



# Merger of Akebia Therapeutics, Inc. and Keryx Biopharmaceuticals, Inc.

***Creating a Fully Integrated Company Focused on  
the Development and Commercialization of  
Therapeutics for Patients with Kidney Disease***

**July 11, 2018**

# Forward-Looking Statements

These materials contain forward-looking statements within the meaning of the federal securities law. Such statements are based upon current plans, estimates and expectations that are subject to various risks and uncertainties. The inclusion of forward-looking statements should not be regarded as a representation that such plans, estimates and expectations will be achieved. Words such as “anticipate,” “create,” “expect,” “project,” “intend,” “believe,” “may,” “will,” “should,” “plan,” “could,” “target,” “contemplate,” “estimate,” “position,” “predict,” “potential,” “opportunity” and words and terms of similar substance used in connection with any discussion of future plans, actions or events identify forward-looking statements. All statements, other than historical facts, including statements regarding the expected timing of the closing of the merger; the ability of the parties to complete the merger considering the various closing conditions; the potential benefits of vadadustat; expected timing of exercise of Akebia’s Otsuka funding option; the timing of availability of top-line results from Akebia’s clinical trials of vadadustat; the potential to establish a new standard of care; the expected timing of enrollment in clinical trials; revenue growth; the market opportunity, commercial momentum and growth potential of Auryxia; the expected benefits of the merger, such as efficiencies, the expected management team, cost savings and the expected timing thereof, synergies, the ability to deliver value, the potential to maximize sales, the ability to build launch momentum for vadadustat in the U.S., enhanced revenues, growth potential, market profile, financial strength, and financial flexibility, the potential for accelerating profitability and reducing capital needs; the competitive ability and position of the combined company; the strategy of the combined company; the potential of the combined company to address common forms of anemia in CKD, deliver innovative therapies, improve patient outcomes, and identify, develop and commercialize new therapeutic options; the potential market opportunity of the combined company; the expected cash runway of the combined company; and any assumptions underlying any of the foregoing, are forward-looking statements. Important factors that could cause actual results to differ materially from Akebia’s and Keryx’s plans, estimates or expectations could include, but are not limited to: (i) Akebia or Keryx may be unable to obtain stockholder approval as required for the merger; (ii) conditions to the closing of the merger may not be satisfied; (iii) the merger may involve unexpected costs, liabilities or delays; (iv) the effect of the announcement of the merger on the ability of Akebia or Keryx to retain and hire key personnel and maintain relationships with customers, suppliers and others with whom Akebia or Keryx does business, or on Akebia’s or Keryx’s operating results and business generally; (v) Akebia’s or Keryx’s respective businesses may suffer as a result of uncertainty surrounding the merger and disruption of management’s attention due to the merger; (vi) the outcome of any legal proceedings related to the merger; (vii) Akebia or Keryx may be adversely affected by other economic, business, and/or competitive factors; (viii) the occurrence of any event, change or other circumstances that could give rise to the termination of the merger agreement; (ix) risks that the merger disrupts current plans and operations and the potential difficulties in employee retention as a result of the merger; (x) the risk that Akebia or Keryx may be unable to obtain governmental and regulatory approvals required for the transaction, or that required governmental and regulatory approvals may delay the transaction or result in the imposition of conditions that could reduce the anticipated benefits from the proposed transaction or cause the parties to abandon the proposed transaction; (xi) risks that the anticipated benefits of the merger or other commercial opportunities may otherwise not be fully realized or may take longer to realize than expected; (xii) the impact of legislative, regulatory, competitive and technological changes; (xiii) expectations for future clinical trials, the timing and potential outcomes of clinical studies and interactions with regulatory authorities; and (xiv) other risks to the consummation of the merger, including the risk that the merger will not be consummated within the expected time period or at all. Additional factors that may affect the future results of Akebia and Keryx are set forth in their respective filings with the SEC, including each of Akebia’s and Keryx’s most recently filed Annual Report on Form 10-K, subsequent Quarterly Reports on Form 10-Q, Current Reports on Form 8-K and other filings with the SEC, which are available on the SEC’s website at [www.sec.gov](http://www.sec.gov). See in particular Item 1A of Akebia’s Quarterly Report on Form 10-Q for the quarter ended March 31, 2018 under the heading “Risk Factors” and Item 1A of Keryx’s Quarterly Report on Form 10-Q for the quarter ended March 31, 2018 under the heading “Risk Factors.” The risks and uncertainties described above and in Akebia’s most recent Quarterly Report on Form 10-Q and Keryx’s most recent Quarterly Report on Form 10-Q are not exclusive and further information concerning Akebia and Keryx and their respective businesses, including factors that potentially could materially affect their respective businesses, financial condition or operating results, may emerge from time to time. Readers are urged to consider these factors carefully in evaluating these forward-looking statements, and not to place undue reliance on any forward-looking statements. Readers should also carefully review the risk factors described in other documents that Akebia and Keryx file from time to time with the SEC. The forward-looking statements in these materials speak only as of the date of these materials. Except as required by law, Akebia and Keryx assume no obligation to update or revise these forward-looking statements for any reason, even if new information becomes available in the future.

# Additional Information

## *About Akebia Therapeutics, Inc.*

Akebia Therapeutics, Inc. is a biopharmaceutical company headquartered in Cambridge, Massachusetts, focused on delivering innovative therapies to patients with kidney disease through hypoxia-inducible factor biology. For more information, please visit our website at [www.akebia.com](http://www.akebia.com), which does not form a part of this release.

## *About Keryx Biopharmaceuticals, Inc.*

Keryx Biopharmaceuticals, Inc., headquartered in Boston, Massachusetts, is focused on the development and commercialization of innovative medicines that provide unique and meaningful advantages to people with kidney disease. The Keryx team works with passion to advance the care of people with this complex disease. This dedication has resulted in two FDA-approved indications for Keryx's first medicine, Auryxia (ferric citrate) tablets. For more information about Keryx, please visit [www.keryx.com](http://www.keryx.com).

## *Additional Information and Where to Find It*

In connection with the proposed merger, Akebia Therapeutics and Keryx Biopharmaceuticals plan to file with the SEC and mail or otherwise provide to their respective shareholders a joint proxy statement/prospectus regarding the proposed transaction. BEFORE MAKING ANY VOTING DECISION, AKEBIA'S AND KERYX'S RESPECTIVE shareholders ARE URGED TO READ THE JOINT PROXY STATEMENT/PROSPECTUS IN ITS ENTIRETY WHEN IT BECOMES AVAILABLE AND ANY OTHER DOCUMENTS FILED BY EACH OF AKEBIA AND KERYX WITH THE SEC IN CONNECTION WITH THE PROPOSED MERGER OR INCORPORATED BY REFERENCE THEREIN BECAUSE THEY WILL CONTAIN IMPORTANT INFORMATION ABOUT THE PROPOSED TRANSACTION AND THE PARTIES TO THE PROPOSED TRANSACTION. Investors and shareholders will be able to obtain a free copy of the joint proxy statement/prospectus and other documents containing important information about Akebia and Keryx, once such documents are filed with the SEC, through the website maintained by the SEC at [www.sec.gov](http://www.sec.gov). Akebia and Keryx make available free of charge at [www.akebia.com](http://www.akebia.com) and [www.keryx.com](http://www.keryx.com), respectively (in the "Investors" section), copies of materials they file with, or furnish to, the SEC.

## *Participants in the Solicitation*

This document does not constitute a solicitation of proxy, an offer to purchase or a solicitation of an offer to sell any securities. Akebia Therapeutics, Keryx Biopharmaceuticals and their respective directors, executive officers and certain employees and other persons may be deemed to be participants in the solicitation of proxies from the shareholders of Akebia and Keryx in connection with the proposed merger. Security holders may obtain information regarding the names, affiliations and interests of Akebia's directors and officers in Akebia's Annual Report on Form 10-K for the fiscal year ended December 31, 2017, which was filed with the SEC on March 12, 2018 and its definitive proxy statement for the 2018 annual meeting of shareholders, which was filed with the SEC on April 30, 2018. Security holders may obtain information regarding the names, affiliations and interests of Keryx's directors and officers in Keryx's Annual Report on Form 10-K for the fiscal year ended December 31, 2017, which was filed with the SEC on February 21, 2018, and the Amendment No. 1 on Form 10-K/A, which was filed with the SEC on April 30, 2018, and its definitive proxy statement for the 2018 annual meeting of shareholders, which was filed with the SEC on May 31, 2018. To the extent the holdings of Akebia's securities by Akebia's directors and executive officers or the holdings of Keryx securities by Keryx's directors and executive officers have changed since the amounts set forth in Akebia's or Keryx's respective proxy statement for its 2018 annual meeting of shareholders, such changes have been or will be reflected on Statements of Change in Ownership on Form 4 filed with the SEC. Additional information regarding the interests of such individuals in the proposed merger will be included in the joint proxy statement/prospectus relating to the proposed merger when it is filed with the SEC. These documents (when available) may be obtained free of charge from the SEC's website at [www.sec.gov](http://www.sec.gov), Akebia's website at [www.akebia.com](http://www.akebia.com) and Keryx's website at [www.keryx.com](http://www.keryx.com).

# Fully Integrated, Kidney Disease Therapeutics Company Positioned to Deliver Substantial Value Long Term



**Establishes a Leading Renal Company with Enhanced Position and Large Market Opportunity**

**Creates Potential for Accelerated Growth and Organizational Synergies**

**Combines Experienced Renal Management Teams**

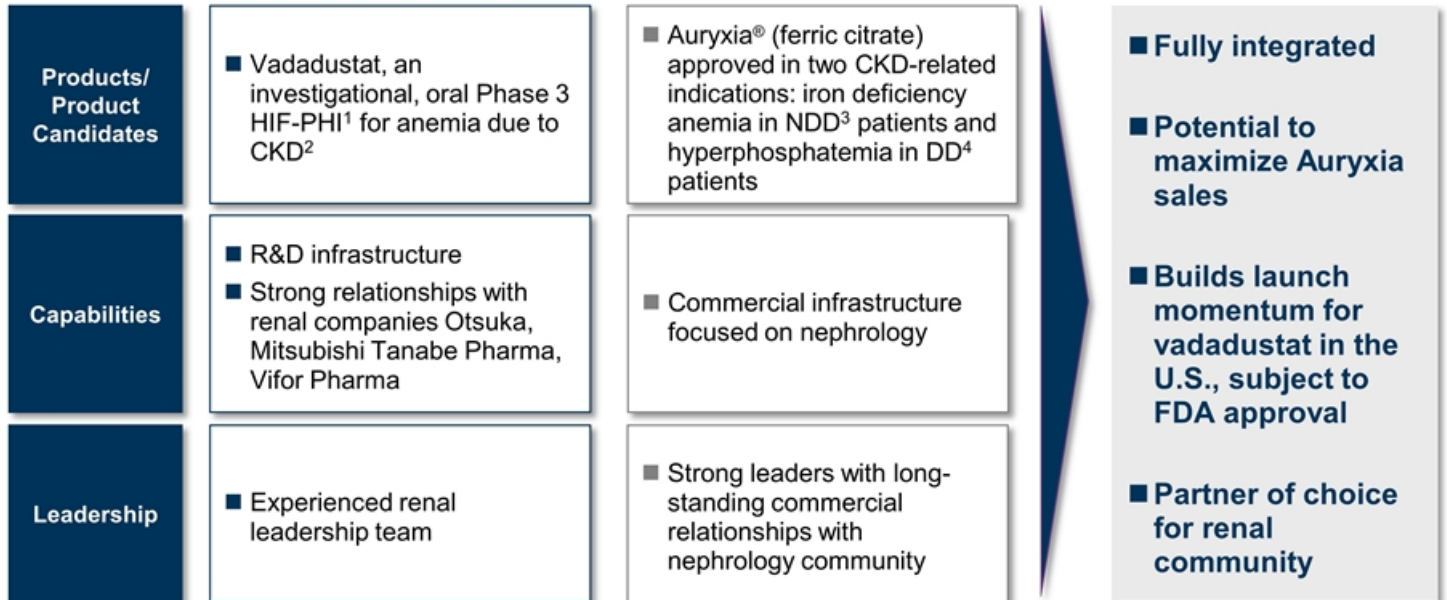
**Strengthens Financial Profile**

**Potential Cost Savings of >\$250M to Be Realized Five Years Following Closing**

# Combination Highlights

<b>Terms</b>	<ul style="list-style-type: none"> <li>• Stock for stock merger</li> <li>• Each share of Keryx will be converted into 0.37433 shares of Akebia</li> </ul>
<b>Ownership</b>	<ul style="list-style-type: none"> <li>• Akebia shareholders to own 49.4% of the pro forma company and Keryx shareholders to own 50.6%, on a fully diluted basis</li> </ul>
<b>Cash Position</b>	<ul style="list-style-type: none"> <li>• Pro forma company has \$453M of cash as of March 31, 2018</li> <li>• The Baupost Group, Keryx's largest shareholder, will convert its \$165M convertible bond prior to closing of the transaction; conversion to common will provide financial flexibility</li> </ul>
<b>CEO &amp; Board of Directors</b>	<ul style="list-style-type: none"> <li>• CEO: John P. Butler</li> <li>• Chairperson to be appointed by Keryx</li> </ul>
<b>Closing Conditions</b>	<ul style="list-style-type: none"> <li>• Subject to approval of Akebia and Keryx shareholders</li> <li>• Subject to other customary closing conditions</li> </ul>
<b>Voting Agreements</b>	<ul style="list-style-type: none"> <li>• The Baupost Group, holder of 21.4% of outstanding Keryx common stock</li> <li>• Muneer A. Satter, Chairperson of Akebia's Board of Directors and holder of 5.3% of outstanding Akebia common stock</li> </ul>
<b>Transaction Close</b>	<ul style="list-style-type: none"> <li>• Expected by the end of 2018</li> </ul>

# Creating a Leader in Kidney Disease Therapies

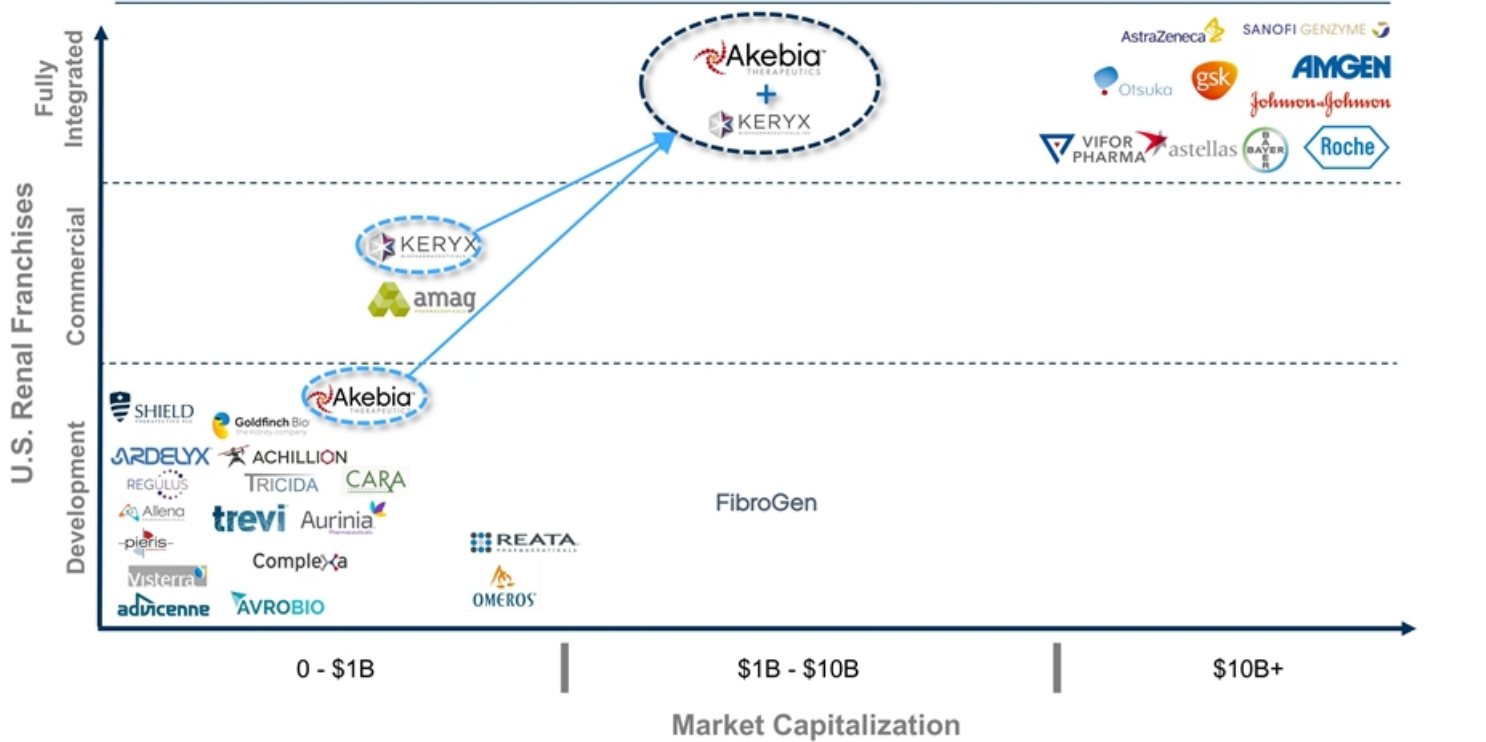


1. Hypoxia Inducible Factor - Prolyl Hydroxylase Inhibitor  
 2. Chronic Kidney Disease  
 3. Non Dialysis Dependent  
 4. Dialysis Dependent

# Renal Portfolio and Scale Create a Well-Positioned Renal Company

The combined company will have significant financial strength and flexibility with a highly complementary nephrology portfolio

## Renal Competitive Landscape



# Strong Financial Pro Forma Company

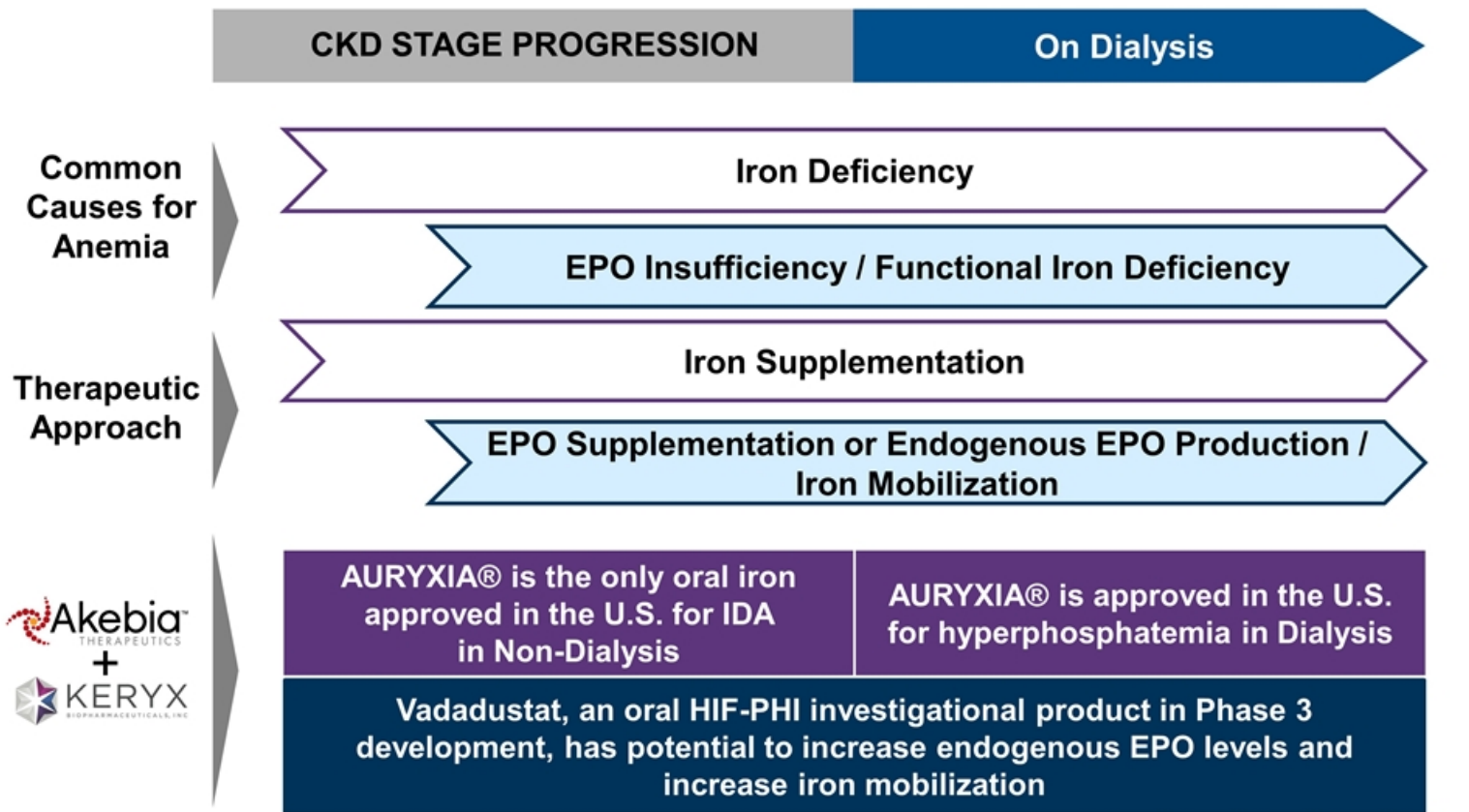
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- Pro forma cash on hand \$453M as of March 31, 2018
- Pro forma funded into Q1 2020, unchanged vs Akebia stand alone
- Vadadustat brings Phase 3 data catalysts and large revenue opportunity, vs Keryx stand alone
- Auryxia revenue has the potential to accelerate profitability and reduce capital needs, vs Akebia stand alone
- Vadadustat R&D funding option<sup>1</sup> exercisable in 1H 2019, covering vast majority of Phase 3 cost
- Financial flexibility following full conversion of \$165M of Baupost debt into common stock
- Cost synergies expected to start in 2019 and ramp up with launch preparation for vadadustat

1. Describes R&D Funding Option (Otsuka to pay 80% of development costs across its two collaborations with Akebia going forward)



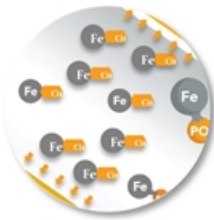
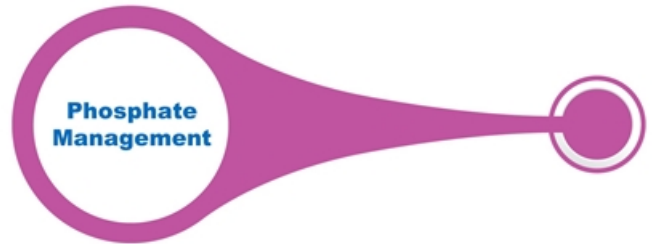
# Combined Company Has Potential to Address Common Forms of Anemia in CKD



# Auryxia: One Mechanism of Action to Treat Two Common Complications Associated with CKD



The ferric iron component binds to dietary phosphate in the GI tract and precipitates as ferric phosphate. This compound is insoluble and excreted in the stool.



The ferric iron/citrate component remains soluble in the GI tract, enabling absorption and transport of the iron and eventual incorporation into hemoglobin.



# Auryxia: Approved in Two Indications in the U.S.: Iron Deficiency Anemia in Non-Dialysis & Hyperphosphatemia in Dialysis



## AURYXIA® (ferric citrate) tablets

①

A phosphate binder indicated for the control of serum phosphorus levels in adult patients with chronic kidney disease on dialysis

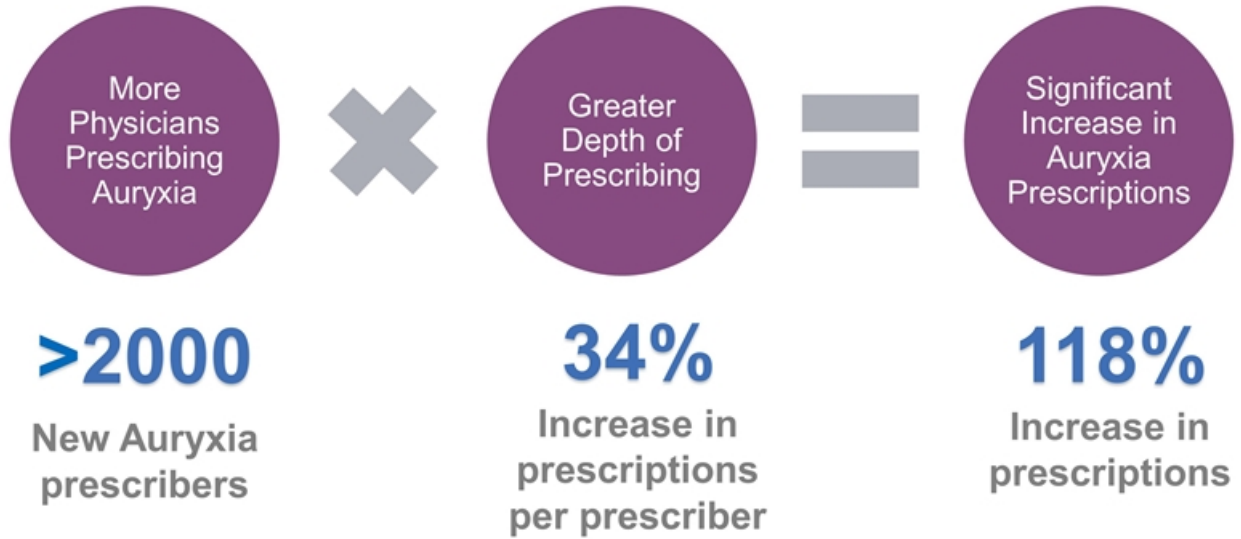
AND

②

An iron replacement product indicated for the treatment of iron deficiency anemia in adult patients with chronic kidney disease not on dialysis

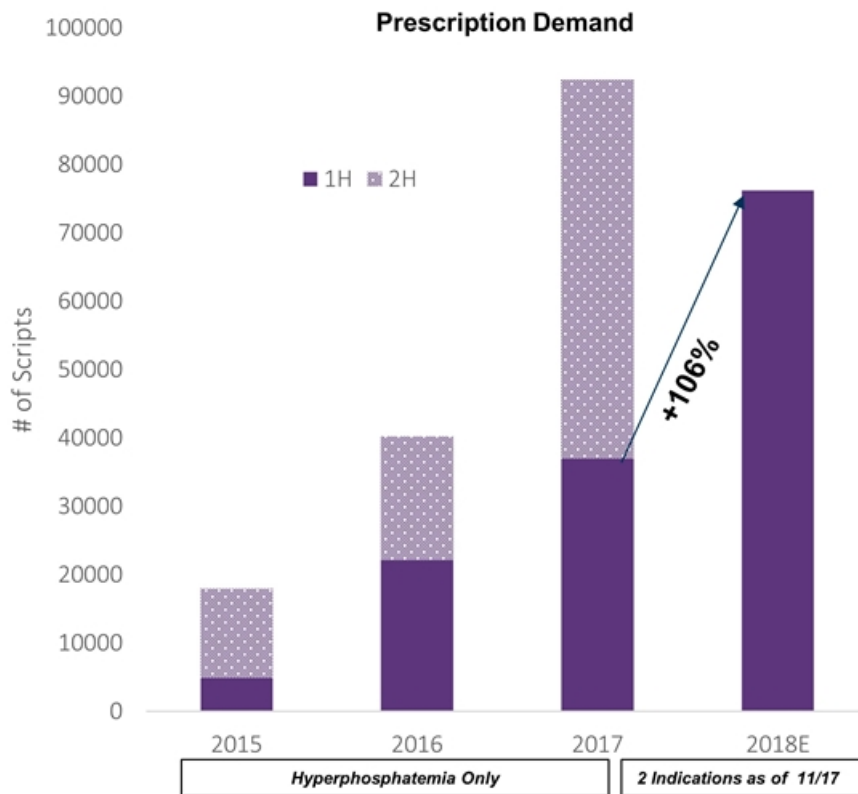
# Auryxia Commercial Progress Yielded >2X Scripts Year-Over-Year

(Q1 2018 vs. Q1 2017)



\*Source: Keryx data on file based on IMS and specialty pharmacy demand data

# Snapshot of Increasing Prescription Demand

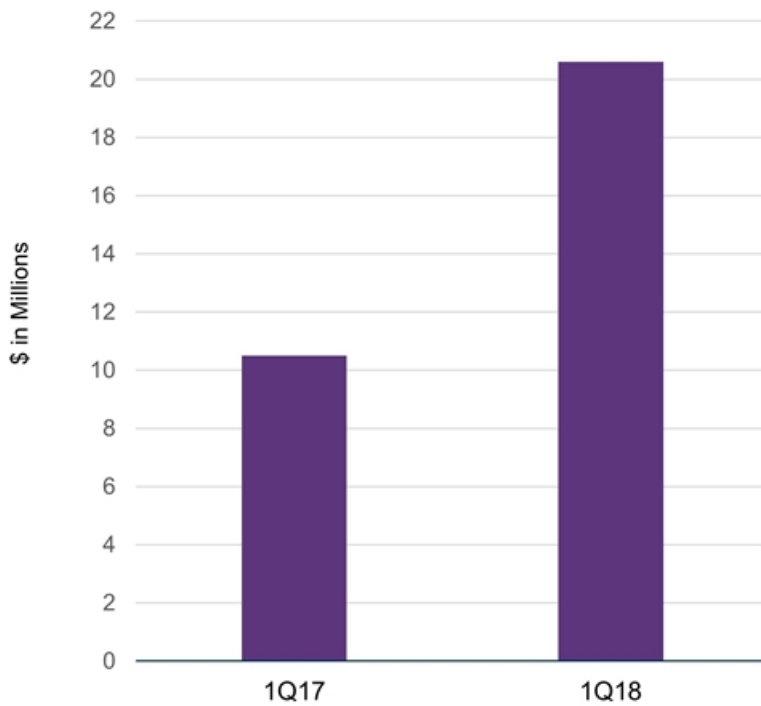


## 2018 IDA Launch Highlights:

- ~90% of surveyed nephrologists\* are aware of Aurxyia's indication for iron deficiency anemia
- Majority of surveyed nephrologists\* who have used Aurxyia as a treatment for IDA report that they are satisfied with Aurxyia
- Continued growth in hyperphosphatemia while gaining traction in IDA

# Auryxia Revenue Nearly Doubled Year-Over-Year

First Quarter Net Auryxia U.S. Product Sales



**Strong momentum accelerating in 2018**

- **Broad formulary access with Medicare Part D and commercial insurers**
- **Highest Rx market share growth in hyperphosphatemia in the U.S. YTD<sup>1</sup>**
- **High disease awareness<sup>2</sup> and Auryxia differentiation accelerate uptake in IDA**

\*Sources:

1. Keryx data on file based on IMS and specialty pharmacy demand data
2. Spherix Global Anemia 1Q Pulse (2018); aided awareness data

# Significant Growth Potential for Auryxia

## Strong commercial momentum in 2018

- Broad formulary access with Medicare Part D and commercial insurers
- Highest Rx market share growth in hyperphosphatemia in the U.S. YTD<sup>1</sup>
- High disease awareness<sup>2</sup> and Auryxia differentiation accelerate uptake in IDA

## Recent ERA-EDTA data<sup>3</sup>

- Single center, open label, IST<sup>4</sup> compared ferric citrate vs. SOC in 200 pre-dialysis patients
- Analysis showed an effect on TSAT & ferritin, hemoglobin, serum phosphate, and FGF23 in non-dialysis period

## Additional growth opportunities driven by portfolio synergies

- Operational efficiencies including leveraging combined set of relationships and leadership
- Education about anemia in CKD and development with HIF-PHI opportunities

\*Sources:

1. Keryx data on file based on IMS and specialty pharmacy demand data
2. Spherix Global Anemia 1Q Pulse 2018; aided awareness data
3. Block et. al., A Randomized Trial of the Effects of Ferric Citrate in Patients with Advanced Chronic Kidney Disease; late breaker at ERA, 2018
4. IST = Investigator Sponsor Trial. Funded by Keryx

# Vadadustat, an Investigational HIF-PHI, Represents an Innovative Potential Approach to Treatment of Anemia Due to CKD

- **iESAs\*:** standard of care for anemia due to CKD for over 20 years
- **iESAs are associated with significant safety concerns:**
  - A proportion of NDD patients are not treated with iESAs due to safety and administration considerations<sup>1</sup>
  - DD patients rely on iESAs for treatment

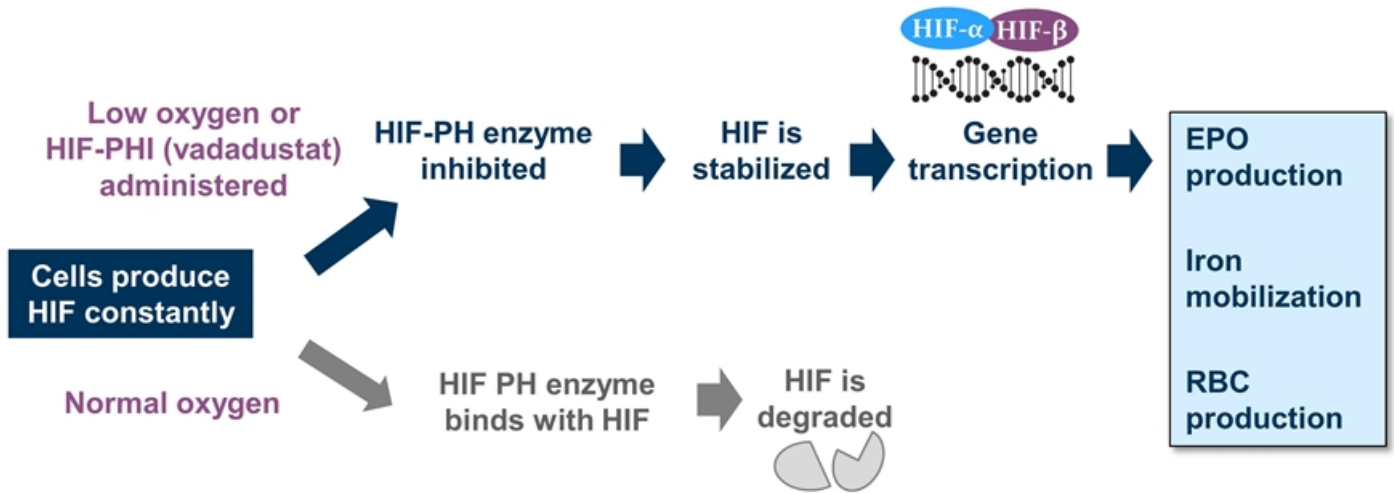
- **HIF-PHIs represent opportunity for a new class of treatment:**
  - Have potential to be oral alternatives to iESAs
  - Rely on the same pathway the body uses to adapt to lower oxygen availability
  - Potential for a differentiated profile

\*Injectable erythropoiesis-stimulating agents

1. Thamer et. al. Am J Kidney Dis. 2014 Nov; 64(5):706-13, Akebia market research



# Vadadustat Has Potential to Stimulate Endogenous EPO Production and Mobilize Iron by Inhibiting HIF-PH



Vadadustat is a Phase 3, investigational, oral HIF-PHI that is not approved by the FDA

EPO, erythropoietin; PH, prolyl hydroxylase; RBC, red blood cell.  
Maxwell PH, Eckardt K-U. HIF prolyl hydroxylase inhibitors for the treatment of renal anemia and beyond. *Nat Rev Nephrol.* 2015;12(3):157-168.

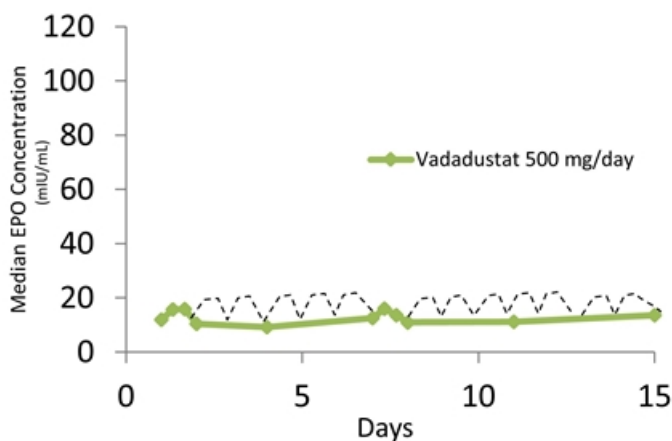
# Vadadustat Avoided Supra-Physiological EPO Levels

## EPO vs. Time by Study

### Vadadustat (Oral)

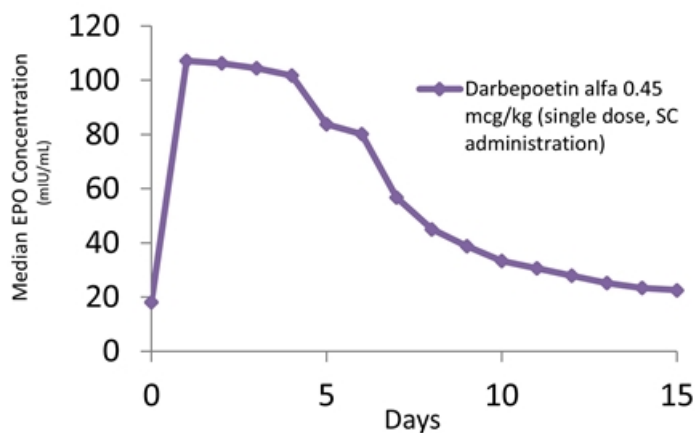
Phase 1 Study in Healthy Volunteers<sup>1</sup>

Dashed line represents simulated EPO data



### Darbepoetin Alfa (SC)

PK-PD Model in CKD Patients<sup>2</sup>



Not a head-to-head comparison

#### Vadadustat is a Phase 3, investigational, oral HIF-PHI that is not approved by the FDA

1. Akebia Therapeutics, Inc. Data on File (2010). Data from Phase 1 study in healthy volunteers with vadadustat once daily dosing. Pre-dose EPO concentrations were evaluated on Days 1, 4, 7, 11, 15 and 22. Post-dose data to assess acute rise in EPO following vadadustat dosing was only completed on Day 1 and Day 7 (8 and 16 hours post-dose). Dashed line represents estimated EPO levels based on post-dose data from Day 1 and Day 7.
2. Doshi S et al. Journal of Clinical Pharmacology, 2010;50:75S-90S. Original figure redrawn to depict darbepoetin alfa serum concentration (ng/mL/(mcg/kg)) converted to mIU/mL. Data from 6 clinical studies conducted with extensive PK sampling in CKD patients following subcutaneous (SC) administration of a single dose or first dose of a monthly dosing regimen ranging from 0.4-0.6mcg/kg, dose normalized to 0.45 mcg/kg.

# Vadadustat Development Program Informed By Key Unmet Needs In Anemia Due to CKD

## Unmet Needs

Less variability in **hemoglobin levels**

Lower risk of **CV events**

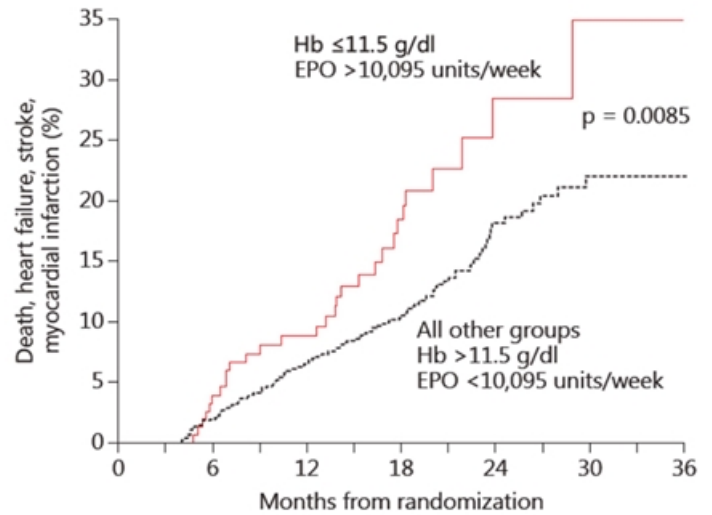
Lower risk of **hypertension**

Efficacy in **hyporesponders**

More **convenient dosing** for NDD

## Kaplan-Meier Survival Curves<sup>1</sup>

Death, Heart Failure, Stroke, Myocardial Infarction (%)



<sup>1</sup> McCullough P.A., et al. Am J Nephrol 2013;37:549-558 (DOI:10.1159/000351175); Permission granted by S. Karger AG, Basel.

# Vadadustat Phase 3 Global Development Program

- Global, ~7,000 patients, active-controlled, open-label, non-inferiority, cardiovascular outcome studies ongoing
- 17 Phase 1 and Phase 2 trials provide foundation for the Phase 3 program globally
- Collaborations with Otsuka and Mitsubishi Tanabe

## Non-Dialysis Dependent (NDD)

PRO<sub>2</sub>TECT

CORRECTION

Not ESA Treated  
Vadadustat vs  
Darbepoetin Alfa

PRO<sub>2</sub>TECT

CONVERSION

ESA Treated  
Vadadustat vs  
Darbepoetin Alfa

## Dialysis Dependent (DD)

INNO<sub>2</sub>VATE

CORRECTION  
CONVERSION

New-Onset Dialysis\*  
Vadadustat vs  
Darbepoetin Alfa

\* ≤16 weeks of dialysis treatment,  
with or without prior ESA treatment

INNO<sub>2</sub>VATE

CONVERSION

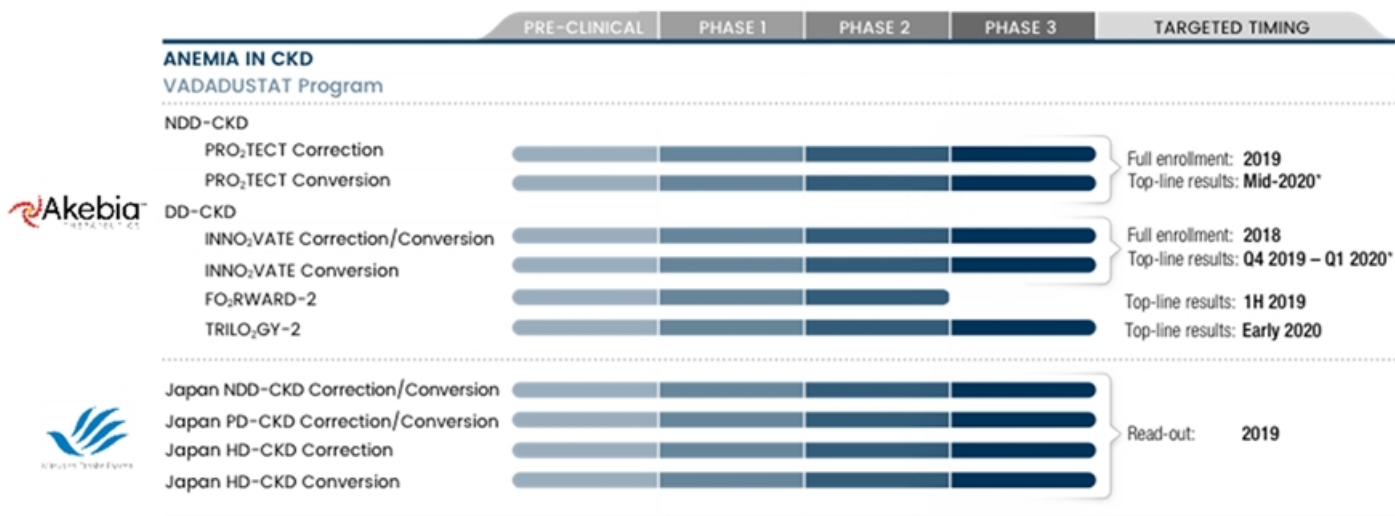
ESA Treated  
Vadadustat vs  
Darbepoetin Alfa

Primary Efficacy Endpoint: Change in hemoglobin (Hb) from baseline  
Primary Safety Endpoint: Major Adverse Cardiovascular Events (MACE)

Top-Line Results Expected Mid-2020,  
subject to MACE

Top-Line Results Expected Q4 2019 to Q1 2020,  
subject to MACE

# Upcoming Vadadustat Milestones Include Multiple Data Readouts in 2019, Including From Phase 3 Japan Studies

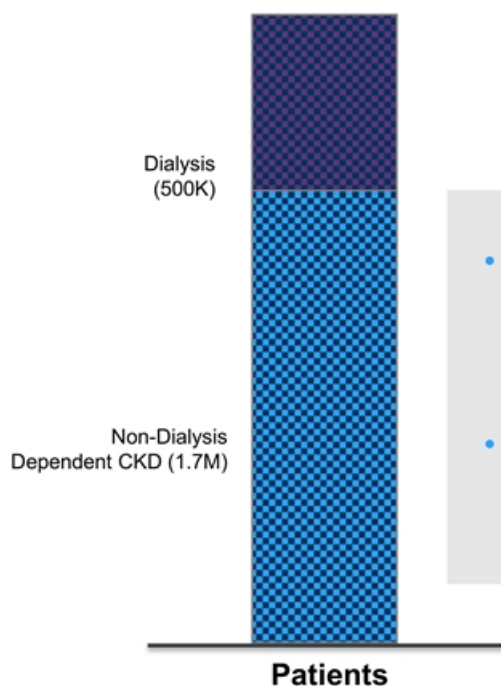


\*Subject to the accrual of MACE events

Note: NDD-CKD denotes non-dialysis-dependent chronic kidney disease and DD-CKD denotes dialysis-dependent chronic kidney disease.

# ~\$4B Market<sup>1</sup> Today with Major Growth Opportunity Driven by Potential to Establish New Standard of Care in NDD

## 2.2M Patients under Nephrologist Care<sup>2</sup>



- Only 16% of NDD patients are treated with ESAs<sup>3</sup>
- Less than 20% of NDD patients have an optimal response to OTC iron<sup>3</sup>

**2-3x<sup>4</sup> Growth Potential in Anemia in NDD**

- Auryxia, Hyperphosphatemia in DD
- Auryxia, Iron Deficient Anemia in NDD
- Vadadustat, Anemia Due to CKD in DD & NDD, if approved by the FDA

1. Company reported US ESA sales 2017 based on SEC filings for each ESA company, EvaluatePharma reported phosphate binder sales in the US in 2017, and estimated iron sales related to CKD in the US based on EvaluatePharma  
 2. US Census Bureau 2017; NHANES 2009-2014  
 3. Spherix Global Insights – Real World Dynamix - Feb 2018  
 4. Akebia primary market research, internal analysis of chart-based nephrologists surveys (Decision Resources ChartTrends Renal Anemia in ND-CKD, 2014, Decision Resources ChartTrends Nephrology in Dialysis, 2015), and management assumptions

# Potential to Deliver Innovative Therapies to Advance Care and Improve Outcomes for Kidney Disease Patients



## Approved and Target Indications

- ✓ Iron deficiency anemia (NDD) – Auryxia
- ✓ Hyperphosphatemia (DD) – Auryxia
- ✓ Anemia associated with CKD (DD&NDD)  
– **Vadadustat** *In development, subject to FDA approval*

The combined company will continue to identify, develop and commercialize new therapeutic options to address the needs of patients with kidney disease

# Fully Integrated, Kidney Disease Therapeutics Company Positioned to Deliver Substantial Value Long Term



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**Creates Potential for Accelerated Growth and Organizational Synergies**

**Combines Experienced Renal Management Teams**

**Strengthens Financial Profile**

**Potential Cost Savings of >\$250M to be Realized Five Years Following Closing**