

Developing Highly Differentiated Antibody Therapeutics

June 2017



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Company Highlights

Differentiated therapeutic antibodies pioneering in severe autoimmune diseases & cancer



Novel concept in auto-immunity

- **ARGX-113:** potential first-in-class FcRn antagonist targeting array of IgG mediated AI diseases
 - Phase 1: favorable safety profile; IgG reduction up to 85%
 - Phase 2: ongoing in myasthenia gravis and immune thrombocytopenia



Deep pipeline with multiple shots on goal

- **ARGX-110:** potential first-in-class CD70 antagonist in Phase 1/2 in CTCL and AML
- 4 clinical stage programs; 3 preclinical programs; Innovative Access Program



Powerful technology suite

- **SIMPLE Antibody™:** Human V-regions sourced from llama unlock **novel & complex targets**
- **NHance®, ABDEG™, POTELLIGENT®:** Fc engineering to augment natural properties of antibodies



Validating selective partnerships

- **abbvie:** **ARGX-115** (IO-focused novel target GARP)
 - \$40mm upfront and up to \$625mm in potential milestone payments
- 4 additional partnerships designed to maximize value of our platform in non-core areas



Well financed to execute plan

- **Strong cash position:** €85.0mm March 2017 + €102mm Nasdaq IPO May 2017
- Blue chip investor base: more than 50% US shareholders

Pipeline

- ARGX-110 Phase 2 study in CTCL patients initiated (April '17)
- ARGX-113 Phase 2 study in MG patients 50% recruited (May '17)



Partnerships


- ARGX-115: 1st \$10mm preclinical milestone payments received from AbbVie (May '17)
- ARGX-112: 2nd undisclosed preclinical milestone received from LEO Pharma (June '17)

Financing

- Successful IPO on Nasdaq (ticker: ARGX) (May '17)
- ~\$115mm raised (increased from \$65mm initial base size)
- US shareholding expanded above 50%
- US analyst coverage expanded
 - B. Peaker (Cowen), T. Tenthoff (Piper Jaffray), M. King (JMP), D. Nierengarten (Wedbush)
- Use of proceeds
 - Clinical development of ARGX-113 for the treatment of autoimmune diseases
 - Expand applications of ARGX-113 to develop a subQ formulation & explore additional indications
 - Clinical development of ARGX-110 for the treatment of hematological malignancies



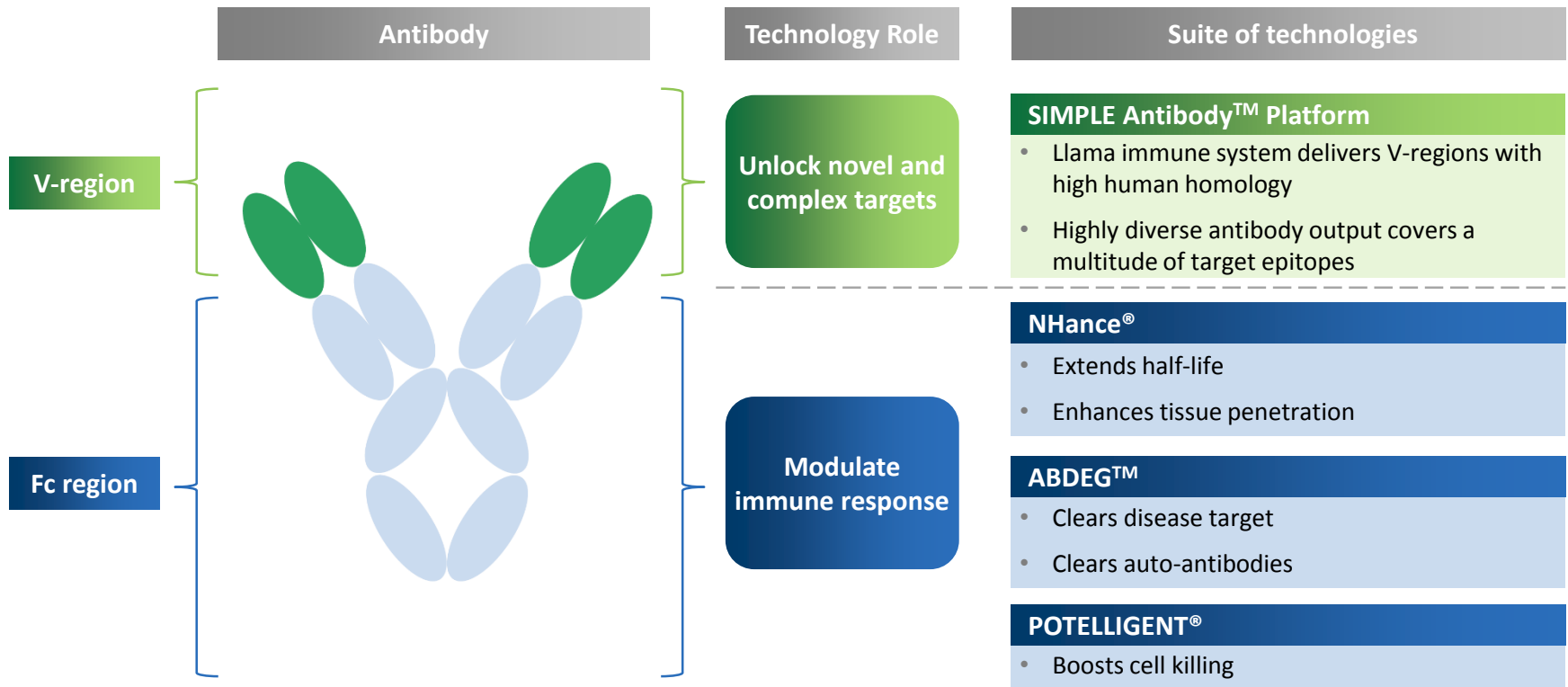
Product Candidate	Target	Indication	Preclinical	Phase 1	Phase 2	Phase 3	Next Milestone / Commentary
Wholly-Owned Product Candidates							
ARGX-113 (efgartigimod)	FcRn	Myasthenia Gravis	<div></div>				1Q18: Announce Phase 2 topline results
		Immune Thrombocytopenia	<div></div>				2H18: Announce Phase 2 topline results
		Chronic Autoimmune Diseases	<div></div>				2H17: Initiate Phase 1 clinical trial
ARGX-110 (cusatuzumab)	CD70	T-Cell Lymphoma	<div>Phase 1/2</div>				2H18: Announce Phase 2 topline results CTCL
		Acute Myeloid Leukemia	<div>Phase 1/2</div>				YE17: Interim update Phase 2 CTCL and Phase 1 dose-escalation in AML/MDS
ARGX-111	c-MET	Solid Tumors / Blood Cancer	<div></div>				Intend to partner
Partnered Product Candidates							
ARGX-109 (gerilimzumab)	 IL-6	Rheumatoid Arthritis	<div></div>				Eligible for up to €32.5mm in milestones, royalties & additional shares of Bird Rock stock
ARGX-112	 IL-22R	Skin Inflammation	<div></div>				Eligible for up to ~€100mm in milestones and tiered royalties
ARGX-115	 GARP	Cancer Immunotherapy	<div></div>				Received \$50mm so far; eligible for up to \$625mm milestones & tiered royalties
ARGX-116	 ApoC3	Dyslipidemia	<div></div>				Eligible for double-digit royalties and exclusive option to license the program

- In March 2017 we obtained the exclusive license option for the rights for an antibody against a novel complement target
- argenx has an antibody discovery alliance with  **Shire** focused on multiple rare disease targets

Our Suite of Technologies



A brown and white llama stands in a grassy field, looking towards the left. The background shows a blurred mountain range under a clear blue sky. The llama's fur is a mix of brown and white, with its head turned slightly to the left. The grass in the foreground is green and slightly out of focus.



We apply our unique suite of technologies to create differentiated product candidates against novel targets

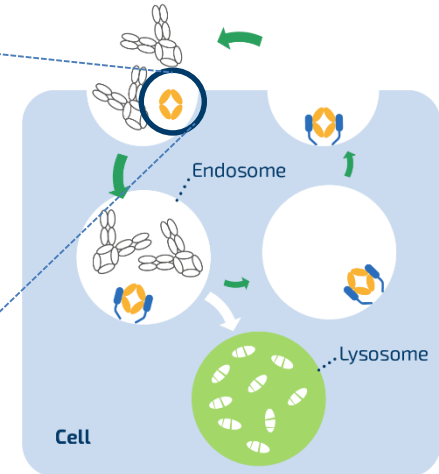
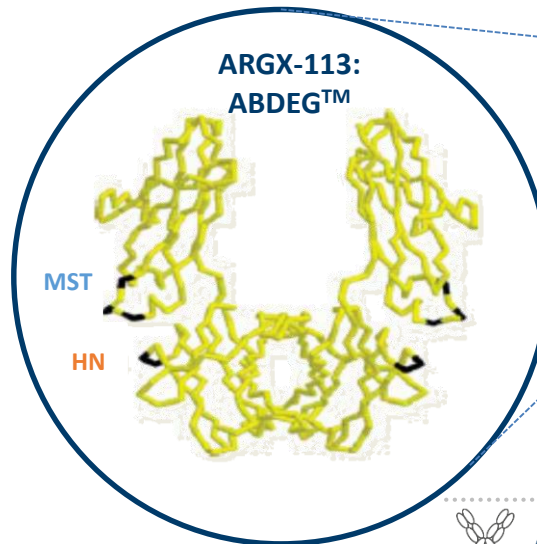
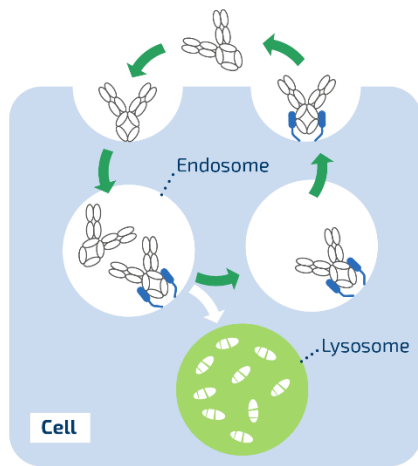


ARGX-113: Advancing to Clinical Proof-of-Concept

ARGX-113: Lead Program Based On Novel Target FcRn

An innovative approach to eliminate IgG auto-antibodies

IgG antibodies recycle through FcRn⁽¹⁾... ..ARGX-113 potently blocks FcRn... ..leading to IgG elimination



- ARGX-113 is a human IgG1 Fc-fragment that utilizes ABDEG™ Fc engineering technology⁽²⁾⁽³⁾
- ARGX-113 targets and binds to FcRn blocking the recycling of IgG leading to an elimination of IgG antibodies
 - Demonstrated 50% to 85% reduction of circulating IgG antibody levels in Phase 1 trial
- Pathogenic IgG antibodies mediate multiple autoimmune diseases
 - 30% pathogenic IgG reduction believed to be clinically meaningful in MG
- Phase 2 focus on myasthenia gravis (MG) and immune thrombocytopenia (ITP), data est. 1Q2018/2H2018

(1) Roopenian et al. 2007, Nat Rev Immunol.

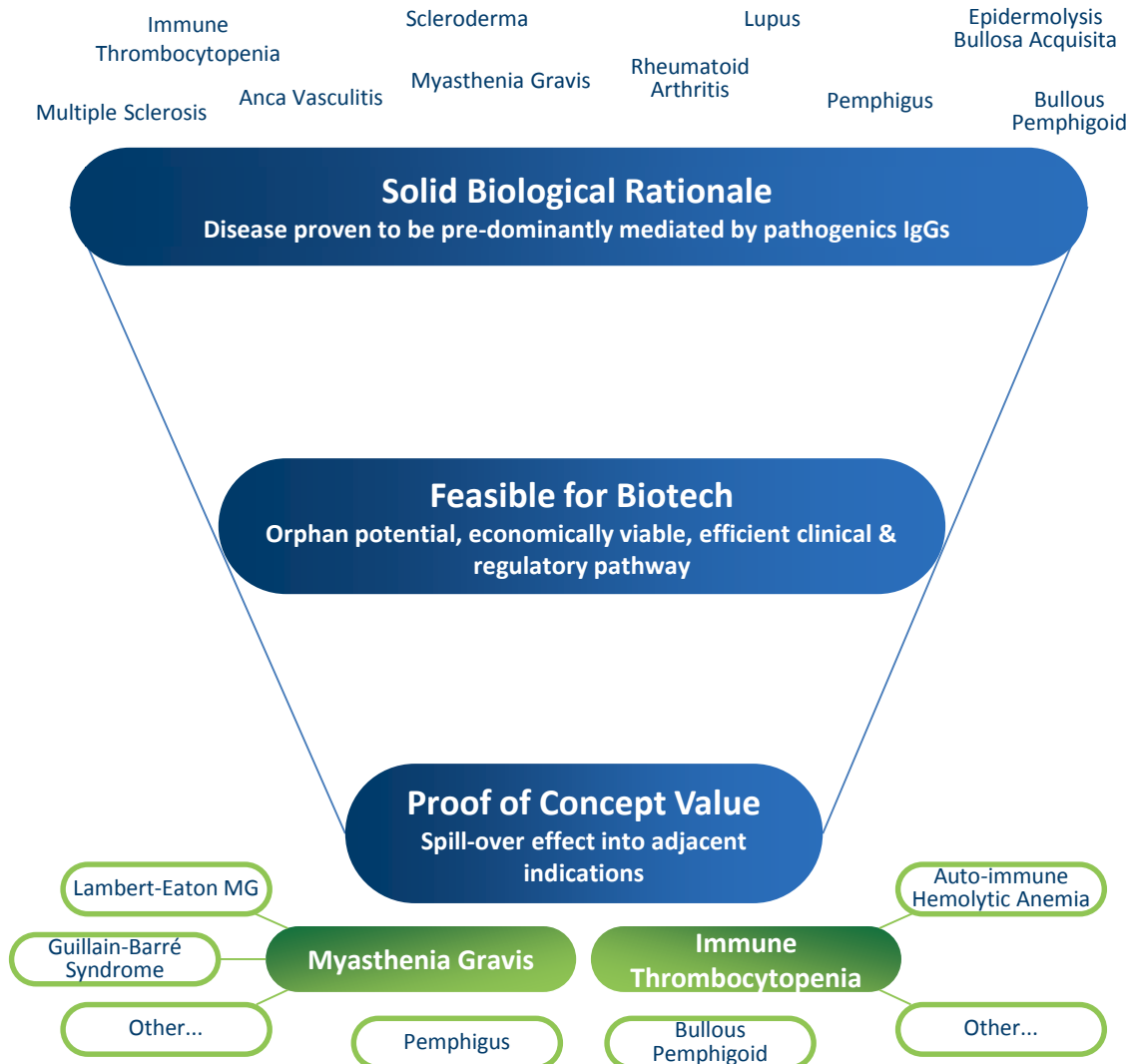
(2) Vaccaro et al. 2005, Nat Biotech.

(3) argenx data

ARGX-113: Pipeline-In-Product Opportunity

Prioritizing IgG auto-antibody mediated diseases

Landscape of IgG severe auto-immune diseases (selection)



What is Myasthenia Gravis?

- Rare auto-immune disorder; 64,000⁽¹⁾ patients in U.S., 55,000⁽²⁾ with generalized MG, affecting all ages and both genders
- MG is associated with muscle weakness, it can be life threatening if respiratory muscles are affected
- Symptoms include: **Life-threatening chocking**, **muscle dislocation**, **eyelid fatigued**, **pain**, problems: seeing, **talking**, **tired**, **trouble walking**

Limited current treatment options

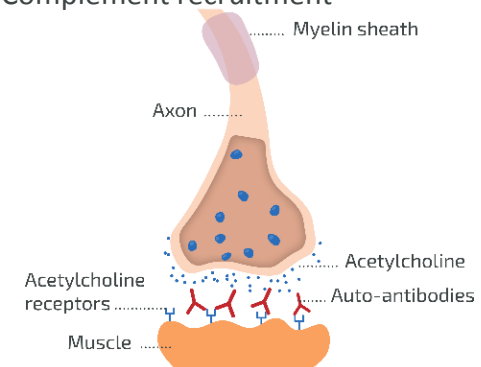
- Limited treatment options
 - Cholinesterase inhibitors
 - Corticosteroids
 - Immunosuppressants
 - IVIg or Plasmapheresis (exacerbations or rescue)
 - Thymectomy (minority of patients)
- Severe side effects of current treatment options:
Injury, **liver malignancy**, **osteopenia**, **osteoporosis**, **cataracts**,
Depression, **hypertension**, **hematological suppression**,
headache, **disfigurement**, **hypertension**, **infection**, **thrombosis**
- IVIg and Plasmapheresis place a heavy cost burden on healthcare system in the acute setting (~\$79,000⁽³⁾ and ~\$101,000⁽³⁾ respectively)



Myasthenia Gravis Cause

Auto-antibodies (IgG type) destroy neuromuscular junction:

- Blocking of Acetylcholine Receptors (AChRs)
- Cross-linking + internalization of AChRs
- Complement recruitment



(1) Philips et al. 2003, Ann N Y Acad Sci.
(2) Drachman et al. 1993, New Eng J Med.
(3) Heatwole et al. 2011, J Clin Neuromuscul Dis.

Auto-Antibody Levels (IgGs) Correlate With MG Disease Score

>30% auto-antibody reduction clinically meaningful

Treatment*	Plasmapheresis	Immuno-adsorption	IVIg
Decrease in auto-antibody levels (%) after treatment	62.6 ± 0.9	55.1 ± 3.2	28.9 ± 3.8
Decrease in disease score (%) after treatment	60.8 ± 3.5	42.4 ± 4.2	23.8 ± 3.7
Clinical efficacy rate after 14 days**	12/15	7/10	6/15
Duration of hospital stay (days)	12.80 ± 0.28	13.50 ± 0.50	16.00 ± 0.50

* Comparison between 3 cycles of Plasmapheresis/Immunoabsorption every 24h-48h and 5 cycles of IVIg every 24h

** Clinically effective if disease score has improved by >50% 14 days after treatment

Degree of auto antibody reduction correlates with clinical improvement and reduced hospital stay

What is Immune Thrombocytopenia?

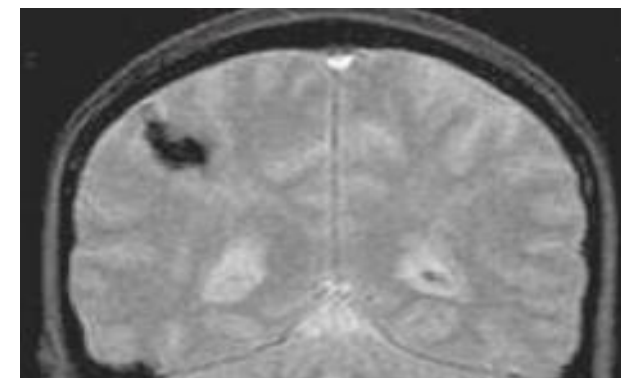
- Rare bleeding disease; estimated 72,000⁽¹⁾ patients in US, more frequent in females and patients over 60
- Symptoms rare from mild bruising to severe bleeding
- Symptoms include: **Fatigue**, emotional strain, **impact on work**, **Fear of bleeding**, impact on social activities, **Bruising**

Limited current treatment options

- Limited treatment options
 - Multiple iterations on corticosteroids & IVIg
 - Immunomodulatory agents
 - TPO-mimetics & splenectomy
- Severe side effects from current treatments:

Anaphylaxis, **anorexia**, **backache**, **cancer**, **cataracts**, **Depression**, **Diabetes**, **fatal hemolysis**, **hepatitis**, **Hypertension**, **infections**, **infusion relation reaction**, **leukoencephalopathy**, **nausea**, **osteoporosis**, **psychosis**, **sweating**, **neutropenia**, **thrombosis**, **vomiting**, **weakness**

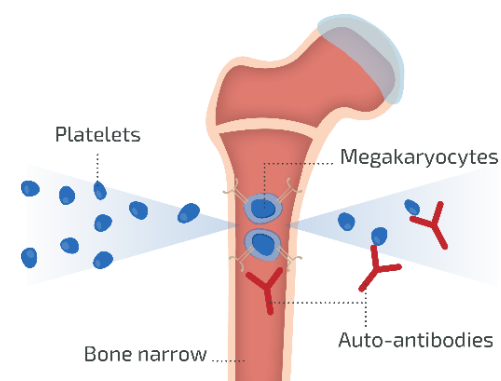
- Romiplostim and Eltrombopag, last-line therapy for ITP and have generated global revenues of \$584 million⁽²⁾ and \$635 million⁽³⁾ in 2016



Immune Thrombocytopenia Cause

Auto-antibodies (IgG type) destroy blood platelets:

- Increased platelet removal
- Reduced platelet production

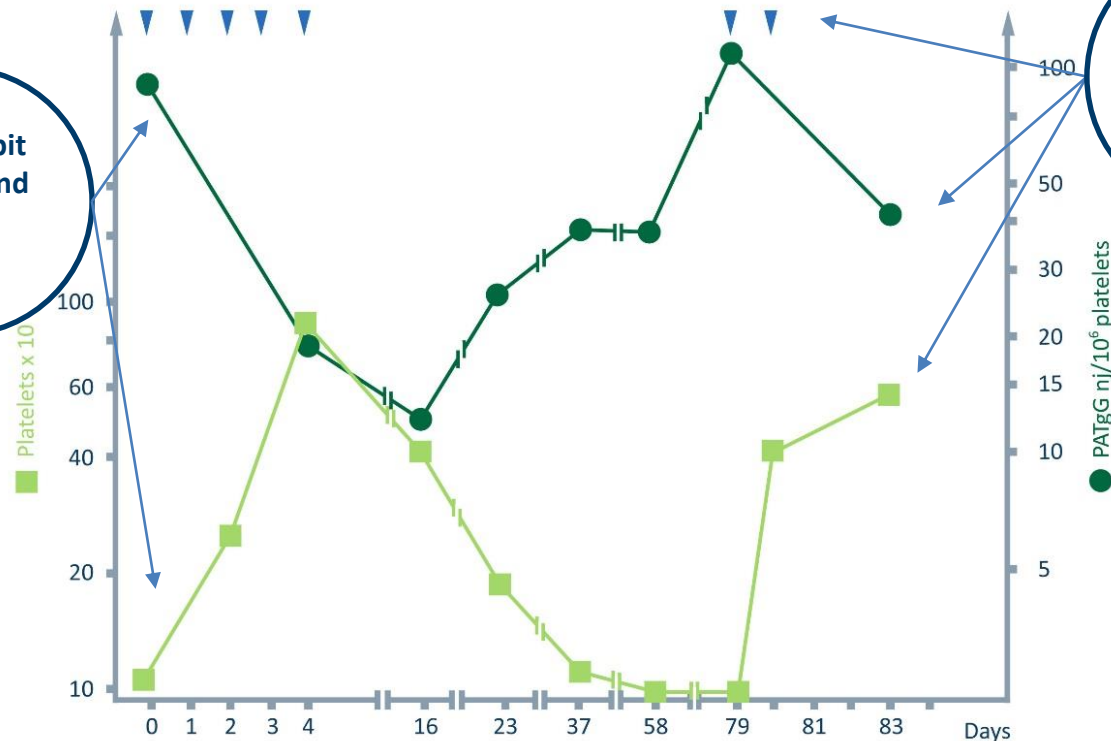


(1) Saleh et al. 2015, Curr Med Res Opin.; Terrell et al. 2012, Am J Hematol.; Grace et al. 2012, Pediatr Blood Cancer.

(2) Amgen Inc. 2016, Form 10-K.

(3) Novartis Annual Report 2016

Platelet levels during IVIg treatment in Adult Immune Thrombocytopenia



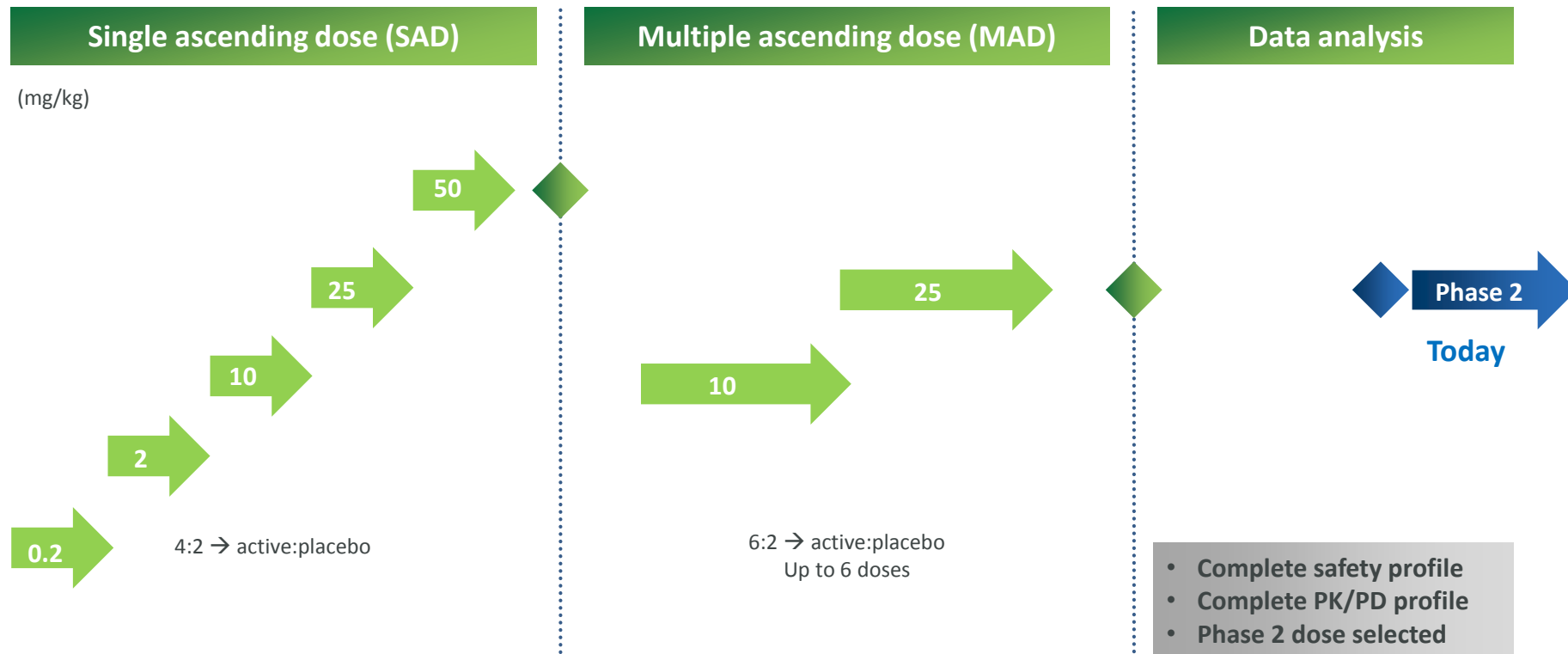
Auto-antibodies inhibit platelet production and accelerate platelet destruction

Therapy aimed at reducing auto-antibodies like IVIg (shown), plasmapheresis and immunoadsorption results in platelet increase

▼ = IVIg treatment ● = Auto-antibody level ■ = Platelet counts

ARGX-113: Favorable Safety & Tolerability Profile

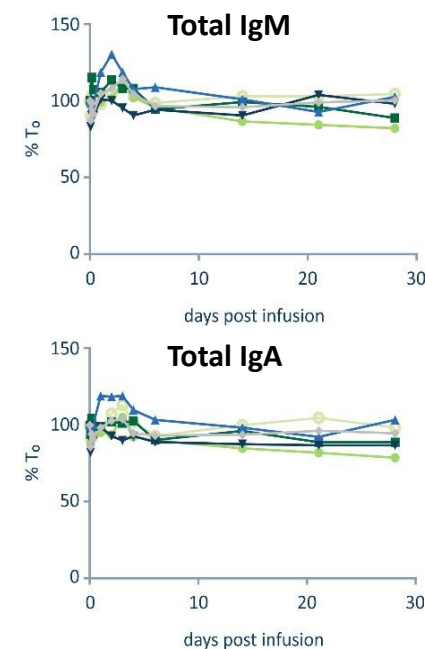
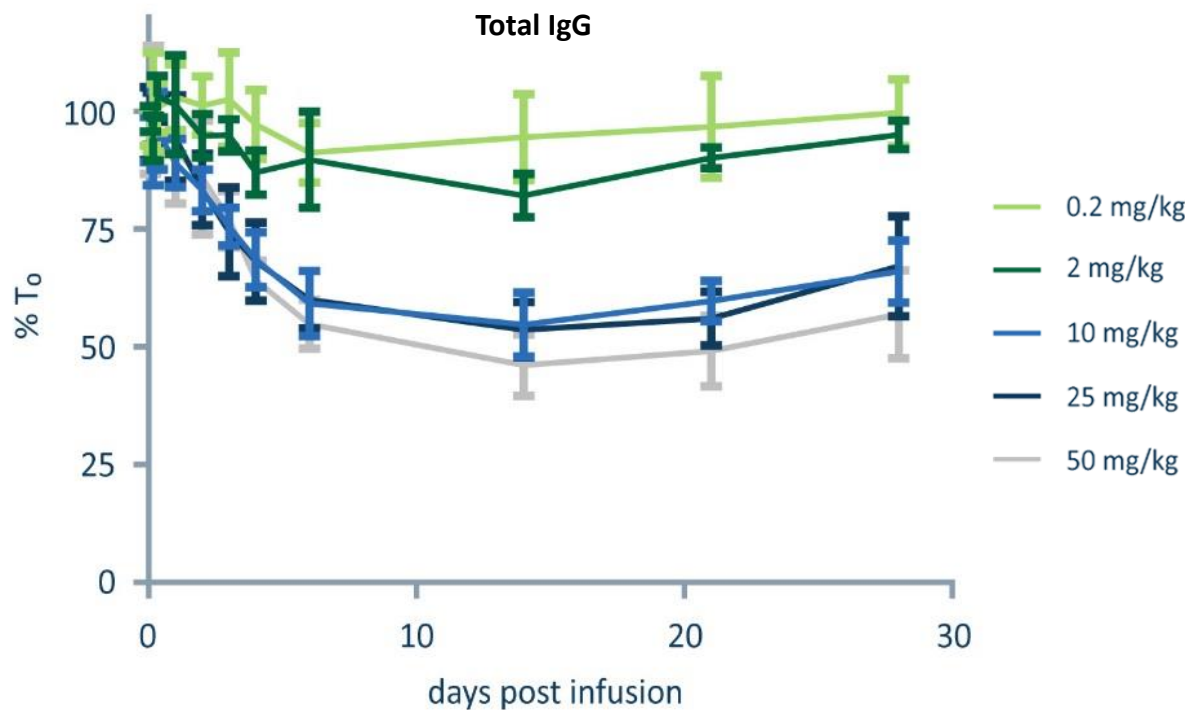
Phase 1 design: Double-blind, placebo-controlled trial in healthy volunteers



- SAD & MAD studies completed according to plan (62 healthy volunteers in total)
- Reported to be well tolerated in single and multiple doses of up to 25 mg/kg

ARGX-113: Selective IgG Reduction

Single ascending dose escalation study (SAD) in healthy volunteers (single 2hr infusion)

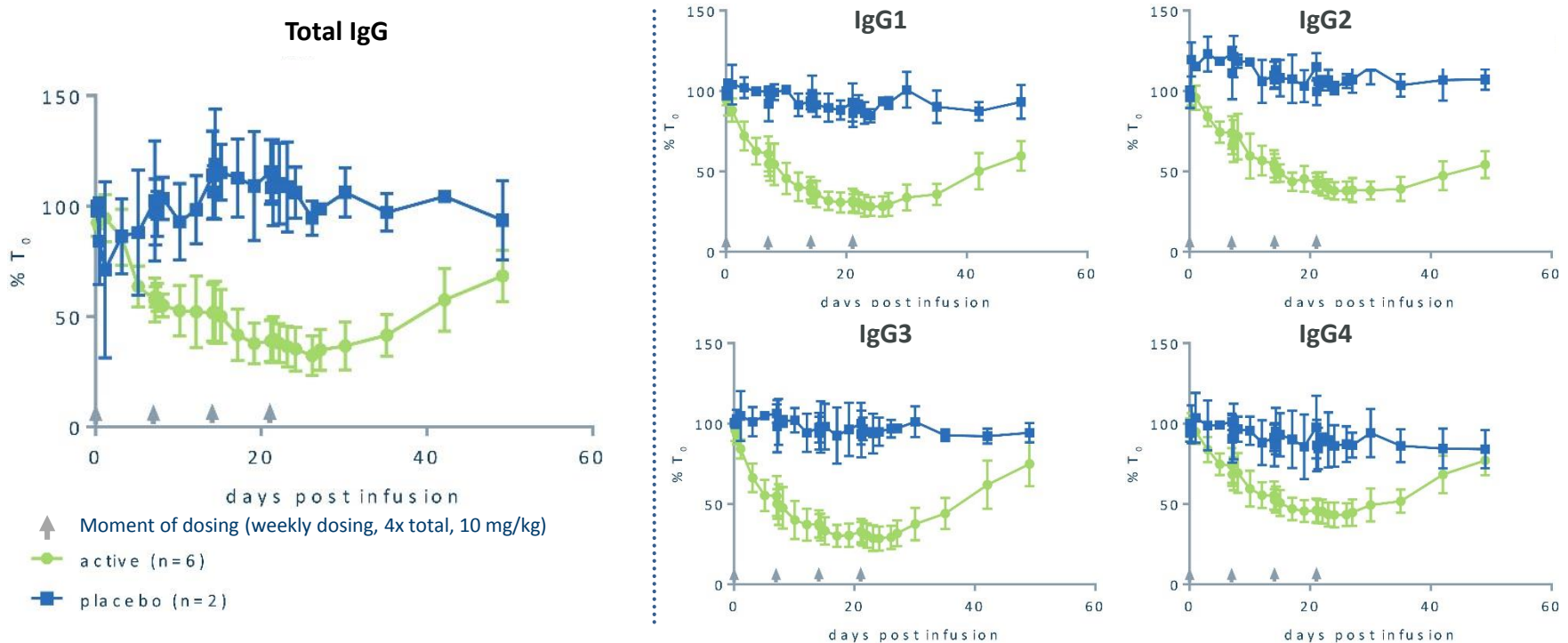


- ~50% IgG reduction (maximal PD effect) as of 6 days after infusion
- Selective IgG reduction, no significant reductions in IgM/IgA and albumin levels
- Low IgG levels maintained for more than four weeks after the last dose
- Saturation of PD effect observed at 10 mg/kg dose

ARGX-113: Potent and Lasting IgG Reduction

PD data multiple ascending dose (MAD) study in healthy volunteers

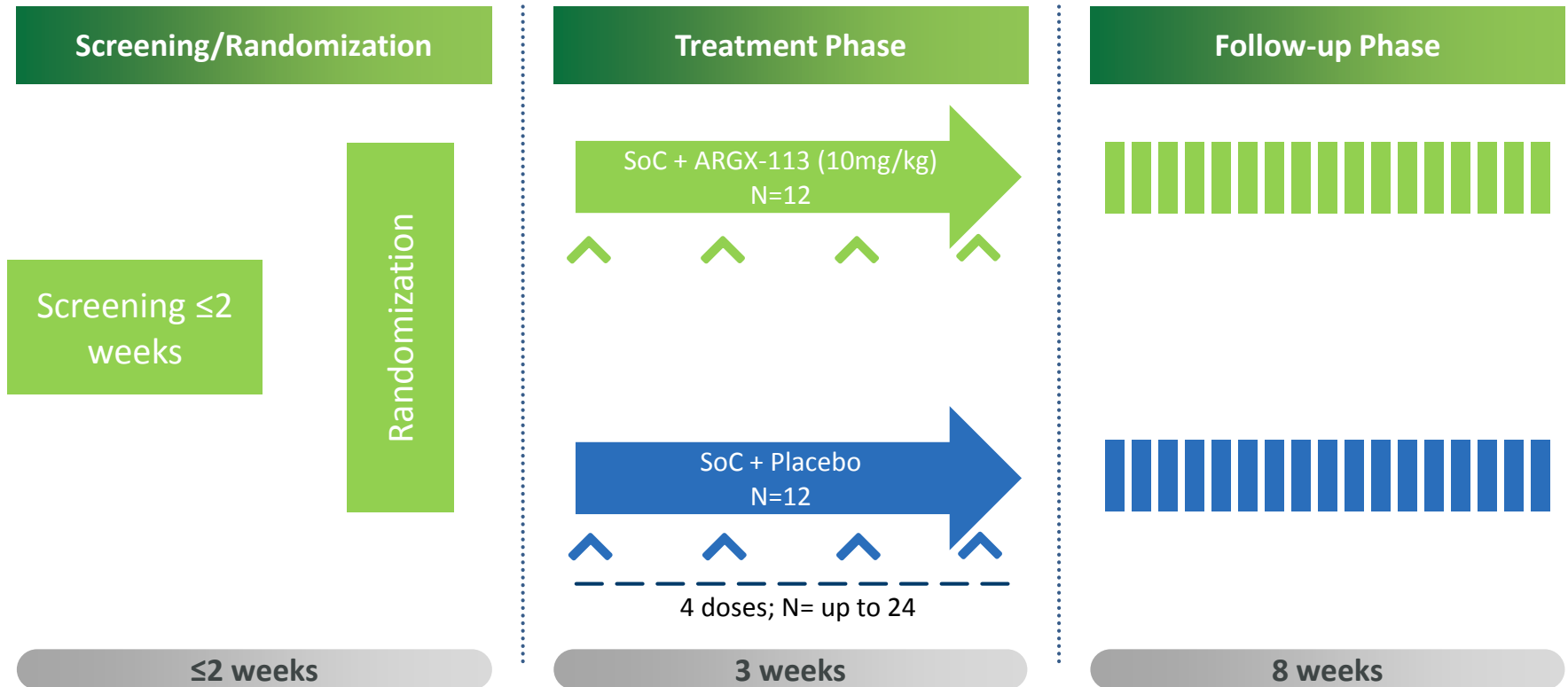
Dosing 10 mg/kg; every 7 days



- Potent IgG reduction across isotypes
- IgG reduction: 50% achieved in 1 week; up to 85% maximum reduction
- After last dose, IgG levels remain reduced by 50% or more for ~3 weeks, return to baseline after > 1 month
- Comparable data for 25 mg/kg, every 7 days (data not shown)

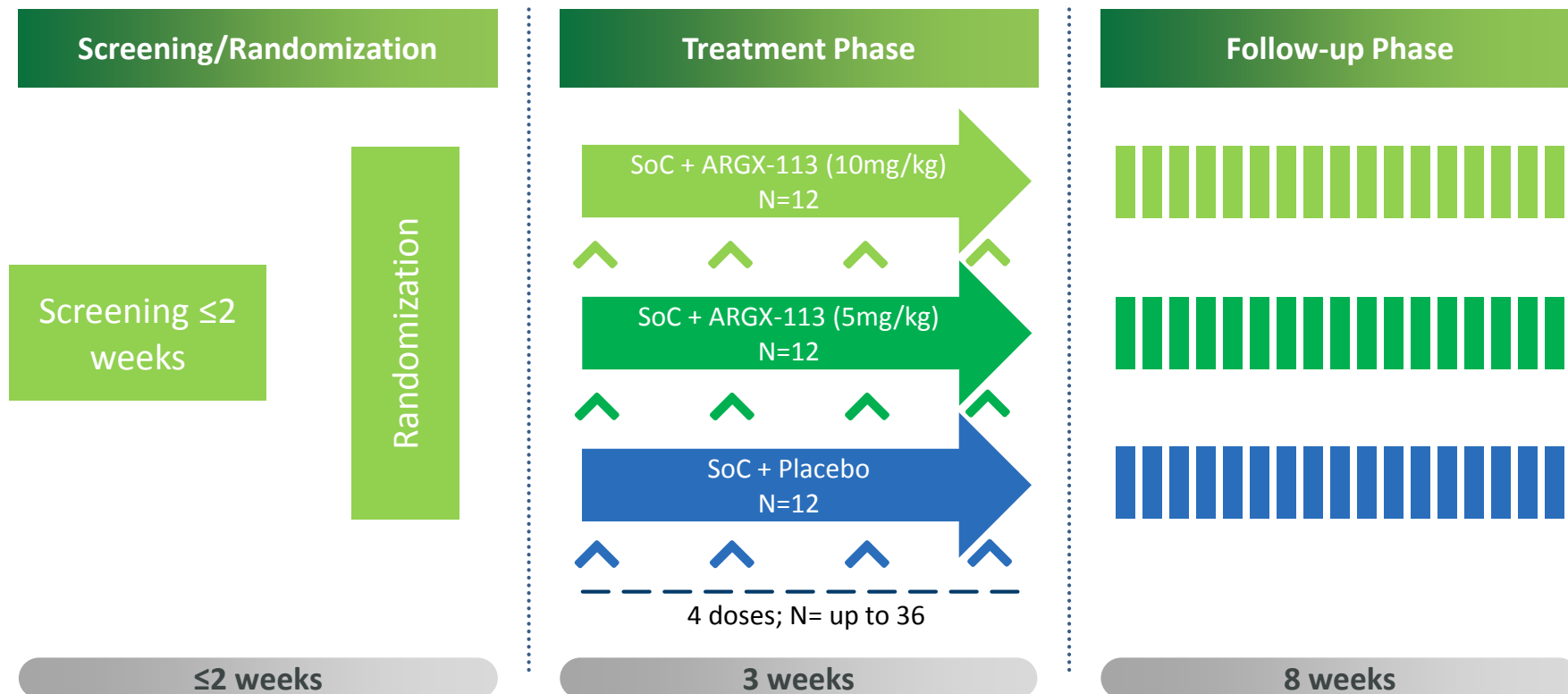
ARGX-113 in MG: Phase 2 Trial Design

Study
50% recruited
May 2017



- Population: MG patients with generalized muscle weakness with total MG-ADL score $\geq 5^*$
- Primary Objectives: Evaluate safety and tolerability
- Secondary Objectives:
 - (i) Evaluate efficacy, impact on quality of life and immunogenicity
 - (ii) Assess pharmacokinetics (PK) and pharmacodynamics (PD) markers



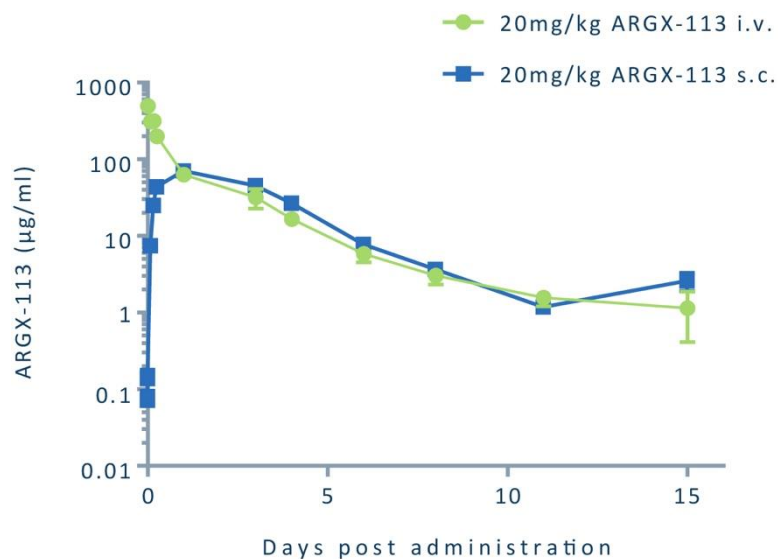


- Population: ITP patients with platelet levels $< 30 \times 10^9/L$
- Primary Objectives: Evaluate safety and tolerability
- Secondary Objectives: (i) Evaluation of efficacy based on platelet counts, use of rescue treatment & bleeding events
(ii) Assess pharmacokinetics (PK) and pharmacodynamics (PD) effect
(iii) Evaluate immunogenicity

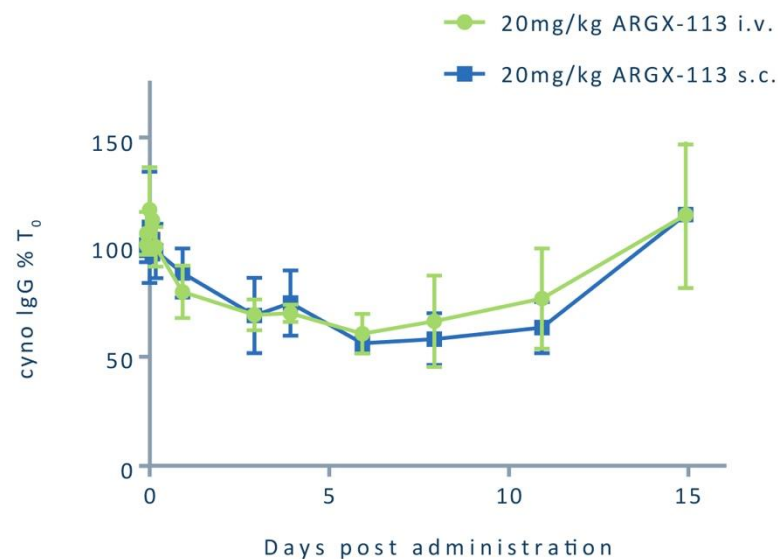
ARGX-113: Feasibility of SubQ Dosing

Exploring SubQ formulations for larger patients populations (chronic, ex hospital)

PK single dose administration: IV vs SubQ (in cyno)



PD single dose administration: IV vs SubQ (in cyno)



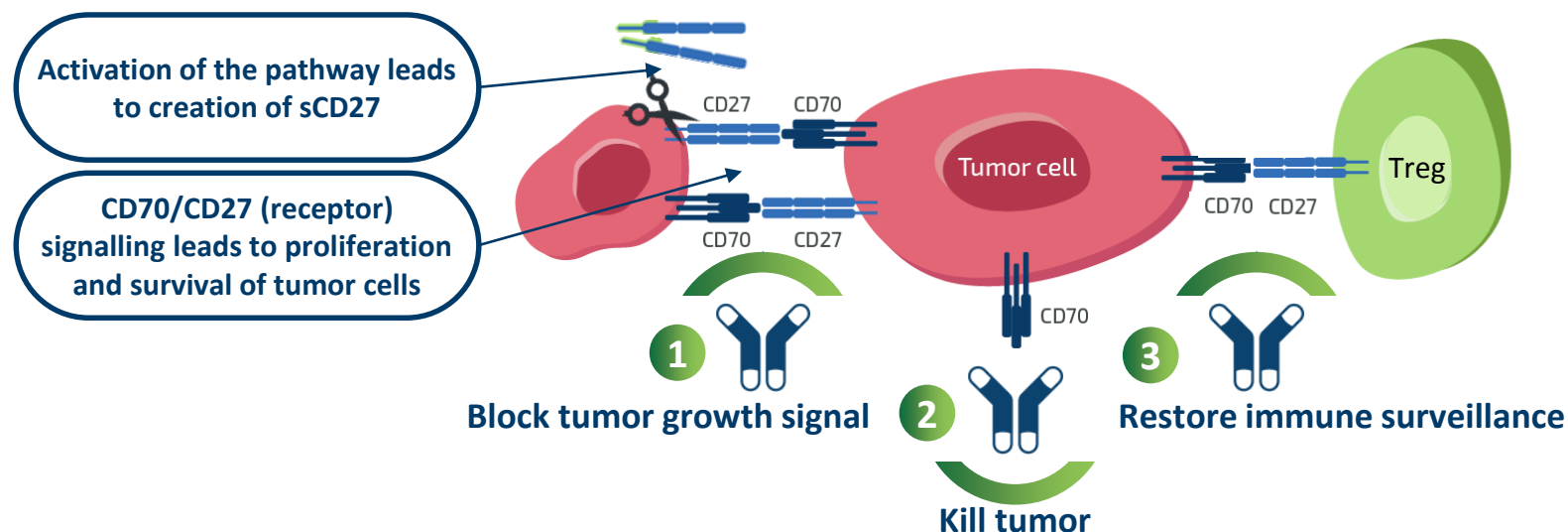
- Comparable PK and PD of IV versus SubQ dosing in preclinical studies demonstrated:
 - Comparable half life
 - Favorable bio-availability of the compound in SubQ dosing (> 75%)
 - Comparable reduction of IgGs with single dose; up to 50%



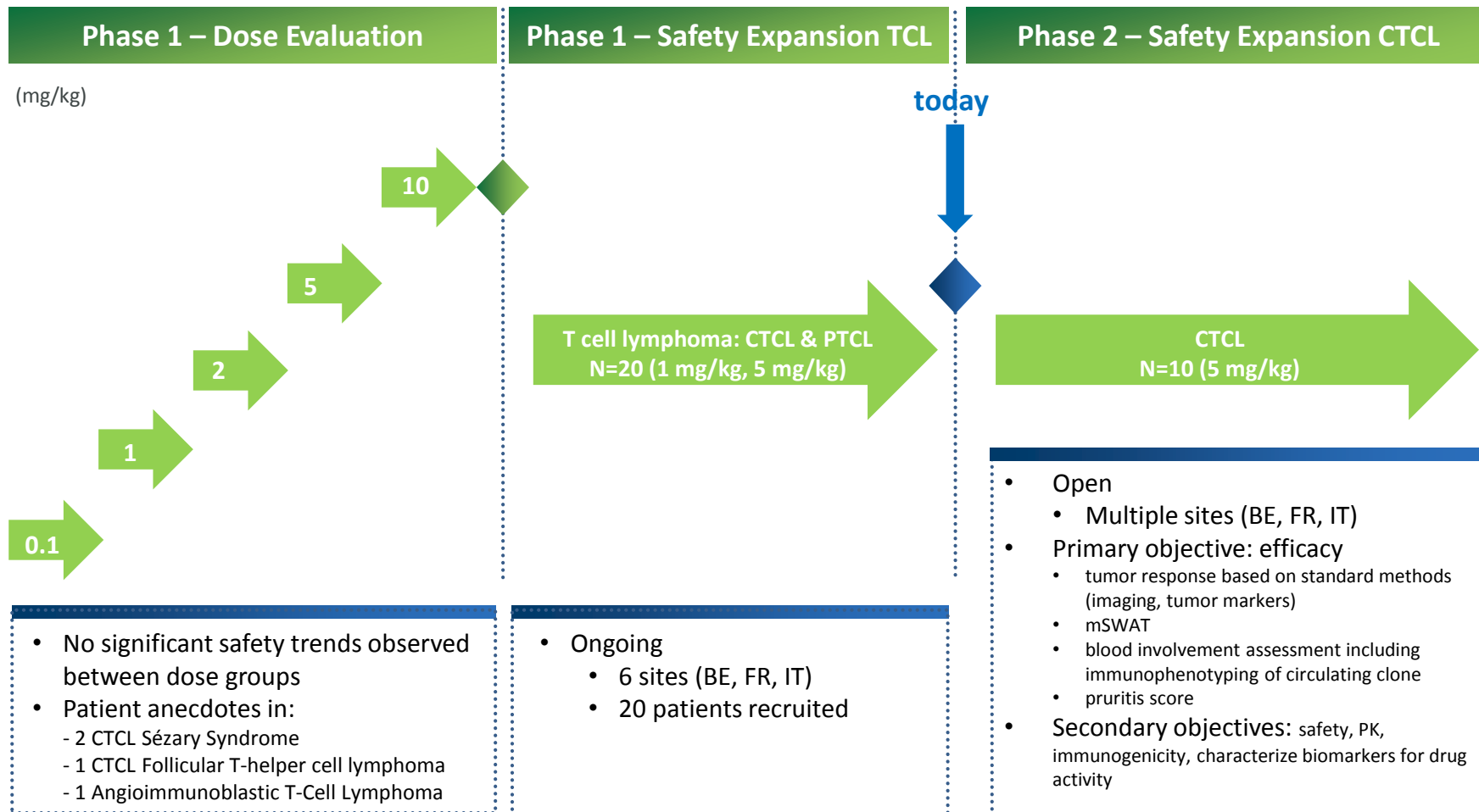
ARGX-110: Phase 1 / 2 Mono & Combo Therapy

ARGX-110: Lead Cancer Program Based On Novel Target CD70

Three distinct modes of action to target CD70+ tumor cells



- ARGX-110 is a SIMPLE Antibody™, equipped with the POTELLIGENT® Fc engineering technology
- ARGX-110 targets CD70 to block CD27 interaction, kill CD70 expressing cells and restore immune surveillance
- Soluble CD27 is a biomarker
- Phase 1: encouraging safety & tolerability profile and promising preliminary signs of efficacy in CTCL
- Focus on two rare & aggressive hematological tumors: CTCL and newly diagnosed AML / high-risk MDS
 - Interim results from dose escalation part of Phase 1/2 AML/MDS trial expected YE:2017
 - Interim POC data from Phase 2 CTCL trial expected YE:2017



What is cutaneous T-Cell Lymphoma?

- Rare and incurable sub-type of T-cell lymphoma
- Prevalence (US & Canada): ~ 30,000 & Incidence (US): ~ 3,000⁽¹⁾
- Patients typically diagnosed in their 60ies
- Mycosis fungoides (50%), Sézary syndrome most common types⁽²⁾
- Symptoms include: **Severe Rash, itching,**
- Skin infection often cause of death **tumor Skin Infections**

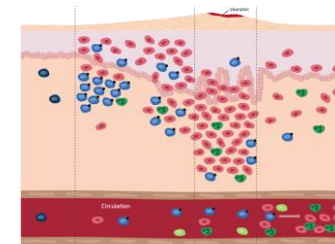
Limited current treatment options

- Initially diagnosed and treated by dermatologist using topical agents (corticosteroids, PUVA, e-beam therapy)
- More advanced stage patients are treated by oncologist using systemic agents which are only moderately effective and not curative
 - Targretin bexarotene (oral) 1st line option – ease of administration
 - Istodax romidepsin (ORR: 34%, mDoR: 13-15 mos)⁽³⁾ 2nd line – complicated dosing and myelosuppression
 - Antifolates (methotrexate, pralatrexate), Campath, chemo (Doxil, CHOP, etc)
- Heavily pre-treated, elderly patients are unfit for aggressive chemotherapy or stem cell transplantation
- Significant unmet need for effective, tolerable, long-lasting CTCL treatments



Cutaneous T-Cell Lymphoma Cause

- Disease aetiology unknown
- Potentially caused by aberrant stimulation of CD4+ T-cells by Langerhans cells, specialized antigen presenting cells in skin
- Malignant T-cells become independent of stimulation by LCs and invade other tissues
- Sézary syndrome is a leukemic variant of CTCL



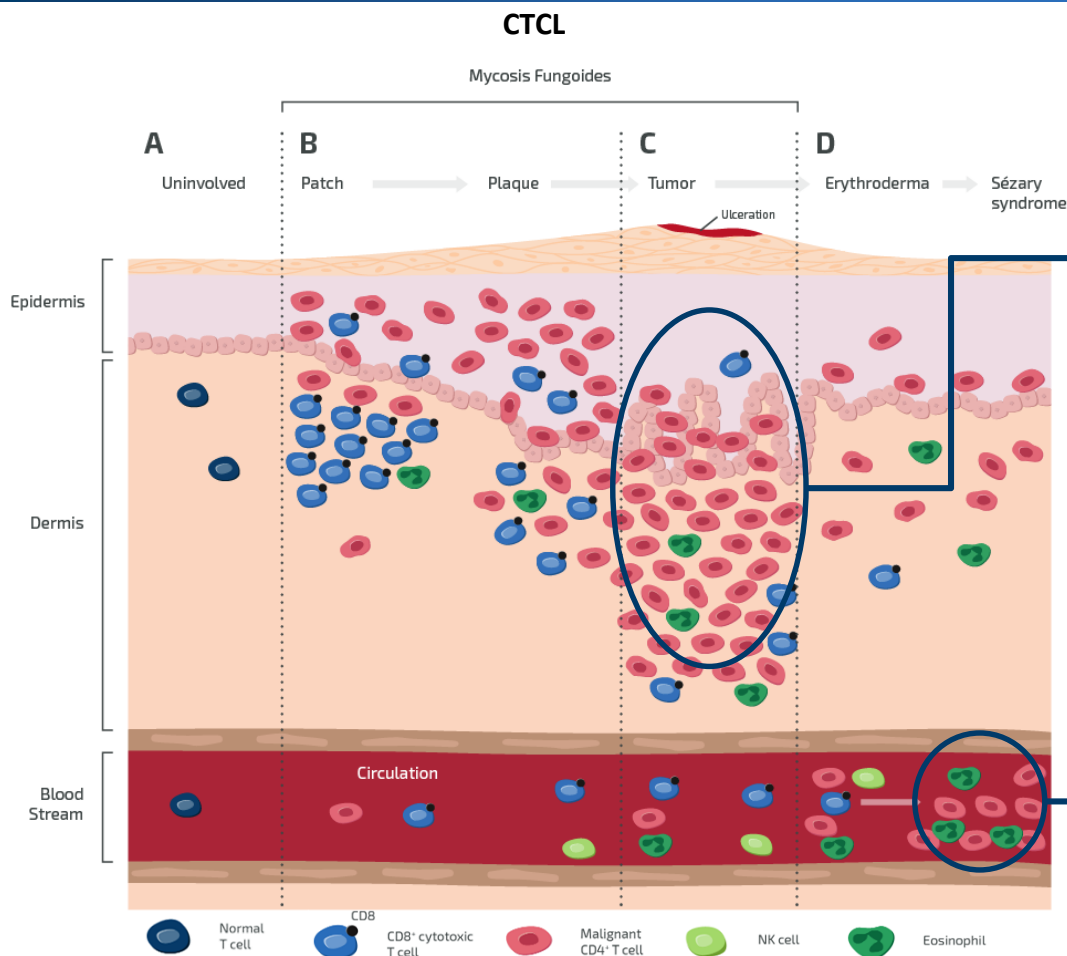
(1) Cutaneous Lymphoma Online Learning Center: <http://www.clfoundation.org/online-learning-center/disease/faq/who-gets-cutaneous-lymphoma-how-many-people-have-it>.

(2) Lymphoma Research Foundation: <http://www.lymphoma.org/site/pp.asp?c=bkLTkaOQLmK8E&b=6300151>

(3) <http://www.istodax.com/hcp/ctcl/study-design/efficacy>

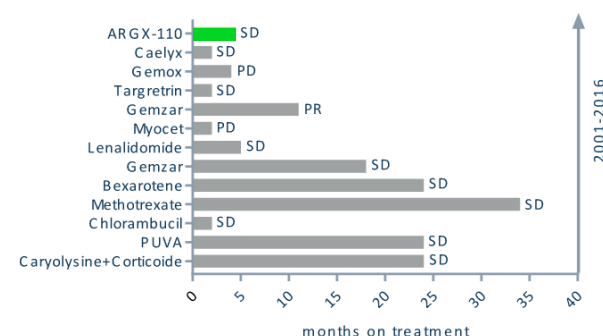
ARGX-110 In Cutaneous TCL

Phase 1-2: Typical patients are elderly and failing multiple lines of previous treatment



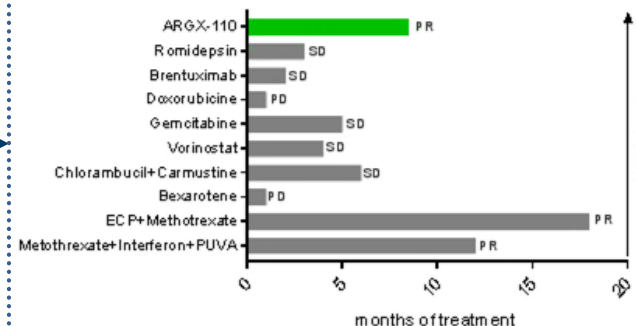
Example Mycosis fungoides (MF) patient

Treatment & best response



Example Sézary Syndrome (SS) patient

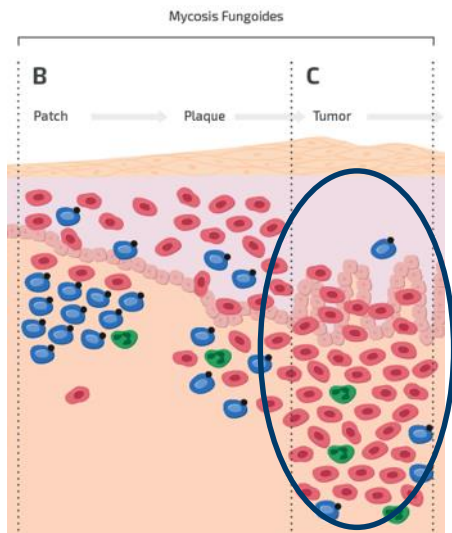
Treatment & best response



ARGX-110: Effect On Malignant Cells In Skin

Patient example 1: Cutaneous TCL – mycosis fungoides (MF)

Typical Mycosis fungoides (MF) patient



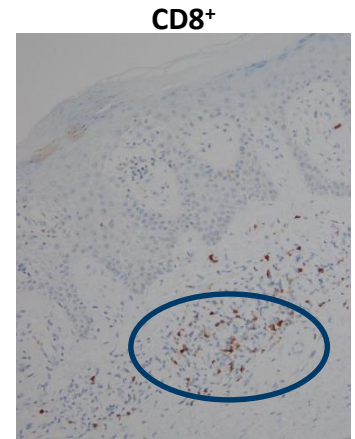
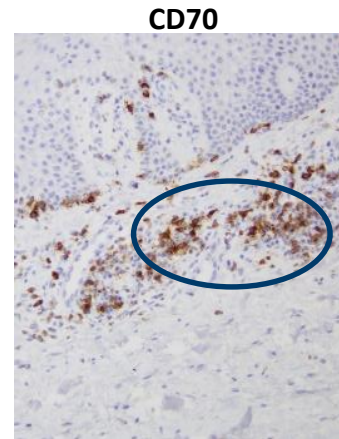
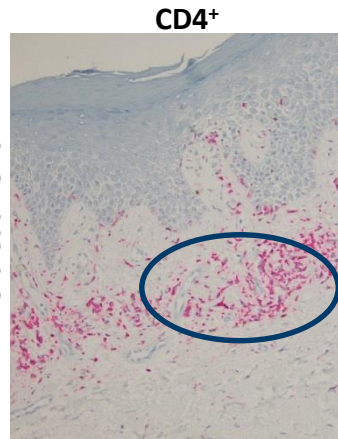
Patient	67 year old male CTCL-MF, diagnosed 2001
Tumor	Skin T4, Nx, M0, B0 (Stage IIIA)
Doses	6

Decrease of CD4+ malignant T-cells

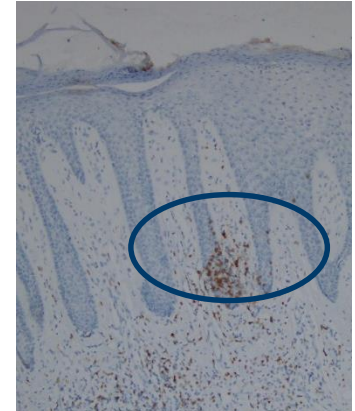
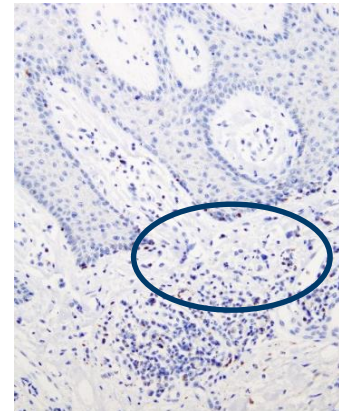
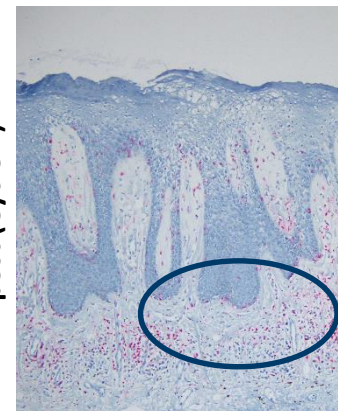
Depletion of CD70+ malignant T-cells

Infiltration of CD8+ T-cells

Pre-treatment



post (Cycle 2)



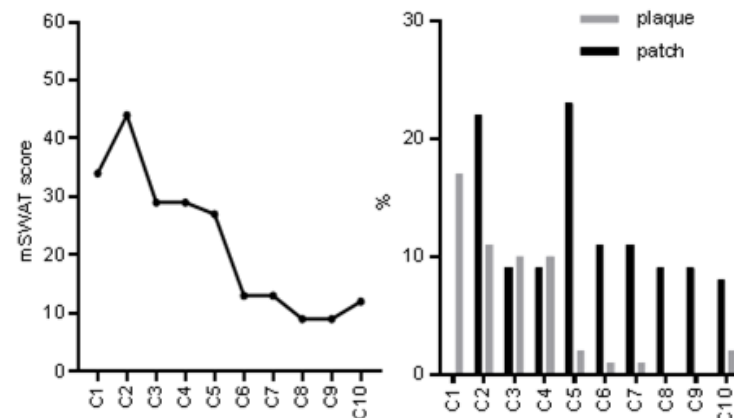
ARGX-110: Improved mSWAT & Skin Lesions

Patient example 2: Cutaneous TCL – mycosis fungoides (MF)

Pre treatment



At cycle 6

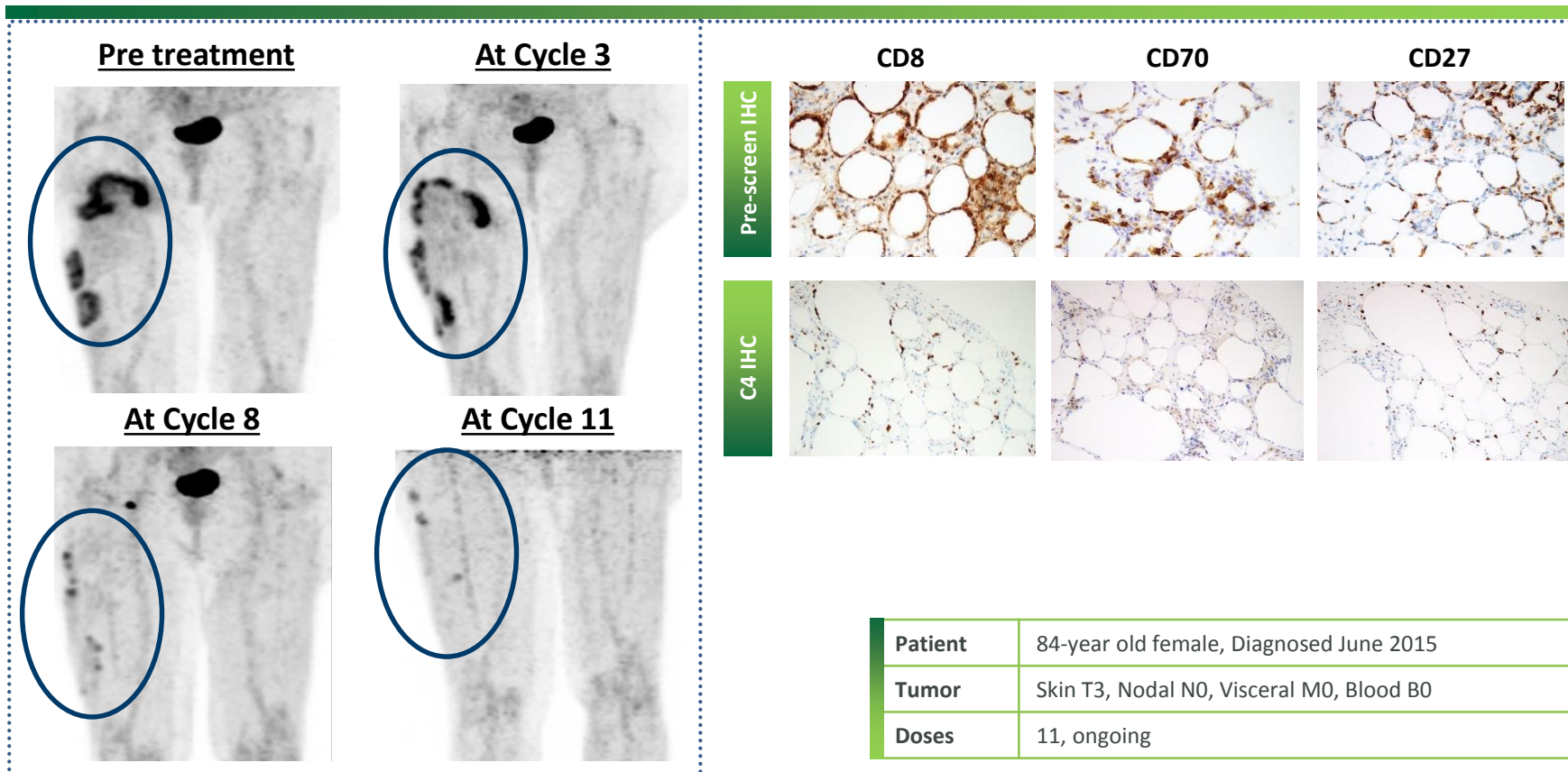


- 60% reduction of mSWAT score constitutes a partial response (PR)
- Decrease in surface area of cutaneous tumor lesions
- Lesions improve from plaques to patches
- Some lesions completely resolved

Patient	79 year old female with CTCL-MF, diagnosed 2007
Tumor	Skin T2, N0, M0, B0 (stage IB)
Doses	16, 1 mg/kg q3w

ARGX-110: Partial Response Confirmed by PET/CT

Patient example 3: CTCL (panniculitis like TCL type)



- Partial Response after 5 doses (dose 1 mg/kg)
- Further improvement through cycle 8 to 11 cycles (dose increased to 5 mg/kg)
- The patient is now on a maintenance dose of 5 mg/kg q6wk

ARGX-110 Shows Activity Across CTCL Types & Disease Stages

Interim Phase 1b data in CTCL

June 1, 2017

Indication ⁽¹⁾	Stage	C1	C2	C3	C4	C5	C6	C7	C8	C9	C10	C11	C12	C13	C14	C15	C16	C17	C18	C19	C20	Best Response ⁽²⁾
CTCL with circ clone	Not known																					Stable Disease
CTCL TFH like	Not known																					Stable Disease
CTCL-SS	Not known																					Progressive disease
CTCL TFH like	T2, N0, M0, B0																					Progressive Disease
CTCL panniculitis type*	T3, N0, M0, B0																					Partial Response
CTCL-MF/SS (+PTCL-NOS ⁽³⁾)	T4, N3, M0, B0																					Progressive disease
CTCL-MF	T4, N0, M0, B0																					Stable Disease
CTCL-MF	T4, Nx, M0, B0																					Stable Disease
CTCL-MF	T2, N0, M0, B0																					Partial Response
CTCL-MF	T4, Nx, M0, B0																					Progressive Disease
CTCL-SS	T4, N3, M0, B1																					Progressive Disease
CTCL-SS	T4, Nx, M0, B2																					Partial Response
CTCL-SS	T2, Nx, M0, B2																					Progressive Disease
CTCL-MF	T3, N0, M0, B1																					Stable Disease
CTCL-MF*	T1, Nx, M0, B0																					Stable Disease
CTCL-MF	T3, Nx, M0, B0																					Stable Disease

- Encouraging signs of clinical activity
- Heavily pre-treated patients on study dosed up to 16 cycles
- Itching often disappears after first cycle(s)

argenx data (uncleaned)

* Still on study

(1) CTCL = cutaneous T-cell lymphoma; MF = mycosis fungoides; SS = Sézary syndrome.

(2) Based on modified Severity Weighted Assessment Tool (mSWAT) scoring, a common method of scoring skin lesions in CTCL; assess number and severity of lesions as and total body surface area affected. Stable disease = mSWAT score does not increase by >25%; partial response = at least 50% reduction in mSWAT score; complete response = 100% reduction in mSWAT score.

(3) NOS: not other specified. PTCL-NOS is the most common TCL subtype.

Number of cycles on study, one cycle = 3 weeks, 17 cycles = ~1 year

1 mg/kg 5 mg/kg



What is Acute Myeloid Leukemia?

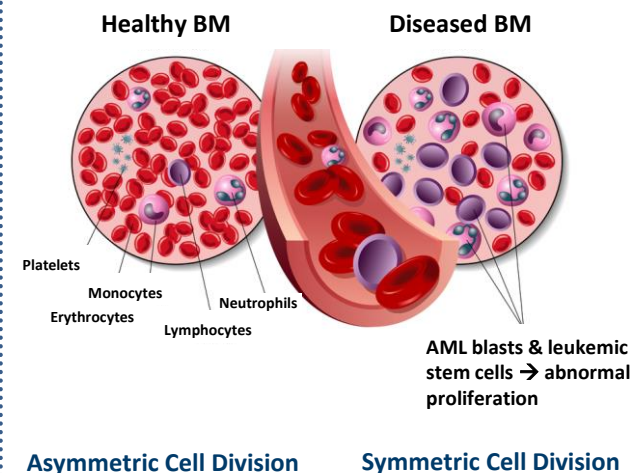
- Rare hematologic cancer characterized by **excessive proliferation of myeloid stem cells** and their **failure to properly differentiate into mature white blood cells**
- Symptoms include: **Fatigue**, shortness of breath, **easy bruising**, **bleeding**, **progresses rapidly**
- AML progresses rapidly and is fatal if left untreated
- ~22,000⁽¹⁾ p/a new cases the U.S. — **2nd most common leukemia subtype** in adults
- Generally a **disease of the elderly** — 60% of diagnosed patients are older than 60

Limited current treatment options

- **Older patients** are typically unfit for chemotherapy — **receive palliative treatment with hypomethylating agents**
 - Median survival of 7 – 10 months
 - ~6%⁽²⁾ five year survival rate for patients over 65
- Current first-line treatments for younger patients (<45yr) typically involve aggressive chemotherapy to induce remission followed by stem cell transplant (7 + 3 regimen / transplant)
 - 5-year survival is ~57%
- **Significant need for safer and more effective treatment options**



Effects of AML on Bone Marrow

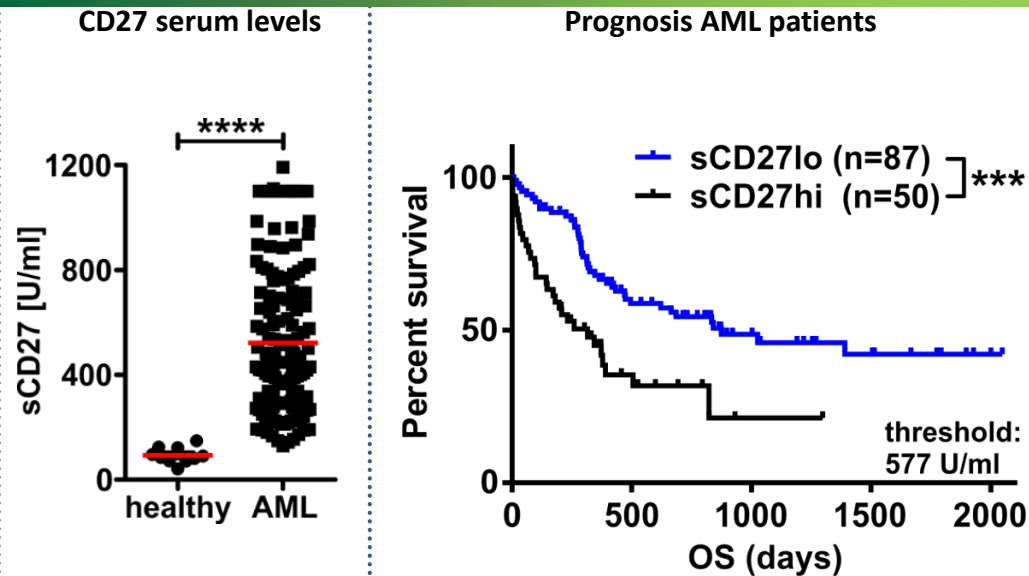


(1) American Cancer Society: <http://www.cancer.org/cancer/leukemia-acutemyeloidaml/detailedguide/leukemia-acute-myeloid-myelogenous-key-statistics> .
(2) National Cancer Institute: Howlader et al. (eds). SEER Cancer Statistics Review, 1975-2013, National Cancer Institute. Bethesda, MD, http://seer.cancer.gov/csr/1975_2013/, based on November 2015 SEER data submission, posted to the SEER web site, April 2016. Table 13.16

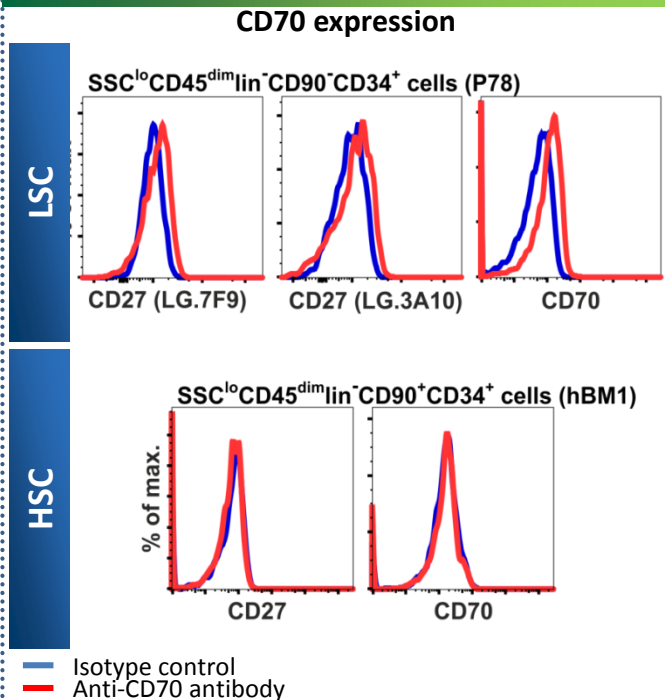
CD70 Unifying Rationale Across Risk & Age Classes in AML

Potential to selectively target leukemic stem cells in AML patients

Elevated sCD27 serum levels correlate with poor prognosis



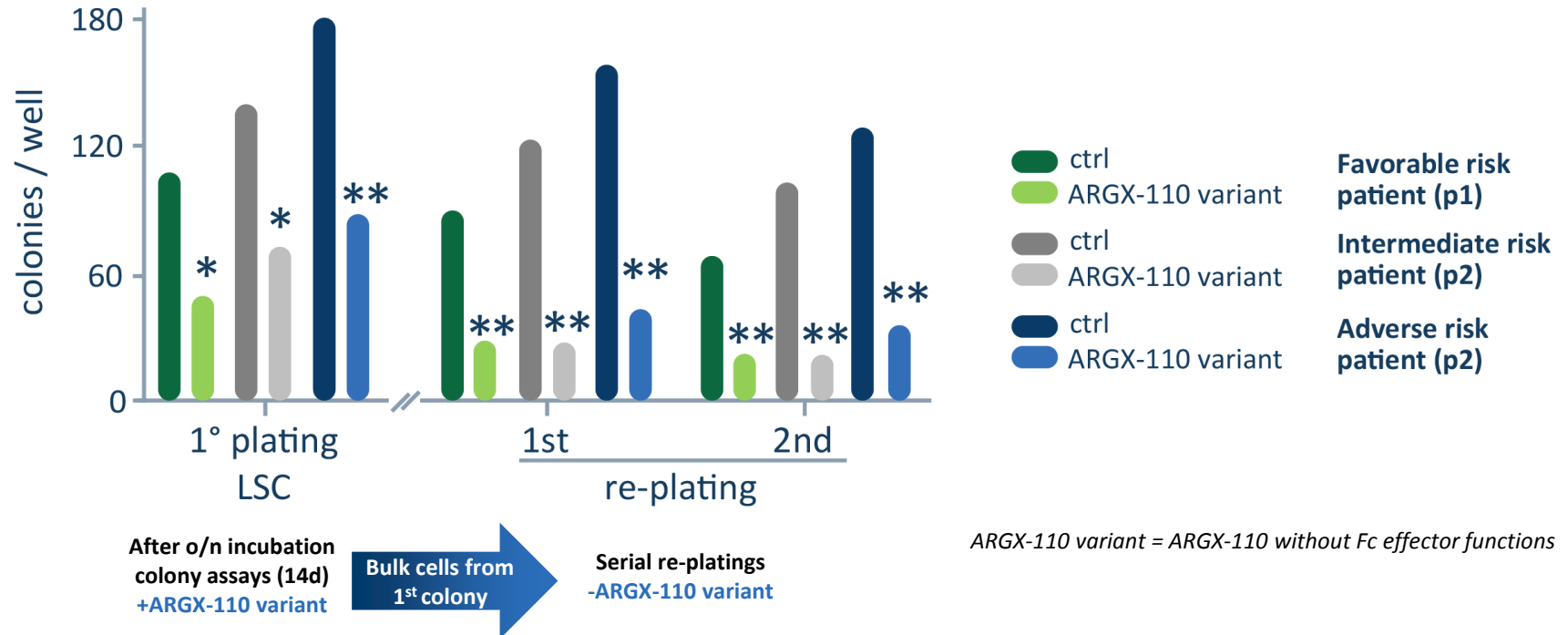
CD70 is selectively overexpressed on LSCs



- Elevated sCD27 serum levels in all newly diagnosed AML patients, **regardless risk or age categories**
- sCD27 levels are an independent **negative prognostic marker** in all newly diagnosed AML patients
- **CD70 expressed on ~100% of AML blasts**, majority of malignant cells are CD70/CD27 double-positive
- **CD70/CD27 selectively overexpressed on Leukemic Stem Cells (LSCs)**, not on Hematopoietic Stem Cells (HSC)

ARGX-110: Inhibits LSC Proliferation In Lasting Fashion

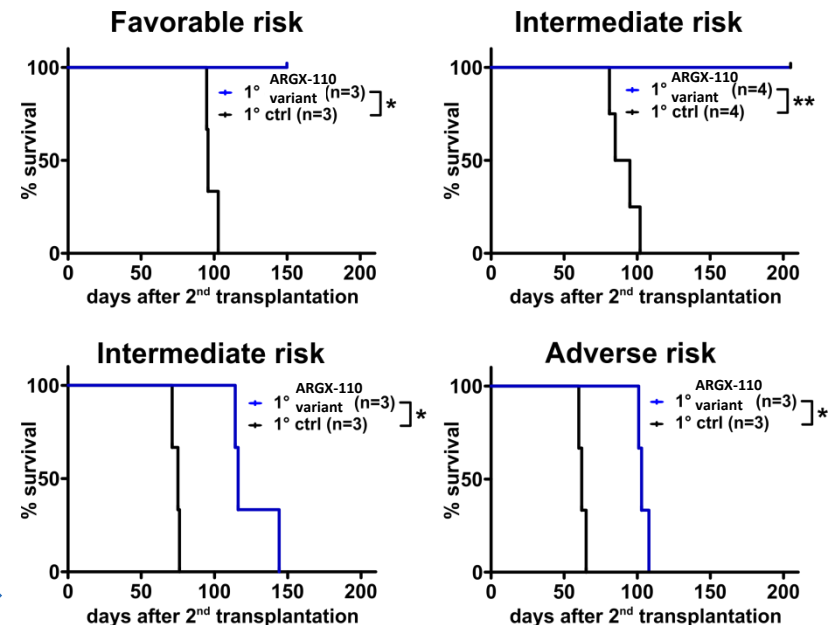
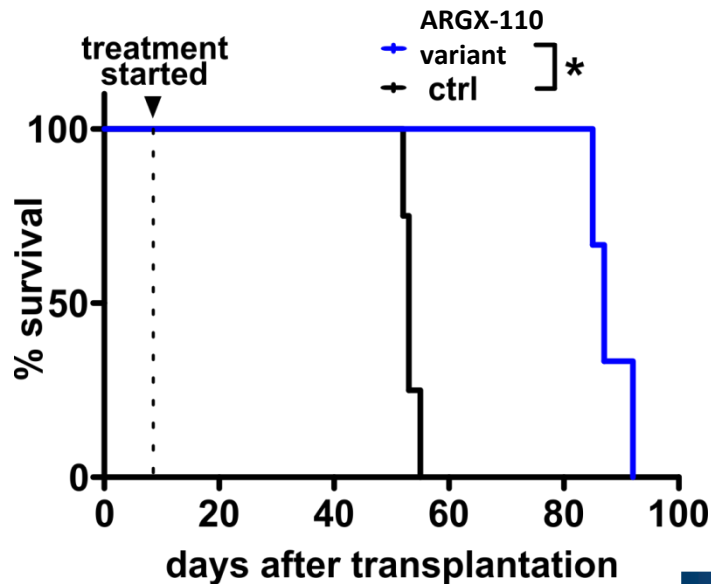
Long-term effects *ex vivo*



- Reduces LSC colony formation across patient risk categories (favorable/intermediate/adverse risk)
- Reduces LSC numbers as determined in serial re-plating experiments
- Blocking CD70 results in: (1) lasting down regulation of stem cell genes (2) **increasing myeloid differentiation**

ARGX-110: Curative Potential Of Monotherapy In Mouse Model

Shown to reduce LSCs, increasing survival in AML model

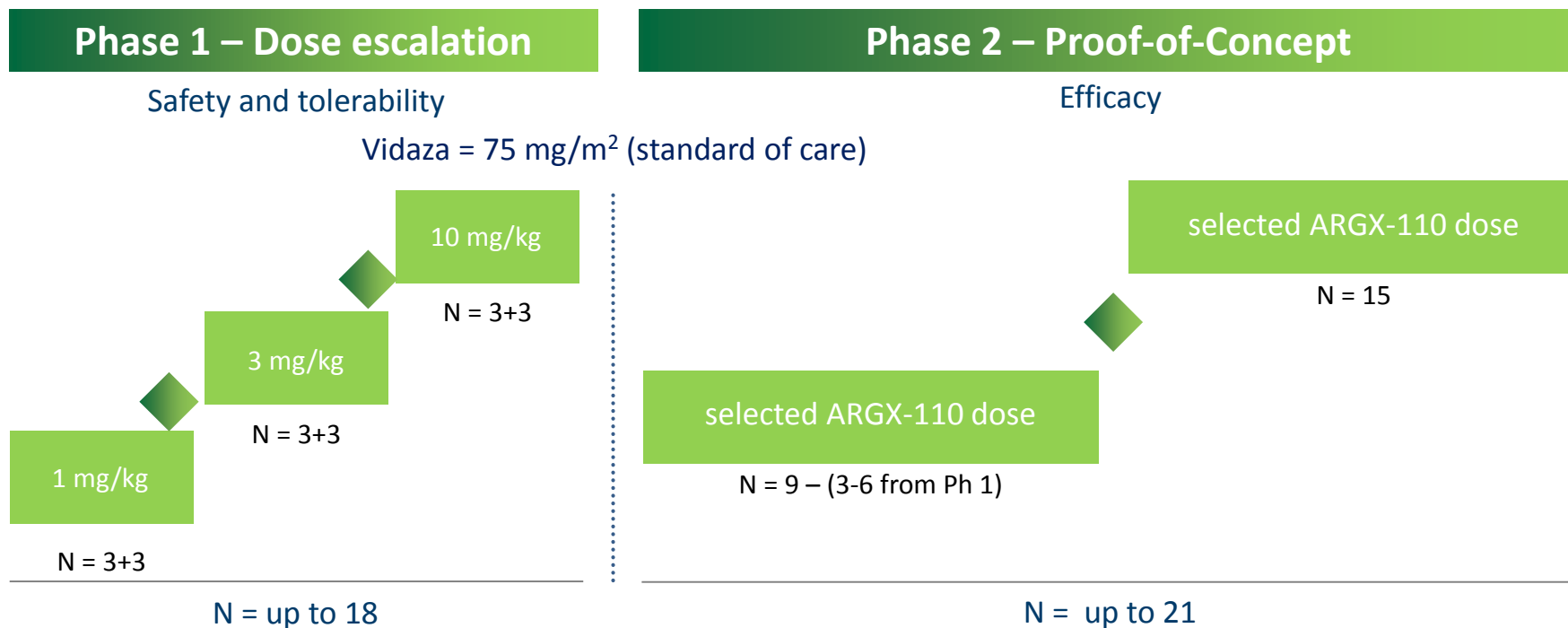


Initial in vivo treatment
+ARGX-110 variant

Grafting Whole Bone Marrow cells from treated
into new mice (14d after start of treatment)

Transplant, evaluation
-ARGX-110 variant

- Increased survival after secondary transplantation of AML BM cells from primary recipients transiently treated with ARGX-110 variant
- Increased survival observed for AML blasts taken from all 3 AML risk categories (fav/int/adv)



- Hypomethylation agents such as Azacitidine increase CD70 expression¹
- Population: untreated AML & high risk of myelodysplastic syndrome (MDS)*, eligible for AZA
- Design: open-label, non-controlled, non-randomized



*Some Myelodysplastic Syndrome (MDS) patients are at high risk of developing AML; MDS affects bone marrow cells, reducing their ability to produce red & white blood cells
(1) Zhou et al. 2011, Lupus.

Business development & financials



Strategic Antibody Collaboration Details





- **GARP** is a protein specifically present on the surface of activated regulatory T-cells (Tregs)
- **AbbVie** has option to
 - obtain exclusive, worldwide license to develop and commercialize ARGX-115
 - fund further GARP-related research by argenx beyond ARGX-115
- **argenx** conducts and funds all R&D through completion of IND-enabling studies
- **argenx** retains rights to combine ARGX-115 with its pipeline programs

Financial Highlights

- **\$40mm upfront payment**
- **Eligible for two near-term \$10mm preclinical milestones;** first of which already **achieved**
- **\$625mm in potential** development, regulatory and commercial milestones
- **Tiered royalties** on sales at percentages ranging from **mid-single digits to low teens**
- **Co-promotional** rights for ARGX-115-based products in the **European Economic Area and Switzerland**



Additional Strategic Collaborations

Partner	Asset	Key commentary
	ARGX-109 (Gerilimzumab)	<ul style="list-style-type: none"> Focused on developing an anti-IL-6 antibody for Rheumatoid Arthritis Bird Rock responsible for all costs incurred in R&D and commercialization
	ARGX-112	<ul style="list-style-type: none"> Focused on inflammation-based dermatological indications LEO Pharma fund >50% of all development costs up to CTA approval and all development post-approval of first Phase 1 trial in Europe argenx is eligible for ~€100mm in aggregate milestone payments + tiered royalties
	ARGX-116	<ul style="list-style-type: none"> Focused on developing an anti-ApoC3 antibody for dyslipidemia Jointly responsible for conducting dyslipidemia research — Staten responsible for additional clinical development argenx eligible for royalties in the low twenties
Broteio Pharma	Undisclosed	<ul style="list-style-type: none"> Focused on developing a differentiated antibody against a novel complement target Potential to act synergistically with ARGX-113 Jointly responsible for development expenses until preclinical POC — argenx granted exclusive option to license program after achieving preclinical POC
	Discovery Programs	<ul style="list-style-type: none"> Focused on novel rare disease targets Provides Shire access to SIMPLE Antibody platform + Fc engineering technologies argenx has received \$12mm in aggregate upfront and milestone payments and R&D fees over the course of the collaboration Shire purchased €12mm of argenx ordinary shares through participation in July 2014 IPO

Financial Profile and Key Investor Composition

Shareholder base > 50% US investors

Historical Financial Performance

(In thousands of euros)	1Q ended March 31	
	2016	2017
Total Operating Income	2,835	7,211
R&D Expense	(4,408)	(12,196)
G&A Expense	(1,401)	(3,411)
Total comprehensive loss	(2,970)	(8,410)
Cash, Cash Equivalents & Current Financial Assets	53,847	84,977

Additional Key Statistics

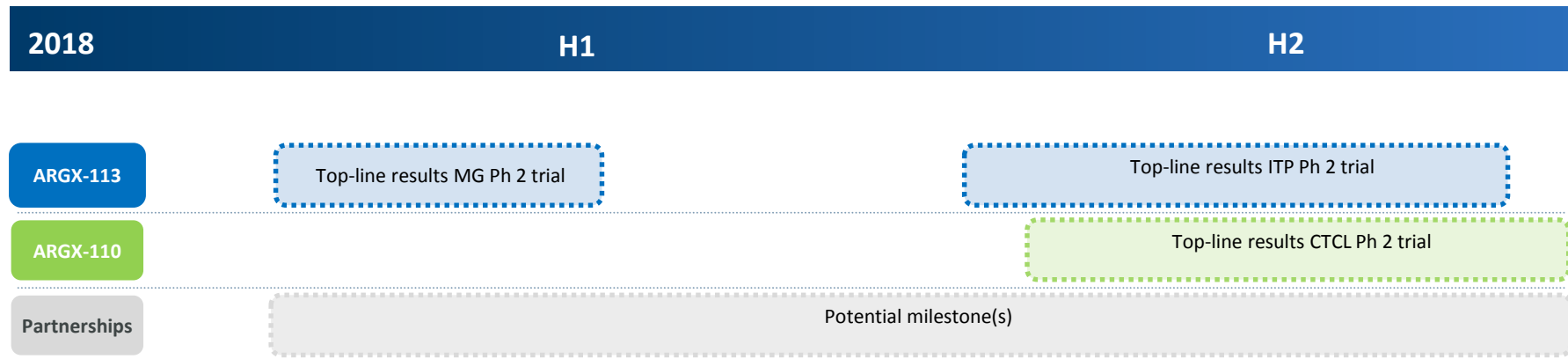
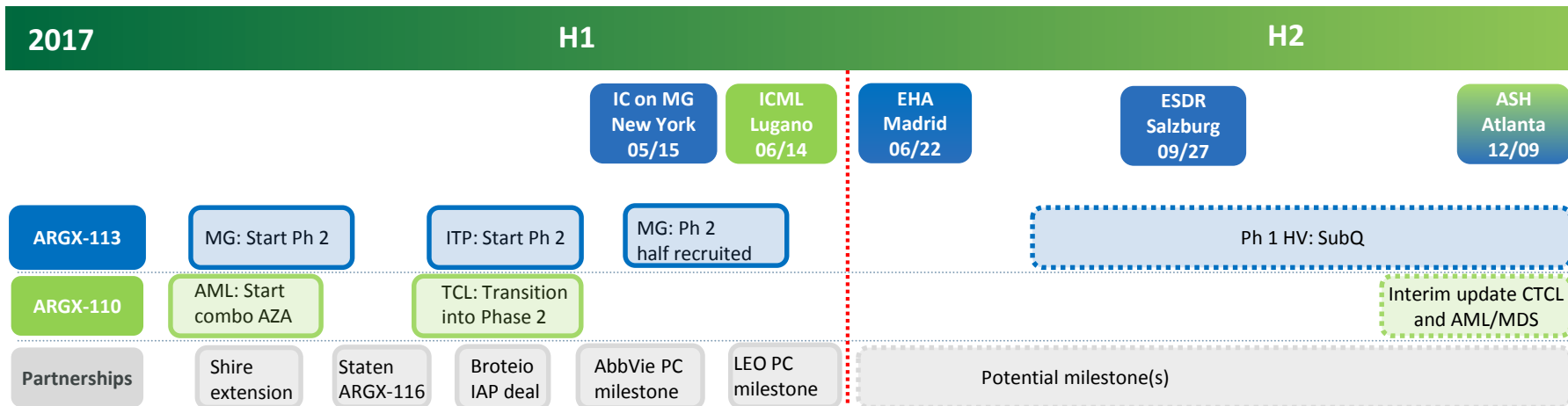
- **Capital raised since inception⁽¹⁾: €236mm (ex. grants)**
 - 2016: raised €46mm across two PIPEs
 - 2017: raised \$115mm (€102mm) in NASDAQ IPO
- **Non-dilutive funding since inception⁽¹⁾: €73mm**
 - 2016: \$40mm AbbVie upfront payment
 - 2017: \$10mm preclinical milestone
- **69 employees⁽¹⁾ — 54 R&D, 15 G&A**

Blue-Chip Investor Base > 50% US investors



(1) As of June, 2017

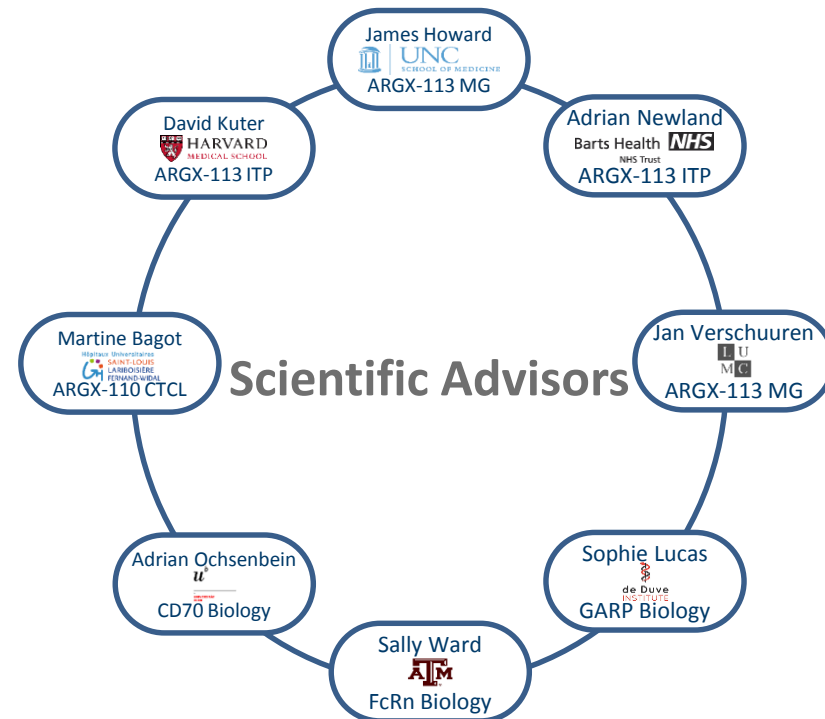
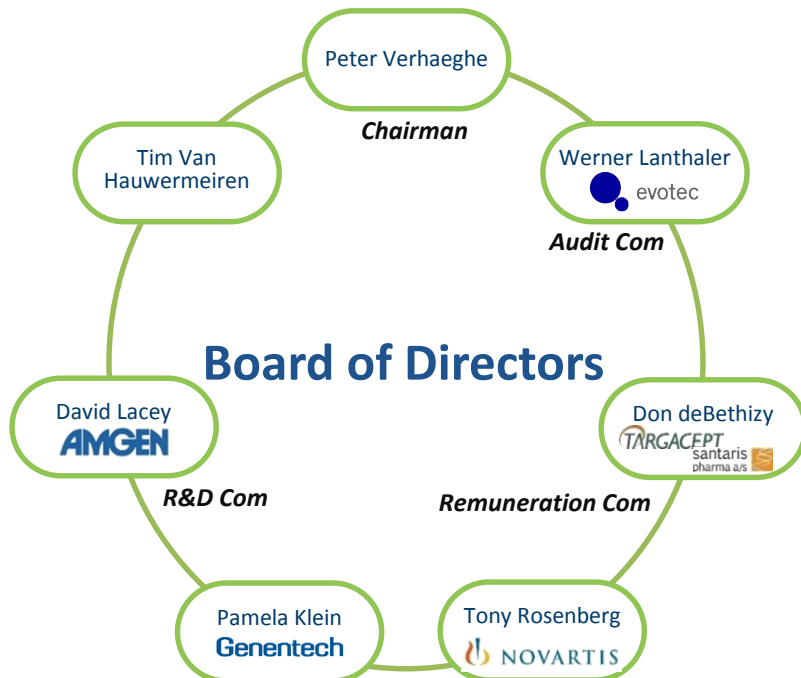
Key Upcoming Milestones & Communications



Appendix

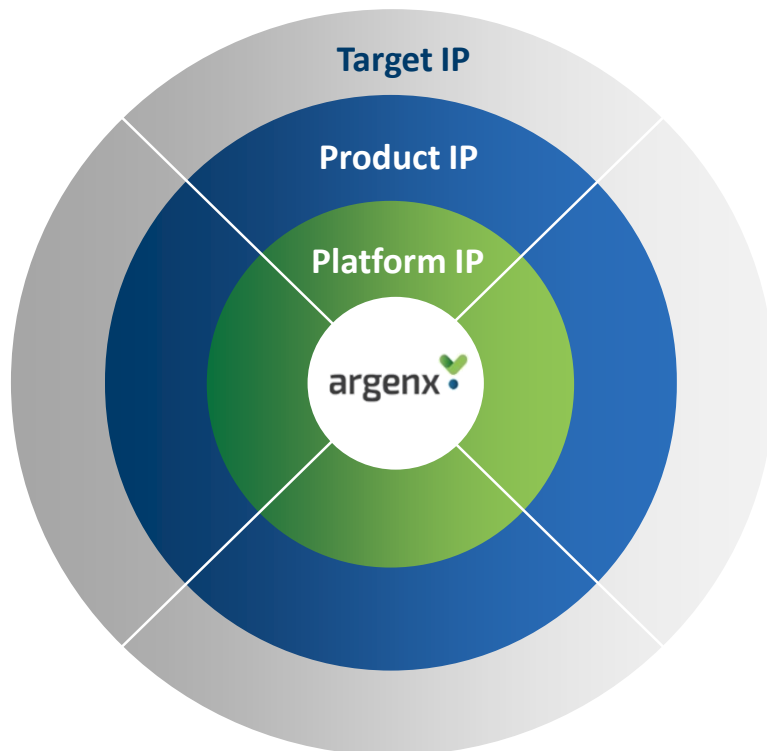


Management



Robust IP Portfolio

Multi-layered protection for key programs



Key Highlights of IP Portfolio

Targets

- Antibody epitope, functionality claims
- Issued and pending patents in US/J/EU/...
- Expiring in 2032-2037 timeframe excl. PTE

Product Candidates

- Composition of matter & method of treatment claims
- ~70 Issued and pending patents in the US/J/EU/...
- Expiring in 2032-2037 timeframe excl. PTE

Suite of Technologies

- *SIMPLE Antibody™ Platform*
 - Composition of matter and process & tools claims
 - ~50 Issued and pending patents in the US/J/EU/...
- *Fc Engineering Technologies*
 - Exclusively licensed patents for ABDEG™ and NHance® with 2027-2034 expirations in the US

MG Market Segmentation Hypothesis

Potential beach heads and expansion opportunities

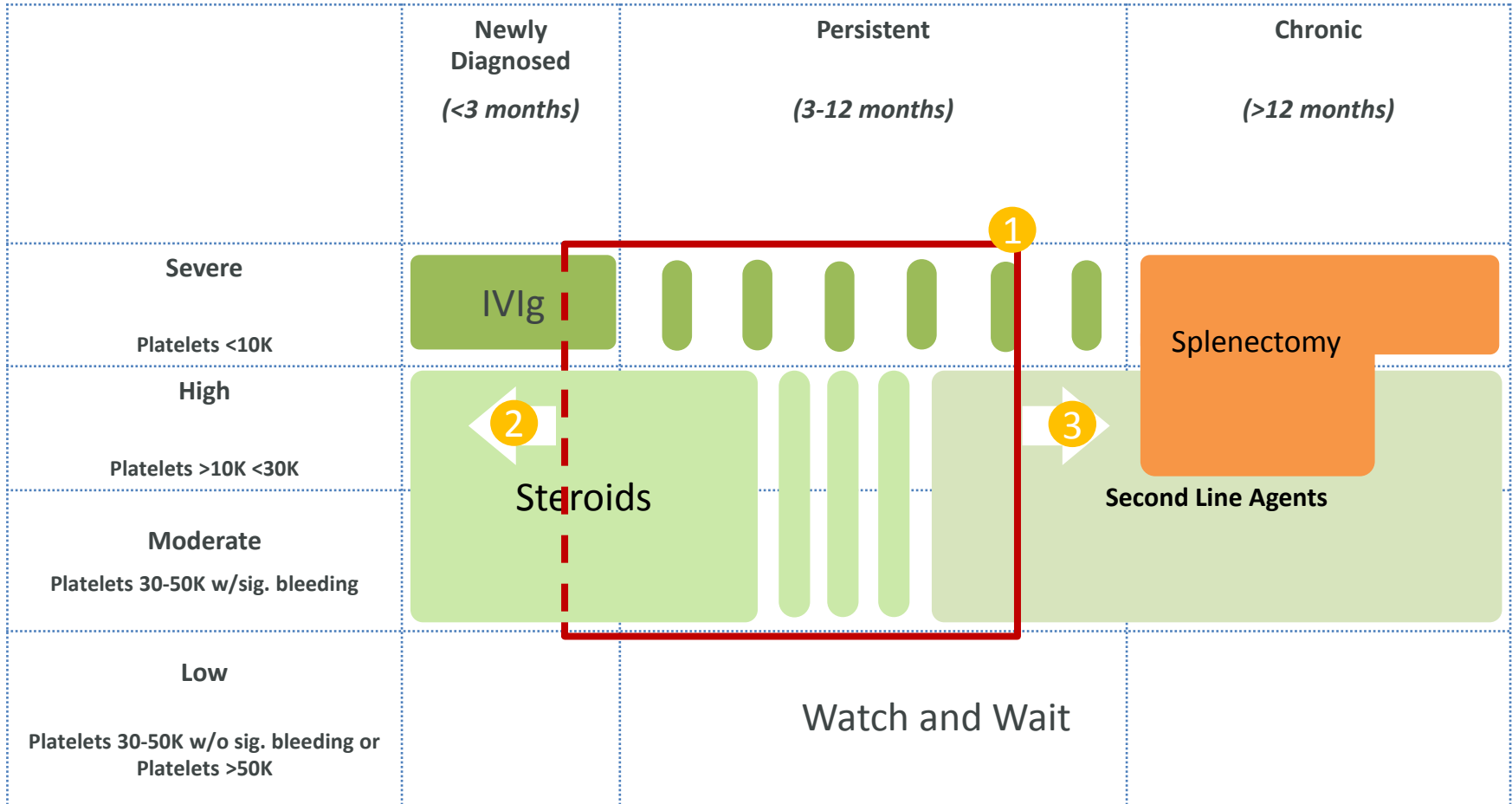
Patients	Newly Diagnosed		Chronic		Refractory		
	Mild/Mod. Patients / Symptomatic Treatment	Severe, MuSK-Ab+, or Pyridostigmine Refractory	Chronic Immunosuppressant Therapy or Prednisone Intolerant	Thymo-ma	No Thymo-ma	Bridge / Remission Therapy	Experimental
Academic Tertiary Care	Pyridostigmine	Prednisone	Azathioprine Mycophenolate Mofetil Methotrexate Tacrolimus/ Cyclosporine	Preop IVIg Thymectomy		PLEX or IVIg	Experimental agent Rituximab Other Immunosuppressants
	<div> <div>←</div> <div>② Myasthenic Crisis PLEX / IVIg</div> <div>→</div> </div>						
Community						IVIg	

- Use ARGX-113 in patients with inadequate response to previous therapy and **before second line choices**
- Expand to **initial and ongoing therapy** to replace IVIg for MG crises
- Expand into management of **chronic/refractory** MG with a sub-cutaneous dosage form

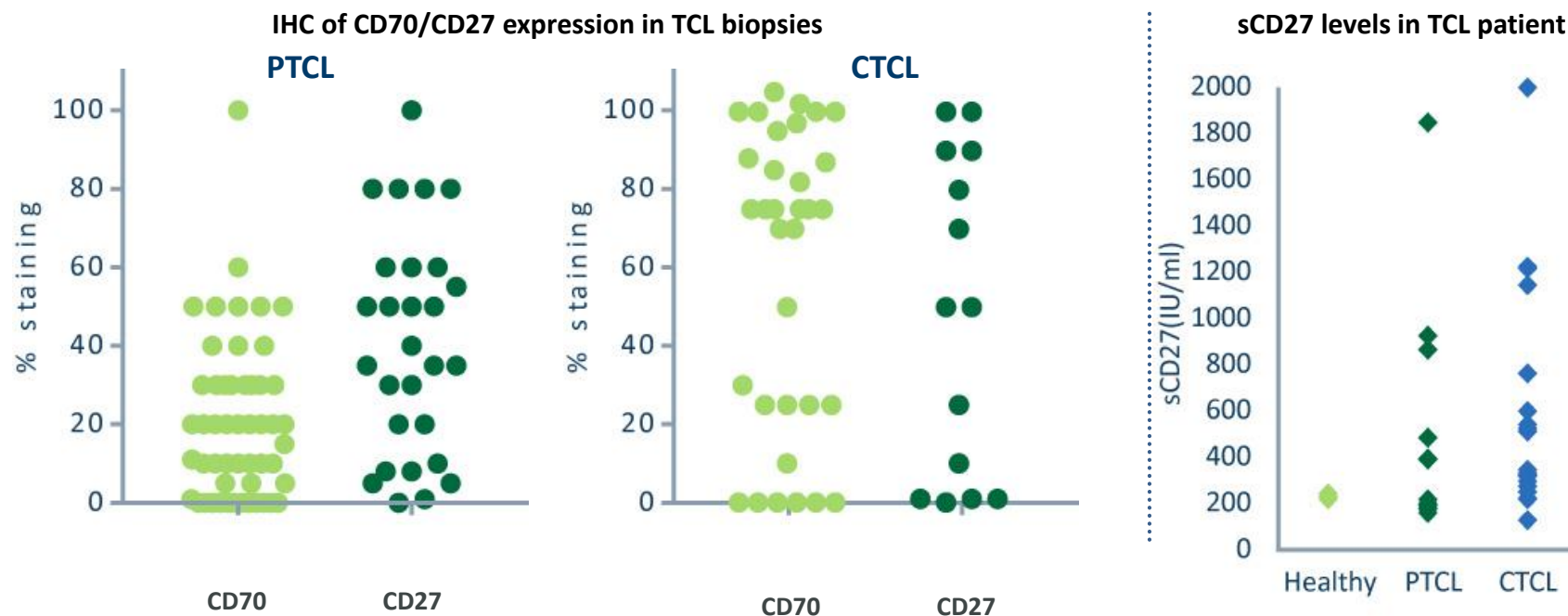


ITP market segmentation hypothesis

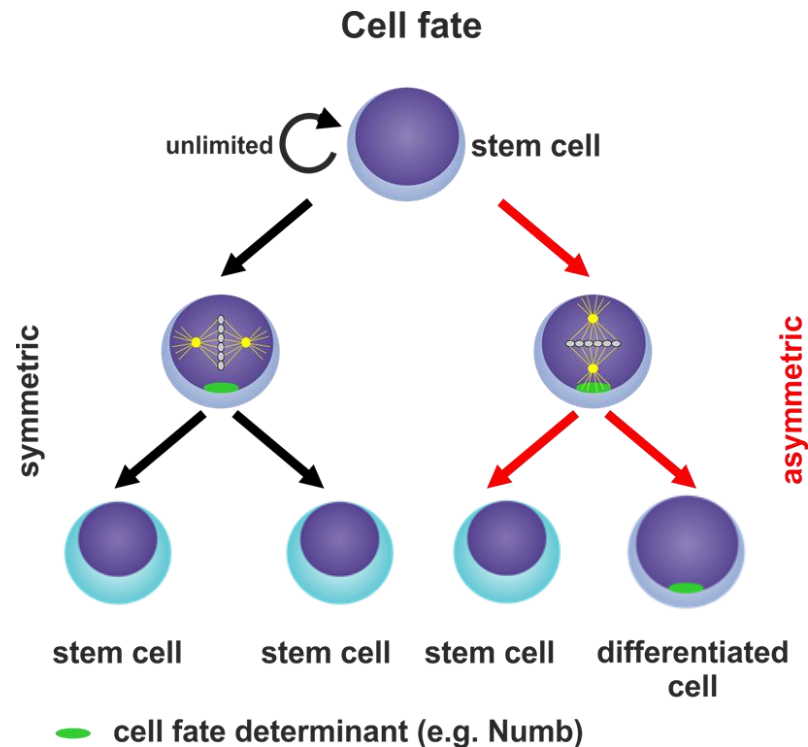
Potential beach heads and expansion opportunities



- Use ARGX-113 in patients with inadequate response to previous therapy and **before second line choices**
- Expand to **initial therapy** to replace IVIg and combine with steroids
- Expand into management of **persistent/chronic** ITP with a subcutaneous dosage form



- CD70/CD27 strongly overexpressed across different TCL types
- Elevated sCD27 levels suggest strong pathway activity in TCL

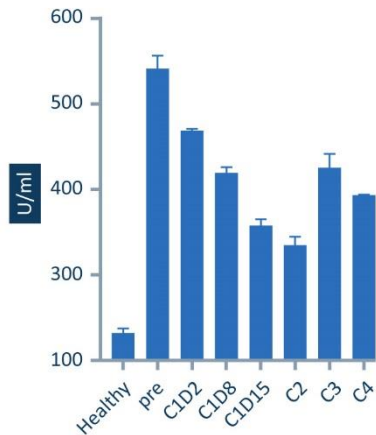


- Accumulation of blasts in bone marrow and blood → drop in red blood cells, platelets, and normal white blood cells
- Leukemic stem cells (LSCs = AML stem/progenitor cells) are responsible for disease relapse

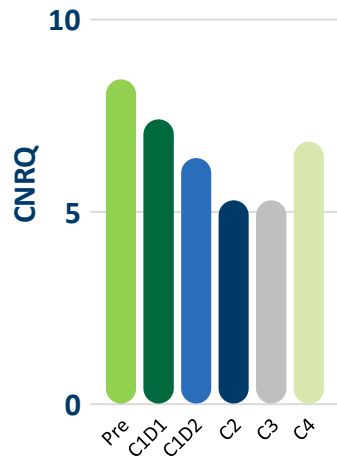
Decreased sCD27 and Sézary Clone In Blood

Patient example 3: Cutaneous TCL patient – Sézary-syndrome (SS)

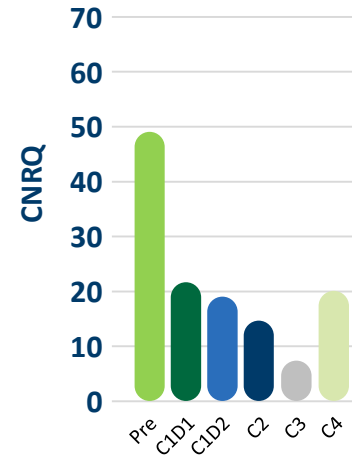
sCD27 levels in serum



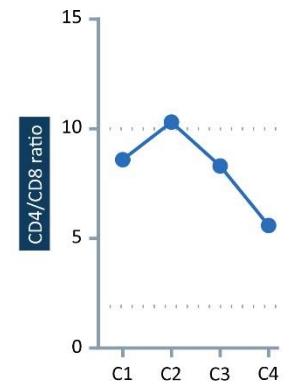
RNA CCR4



RNA TWIST1



CD4/CD8 cells in blood



- Decrease in sCD27 first 2 cycles; then stabilization
- Sézary clone levels decrease as shown by CD4/CD8 ratio and CCR4 & TWIST1 levels (RNA)

ARGX-111 Overview

- Potential best-in-class anti-MET antibody targeting MET driven metastasis
- Enables novel routes of intervention in MET-driven cancers via multiple modes of action
- **Differentiated design** — complete blockade of MET signaling and enhanced tumor cell killing
- **Demonstrated biological activity MET amplified tumors in Phase 1b trial** — we plan to partner due to indication scope

SIMPLE Antibody

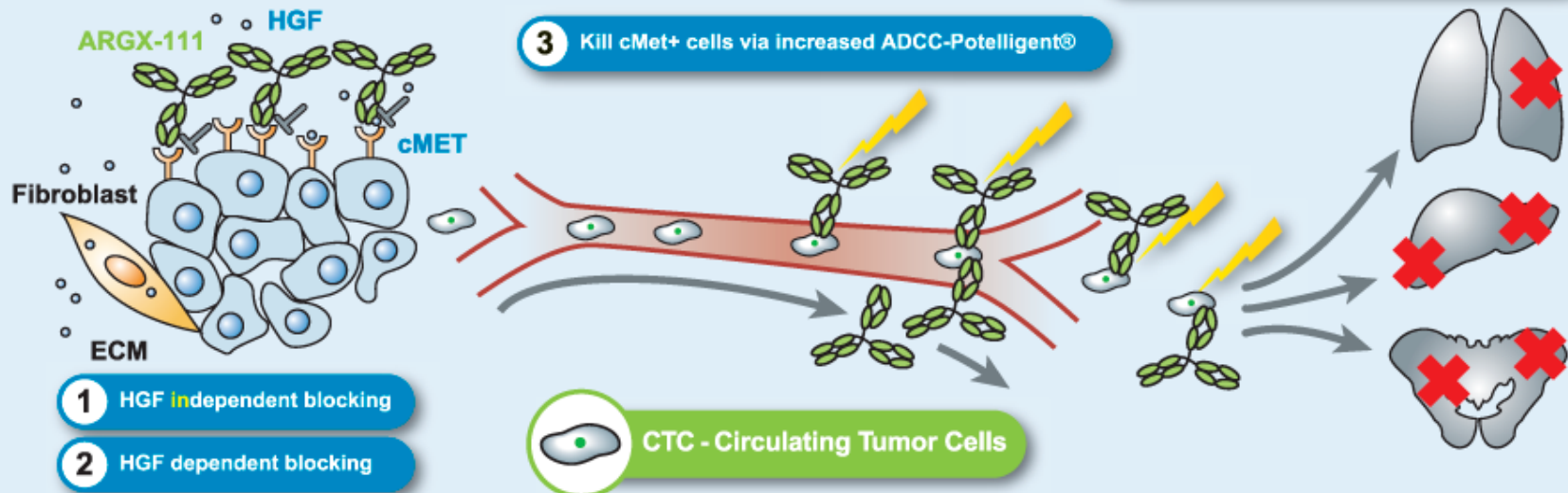


NHance

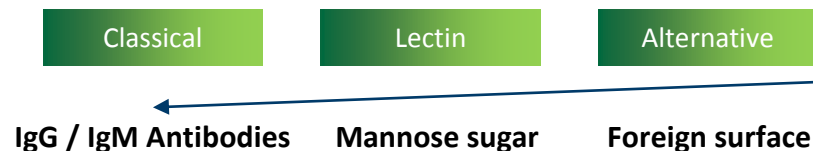
POTELLIGENT

4 modes of action to attack MET+ tumor cells

4 Increased tissue penetration-NHance®

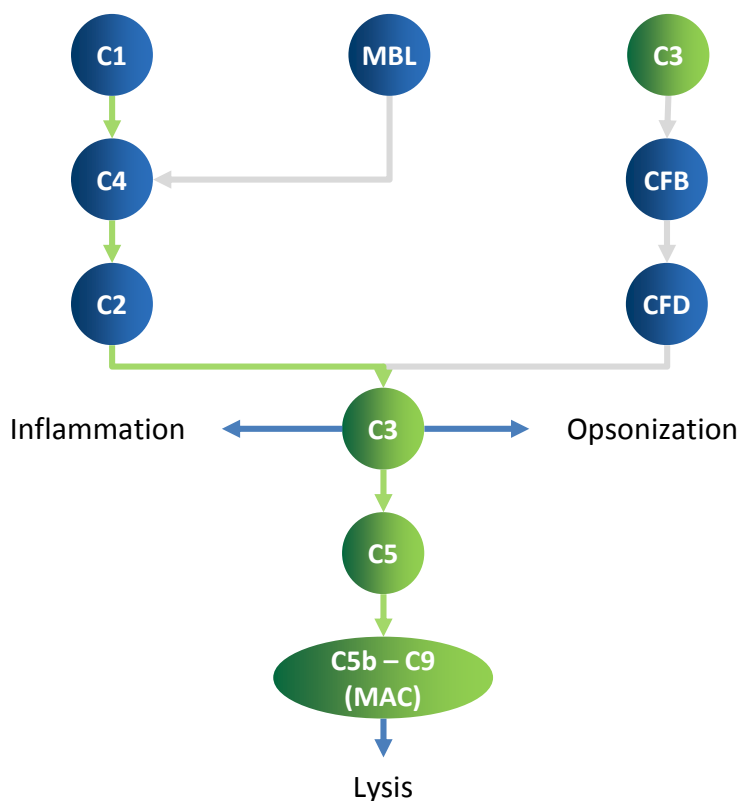


Innovative Access Program: Securing Novel Complement Target



Potential focus of IAP program

Focus of ARGX-113



Innovative Access Program collaboration with Broteio Pharma:

- Antibody against a **novel target** in the classical complement cascade
- Therapeutic potential in complement driven disease triggered by auto-IgM antibodies: auto-immune haemolytic anemia, antibody-mediated rejection, ...
- We have an exclusive option to license the rights to further develop and commercialize the program
- Strategic fit with ARGX-113: potential to create a franchise in severe auto-immune diseases driven by pathogenic antibodies



Thank you!
