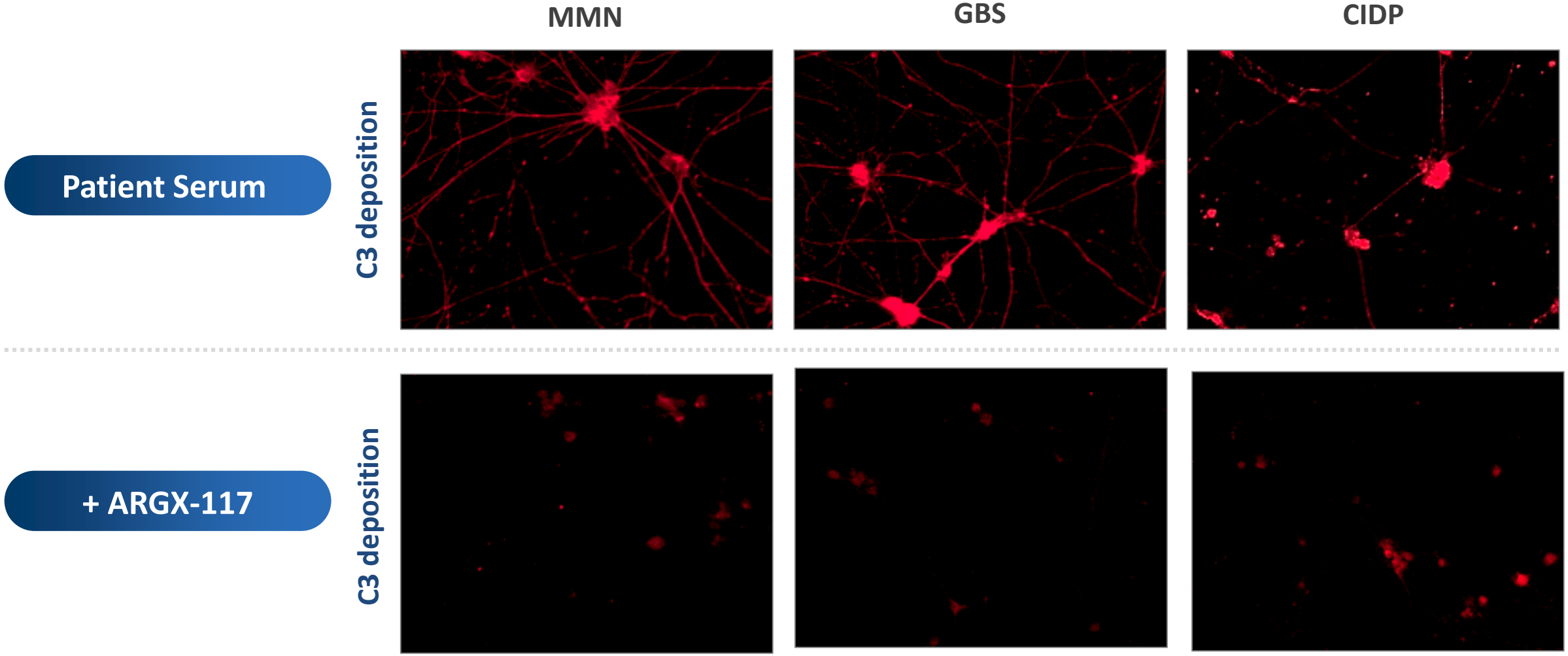
CD59 staining



CD59	Normal	Stripped
C3	✓	✓
MAC	✗	✓
Cell lysis	✗	✓

C3 activation confirmed MOA in MMN

New Biology Insights Translate Across Motor Neuron Diseases



Feasibility: Efficient Clinical and Regulatory Path

MMN indication has precedent clinical and regulatory path

Journal of the Peripheral Nervous System 18:321–330 (2013)

RESEARCH REPORT

A controlled trial of intravenous immunoglobulin in multifocal motor neuropathy

Angelika F. Hahn[1], Said R. Beydoun[2], Victoria Lawson[3], for The IVIG in MMN Study Team[†], MyungShin Oh[4], Victoria G. Empson[5], Heinz Leibl[6], Leock Y. Ngo[7], David Gelmont[7], and Carol L. Koski[8]

[1]*Department of Neurology, London Health Sciences Centre, London, Ontario, Canada;* [2]*Department of Neurology, University of Southern California, Los Angeles, CA, USA;* [3]*Department of Neurology, The Ohio State University, Columbus, OH, USA;* [4]*Clinical Biostatistics, Baxter Healthcare Corporation, Westlake Village, CA, USA;* [5]*Clinical Scientific Affairs and* [6]*Clinical Research, BioTherapeutics, Baxter Innovations GmbH, Vienna, Austria* [7]*Clinical Research, BioTherapeutics, Baxter Healthcare Corporation, Westlake Village, CA, USA and* [8]*Santa Fe, NM, USA*

Abstract Intravenous immunoglobulin (IVIG) has become the standard treatment for multifocal motor neuropathy (MMN) based on limited data. To critically assess the efficacy, safety, and tolerability of 10% liquid IVIG (IVIG), 44 adults with MMN were randomized

Feasibility: Orphan Potential & Economically Viable Indication

Unmet need for new therapies that slow down progression of disease and reduce reliance on IVIg

MMN

Multifocal Motor Neuropathy

"ALS patient that didn't die"

Slowly progressive

Asymmetric distal limb weakness mainly affecting upper limbs

Patients become dependent

Prevalence

~13,000 patients in the US

Often underdiagnosed

Predominantly men under 50

Diagnosis / Metrics

Anti-GM1 IgM antibody presence

Nerve conduction block

Defined clinical endpoints (i.e. 9-HPT, grip strength, Guy's neurological disability score)

Treatment

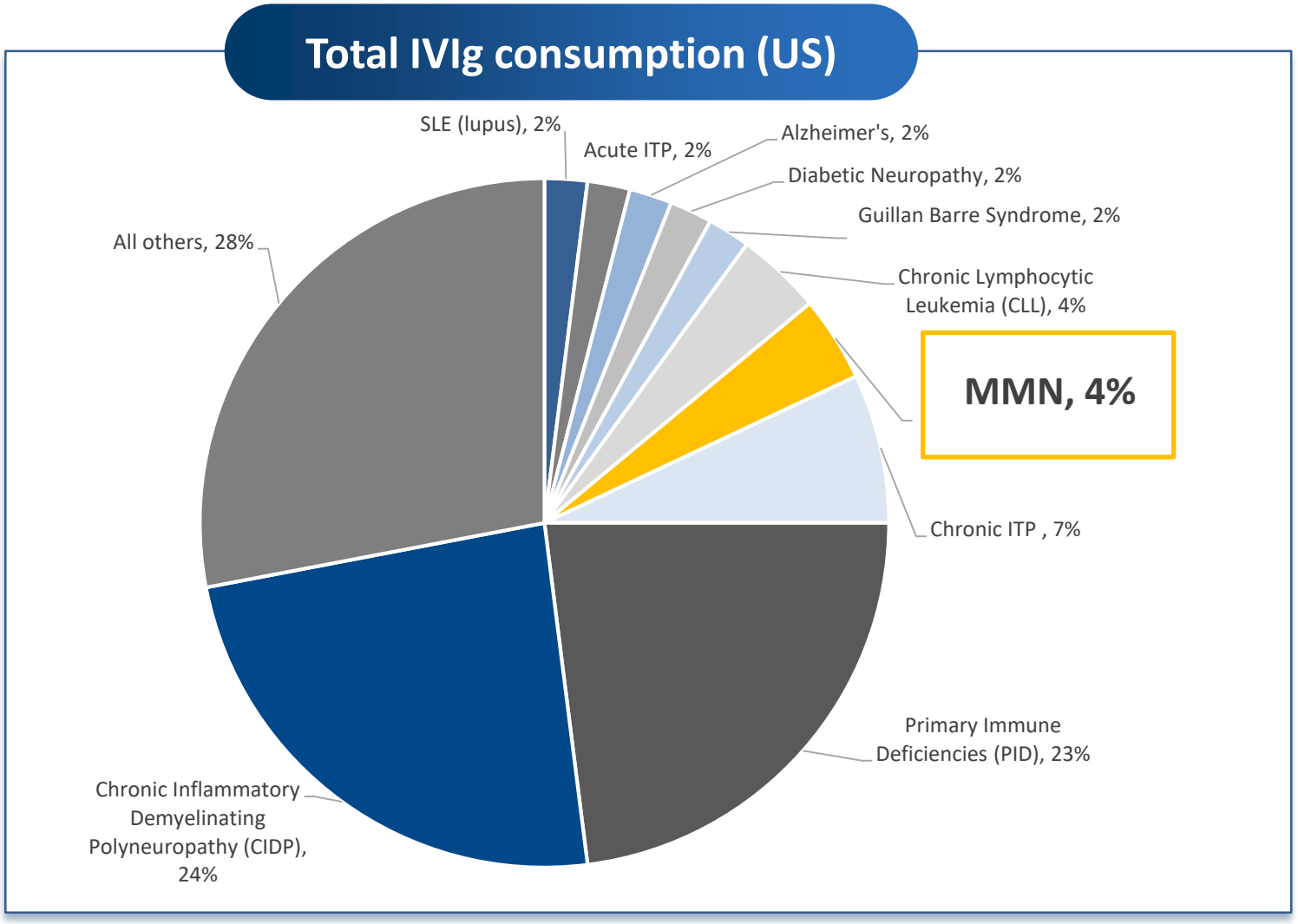
First line therapy is frequent, high dose of IVIg over 2-5 days

Patients unhappy with short duration of effect, disease progression despite strict adherence, side effects of IVIg

Payors aligned in need for new therapies

Feasibility: Economically Viable Indication

MMN represents 4% of total IVIg market by volume



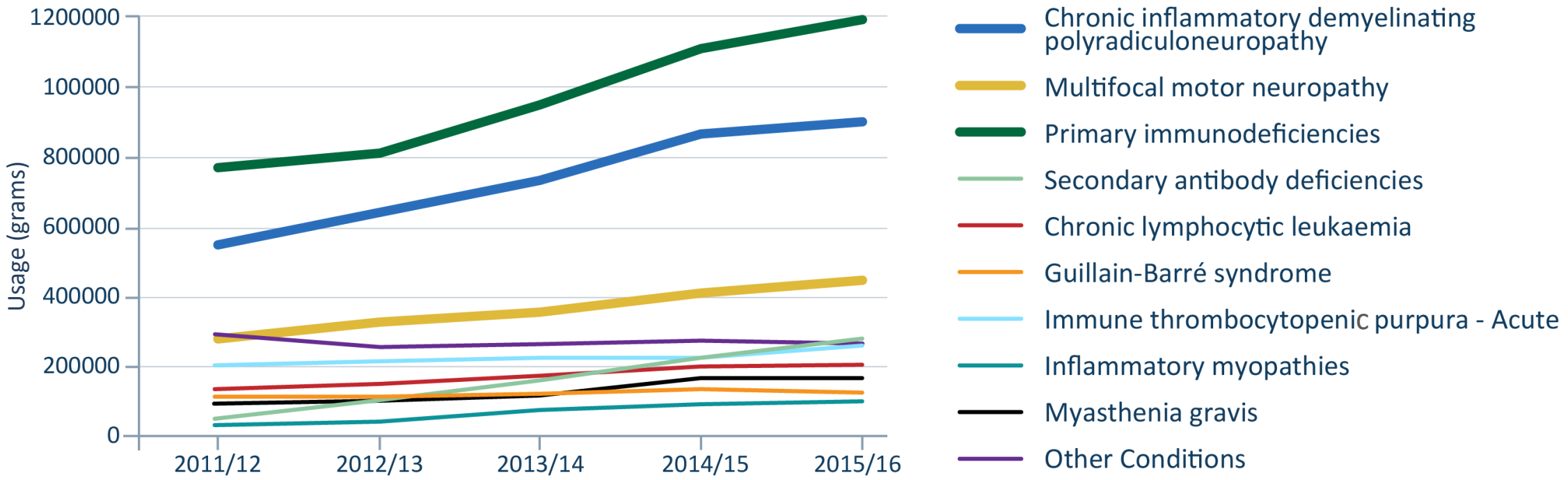
Global IVIg market

- ~$10B with 6-8% CAGR
- Neurology represents 40-50% (CIDP, MG, MMN, GBS, etc.)
- Neurology showing rapid growth outside of PID
- MMN represents 4% of total IVIg market

MDSAS: Immunoglobulin Database Report 2015/16

Neuropathies Highest Growth Indications for IVIg

Recorded yearly immunoglobulin use for the top 10 diagnoses (UK)



MDSAS: Immunoglobulin Database Report 2015/16

ARGX-117: Pipeline-in-a-Product Opportunity

Classical and Lectin Pathway-mediated Severe Autoimmune Diseases

Solid Biology Rationale

Feasible for Biotech
Orphan potential, economically viable, efficient clinical & regulatory pathway

Neuromuscular	Hem/Onc	Kidney
MMN CIDP - MFS MG GBS MGUS anti-MAG	CAD Autoimmune hemolytic anemia Stem cell transplant ITP	AMR/Ischemia reperfusion injury Membranous nephropathy IgAN Lupus nephritis

Role of Complement in Neurology and Kidney Indications

Prof. Ludo van der Pol, M.D., Ph.D.

- Neurologist, Associate Professor of Neurology
- Head of Netherlands SMA Center at the University Medical Center Utrecht, The Netherlands

Dr. Rafael Villicana, M.D.

- Associate Professor of Medicine
- Medical Director of Kidney Transplantation at Loma Linda University Medical Center, U.S.

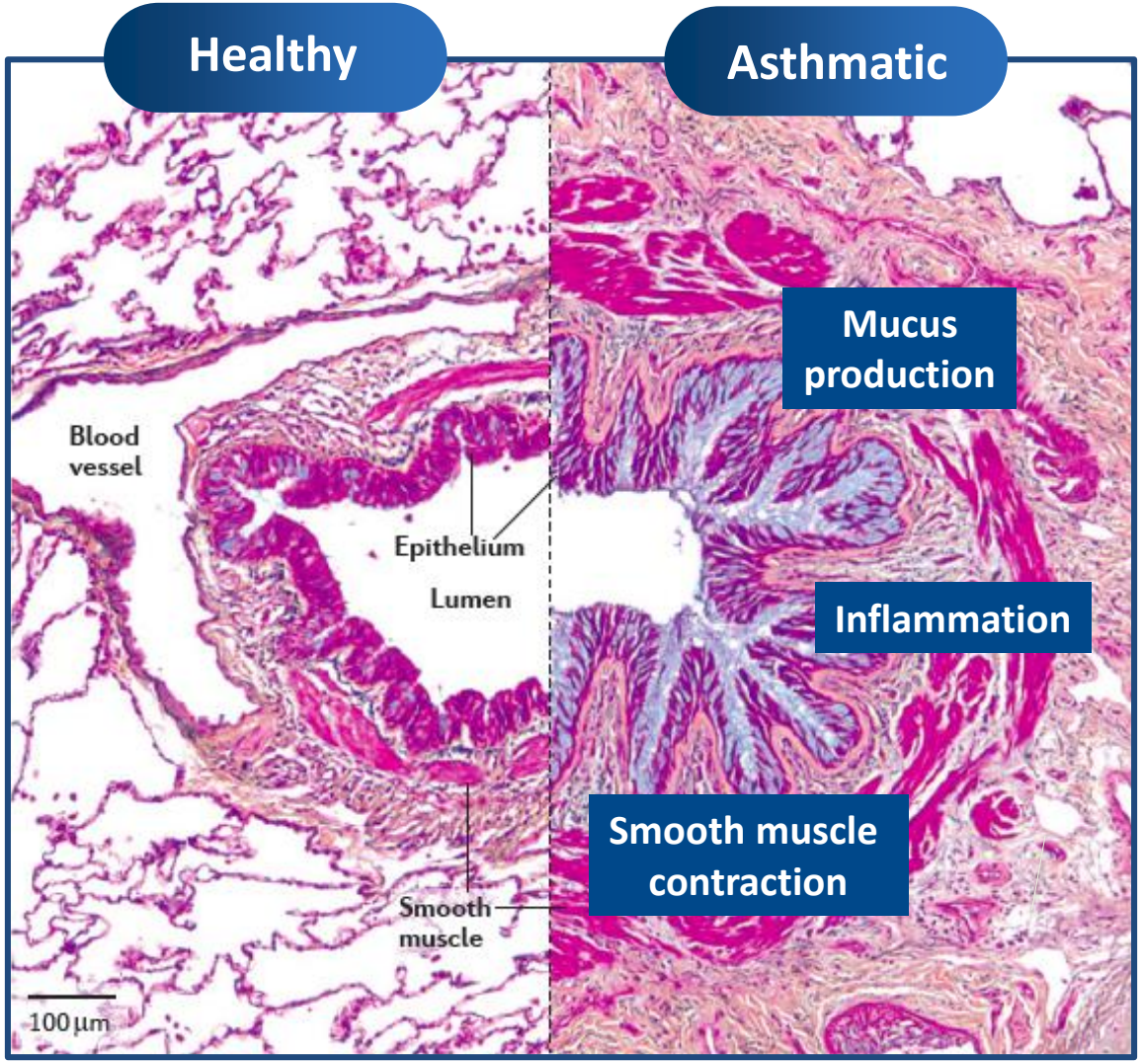
ARGX-118: Immunology Breakthrough for Airway Inflammation

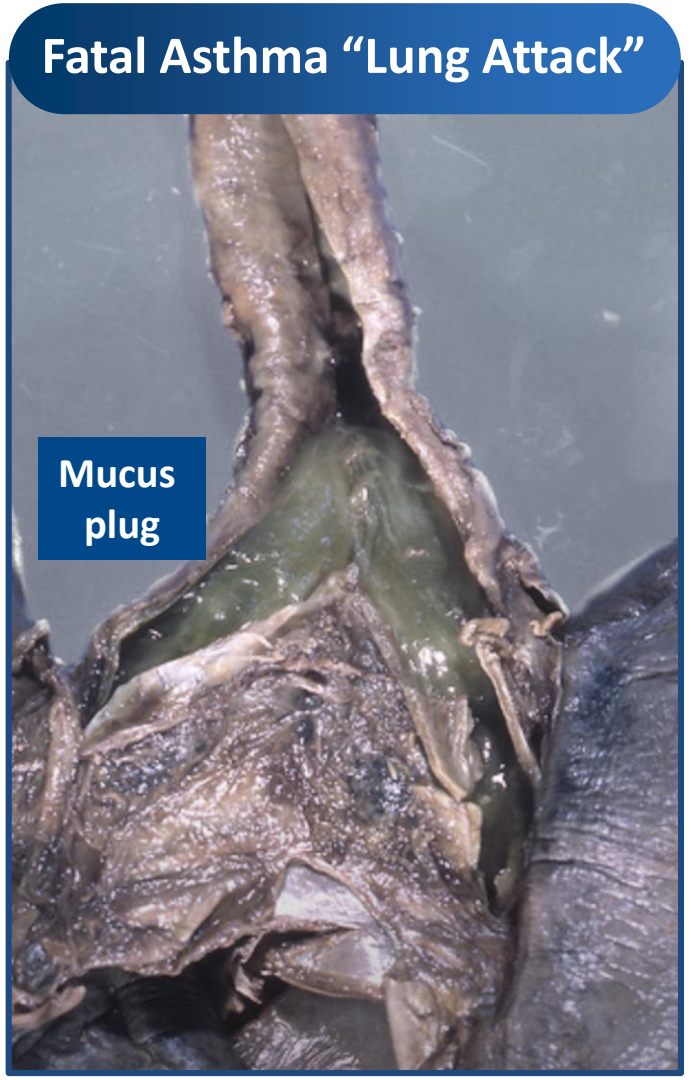
Bart Lambrecht | Prof. VIB-UGent

Professor Bart Lambrecht, M.D., Ph.D.

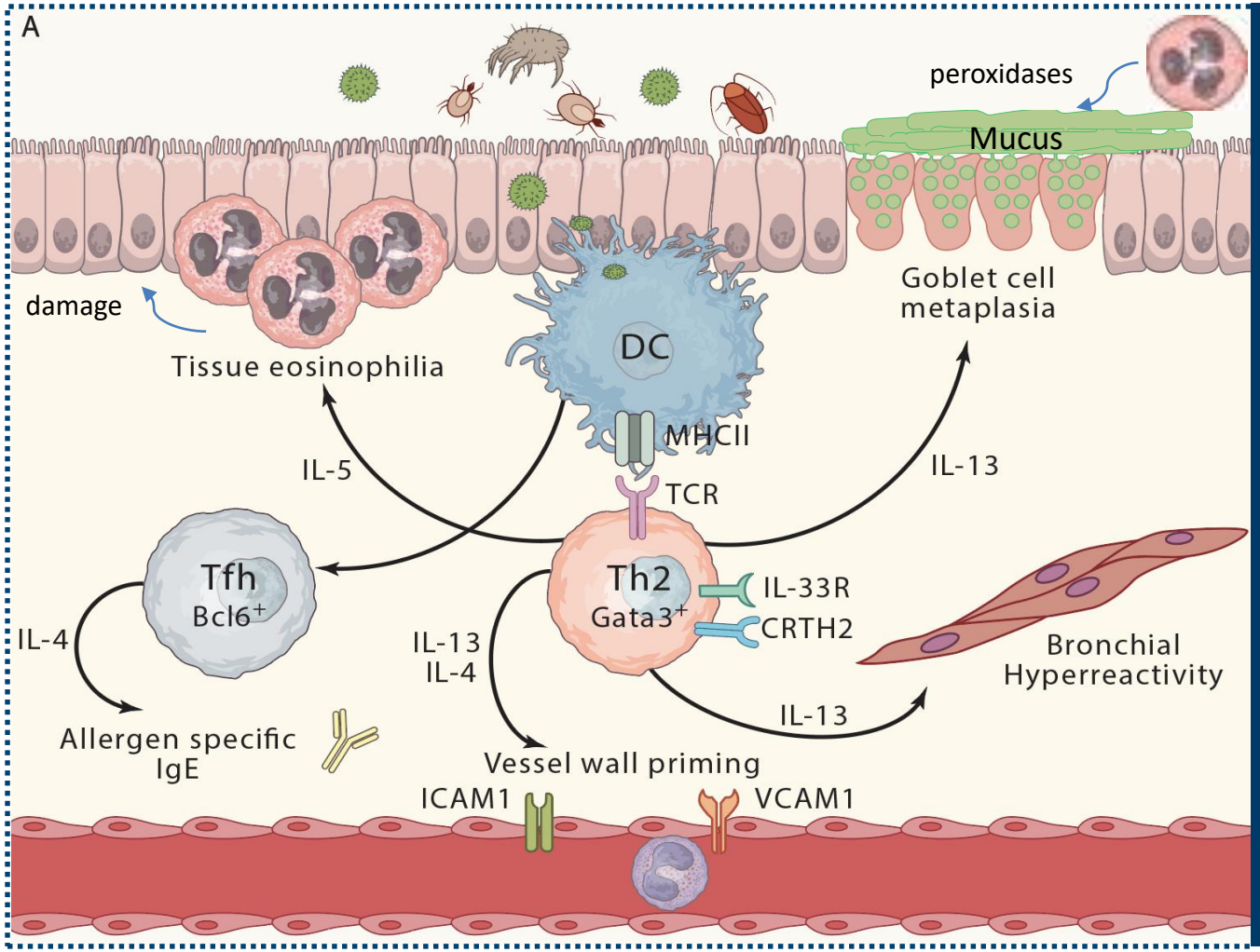
- Director of VIB Inflammation Research Center, UGent (350 scientists)
- Prof. of Pulmonary Medicine at Ghent University and Erasmus MC
- Research interests: immunology of asthma; basic science and translational research
- Theory on mechanisms of allergic sensitization are the basis of many research efforts globally, enabling new preventive and therapeutic avenues for asthma
- Author of 330 papers and 25 book chapters on asthma and allergy

Mucus Plugs Remain Largest Unmet Need in Asthma





Eosinophils Drive Severe Asthma Pathology and Mucus Production

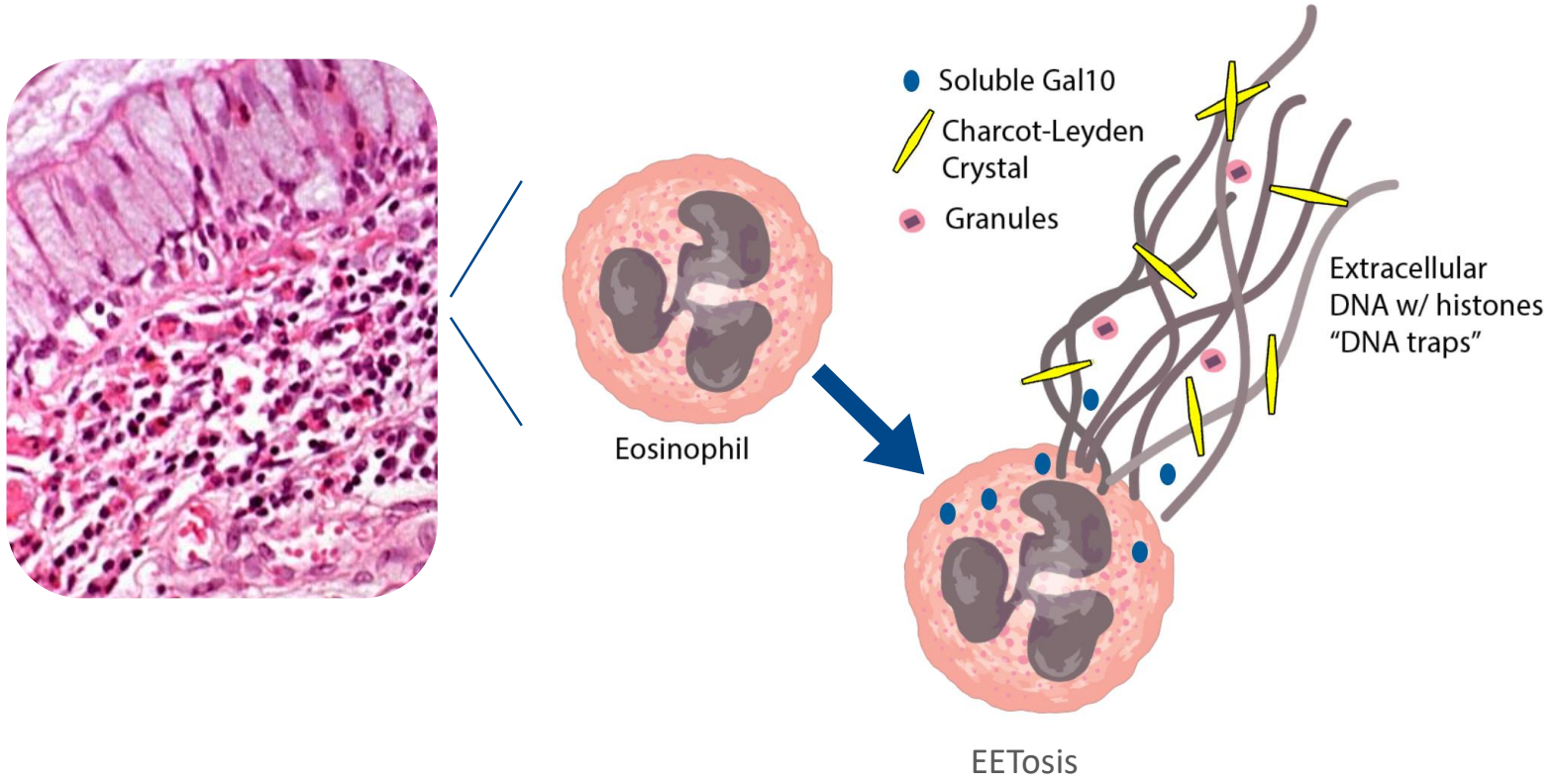


- Current therapies treat
  - Inflammation
  - Smooth muscle contraction
- Currently no drugs to treat the mucus build-up
- ARGX-118: potential first-in-class drug for mucus plugging and persistent airway obstruction

Lambrecht, Hammad and Fahy, Immunity, 2019

Charcot-Leyden Crystals (CLCs) and Galectin-10 Emerge From Eosinophil Burst

Asthma Biopsy

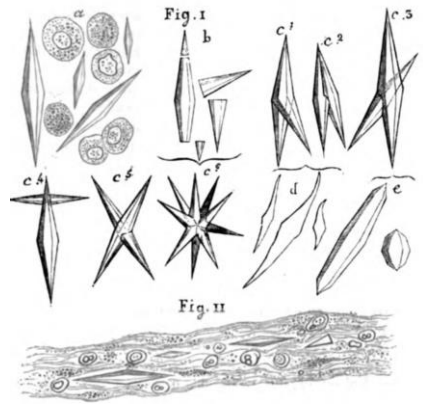


- Eosinophils seen in about 70% of severe asthmatics
- Galectin-10 makes up 10% of all eosinophil protein content
- EETosis = burst of eosinophils: releasing intracellularly formed CLCs and soluble Galectin-10; contributing to formation of large extracellular CLCs

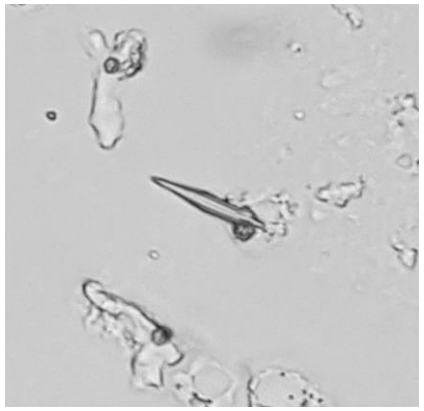
Ueki, Blood, 2018

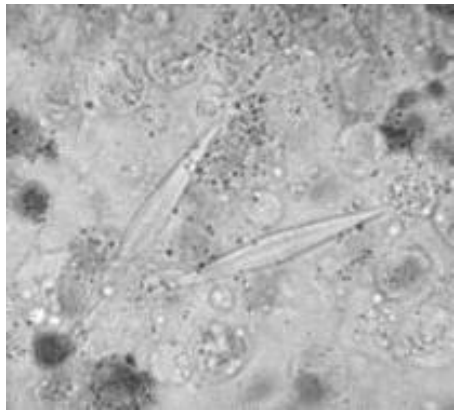
CLCs Intuitive Yet Overlooked Asthma Target

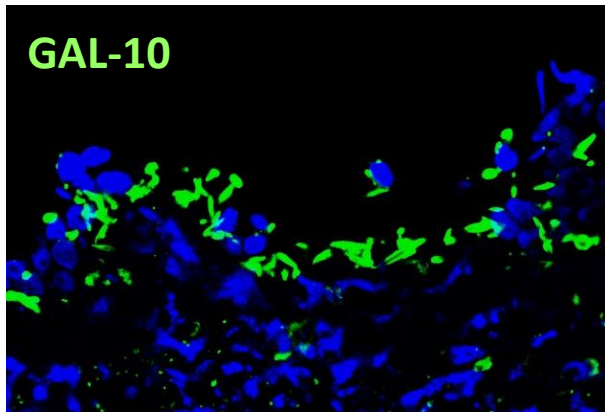
- Described in 1853 in sputum and mucus plugs of severe asthmatics
- Comprised of protein GAL-10
- GAL-10/CLCs serve as biomarker of mucus plugs
- Crystals present in “allergic mucin” in many diseases including:
  - Asthma and cystic fibrosis
  - Chronic rhinosinusitis (CRS) with nasal polyps



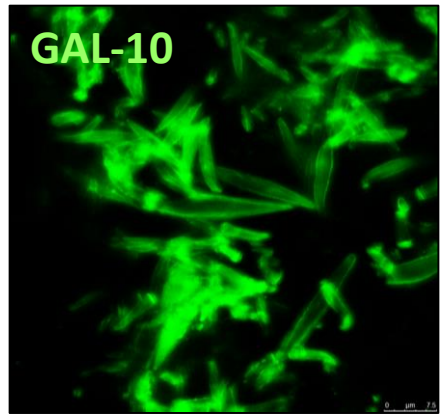
Charcot & Vulpian, Gaz. Hebd, 1870


Fresh sputum from asthma patient


Mucus plug from ABPA patient



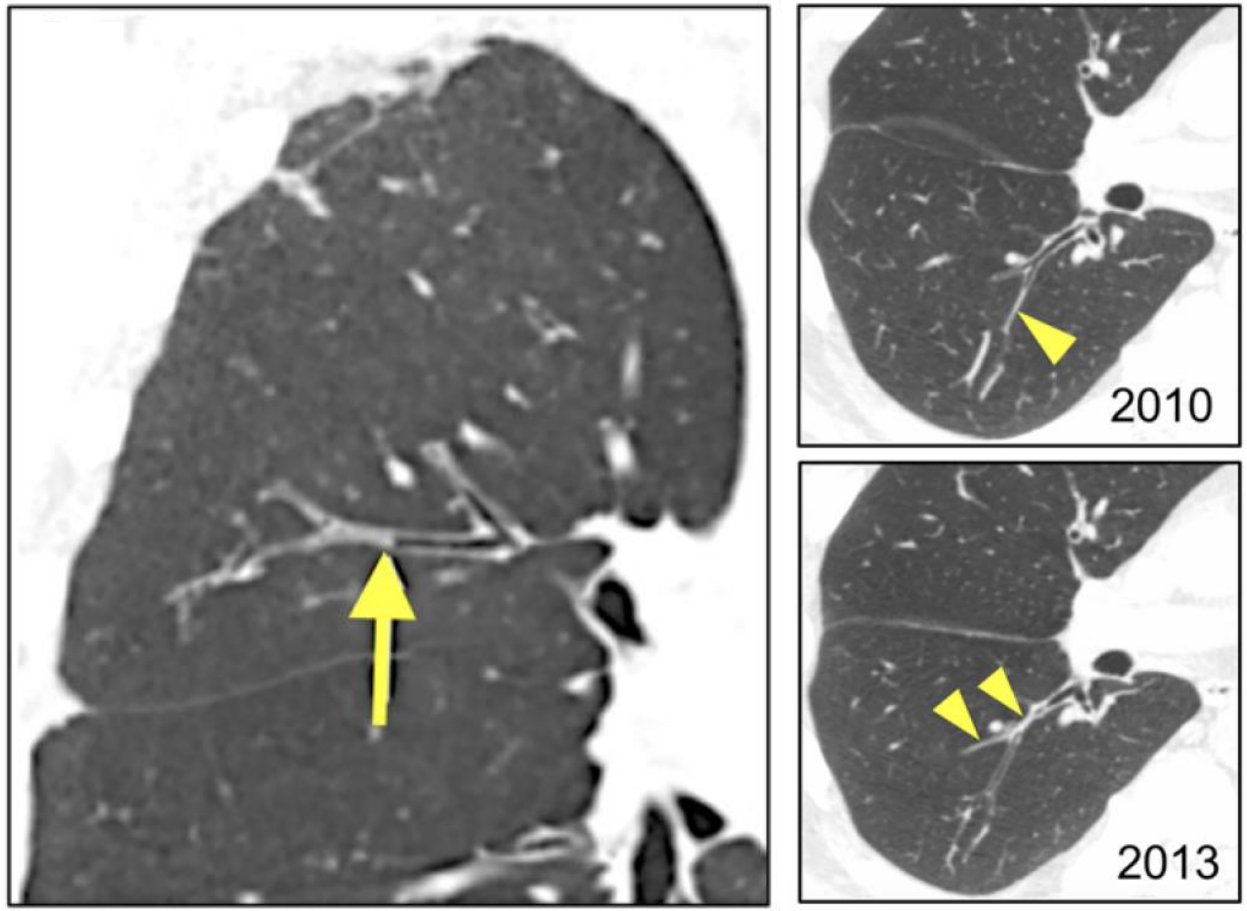
Mucosal biopsy from CRS with nasal polyps



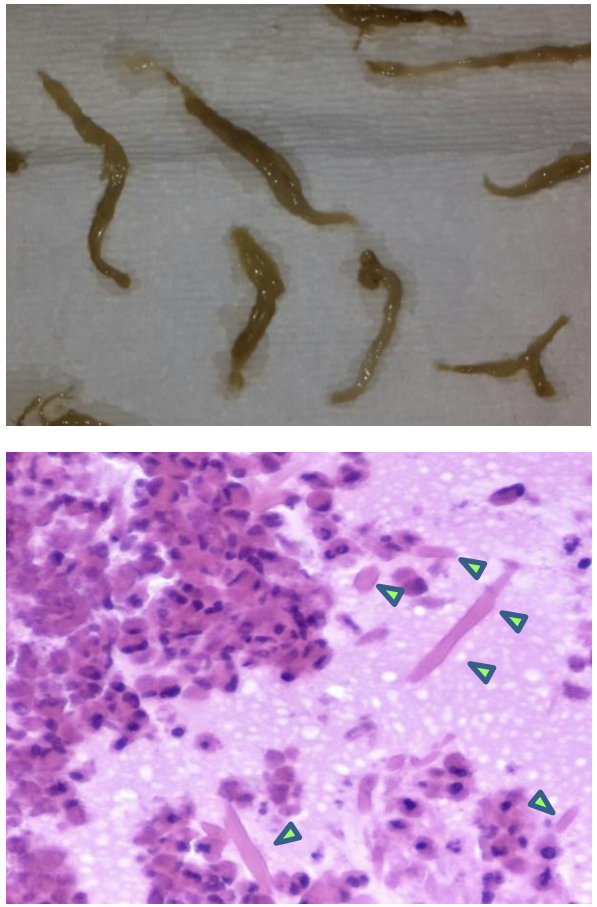
”Peanut butter”

Mucus Plugging Seen in 70% of Severe Asthmatics with Persistently Low Lung Function

Mucus plugs on chest CT scan

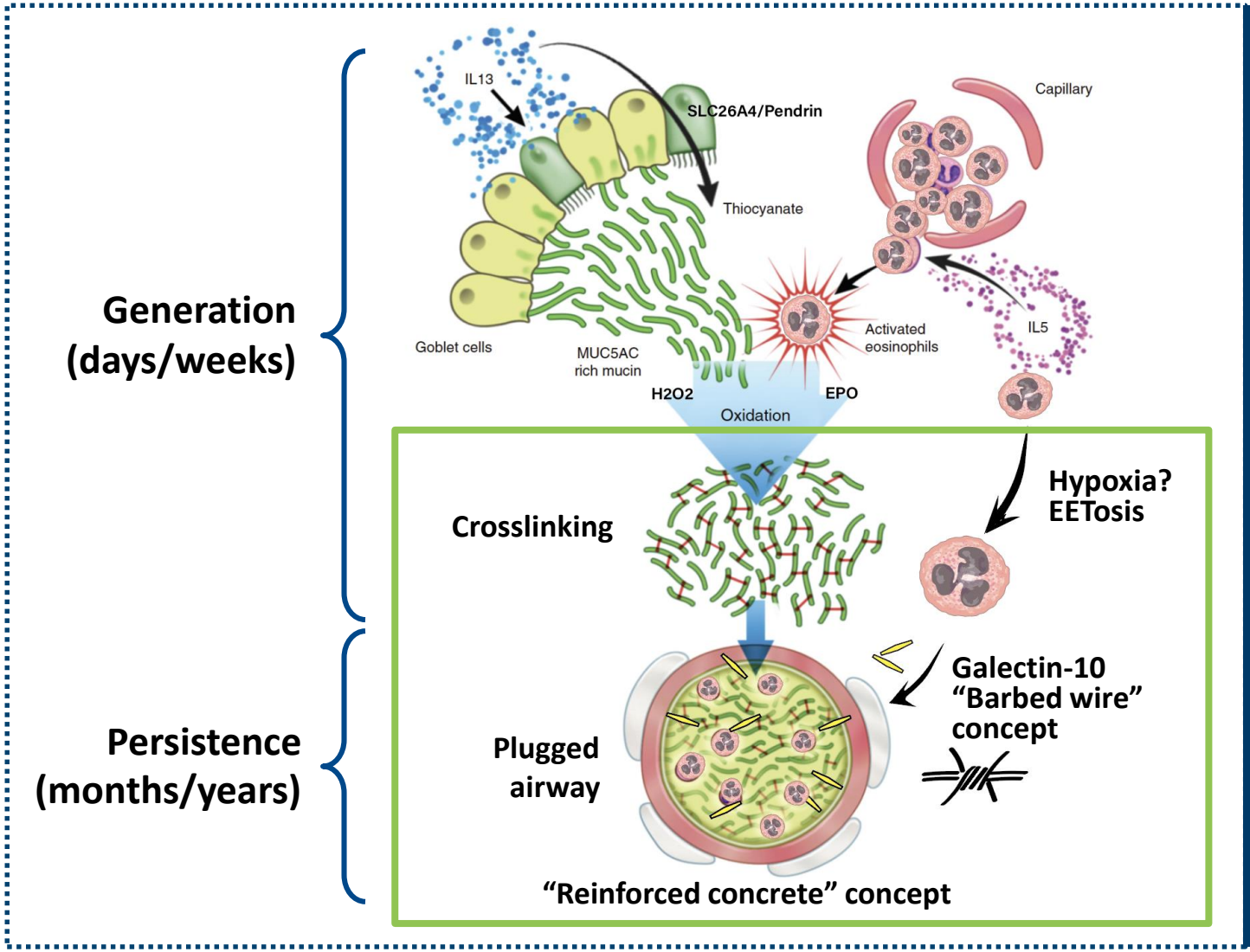


Asthma mucus plugs contain numerous CLCs



Dunican & Fahy J., Journal of Clin. Invest. , 2018

CLCs and Galectin-10 Are Basis of Mucus Plug Persistence

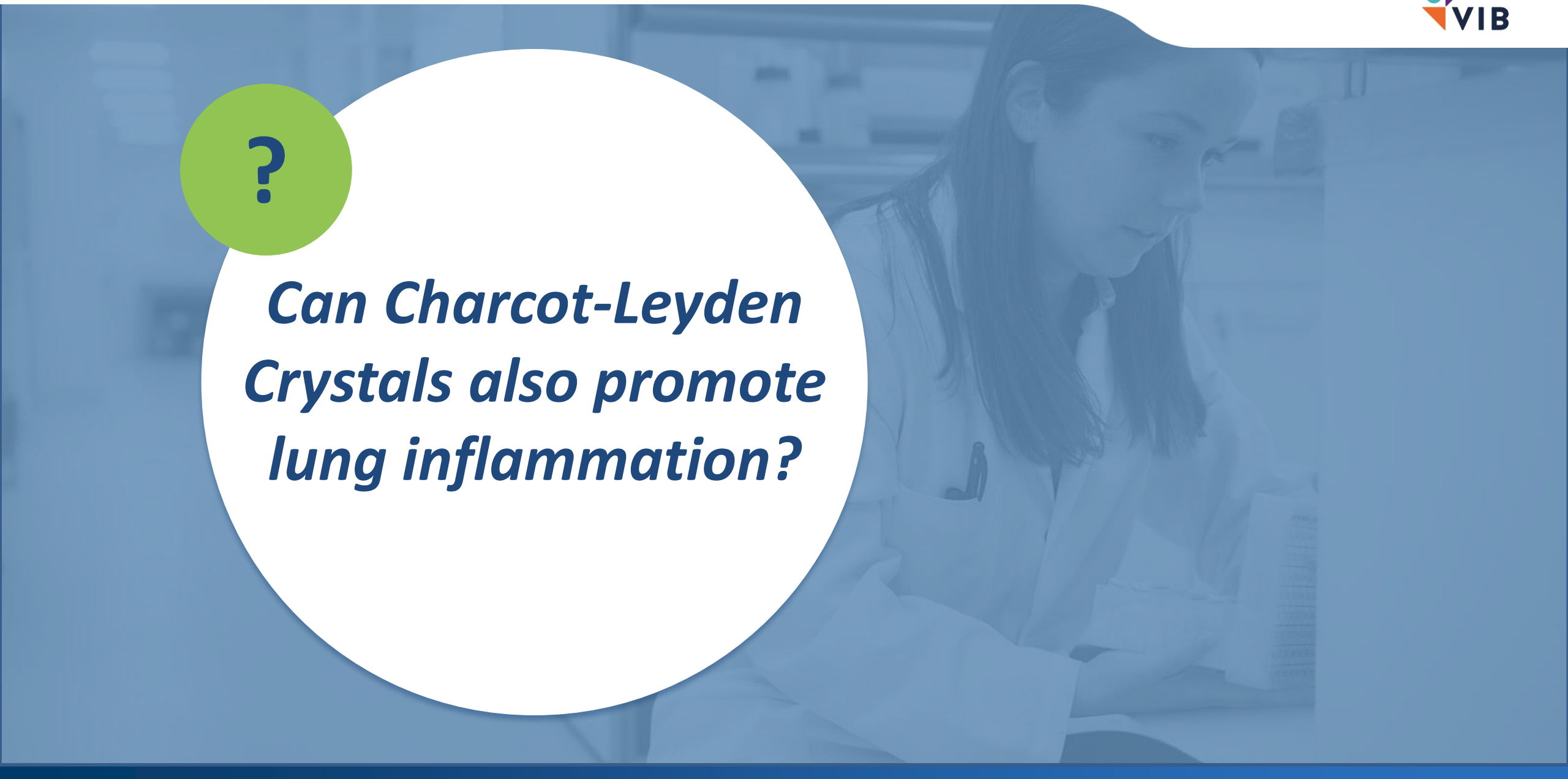


- CLCs together with crosslinked mucin form "barbed wire" which makes the plug stick in the airways
- Once mucus plugs are formed, they are persistent for years
- High unmet need for treatment that can resolve plugs
  - Only interventional therapies available

Fahy and Lambrecht

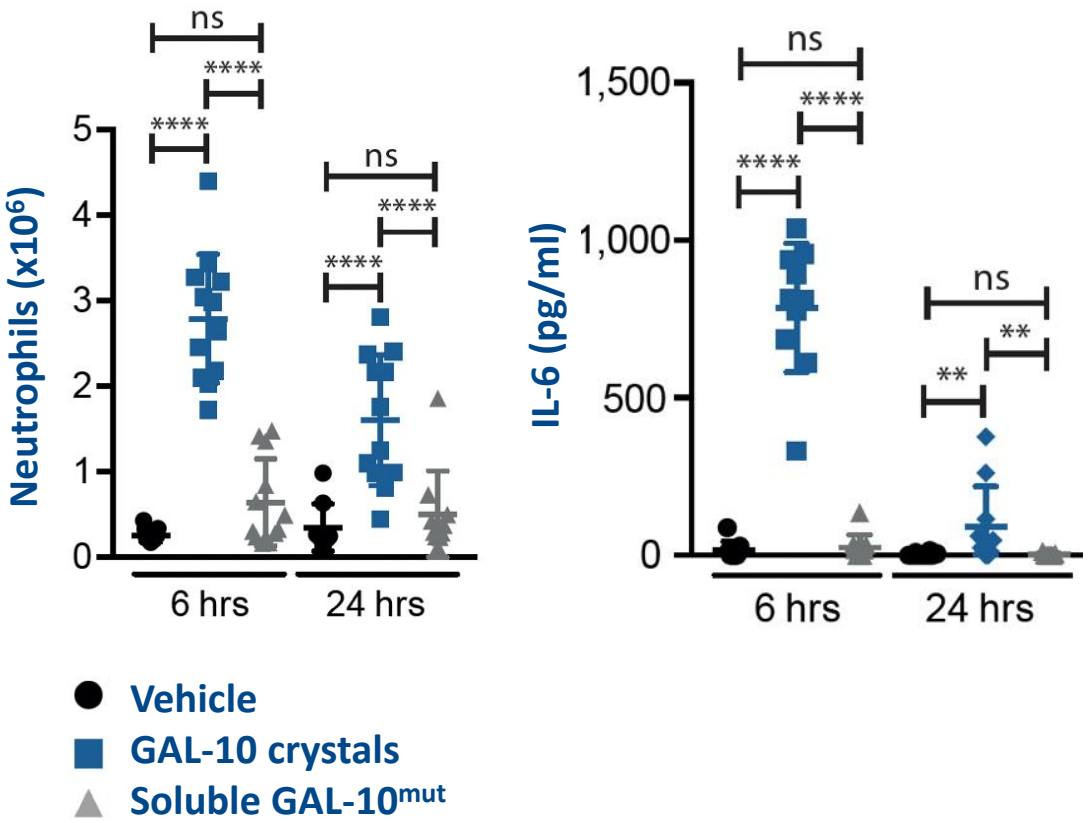
?

Can Charcot-Leyden Crystals also promote lung inflammation?

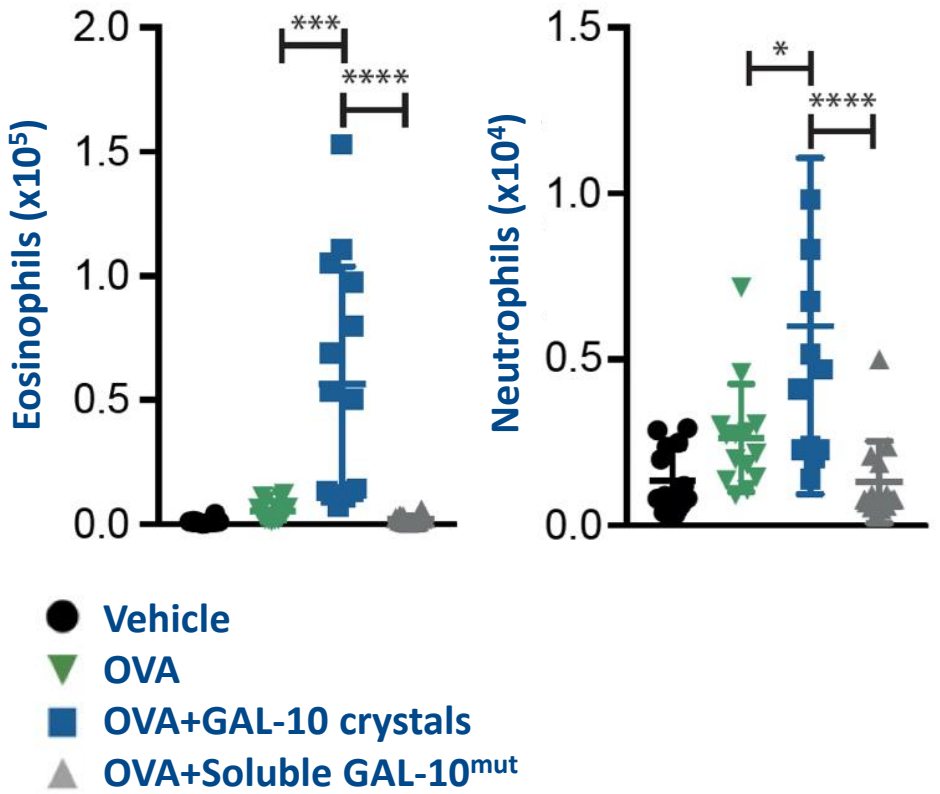


CLCs Induce Inflammation and Promote Development of New Allergies *in vivo*

CLCs induce inflammation when injected in lungs of mice

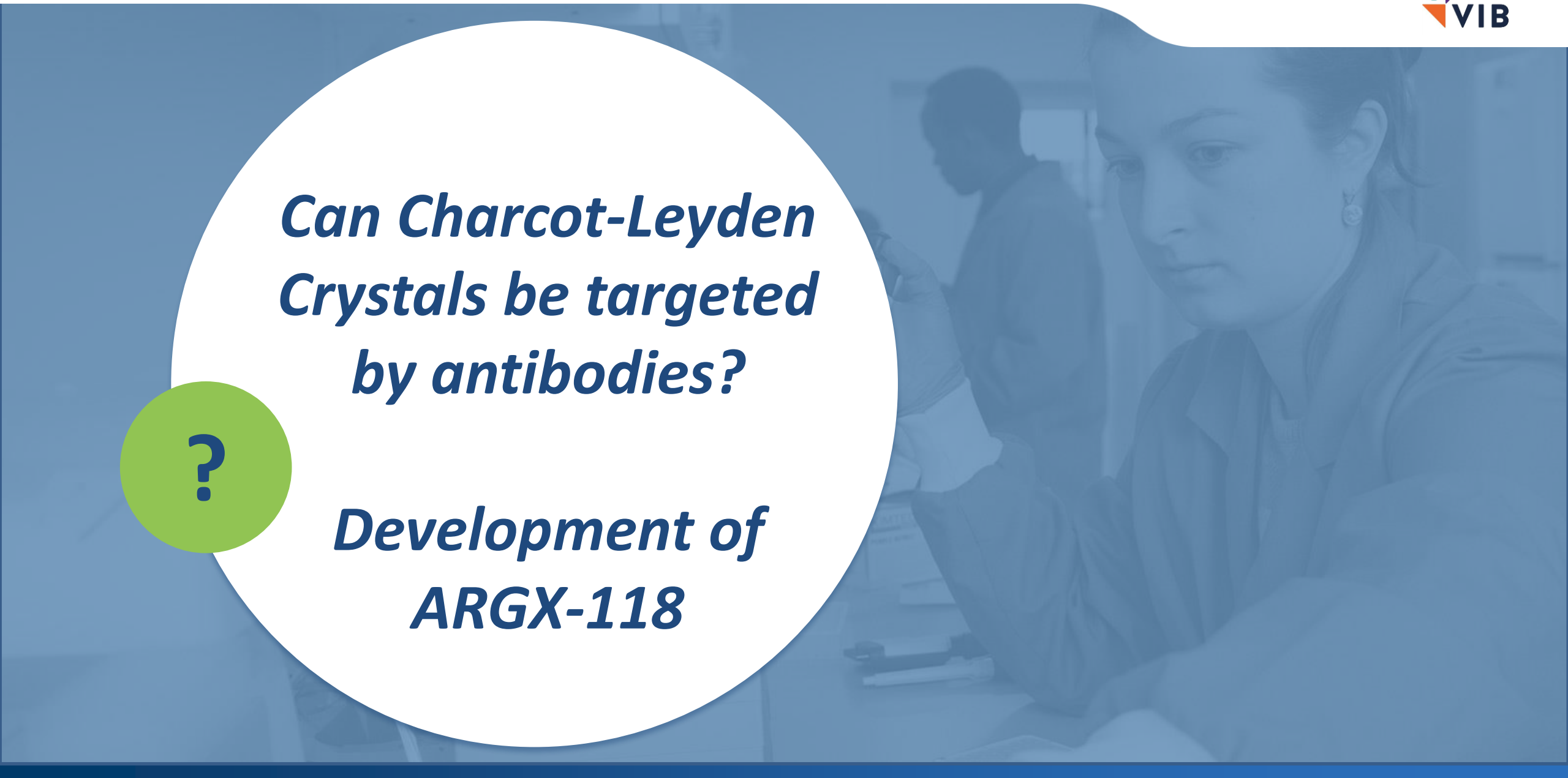


CLCs promote development of new allergies in mice



OVA: Ovalbumin

Persson et al., *in press*

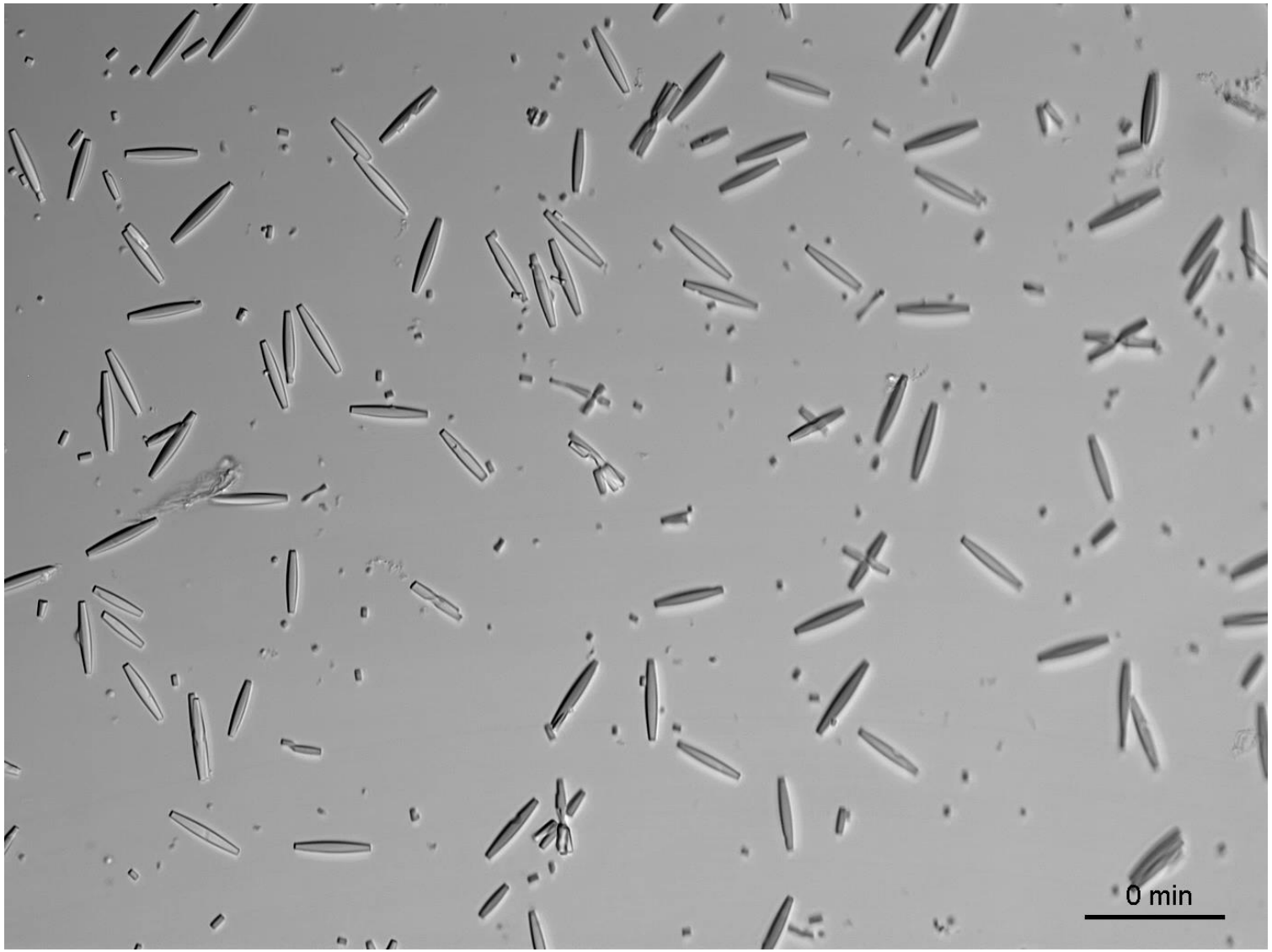


Can Charcot-Leyden Crystals be targeted by antibodies?

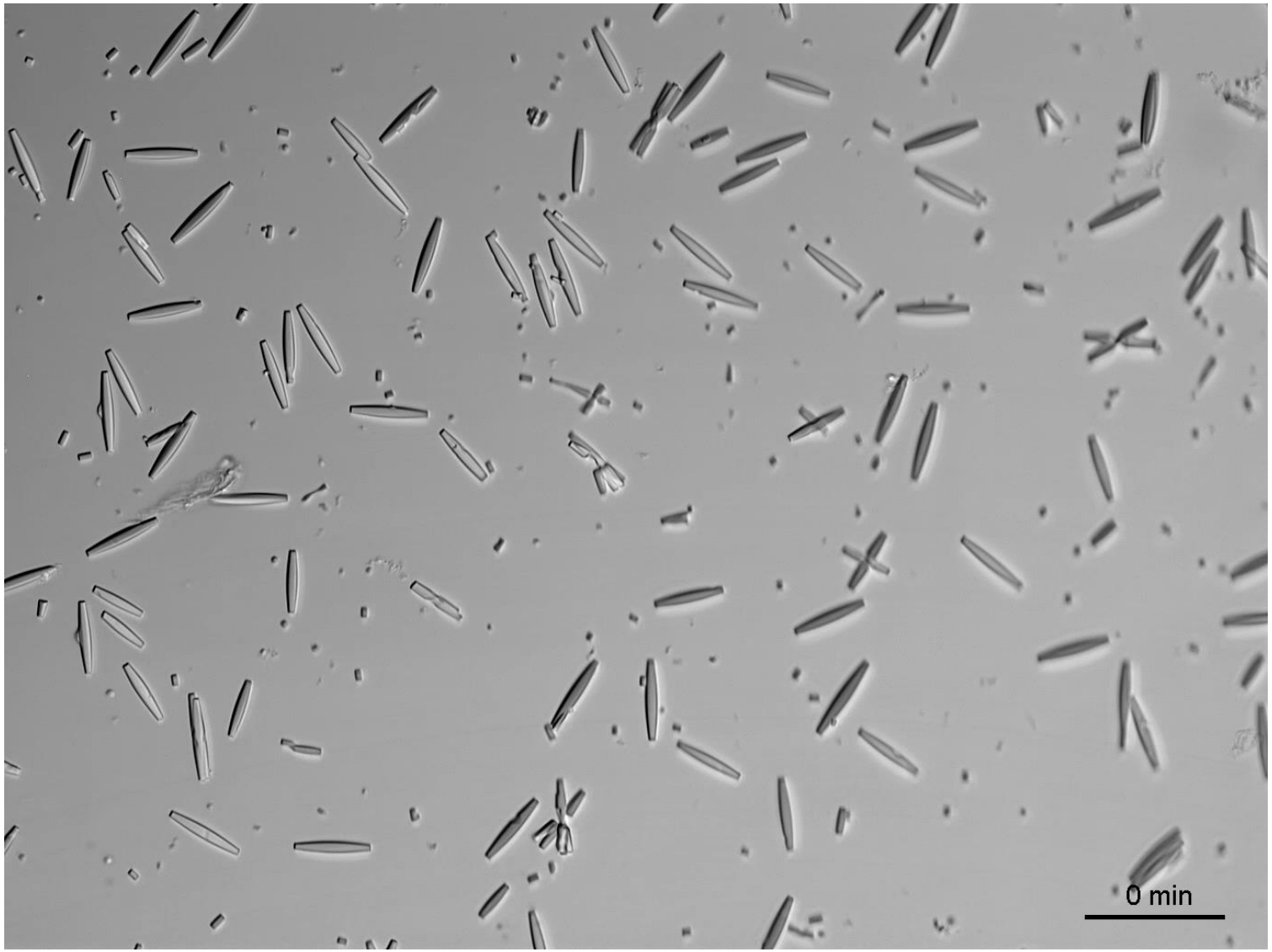
?

Development of ARGX-118

ARGX-118 Solubilizes CLCs



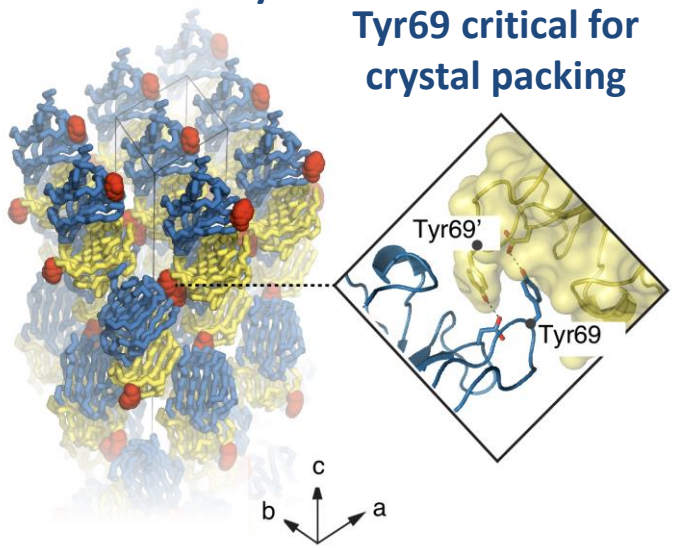
ARGX-118 Solubilizes CLCs



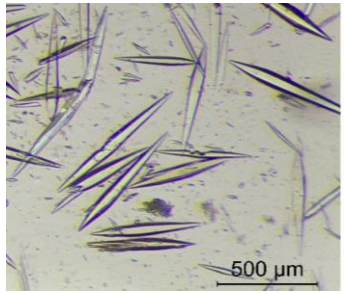
ARGX-118 Binds Critical Residue for Crystal Packing

Galectin-10 crystal

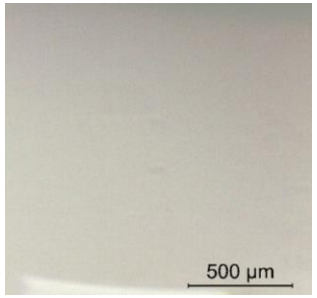
Tyr69 critical for crystal packing



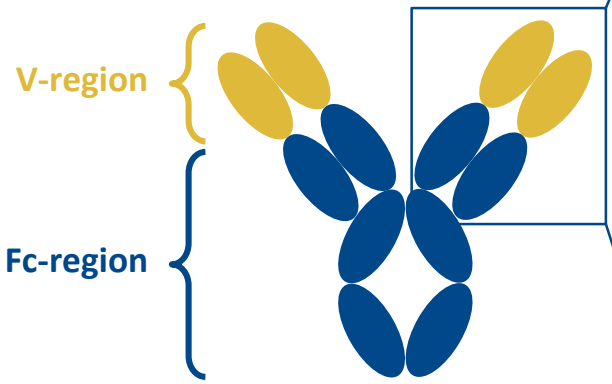
GAL-10-WT

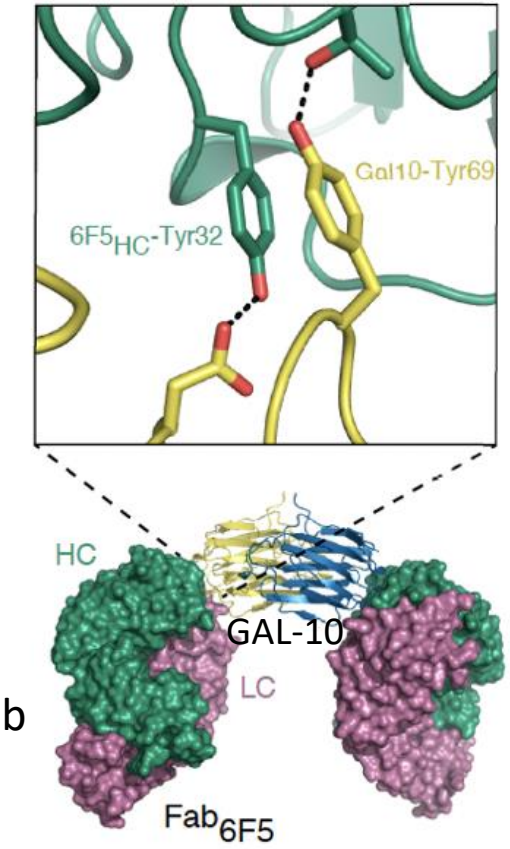


GAL-10-Tyr69GW



ARGX-118 binds Galectin-10 Tyr69

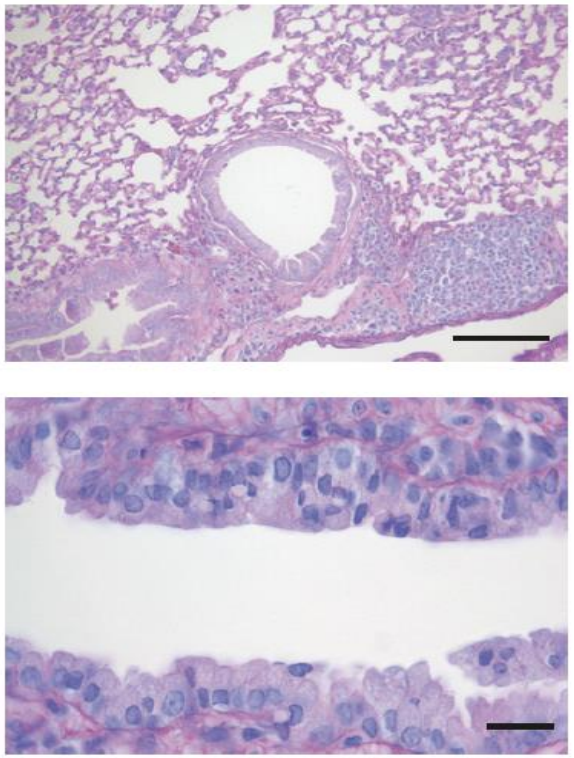




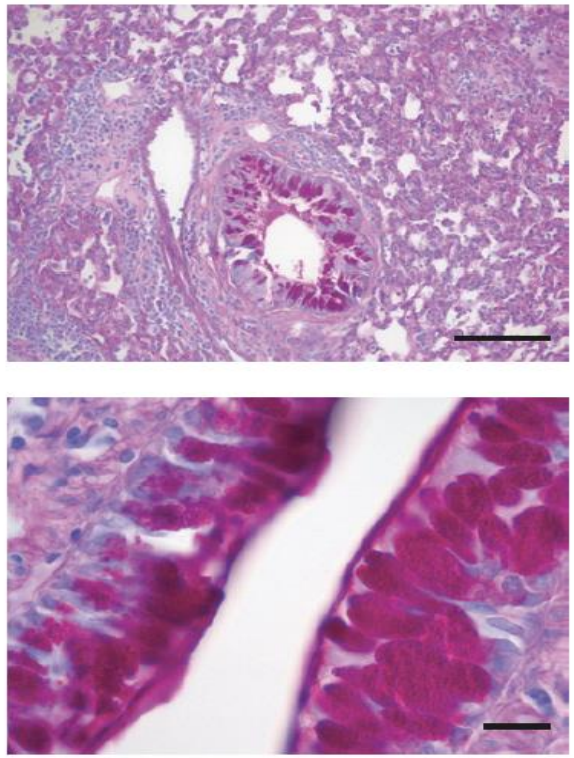
Targeting Tyr69 solubilizes the crystal

ARGX-118 Reverses Goblet Cell Metaplasia Caused by CLCs

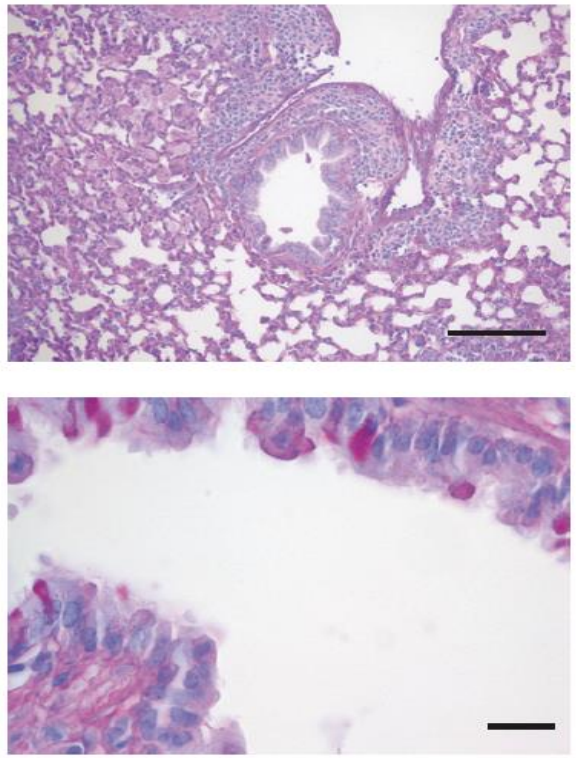
HDM Isotype Ab



HDM + CLC Isotype Ab



HDM + CLC ARGX-118

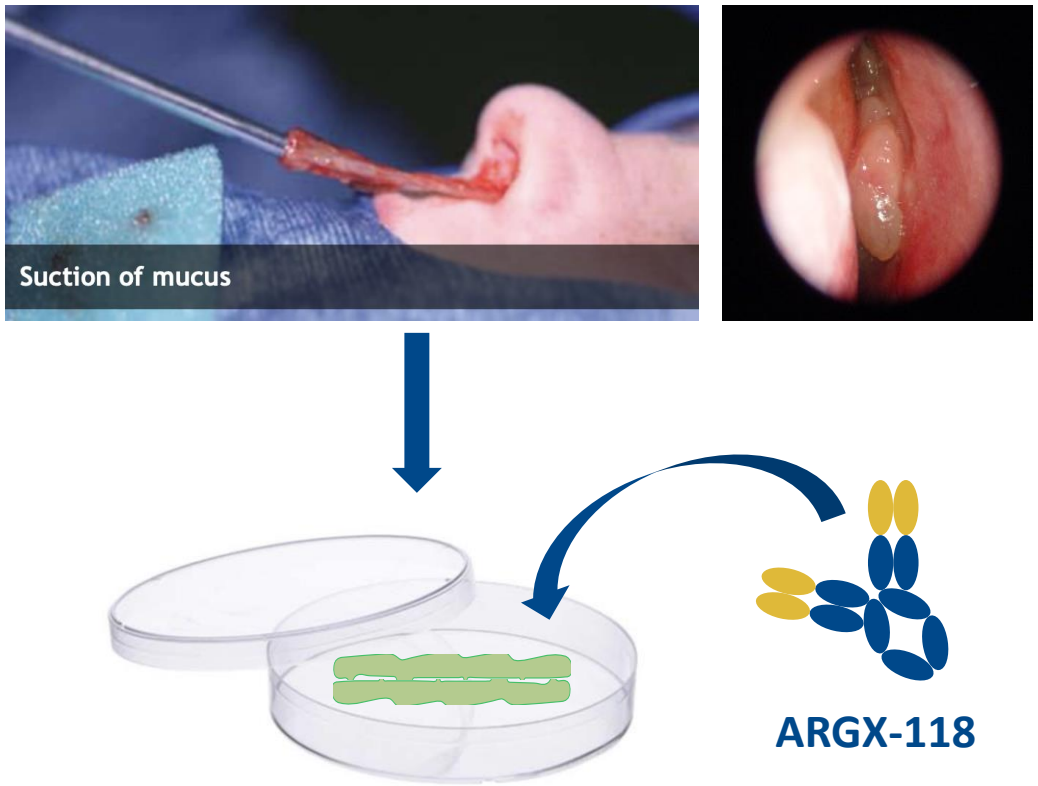


HDM= House Dust Mice Model

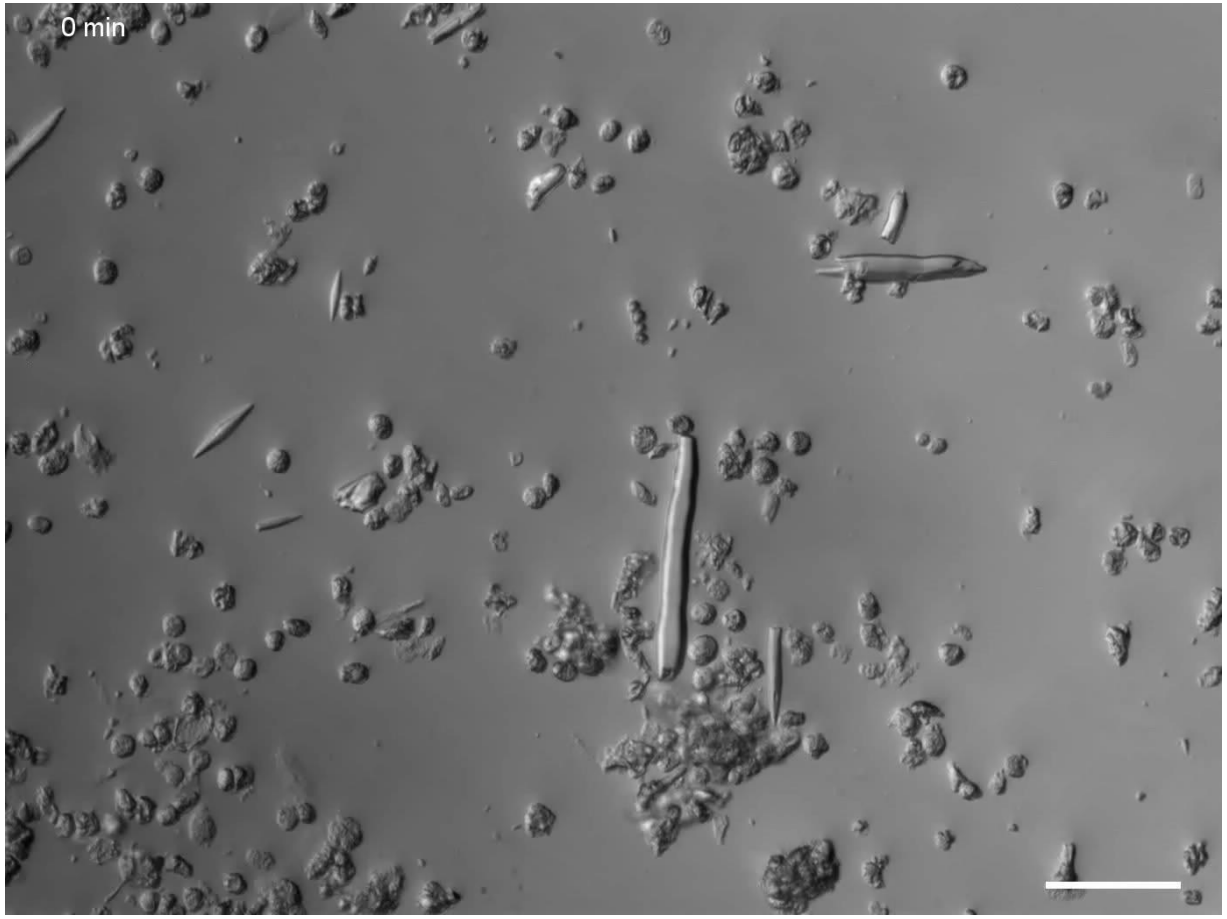
Persson et al., *in press*

ARGX-118 Dissolves CLCs Within Native Patient Mucus

Allergic mucin protruding from nasal sinus

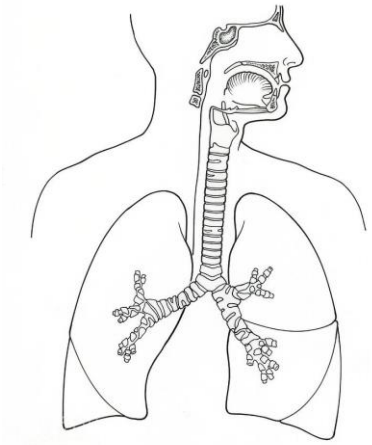


Persson et al., *in press*



Video shot over 16 hours

Multiple Crystallopathies Induced by CLCs



ARGX-118 targets GAL-10, sole component of CLCs

- **Stimulate innate and adaptive immunity**
- **Act as a type 2 adjuvant when administered to airways**
- **Stimulate mucus production**

Biology rationale supports potential role of CLCs as disease drivers in eosinophilic diseases

Lung attack or asthma exacerbation

- Acute intratracheal treatment for asthma attack by bronchoscopy
- Chronic nebulization for persistent plugging
- MOA: reverse mucus plugging

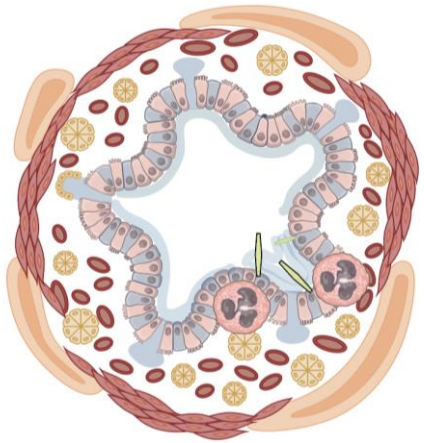
ABPA

- Chronic treatment
- Nebulization
- MOA: block type 2 immunity trigger (IgE) plus mucus plugging

CRS with Nasal Polyps

- Chronic treatment
- Nebulization
- Following sinus surgery
- MOA: block type 2 immunity trigger plus mucus stickiness

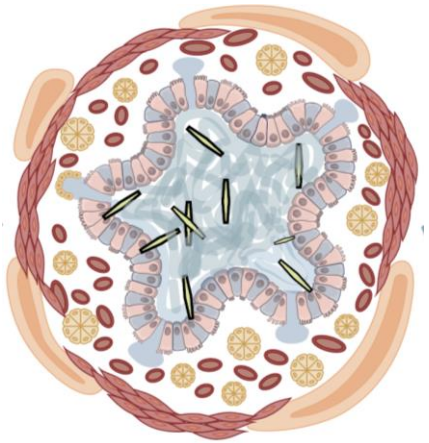
Mucus Plugging Requires Intervention or Chronic Therapy



Bronchus is narrowed
Exacerbation

Prevention by biologicals

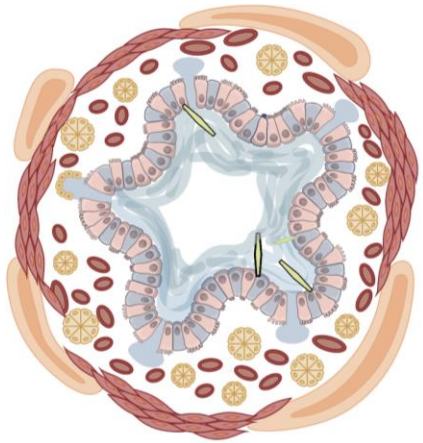
Mepolizumab
Benralizumab
Dupilumab



Acute occlusion
by mucus plugging

Bronchoscopic mucolysis

Potential for ARGX-118



Healing with persistent airway obstruction caused by residual mucus

Inhaled mucolytic therapy

Potential for ARGX-118

Innovative Access in Action

argenx

Antibody Expertise

VIB - UGent

Disease Biology Expertise

Development of unique Simple Antibody™; track record in drug development

High risk-high gain: early collaboration focused on science, feeling of ownership on both ends

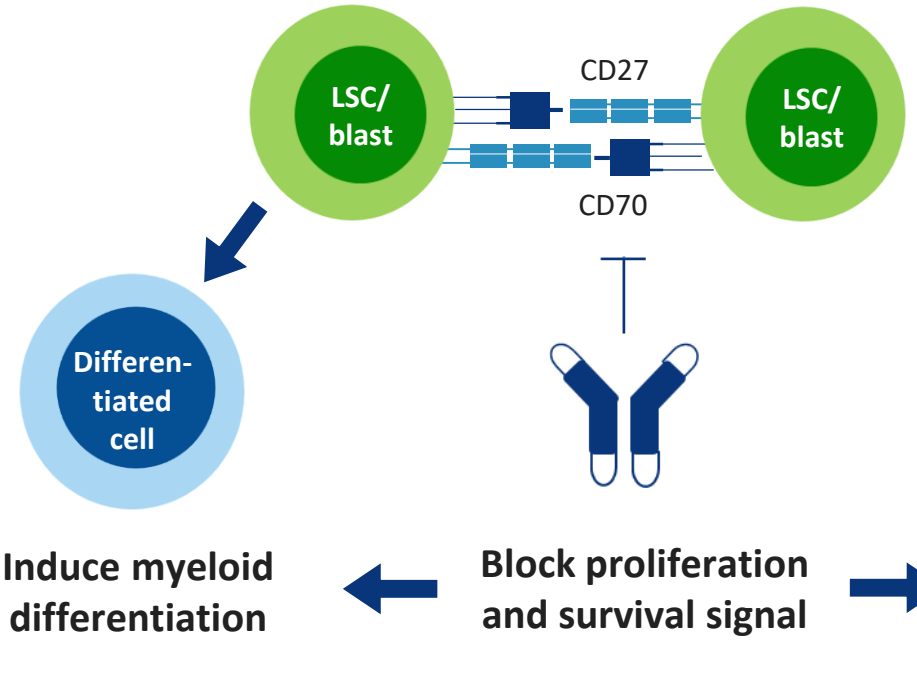
Open communication; fair economics; aligned expectations

Cusatuzumab Development Plan

Wim Parys | Chief Medical Officer

Cusatuzumab: Potential Foundational, Novel Therapy for Acute Myeloid Leukemia

Multiple MOA of Cusatuzumab



- Novel target and mechanism of action[1] (inhibition of CD70 pathway)
- Intrinsic activity shown as a single-agent in AML
- Potential for combination therapy[2]
- Phase 1/2 study: 92% ORR with 10/12 patients with CR/CRi after cusatuzumab treatment in combination with azacitidine (AZA) in newly diagnosed AML patients[3]
- IAP, Bern University – Prof. Ochsenbein

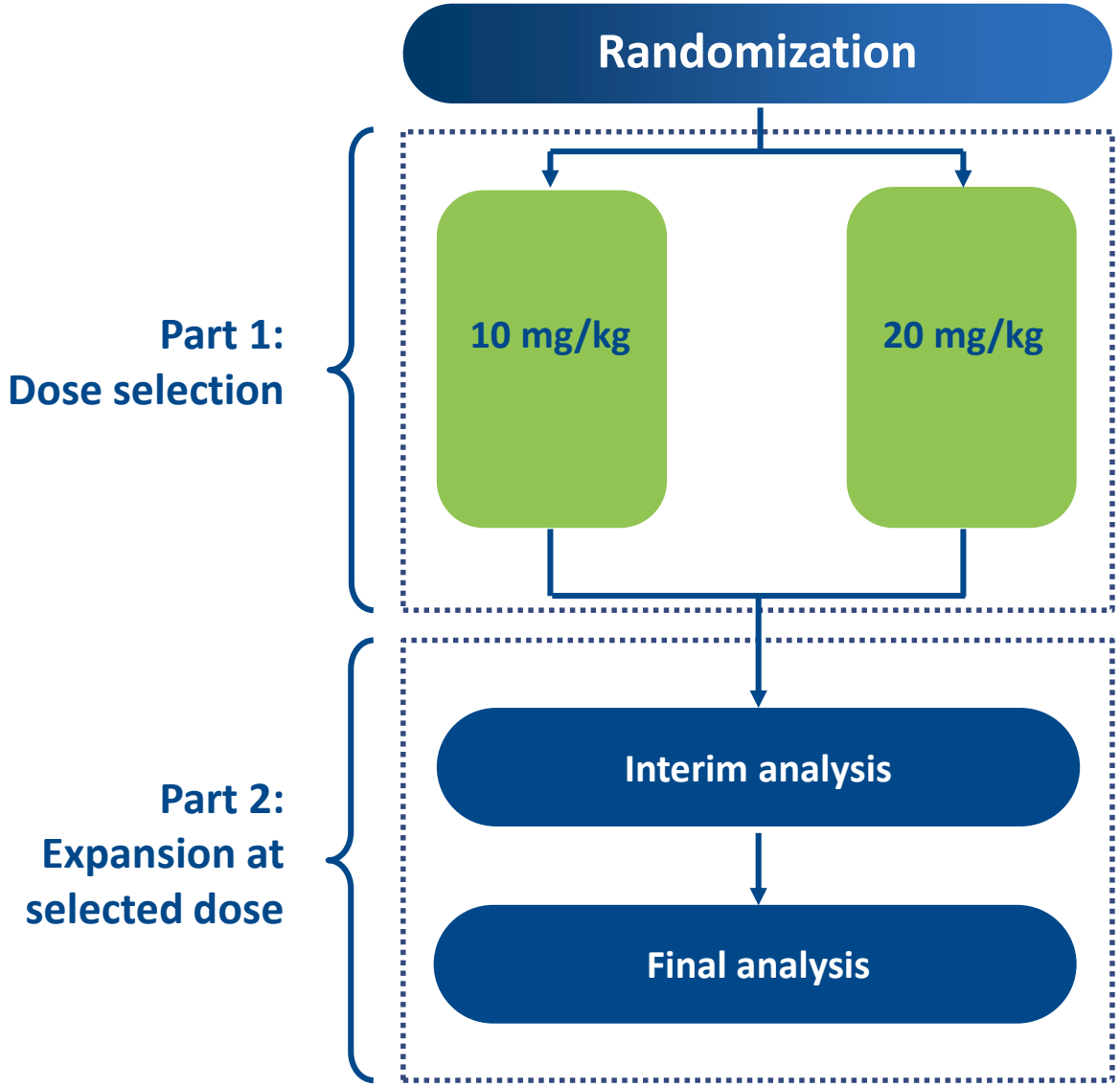
LSC, leukemic stem cells; sCD27, soluble CD27

1. Silence K, et al. MAbs. 2014;6:532232 | 2. Riether C, et al. J Exp Med. 2017;214:359–80 | 3. argenx ASH 2018 investor meeting

Cusatuzumab Strategic Alliance with Janssen Pharmaceuticals

argenx objectives	Janssen alliance
Accelerate & broaden development plan	Joint development plan focused on AML, MDS and other heme malignancies
Secure strong financials	Upfront $300M + $200M equity @ 20% premium, $1.3B in milestones, double digit royalties OUS
Retain commercial upside	50% of US economics on a royalty basis, up to 50% commercial efforts

Phase 2 Study in Newly Diagnosed, Unfit AML Patients



Combination Therapy: Cusatuzumab + Azacitidine

Patient Population: Newly diagnosed AML patients unfit for intensive chemotherapy (n= up to 150)

Primary Objective: To determine the efficacy (CR rate)

Secondary Objectives:

- ORR = (CR + CRh + CRi)
- Time to response and duration of response
- Event-free survival
- Overall survival
- Safety
- PK/immunogenicity
- MRD

Anticipated Phase 2 study start in US: second half 2019

argenx 2021: Global Commercial Vision

Keith Woods | Chief Operating Officer

Agenda

WHAT

product franchise / pipeline

WHY

unmet medical need / market opportunity

HOW

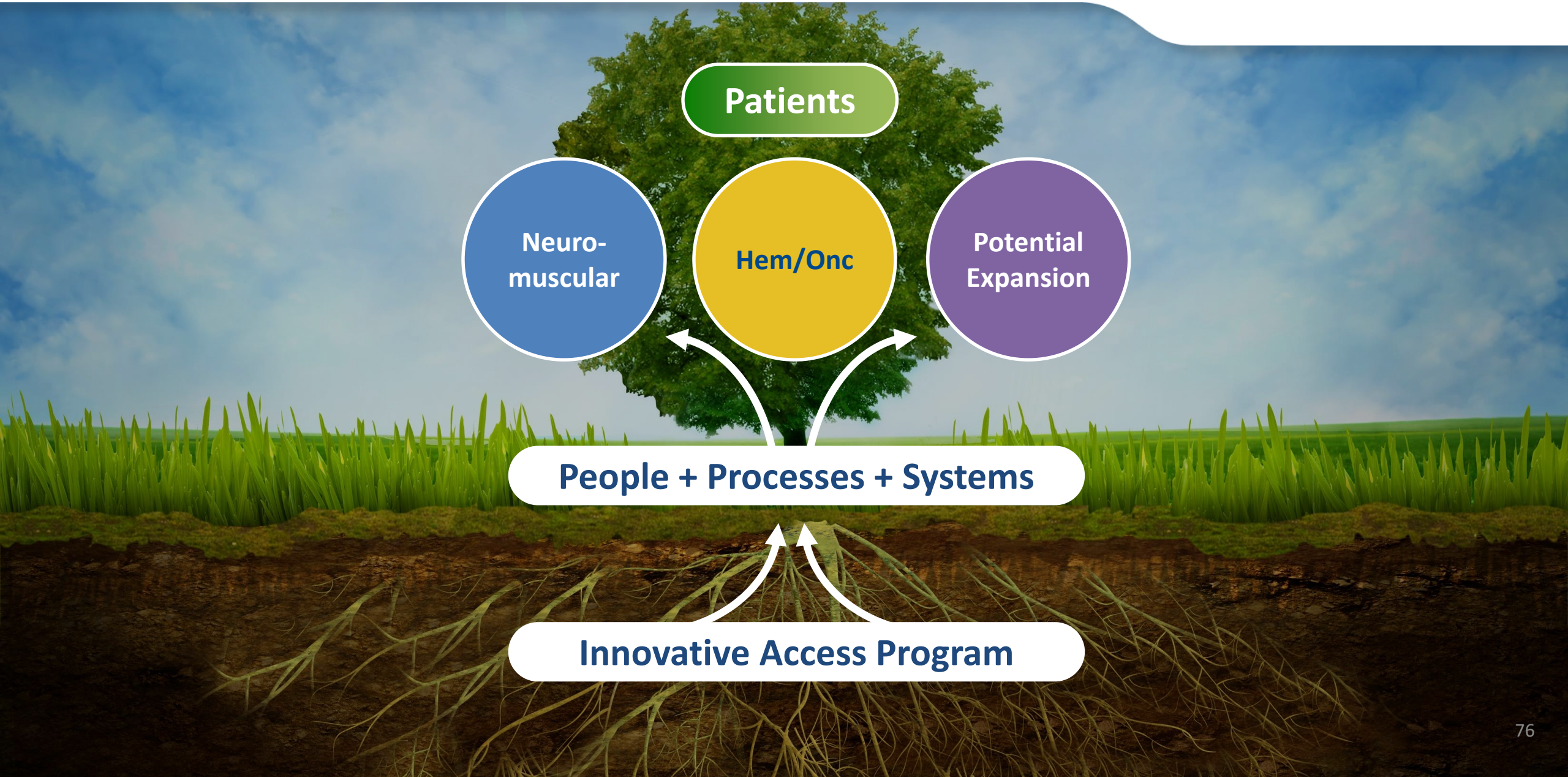
building commercial capabilities

01

WHAT

product franchise / pipeline

argenx 2021:
Becoming an Integrated Immunology Global Biotech



Building Immunology Franchises

Neuro Inflammation | Poly Neuropathies | Myositis

Expansion Opportunities

MG | CIDP | MMN

Neuro-muscular

BMT | AIHA | ANCA | Leukemia Lymphomas

Expansion Opportunities

MALIGNANT HEME

AML | MDS

BENIGN HEME

ITP

janssen

50/50

Hem/Onc

Building Immunology Franchises

Complement or IgG-mediated Diseases in Skin and Kidney

- Pemphigus
- Dermatomyositis
- Bullous Pemphigoid

SKIN

- Lupus Nephritis
- AMR
- Nephropathies
- IRI

KIDNEY

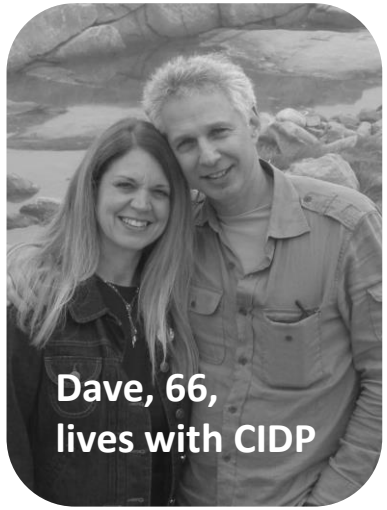
Potential Expansion

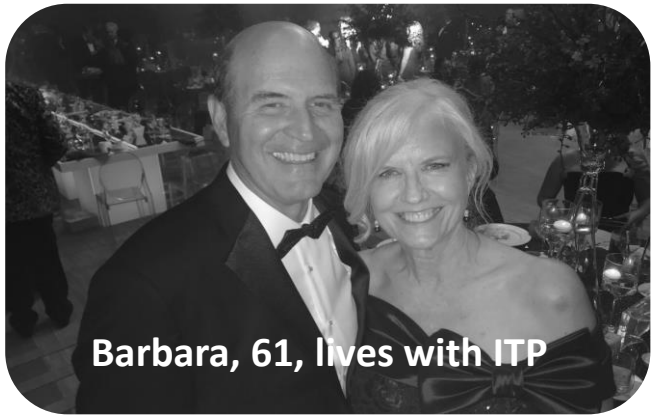
02

WHY

unmet medical need / market opportunity

High Unmet Medical Need



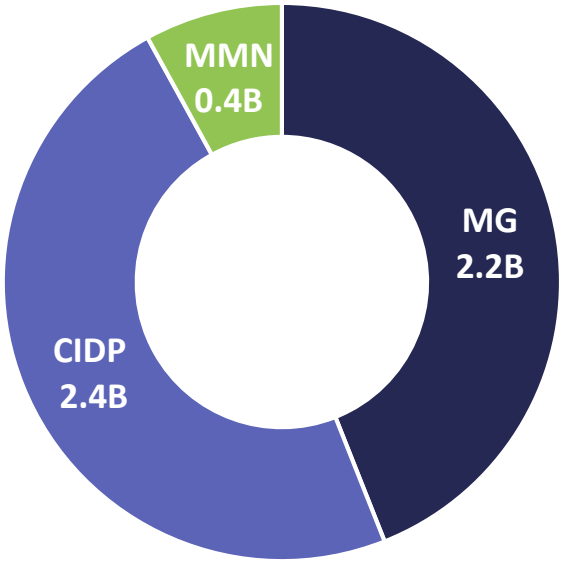


…the …ly a …oint; it's not solving my issues, but rather just patching them."

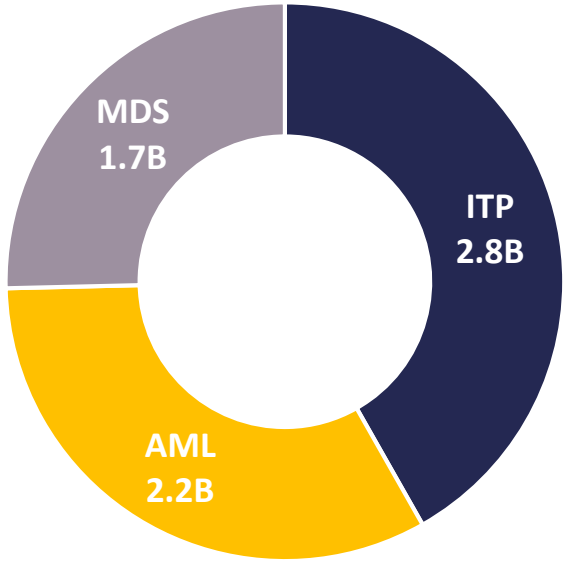
— Lauren, 32, lives with generalized MG

Our Franchises Sit in High-Value Rapid-Growth Markets

Neuromuscular
>$5B (CAGR ~10%+)



Hem/Onc
~$7B (CAGR ~10%)



- ***~370,000 patients***
- Double digit CAGR

Efgartigimod ARGX-117 Cusatuzumab

03

HOW

building commercial capabilities

Building Efgartigmod/MG Franchise

Building Efgartigmod/MG Franchise

Building Efgartigmod/MG Franchise

Building the Experienced, Diverse Organization

Business Analytics

Distribution

Finance

Patient Advocacy

Human Resources

Legal / Compliance

Marketing

Market Access

Pharmacovigilance

Strategic Insight

Sales Leadership

Regulatory Affairs

Medical Affairs

Japan GM

EU Commercial Dev Leader

2019 R&D Day

May 22, 2019 - New York

5 LAUNCHES IN 5 YEARS

2019 R&D Day

May 22, 2019 - New York