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CAUTION: We have not received approval from the FDA, or any other regulatory entity, to market our therapeutic candidates in the United States or in any other jurisdictions. Our therapeutic candidates, including Pulmazole, PUR1800, and PUR0200 are classified by the FDA as investigational drugs and are limited by Federal (or United States) law to investigational use only that require additional studies to make definitive conclusions and claims about such candidates’ safety or efficacy.
Pulmatrix: Building Value by Meeting Important Unmet Medical Needs in ABPA and AECOPD

**LEAD PROGRAM:** iSPERSE™ Pulmazole Anti-Fungal
- Pulmazole is inhaled Itraconazole for allergic bronchopulmonary aspergillosis (ABPA) – seeking to be the first to market inhaled anti-fungal in US and EU
- Reduced development risk as oral itraconazole (Sporanox) has been on the market for over 25 years
- Itraconazole use in ABPA, though effective, has limited utility given poor bioavailability and toxicity concerns
- Phase 1/1b study results demonstrated Pulmazole safety/tolerability and ability to achieve significantly higher lung exposure and lower plasma exposure than oral Sporanox, supporting the belief that Pulmazole could significantly improve upon known efficacy and safety profile of Sporanox
- Qualified Infectious Disease Product (QIDP) designation obtained in 2017 for ABPA and CF

**Second PROGRAM:** iSPERSE PUR1800 Kinase Inhibitor
- PUR1800 is a novel narrow spectrum kinase inhibitor (NSKI) for COPD first in class (US/EU) inhaled non-steroidal anti-inflammatory to treat acute exacerbations in COPD (AECOPD)
- NSKI in PUR1800 demonstrated anti-inflammatory activity, safety and tolerability in stable COPD patients

**Underlying iSPERSE Platform Technology Supports Future Value Growth**
- iSPERSE dry powder technology significant patient experience and product opportunities
- iSPERSE intellectual property carries well into 2030’s on platform and specific products

ABPA = Allergic Bronchopulmonary Aspergillosis; CF = Cystic Fibrosis; AECOPD = Acute Exacerbations in Chronic Obstructive Pulmonary Disease
### iSPERSE™ Differentiation

- **Small, dense, dispersible**, respirable particles
- **Highly efficient** inhaled delivery
- **Flow rate independent** performance (consistent delivery of high drug loads)
- **Scalable** platform
- **Broad IP** portfolio into 2030s

### Potential iSPERSE™ Advantages

- **Can be used with a broad range of drugs**, small molecule to biologic
- **Can be used with any device** (i.e., metered-dose, reservoir, or blister-based inhalers)
- **Requires low inspiratory flow** for penetration deep into lung based on high dispersibility
- **Can deliver large doses into lungs** (tens of milligrams) with high lung delivery efficiency
- **Avoids first-pass effect and systemic side-effects** with improved PK profile compared to oral delivery

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**iSPERSE™ Enables Sick Patients to Get More Effective Doses**
# Robust Pipeline with Projected Significant Value Catalysts*

<table>
<thead>
<tr>
<th>Product Pipeline</th>
<th>Indication</th>
<th>2H 2018</th>
<th>2019</th>
<th>1H 2020</th>
<th>Milestone</th>
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<tbody>
<tr>
<td><strong>Pulmazole</strong></td>
<td>Anti-fungal</td>
<td></td>
<td></td>
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<tr>
<td></td>
<td>Allergic Bronchopulmonary Aspergillosis (ABPA) in Patients with Asthma</td>
<td></td>
<td><strong>Phase 2</strong></td>
<td></td>
<td>Phase 2 4Q 2019</td>
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<tr>
<td><strong>PUR1800</strong></td>
<td>NSKI</td>
<td></td>
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<tr>
<td></td>
<td>Acute Exacerbations of COPD (AECOPD)</td>
<td></td>
<td><strong>Phase 2a</strong></td>
<td></td>
<td>Phase 2a 2Q 2020</td>
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<tr>
<td><strong>PUR5700</strong></td>
<td>NSKI</td>
<td></td>
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<tr>
<td></td>
<td>Idiopathic Pulmonary Fibrosis (IPF)</td>
<td></td>
<td><strong>Pre-Clinical</strong></td>
<td></td>
<td>Pre-clinical</td>
</tr>
</tbody>
</table>

## Potential Future Revenue Opportunities

<table>
<thead>
<tr>
<th>Product Pipeline</th>
<th>Indication</th>
<th>2H 2018</th>
<th>2019</th>
<th>1H 2020</th>
<th>Milestone</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>PUR0200-US</strong></td>
<td>LAMA</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td></td>
<td>Chronic Obstructive Pulmonary Disease (COPD)</td>
<td></td>
<td><strong>Phase 1</strong></td>
<td></td>
<td>Out-Licensed to Vectura for U.S. 09/2017</td>
</tr>
</tbody>
</table>

LAMA = Long-Acting Muscarinic Antagonist; * Estimated Milestones
Pulmazole – Inhaled Antifungal

Inhaled Itraconazole to Treat Allergic Bronchopulmonary Aspergillosis (ABPA) in Asthmatics

“I am very encouraged by the phase 1 results demonstrating that it is both feasible to administer itraconazole by inhalation and further that high levels of the drug may be achieved within the airways. I believe that Pulmazole has the potential to significantly improve upon both the efficacy and safety profile of oral Sporanox, and I look forward to working with the other members of the advisory board to assist Pulmatrix in advancing the development of Pulmazole to treat patients with fungal asthma, focusing initially on ABPA.”

– David Denning, MD
Professor of Infectious Diseases in Global Health and Director of the National Aspergillosis Centre, Manchester, UK
Clinical Stage Antifungal

- Pulmazole is an inhaled reformulation of itraconazole in Ph2 for airway fungal infections
- Ph1/1b study demonstrated safety/tolerability in healthy normal volunteers dosed up to 14 days and asthmatics following single dose
- Ph1/1b results support belief that Pulmazole could significantly improve upon known efficacy and safety profile of Sporanox

Attractive Market Opportunity

- Up to $1.5B U.S. in Asthma-ABPA and in other Aspergillus pulmonary disease (ex. Cystic Fibrosis – ABPA)*
- Following 505(b)(2) pathway for registration as Sporanox has been on market for 25+ years
- Additional 5 years exclusivity with QIDP status

Clinical Data & Upcoming Milestones

- Phase 2 28-day proof-of-mechanism in ABPA patients scheduled to begin 4Q 2019 (FPI 4Q 2018)
- Phase 2 proof-of-concept efficacy/safety trial in ABPA patients scheduled to begin 2Q 2020

Superior Technology and Team

- iSPERSE best-in-class particle engineering technology for lung delivery
- Clinical stage COPD assets (PUR0200 and PUR1800) with substantial follow on development opportunities
- Experienced management team and board with success in commercializing respiratory products

ABPA = Allergic Bronchopulmonary Aspergillosis; COPD = Chronic Obstructive Pulmonary Disease; FPI = First Patient In

* Peak revenues expected at loss of market exclusivity, ~11 years post launch
### Pulmazole Has Potential to Transform the Standard of Care for ABPA

**Significant Product Opportunity**
- Estimated **300K US** and **5M WW** ABPA patients
- No approved antifungal therapy available
- **Limited/no novel competition** anticipated for ABPA market

**Improving Upon Standard of Care**
- Sporanox (oral itraconazole) use is limited due to poor bioavailability, safety/tolerability issues and drug-drug concerns associated with high oral plasma exposure

**Target Product Profile (TPP)**
- Pulmazole is anticipated to provide **higher lung exposure** and **lower plasma exposure** than Sporanox, potentially improving both **efficacy** and **safety/tolerability**

**Ph1/1b Clinical Study Results**
- Pulmazole was **safe** and **well tolerated** in SAD/MAD study in healthy normal volunteers and SD administration to asthmatics
- Pulmazole demonstrated **significantly higher lung exposure** than Sporanox in asthmatics, despite inhaling **1/10 of the dose** administered orally
- Pulmazole **plasma exposure** in asthmatics and healthy normal volunteers was **significantly lower** than that of Sporanox

**Pulmazole Clinical Value Drivers**
- Investment required to deliver **Phase 2 28-day POM data expected in 4Q 2019**
- Potential registration Phase 2b/3 POC trial start date 2Q 2020

Ref: Clearview Analysis.; IDSA = Infectious Diseases Society of America; SAD/MAD = Single Ascending Dose / Multiple Ascending Dose; POM = Proof of Mechanism; POC = Proof of Concept
**ABPA Represents a Large Addressable Antifungal Patient Population in Aspergillus Related Diseases**

**ABPA is a Debilitating Disease**

- ABPA in asthmatics is caused by hypersensitivity to *Aspergillus* species lung infection
- ~300K adult asthma ABPA patients in U.S. and ~5M worldwide
- Diagnostic criteria include:
  - High IgE (>1000 IU/mL)
  - Skin prick positivity to Aspergillus
  - Eosinophilia (>500 cells/µL)
  - Elevated IgE/IgG Antibodies to Aspergillus
  - Radiographic pulmonary opacities consistent with ABPA

**ABPA Pathophysiology**

*Healthy Lung* vs. *Diseased Lung*

Persistence of *A. fumigatus* in the airways leads to local inflammation, mucus production, reduction in lung function and worsening of asthma symptoms. Untreated ABPA may result in pulmonary fibrosis, respiratory failure and potentially death.

ABPA Progression and Treatment Options

ABPA is Treated by Allergists and Pulmonologists

**ABPA-Asthma Treatment Goals**

- Symptom control of asthma or cystic fibrosis
- Prevent or treat pulmonary exacerbations
- Reduce or remit pulmonary inflammation
- Mitigate progression to end-stage fibrotic or cavitary disease

**Treatment is Limited Primarily to Steroids and Antifungal Therapy**

- Oral steroid therapy is the standard of care for treating ABPA
  - In patients with insufficient treatment response, antifungal therapy is added to steroid therapy for ~4 months
- Antifungals are sometimes used 1st line in conjunction with steroids

**Limited Treatment Options Impede Ability to Achieve Treatment Goals**

- ~50% of ABPA patients have **inadequate response** to oral steroids alone
- ~20% of ABPA patients become **steroid dependent**
- Antifungal agents are believed to **reduce fungal burden** (antigen induces inflammatory response)
- Antifungal treatment **improves clinical outcomes** and can enable a reduction in steroid burden
- While the majority of antifungal use in ABPA is itraconazole, overall antifungal use is limited by safety/tolerability concerns

Literature Establishes Strong Clinical Precedent for Itraconazole Use in ABPA

**Literature Indicates Antifungals Improve ABPA Outcomes**

1991, Denning et al.
Itraconazole improved pulmonary function, decreased corticosteroid use and total IgE in 6 patients with ABPA (3 cystic fibrosis and 3 asthma)

2000, Stevens et al.
Itraconazole improved pulmonary function, and decreased steroid use and total IgE in a randomized double-blind trial with 55 patients

2018, Agarwal et al.
In acute stage treatment naive ABPA patients, monotherapy itraconazole is effective in considerable number with less side-effects compared to prednisolone

2016, Ram et al.
Nebulized amphotericin B reduced exacerbations compared to nebulized budesonide in a study in 21 patients

2003, Wark et al.
Itraconazole decreased total IgE and the exacerbation frequency in a randomized, double-blind trial with 29 patients

Pulmazole is Anticipated to Improve Upon Sporanox Outcomes (FEV₁, Exacerbation Rate, Asthma Control and Steroid Burden) Demonstrated in Clinical Literature

Randomized Placebo Controlled Studies Support Efficacy of Oral Sporanox in Asthma-ABPA

### 2000, Stevens et al.  
New England Journal of Medicine

**Design:**
- Part 1: 200 mg Sporanox BID vs PBO for 16 weeks (n=55)
- Part 2: open label, all patients received 200 mg Sporanox QD (n=50)

**Results:**
- 46% response rate in Sporanox group vs 19% response rate in placebo group (p=0.04)
- Greater improvements in lung function (FEV₁, FVC, PEF) noted in Sporanox treatment group versus placebo

### 2003, Wark et al.  
Journal of Allergy & Clinical Immunology

**Design:** 200 mg oral Sporanox bid vs PBO for 16 weeks (n=29)

**Results:**
- Sputum eosinophils (35% decrease/week first 4 weeks) and serum IgE (6% decrease/week) significantly reduced
- Clinically significant improvement in FEV₁ following 16 weeks of treatment (9.8% treatment difference between groups (p=0.5, likely due to small sample size)
- Fewer exacerbations requiring oral corticosteroids in those treated with Sporanox (p=0.03)

**Stevens & Wark Studies — Key Takeaways**
- Both clinical trials support inclusion of Sporanox into current ABPA treatment guidelines
- Key biomarkers of ABPA activity can be reduced as early as 4 weeks of treatment
- Both studies show improvements in FEV₁ at 16 weeks, a potentially approvable endpoint
- Both studies are the foundation for Pulmazole Phase 2 POM and Phase 2b/3 POC development plan

Pulmazole Potentially Addresses the Significant Limitations of Oral Antifungals in ABPA

**Antifungal Limitations**

- Visual Changes
- Cutaneous Reactions
- Hepatic Abnormality
- Gastro Intestinal Intolerance
- Variable and Poor PK
- Drug to Drug Interactions
- Fatigue

**Physician Feedback Supports Pulmazole as a Preferred Treatment**

- Antifungals reduce both exacerbations and chronic steroid use
- Sporanox has been on the market for 25+ years and is the preferred antifungal
- Oral antifungal dose dependent side-effects can be significant restricting physician use and variability in lung concentration limit the ability to maximize therapeutic effect
- Physicians desire additional antifungal treatment options
- Inhalation of itraconazole directly to the airways should allow for higher exposure where Aspergillus is present
- Pulmazole profile would lead to significant treatment adoption

Pulmazole Has Up to $1.5B Peak Revenue Potential in the U.S.

Payers and Physicians Interviewed See Logic in Pulmazole Value Proposition

- Payers suggested ~70% market access***
- Payers indicated ~$40K/year asthma biologic price with restrictions to specialty use and label
- Physicians indicated 67% addressable market penetration with Pulmazole**

1st Line Antifungal Usage May More than Double

% of Antifungal Use 1st Line with Steroids

<table>
<thead>
<tr>
<th>Current Use of Antifungal 1st Line</th>
<th>Future Use of Antifungal 1st Line</th>
</tr>
</thead>
<tbody>
<tr>
<td>30%</td>
<td>65%</td>
</tr>
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</table>

Pulmazole U.S. Peak Revenue Forecast*

Net Revenue

- ~$1.3B
- ~$200M

U.S. Other Indications

- U.S. ABPA
- Asthma

Source: Physician Interviews; Payer Interviews; ClearView Analysis. *Also includes discount for patient compliance, patient persistence, and gross-to-net adjustment; ** Estimate based on ClearView Analysis, which included qualitative physician surveys and interviews; *** Estimate based on ClearView Analysis, which took into account uninsured patients, patients who are unwilling to pay, and projected access restrictions placed by payers; * Peak revenues expected at loss of market exclusivity, ~11 years post launch
Phase 1/1b Data Highlights Potential of Pulmazole to Improve Upon Clinical Utility of Oral Sporanox

**Ph1/1b Study SUCCESSFULLY Met All Endpoints**

**Phase 1/1b : Safety, Tolerability & PK Study in Healthy Normal Volunteers and Asthmatics**

**Part 1: Single Ascending Dose**
Healthy Normal Volunteers (n=6/cohort): Optional 4th cohort of 35 mg

- Pulmazole (5 mg)
- Pulmazole (10 mg)
- Pulmazole (25 mg)
- Pulmazole (35 mg)

**Part 2: Multiple Ascending Dose**
Healthy Normal Volunteers (n=5-6/cohort)

- Pulmazole (10 mg)
- Pulmazole (20 mg)
- Pulmazole (35 mg)

**Part 3: Single Dose Crossover**
Asthmatics (n=16)

- Sporanox® (200 mg; oral)
- Pulmazole (20 mg)

**Parts 1 and 2 in HNV**
- Demonstrated safety and tolerability of Pulmazole administered up to 14 days
- Following inhalation of Pulmazole, total systemic exposure over 24 hours is ~100-400 fold lower than that expected with Sporanox

**Part 3 SD in Stable Asthmatics**
- Demonstrated safety and tolerability of Pulmazole administered as a single dose in asthmatics
- ~50 fold higher lung exposure following inhalation of Pulmazole compared to Sporanox despite inhaling only 1/10 the dose
- ~85 fold lower plasma exposure than oral Sporanox

SD = Single Dose;
Global Thought Leaders in ABPA and Asthma on the Pulmazole Clinical Advisory Board (CAB)

Ritesh Agarwal, MD, DM – Professor of Pulmonary Medicine, Postgraduate Institute of Medical Education and Research, Chandigarh, India

David Denning, FRCP, FRCPath, FIDSA, FMedSci – Professor of Infectious Diseases in Global Health; President, Global Action Fund for Fungal Infections; Director, National Aspergillosis Centre, Manchester, UK

Cendrine Godet, MD – Doctor of Infectious Diseases and Pulmonary Medicine at Poitiers University Hospital, France; Coordinator of the French National Board for Chronic Fungal Infection and Secretary of the European CPA.net Board for Chronic Pulmonary Aspergillosis

Richard B. Moss, MD – Professor Emeritus, Pediatrics-Pulmonary Medicine at Stanford University School of Medicine, Stanford, CA

David A. Stevens, MD, FACP, FAAM, FIDSA – Professor Emeritus, Infectious Diseases and Geographic Medicine at Stanford University School of Medicine, Stanford, CA; President, California Institute for Medical Research, San Jose, CA

Peter Wark, BMed PhD FRACP ThorSoc – Conjoint Professor, Centre for Healthy Lungs, University of Newcastle & Department of Respiratory and Sleep Medicine John Hunter Hospital, New Lambton NSW, Australia

William J Calhoun, MD – Professor and Vice Chair for Research, Divisions of Pulmonary, Critical Care and Sleep, and Allergy/Immunology. Department of Medicine at University of Texas Medical Branch, Galveston TX

Mario Castro, MD, MPH, FCCP – Professor of Medicine, Pediatrics, and Radiology, Division of Pulmonary and Critical Care Medicine at the Washington University School of Medicine in St. Louis, MO

Jonathan Corren, MD – Associate Clinical Professor of Medicine and Pediatrics, David Geffen School of Medicine at UCLA, Los Angeles, CA

Russell Settipane, MD – Clinical Professor of Medicine at Brown Medical School, Director of the Allergy & Asthma Center, Providence, RI; Associate Editor of Allergy & Asthma Proceedings and the Journal of Precision Respiratory Medicine, President Eastern Allergy and Eastern Pulmonary Conferences

Michael Wechsler, MD, MMSc – Professor of Medicine, Director, NJH Cohen Family Asthma Institute, Dept. of Medicine, National Jewish Health, Denver, CO
Phase 2 Scheduled to Begin 4Q 2018 and Expected to Support Proof of Mechanism in Patients with Asthma-ABPA

28-day Safety, Tolerability, Pulmonary Function and Biomarker Study in Asthmatic ABPA Patients

Randomized, double-blind, placebo controlled study (1:1 randomization; each cohort n = 16).

Primary Endpoint
- Safety & Tolerability

Other Endpoints
- Plasma and sputum PK
- Pulmonary Function (FEV₁)
- Sputum and plasma eosinophils
- IgE and IgG (specific to A. fumigatus antigens) plasma concentrations
- Aspergillus burden in sputum
- Disease control (ACQ-6)
- FeNO

Patient Profile
- Moderate-to-severe asthmatics (M/F, ages 18-65) confirmed/stable ABPA, diagnosed with ABPA
- Exacerbation and steroid use entry criteria

Anticipated Outcomes and Value Catalysts
- Establish safety and tolerability of Pulmazole over 4 weeks in patients with ABPA
- Reductions in relevant biomarkers of inflammation in ABPA increase confidence that longer treatment will achieve clinically significant improvements in Phase 2b/3
- Evaluating impact on microbiology including Aspergillus burden in sputum, further substantiating link between fungal burden and other biomarkers / efficacy
- Demonstrate directional improvement in FEV₁ as a potentially approvable endpoint

FeNO = Fractional Exhaled Nitric Oxide; PK = Pharmacokinetics; ACQ-6 = Asthma Control Questionnaire 6
Phase 2b/3 Proof of Concept Trial
Powered to Show Improvement in FEV\textsubscript{1}

Phase 2b/3: POC Efficacy/Safety Trial in Asthmatic ABPA Patients

Randomized, double-blind, placebo controlled study (n=85 per arm). 16 weeks dosing with 16 weeks follow-up.

**Primary Efficacy Endpoint**
- FEV\textsubscript{1}

**Secondary Endpoint**
- Disease control (ACQ-6)
- Exacerbation (frequency/timing)
- Steroid use

**Exploratory Biomarkers**
- Sputum/plasma eosinophils
- IgE and IgG (specific to A. fumigatus antigens) plasma concentrations
- Aspergillus burden in sputum
- FeNO

**Patient Profile**
- Moderate-to-severe asthmatics (M/F, ages 18-65) confirmed/stable ABPA, diagnosed with ABPA
- Exacerbation and steroid use entry criteria

**Anticipated Outcomes and Value Catalysts**
- Demonstrate clear efficacy signal in FEV\textsubscript{1} and/or exacerbations (clinically relevant endpoints)
- Establish safety and tolerability of Pulmazole over 32 weeks in patients with ABPA
- Substantiate correlation between clinical outcomes and ABPA biomarkers and fungal burden in lungs

**Endpoints:**
- PFT/Biomarkers
- Exacerbations
- Disease Control
- QOL

**ABPA Patients**

- Placebo
- Pulmazole (10 mg)
- Pulmazole (20 mg)
PUR1800 – Narrow Spectrum Kinase Inhibitor (NSKI)

Inhaled P38, Syk, Src Kinase Inhibitor to Treat Acute Exacerbations in COPD (AECOPD)
**The PUR1800 Advantage**

**Target Product Profile**
- SOC* + Placebo vs. SOC* + PUR1800
- Acute treatment, not prevention, paradigm
- 4-week treatment period and 24-week follow up period
- Primary Endpoint: >100ml FEV₁ improvement vs. SOC

**Unmet Need in AECOPD**
- Steroid resistant AECOPD
- Steroidal lack of efficacy in viral and/or bacterial driven AECOPD
- 20%–50% treatment failure or only partial response with oral steroids

**PUR1800 Value Drivers**
- Acting across 3-kinase, expected efficacy in viral, bacterial and eosinophilic driven AECOPD
- Non-steroidal, inhaled anti-inflammatory with low systemic exposure

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**Prescriber Reported PUR1800 Utilization**

<table>
<thead>
<tr>
<th>Treatment Option</th>
<th>Current Use</th>
<th>Expected Use</th>
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</thead>
<tbody>
<tr>
<td>PUR1800 (plus oral corticosteroids and/or antibiotics)</td>
<td>0%</td>
<td>~35%</td>
</tr>
<tr>
<td>Oral Corticosteroids + Antibiotics</td>
<td>~58%</td>
<td>~34%</td>
</tr>
<tr>
<td>Antibiotics Alone</td>
<td>~13%</td>
<td>~10%</td>
</tr>
<tr>
<td>Oral Corticosteroids Alone</td>
<td>~25%</td>
<td>~18%</td>
</tr>
<tr>
<td>No Treatment</td>
<td>~4%</td>
<td>~3%</td>
</tr>
</tbody>
</table>

Source: Physician Interviews; Physician Quantitative Survey; ClearView Analysis. Physician Reported Utilization in Moderate-to-Severe Acute Exacerbations.

* Standard of Care (SOC) is oral corticosteroids with or without antibiotic
PUR1800: Inhaled NSKI Has Up to $2.4B Peak Revenue Potential in the U.S.

Large Addressable AECOPD Burden

- ~16M COPD patients in the U.S.
- 77% experience at least one exacerbation annually
- ~18M moderate-to-severe AECOPD episodes annually
- > 20% corticosteroid treatment failure rate in moderate-to-severe AECOPD patients
- PUR1800 potentially has efficacy across the spectrum of causes of AECOPD

Clinical Precedent with this NSKI in Phase 1/1b COPD

Sputum neutrophils (Log10 cells/g sputum)

Neutrophils

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<tr>
<th>Sc</th>
<th>d12</th>
<th>Sc</th>
<th>d12</th>
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<tbody>
<tr>
<td>Placebo</td>
<td>p=0.68</td>
<td>PUR1800</td>
<td>p=0.02</td>
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PUR1800 Up to $2.4B U.S. Peak Revenue* Opportunity in AECOPD

PUR1800 Market Opportunity

- Up to 35% expected use**, in addition to standard of care (oral corticosteroids plus/minus antibiotic)

Pricing Potential & Market Access

- 70% payer market access*** with minimal use restrictions
- Targeting $650 per incident launch price

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*Also includes discount for patient compliance, patient persistence, and gross-to-net adjustment; ** Peak revenues expected at loss of market exclusivity, ~14 years post launch; *** Estimate based on ClearView Analysis, which included qualitative physician surveys and interviews; NSKI = Narrow Spectrum Kinase Inhibitor; AECOPD = Acute Exacerbations of COPD
PUR1800 Phase 2a Study in Stable COPD (RV1162 to PUR1800 Bridging Study)

Safety/Tolerability and PK Study of iSPERSE PUR1800 (NSKI) in Stable Moderate-Severe COPD Patients

Randomization (1:1)

Patient Profile
- Moderate-to-Severe COPD Patients
- 30% ≤ FEV₁ < 80%; n=60; 12–15 patients per arm

225 mcg iSPERSE PUR1800 ≈ 500 mcg Janssen RV1162 Lactose Blend (based on predicted lung deposition)

*Doses to be administered daily for 28 days and confirmed following 4Q 2018 completion of GLP tox study

Primary Endpoint
- Safety & Tolerability

Other Endpoints
- Single dose PK profile
- Multiple dose PK profile over 14 and 28 days
- Pulmonary function
- Exploratory biomarkers
  - Target engagement
  - Sputum biomarkers

Anticipated Outcomes and Value Catalysts
- Establish safety and tolerability of PUR1800 over 4 weeks in patients with stable COPD
- Evaluate kinase target engagement and biomarker activity to demonstrate anti-inflammatory effect

Phase 2 Start
- 3Q-2019
- 4Q-2019
- 1Q-2020
- 2Q-2020
- 3Q-2020

Phase 2a Data
PUR0200 – Long Acting Muscarinic Agonist (LAMA)

- Inhaled Tiotropium Bromide for Treatment of Chronic Obstructive Pulmonary Disease (COPD)
PUR0200: Out-License Creates Revenue Stream and Further Prioritizes Pulmazole and PUR1800

We Believe PUR0200 is an Improved Version of the Spiriva® HandiHaler®

Equivalent Lung Exposure at 80% Nominal Dose

We Believe PUR0200 is an Improved Version of the Spiriva® HandiHaler®

PUR0200 Improves Lung Function in COPD at Low Doses (6x as Potent as Spiriva®)

PUR0200 Shows Comparable Systemic Exposure to Spiriva® at a Lower Dose

Unmet Needs

- Spiriva® $3.5B WW revenues in 2016
- Spiriva lacks a dry-powder multi-dose inhaler in the U.S.
- In the rest of world, there is a need for therapeutically equivalent, lower cost products

Vectura Deal Structure & Future Activity

- Out-licensed PUR0200 and PUR0200 combination products for U.S.
- $1M milestone and revenue share
- No further investment by Pulmatrix to develop and commercialize
- Pursuing PUR0200 ex-U.S. out-license

Spiriva HH = Spiriva® HandiHaler® (18 µg dose)
Summary
# Robust Pipeline with Projected Significant Value Catalysts*

<table>
<thead>
<tr>
<th>Product Pipeline</th>
<th>Indication</th>
<th>2H 2018</th>
<th>2019</th>
<th>1H 2020</th>
<th>Milestone</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Pulmazole</strong> Anti-fungal</td>
<td>Allergic Bronchopulmonary Aspergillosis (ABPA) in Patients with Asthma</td>
<td></td>
<td></td>
<td></td>
<td>Phase 2 4Q 2019</td>
</tr>
<tr>
<td><strong>PUR1800 NSKI</strong></td>
<td>Acute Exacerbations of COPD (AECOPD)</td>
<td></td>
<td></td>
<td></td>
<td>Phase 2a 2Q 2020</td>
</tr>
<tr>
<td><strong>PUR5700 NSKI</strong></td>
<td>Idiopathic Pulmonary Fibrosis (IPF)</td>
<td></td>
<td></td>
<td></td>
<td>Pre-clinical</td>
</tr>
</tbody>
</table>

## Potential Future Revenue Opportunities

<table>
<thead>
<tr>
<th>Product Pipeline</th>
<th>Indication</th>
<th>Milestone</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>PUR0200-US LAMA</strong></td>
<td>Chronic Obstructive Pulmonary Disease (COPD)</td>
<td>Out-Licensed to Vectura for U.S. 09/2017</td>
</tr>
</tbody>
</table>

*LAMA = Long-Acting Muscarinic Antagonist; * Estimated Milestones
Experienced Leadership Team With Experience to Execute and Deliver Value

Management Team

- Robert Clarke, PhD
  CEO

- Bill Duke, MBA
  CFO

- David Hava, PhD
  CSO

- Ted Raad, MBA
  CBO

- Jim Roach, MD
  CMO

Board of Directors

- Mark Iwicki
  Chairman
  Chairman, Pulmatrix

- Matthew Sherman
  BOD
  President, Accleron

- Terry McGuire
  BOD Investor
  Managing Partner, Polaris Partners

- Steve Gillis, PhD
  BOD Investor
  Managing Partner, Arch Venture Partners

- Michael Higgins
  BOD-Audit Chair
  Chairman, Ironwood

- Amit Munshi
  BOD
  General Partner, Arena Pharmaceuticals

Strong Background in Product Development and Commercialization

- Proven fundraising and capital management ability
- Experienced R&D team focused on pulmonary drug delivery and the role of inhaled particles
- Commercialization and partnership experience

Diverse Collective Experience at Leading Respiratory Companies

- Pulmatrix
- Sunovion
- Genzyme
- Novartis
- Valeritas
- Alkermes
Pulmatrix is Positioned to Create Significant Value in the Near Term

Investment Thesis

- Compelling Ph1/1b data demonstrating high itraconazole lung concentration and low plasma concentration with Pulmazole compared to Sporanox
- Ph2a proof of mechanism clinical study readout in 4Q 2019 with product exceeding $1B in peak revenue potential
- Pulmazole is a “de-risked” 505(b)(2) program
- Rigorous clinical development plans developed in collaboration with global thought leaders in ABPA, asthma and COPD
- Robust product portfolio, including PUR5700 and PUR1800 with billions of peak revenue potential in major pulmonary diseases like COPD and IPF
- Underlying iSPERSE™ intellectual property carries well into 2030’s with additional patent protection through specific drug development
Glossary of Terms

- ABPA – Allergic Bronchopulmonary Aspergillosis
- ACQ-6 – Asthma Control Questionnaire 6
- AECOPD – Acute Exacerbations of Chronic Obstructive Pulmonary Disease
- CAB – Clinical Advisory Board
- CF – Cystic Fibrosis
- COPD – Chronic Obstructive Pulmonary Disease
- FeNO – Fractional Exhaled Nitric Oxide
- FEV₁ – Forced Expiratory Volume in 1 Second
- FPI – First Patient In
- FVC – Forced Vital Capacity
- HNV – Healthy Normal Volunteers
- IgE – Immunoglobulin E Antibodies
- IgG – Immunoglobulin G Antibodies
- IP – Intellectual Property
- KOL – Key Opinion Leader
- MAD – Multiple Ascending Dose
- MAPK – Mitogen-Activated Protein Kinases
- MOA – Mechanism of Action
- NSKI – Narrow Spectrum Kinase Inhibitor
- PBO – Placebo
- PEF – Peak Expiratory Flow
- PK – Pharmacokinetics
- POC – Proof of Concept
- POM – Proof of Mechanism
- Pulmazole – PUR1900
- qPCR – Quantitative Polymerase Chain Reaction
- SAD – Single Ascending Dose
- SD – Single Dose
- Spiriva HH – Spiriva® HandiHaler® (18 µg dose)
- Sporanox – Oral Itraconazole 200mg