Developing Highly Differentiated Antibody Therapeutics

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28-29 November 2017
Forward Looking Statements

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Validating selective partnerships

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

- **ARGX-113**: 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, immune thrombocytopenia and pemphigus vulgaris

Well financed to execute plan

- **Strong cash position**: €162mm Sept 30, 2017
- Blue chip investor base: more than 60% U.S. Shareholders
- 26.9 mio shares outstanding

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

Deep pipeline with multiple shots on goal

- **ARGX-110**: first-in-class CD70 antagonist in Phase 1/2 in CTCL and AML

Company Highlights

Differentiated therapeutic antibodies pioneering in severe autoimmune diseases & cancer
Recent Progress

**Pipeline**

- ARGX-113 Phase 2 study in MG patients: 100% recruited (Oct ‘17)
- ARGX-113 for MG: Orphan drug designation by FDA (Sept ‘17)
- ARGX-113 Phase 2 study in ITP patients: 50% recruited (Sept ‘17)
- ARGX-113 Phase 2 study initiation in PV patients (Sept ‘17)
- ARGX-113 Phase 1 study initiation in healthy volunteers with **subcutaneous formulation** (Oct ‘17)

**Partnerships**

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

**Financing**

- Upsized $115mm IPO on Nasdaq (ticker: ARGX)(May ‘17)
- U.S. shareholding expanded above 60%
- Expanded U.S. analyst coverage
- 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
Disciplined Business Model
Maximizes value of our suite of technologies and capabilities

Generating differentiated antibody candidates...

Novel Targets + argenx Technology Suite = Differentiated mAbs

...capturing value at optimal stages

<table>
<thead>
<tr>
<th>Discovery</th>
<th>Preclinical development</th>
<th>Early &amp; late clinical development</th>
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<tbody>
<tr>
<td>Platform deals</td>
<td>Product deals outside strategic focus</td>
<td>Product deals large indications</td>
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<td>Shire</td>
<td>Bird Rock Bio</td>
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<td>ARGX-109</td>
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<td>Value inflection point</td>
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ARGX-109
ARGX-112
ARGX-116
ARGX-115
ARGX-111
ARGX-113
ARGX-110
Augmenting Intrinsic Therapeutic Properties Of Antibodies

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

Klarenbeek et al. 2015, mAbs
Basilico et al. 2014, J Clin Inv.
We obtained the exclusive license option from **Broteio Pharma** for an antibody against a novel complement target.

We have an antibody discovery alliance with **Shire** focused on multiple rare disease targets.

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### Deep Pipeline In Severe Autoimmune Diseases and Cancer

<table>
<thead>
<tr>
<th>Product Candidate</th>
<th>Target</th>
<th>Indication</th>
<th>Preclinical</th>
<th>Phase 1</th>
<th>Phase 2</th>
<th>Phase 3</th>
<th>Next Milestone / Commentary</th>
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<td><strong>Wholly-Owned Product Candidates</strong></td>
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<td>ARGX-113 (efgartigimod)</td>
<td>FcRn</td>
<td>Myasthenia Gravis</td>
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<td>Immune Thrombocytopenia</td>
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<td>Pemphigus Vulgaris</td>
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<td>2H18: Phase 2 interim data</td>
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<td>Chronic Autoimmune Diseases</td>
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<td>2H18: Phase 1 interim data</td>
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<td>T-Cell Lymphoma</td>
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<td>Phase 1/2</td>
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<td>Acute Myeloid Leukemia</td>
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<td>Phase 1/2</td>
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<td>YE17: Interim update Phase 2 CTCL and Phase 1 dose-escalation in AML/MDS</td>
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<td>ARGX-111</td>
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<td>Solid Tumors / Blood Cancer</td>
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<td>Intend to partner</td>
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| **Partnered Product Candidates** | | | | | | | |
| ARGX-109 (gerilimzumab) | IL-6 | Rheumatoid Arthritis | | | | | Eligible for up to €32.5mm in milestones, royalties & additional shares of Bird Rock stock |
| ARGX-112 | IL-22R | Skin Inflammation | | | | | Eligible for up to ~€100mm in milestones and tiered royalties |
| ARGX-115 | GARP | Cancer Immunotherapy | | | | | Received $50mm so far; eligible for up to $625mm milestones & tiered royalties |
| ARGX-116 | ApoC3 | Dyslipidemia | | | | | Eligible for double-digit royalties and exclusive option to license the program |
ARGX-113: A Pipeline-in-a-Product Opportunity
ARGX-113: Lead Program Based On Novel Target FcRn

An innovative approach to eliminate IgG autoantibodies

- 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

(3) argenx data
ARGX-113: Pipeline-In-Product Opportunity

Prioritizing IgG autoantibody mediated diseases

Landscape of IgG severe autoimmune diseases (selection)

Solid Biological Rationale
Disease proven to be predominantly mediated by pathogenic IgGs

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

Proof of Concept Value
Spill-over effect into adjacent indications

Myasthenia Gravis
Beachhead neuromuscular diseases

Immune Thrombocytopenia
Beachhead heme disorders

Pemphigus vulgaris
Beachhead blistering diseases
**Myasthenia Gravis (MG) Overview**

**What is Myasthenia Gravis?**

- Rare autoimmune disorder; 64,000\(^{(1)}\) patients in U.S., 55,000\(^{(2)}\) with generalized MG, affecting all ages and both genders.
- MG associated with muscle weakness; can be life threatening if respiratory muscles affected.
- Symptoms include: Life-threatening choking; muscle dislocation; eyelid fatigue; pain; problems with vision, speech, mobility, fatigue.

**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, hematologic suppression, headache, disfigurement, infection, thrombosis.
- IVIg and Plasmapheresis place a heavy cost burden on healthcare systems in the acute setting (~$79,000\(^{(3)}\) and ~$101,000\(^{(3)}\) respectively).

**Myasthenia Gravis Cause**

Autoantibodies (IgG type) destroy neuromuscular junctions:
- Blocking of Acetylcholine Receptors (AChRs)
- Cross-linking + internalization of AChRs
- Complement recruitment

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\(^{(1)}\) Philips et al. 2003, Ann N Y Acad Sci.
\(^{(3)}\) Heatwole et al. 2011, J Clin Neuromuscul Dis.
Autoantibody Levels (IgGs) Correlate With MG Disease Score

>30% autoantibody reduction clinically meaningful

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

* Comparison between 3 cycles of Plasmapheresis/Immunoadsorption 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 autoantibody reduction correlates with clinical improvement and reduced hospital stay

Liu et al. 2010, Ther Apher Dial.
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-related reactions, leukoencephalopathy, nausea, osteoporosis, psychosis, sweating, neutropenia, thrombosis, vomiting, weakness
- Romiplostim and Eltrombopag, last-line therapies for ITP and have generated global revenues of $584 million\(^{(2)}\) and $635 million\(^{(3)}\) in 2016

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(2) Amgen Inc. 2016, Form 10-K.
(3) Novartis Annual Report 2016
Autoantibody Levels (IgGs) Correlate With ITP Disease Score

Autoantibodies inhibit platelet production and accelerate platelet destruction

Platelet levels during IVIg treatment in Adult Immune Thrombocytopenia

Low bleeding risk

High bleeding risk

\(\downarrow\) = IVIg treatment  \(\bullet\) = Autoantibody level  \(\square\) = Platelet counts

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

**Pemphigus Vulgaris: Overview**

**What is Pemphigus Vulgaris?**
- Chronic, severe – potentially life-threatening – auto-immune disease
- ~ 17,000 people treated (US)\(^{(1)}\)
- Mucosal and skin blisters leading to pain, difficult swallowing, skin infection
- Disease severity directly correlates to pathogenic IgG levels against desmoglein-1 (skin involvement) and desmoglein-3 (mucosal involvement)\(^{(2)}\)
- Patients cycle through periods of remission and relapse for extended periods

**Limited current treatment options**
- Current disease management comes with significant side effects and impacts QoL
  - High dose of corticosteroids and chronic immunosuppression (AZA, MFM)
  - Rituximab, IVIg, immunoadsorption and plasma exchange used for severe or refractory patients (10%), but not perceived as curative
- Treating physicians require new effective therapies with rapid onset of action that are safe
- Rituximab therapy shows slow onset of action, risk of developing serious adverse events and significant relapse rate \(^{(2)}\) \(^{(3)}\) \(^{(4)}\)

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\(\text{Pemphigus Vulgaris Cause}\)

Diagnosis based on presence of pathogenic autoantibodies targeting desmoglein-1 and -3 in the skin

Auto-antibodies (predominantly IgG4 type) sterically hinder desmosomal adhesion and assembly – no complement involvement or immune effector activation\(^{(2)}\)

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\(\text{15}\)

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\(1\) Collective Acumen study\(\)\(^{(1)}\)\(\)
\(2\) Kasperkiewicz et al. 2017, Nature Reviews\(\)
\(3\) Colliou et al. 2013, Autoimmunity\(\)
\(4\) Joly et al. 2017, Lancet
ARGX-113: Favorable Safety & Tolerability Profile

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

**Single ascending dose (SAD)**

- 0.2 → active:placebo
- 2 → active:placebo
- 10 → active:placebo
- 25 → active:placebo
- 50 → active:placebo

**Multiple ascending dose (MAD)**

- 10 → active:placebo
- 25 → active:placebo
- Up to 6 doses

**Data analysis**

- Complete safety profile
- Complete PK/PD profile
- Phase 2 dose selected

- 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 seen in Phase 1

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 seen in Phase 1

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

- Potent IgG reduction across isotypes (AChR autoantibodies are IgG1/3; MuSK autoantibodies are IgG4)
- 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

- Population: MG patients with generalized muscle weakness with total MG-ADL score ≥ 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

argenx data, * 50% of this score attributed to non-ocular items.
ARGX-113 in ITP: Phase 2 Trial Design

**Screening/Randomization**

- Screening ≤2 weeks
- Randomization

**Treatment Phase**

- SoC + ARGX-113 (10mg/kg) N=12
- SoC + ARGX-113 (5mg/kg) N=12
- SoC + Placebo N=12

**Follow-up Phase**

- 4 doses; N= up to 36

**≤2 weeks**

- Population: ITP patients with platelet levels < 30 X 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

[Study 50% recruited Sept 2017]
ARGX-113 Phase 2 study: IDMC-driven adaptive design

- **Treatment Phase**
  - **Induction**: 3 weeks
    - COHORT 1: N= 4
      - ARGX-113 (10mg/kg)
      - 4 infusions (weekly)
    - COHORT 2 & 3: N= 4 + 4
      - ARGX-113 (adjusted dose)
      - 4 infusions (weekly)
  - **Maintenance**: 6 weeks
    - ARGX-113 (10mg/kg)
    - 2 infusions (w2, w6)

- **Follow-up Phase**: 8 weeks

- **Patients enrollment divided in 3 sequential cohorts**
- **IDMC recommendations for cohorts 2 & 3:**
  - Change of dose (max dose of 25mg/kg)
  - Frequency of administration at maintenance (max 2 extra doses after each cohort)
  - Expansion of maintenance duration
ARGX-113: Feasibility of SubQ Dosing
Exploring SubQ formulations for larger patient populations (chronic, ex-hospital)

- 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%

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

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

1. Block tumor growth signal
2. Restore immune surveillance
3. Kill tumor

- ARGX-110 is a SIMPLE Antibody™, equipped with 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

Silence et al. 2014, mAbs.
What is Cutaneous T-Cell Lymphoma?

- Rare and incurable sub-type of T-cell lymphoma
- Prevalence (US & Canada): ~ 30,000 & Incidence (US): ~ 3,000
- Patients typically diagnosed in their 60s
- Mycosis fungoides (50%), Sézary syndrome most common types
- Symptoms include: severe rash, itching, tumor, skin infections
- Skin infection often cause of death

Limited current treatment options

- Initial treatment includes topical dermatology agents (corticosteroids, PUVA, e-beam therapy)
- Advanced stage patients treated with systemic oncology 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) 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

(2) Lymphoma Research Foundation: http://www.lymphoma.org/site/pp.asp?c=bkLTKaOQLmK8E&b=6300151
(3) http://www.istodax.com/hcp/ctl/study-design/efficacy
ARGX-110 In Cutaneous TCL

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

Example Sézary Syndrome (SS) patient

Treatment & best response

Example Mycosis fungoides (MF) 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

<table>
<thead>
<tr>
<th>Patient</th>
<th>67 year old male CTCL-MF, diagnosed 2001</th>
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</thead>
<tbody>
<tr>
<td>Tumor</td>
<td>Skin T4, Nx, M0, B0 (Stage IIIA)</td>
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<tr>
<td>Doses</td>
<td>6</td>
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</table>

### Decrease of CD4+ malignant T-cells

- Pre-treatment
- Post (Cycle 2)

### Depletion of CD70+ malignant T-cells

- Pre-treatment
- Post (Cycle 2)

### 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

*argenx data – patient anecdotes – uncleaned. Pictures kindly provided by investigator.*
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)
- Patient now on maintenance dose of 5 mg/kg q6wk

Patient 84-year old female, Diagnosed June 2015
Tumor Skin T3, Nodal N0, Visceral M0, Blood B0
Doses 11, ongoing

Patient anecdotes – uncleaned. Pictures kindly provided by investigator.
**ARGX-110 Shows Activity Across CTCL Types & Disease Stages**

Interim Phase 1b data in CTCL

**Indication**

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

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

**Raw Text**

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

**Notes**

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

---

*Still on study as of June 1, 2017*
Acute Myeloid Leukemia (AML) Overview

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: weight loss, fatigue, fever, night sweats, loss of appetite, shortness of breath, easy bruising, infections, bleeding
- Disease progresses very rapidly and is fatal if left untreated
- ~22,000\(^{(1)}\) new cases per year in the U.S. — 2\(^{nd}\) most common leukemia subtype in adults
- Generally a disease of the elderly — 60% of diagnosed patients are older than 60

Limited current treatment options

- Elderly, frail patients are typically unfit for high dose chemotherapy — receive palliative treatment with hypomethylating agents
  - Median survival of 7 – 10 months
  - 5 year survival rate of ~6\(^{(2)}\)% for patients over 65
- Younger patients (<45yr) typically get aggressive chemotherapy (“7+3” regimen) to induce remission followed by stem cell transplant
  - 5 year survival rate of ~57\(^{(2)}\)% for patients under 45
- Significant need for safer and more effective treatment options

Effects of AML on Bone Marrow

- AML blasts & leukemic stem cells ➔ abnormal proliferation

References:

**CD70 Unifying Rationale Across Risk & Age Classes in AML**

Potential to selectively target leukemic stem cells in AML patients

- Elevated sCD27 serum levels in all newly diagnosed AML patients, **regardless of 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 (HSCs)

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 variant = ARGX-110 without Fc effector functions

ARGX-110: Curative Potential Of Monotherapy In Mouse Model

Shown to reduce LSCs, increasing survival in AML model

- 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)

ARGX-110 & Azacitidine For AML/MDS: Phase 1 / 2 Combo

**Phase 1 – Dose escalation**
- Safety and tolerability
- Vidaza = 75 mg/m² (standard of care)
- We are here

- 10 mg/kg
  - N = 3+3

- 3 mg/kg
  - N = 3+3

- 1 mg/kg
  - N = 3+3

N = up to 18

**Phase 2 – Proof-of-Concept**
- Efficacy
- selected ARGX-110 dose
  - N = 15

- selected ARGX-110 dose
  - N = 9 – (3-6 from Ph 1)

N = up to 21

- 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
AbbVie Partnership for Novel Target GARP

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**
- **Received** first of two **$10mm** preclinical milestones
- **$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

Cuende et al., 2015, Science Trans Med.
### Additional Strategic Collaborations

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<th>Partner</th>
<th>Asset</th>
<th>Key commentary</th>
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| Broteio Pharma           | ARGX-109 (Gerilimzumab) | • Focused on developing an anti-IL-6 antibody for **Rheumatoid Arthritis**  
                             • Bird Rock responsible for all costs incurred in R&D and commercialization           |
| LEO                      | ARGX-112 | • Focused on **inflammation-based dermatological indications**  
                             • LEO Pharma funds >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 |
| Staten Biotechnology     | ARGX-116 | • 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 | • 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 |
| Shire                    | Discovery Programs | • 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 |
### Key Upcoming Milestones & Communications

#### 2017

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Appendix
Please Join argenx for a Lunch and Discussion during the ASH Annual Meeting

Advancing ARGX-110 to clinical proof-of-concept in acute myeloid leukemia (AML) & cutaneous t-cell lymphoma (CTCL)

**Monday, December 11, 2017**
12:00 PM - 1:30 PM

**Omni Atlanta Hotel**
South Tower Atrium Terrace
Birch Room
100 CNN Center NW
Atlanta, GA 30303
*Next to Convention Center*

**Please RSVP by Monday, December 4**
Rachel Frank
rachelf@sternir.com
212.362.1200

**Agenda**
- Overview of AML
  - Gail Roboz, MD
  - Weil Cornell Medicine, New York
- **CD70: Novel AML Target**
  - Hans de Haard, PhD, argenx
- Phase 1/2 AML Trial:
  - Proof-of-Biology Data
  - Nicolas Leupin, MD, argenx
- Phase 1/2 CTCL Trial:
  - Data Update
  - Nicolas Leupin, MD, argenx
- Q&A

**Guest Speaker**

Gail Roboz, MD
*Professor of Medicine and Director of Clinical and Translational Leukemia Program*  
Weill Medical College of Cornell University and New York Presbyterian Hospital

Following the presentation, please join us and our colleagues from the University of Bern for an informal mix and mingle.