
Akebia®
THERAPEUTICS
DEDICATED TO ADVANCING
CARE FOR PATIENTS WITH
KIDNEY DISEASE

CAUTIONARY NOTE ON FORWARD-LOOKING STATEMENTS

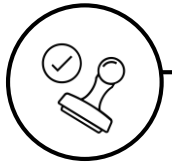
Statements in this presentation regarding Akebia's strategy, plans, prospects, expectations, beliefs, intentions or goals are forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995, as amended, including but not limited to statements regarding the expected period of time our cash resources and estimated product revenue will fund operations; the timing, availability and presentation of clinical trial data and results; the commercial potential, growth potential and market opportunity for our product and, if approved, our product candidates; our strategy, mission and vision; potential for our product candidates to set a new standard of care; the potential benefits of our product candidates; the timing of enrollment, including full enrollment, of our clinical trials; the target enrollments of our clinical trials; the assessments and evaluations we expect from our clinical programs; the potential to be a partner of choice for innovation in renal; and exploring co-development potential for vadadustat and Auryxia. The terms "estimate," "expect," "growth," "momentum," "mission" "opportunity," "positioned," "potential," "vision" and similar expressions are intended to identify forward-looking statements, although not all forward-looking statements contain these identifying words. Each forward-looking statement is subject to risks and uncertainties that could cause actual results to differ materially from those expressed or implied in such statement, including the actual product revenues for Auryxia; the timing of generic entrants for Auryxia, vadadustat or any other product candidates; the rate of enrollment in clinical studies of vadadustat; the risk that clinical trials may not be successful; the risk that existing preclinical and clinical data may not be predictive of the results of ongoing or later clinical trials; manufacturing risks; the quality and manner of the data that will result from clinical studies of vadadustat; the actual funding required to develop and commercialize Akebia's product candidates and operate the company, and the actual expenses associated therewith; efficacy, safety and tolerability of our products and product candidates; the risk that clinical studies need to be discontinued for any reason, including for safety, tolerability, enrollment, manufacturing or economic reasons; early termination of any of Akebia's collaborations or license agreements, and the parties' ability to satisfy their obligations under such agreements; the timing and content of decisions made by regulatory, judicial or similar authorities; the timing of any additional studies initiated for vadadustat; the actual time it takes to initiate and complete research and clinical studies; the success of competitors in developing product candidates for diseases for which Akebia is currently developing its product candidates; the scope, timing, and outcome of any ongoing legal proceedings; changes in the economic and financial conditions of the businesses of Akebia and its partners; and Akebia's ability to obtain, maintain and enforce patent and other intellectual property protection for Auryxia, vadadustat and its other product candidates. Other risks and uncertainties include those identified under the heading "Risk Factors" in Akebia's Quarterly Report on Form 10-Q for the quarter ended June 30, 2019, filed with the SEC, and other filings that Akebia may make with the SEC in the future. These forward-looking statements (except as otherwise noted) speak only as of the date of this presentation, and Akebia does not undertake, and specifically disclaims, any obligation to update any forward-looking statements contained in this presentation. Vadadustat is an investigational drug and has not yet been approved by the U.S. Food and Drug Administration (FDA) or any regulatory authority.

Fully Integrated Biotech Company Focused On Kidney Disease

John P. Butler, President and CEO

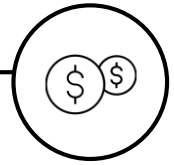
FDA-APPROVED

Product revenue in two FDA-approved indications with substantial growth potential



CASH RESOURCES

Expect cash resources[1] to fund operations into Q3 2020. $136.8 million cash position[2].



MULTIPLE CLINICAL CATALYSTS

Multiple clinical catalysts over next 12 to 18 months for Phase 3 product candidate with multi-billion-dollar global market opportunity

COMMERCIAL OPERATIONS

Experienced nephrology-focused commercial sales and marketing organization

1 Includes committed research and development funding from collaborators | 2 Cash, cash equivalents and available for sale securities as of 6/30/19.

Akebia THERAPEUTICS

Significant Strategic Partnerships

Otsuka

VIFOR PHARMA

Johnson & Johnson

Mitsubishi Tanabe Pharma

JT

PIC/S GMP 藥廠 PBF 寶齡富錦生技 Panion & BF Biotech Inc.

Track record of SOLID EXECUTION

JNDA FILING IN JAPAN FOR VADADUSTAT

First regulatory filing for marketing approval of vadadustat (July)

COMPLETED ENROLLMENT IN GLOBAL PHASE 3 PROGRAM FOR VADADUSTAT

INNO$_2$VATE enrolled 3,923 DD-CKD patients (April)
PRO$_2$TECT enrolled 3,513 NDD-CKD patients (Aug.)

DELIVERED ROBUST GROWTH

Product revenue increased 21% yr/yr and 26% sequentially qtr/qtr

SETTLED AURYXIA ANDA LITIGATION WITH PAR

Settlement reinforces the strength of Auryxia IP

OUR PURPOSE

To Better the Life of Each Person Impacted by Kidney Disease

Vadadustat

AN INVESTIGATIONAL ORAL HIF PH INHIBITOR DESIGNED TO STIMULATE ENDOGENOUS EPO PRODUCTION

with the potential to establish a new standard of care

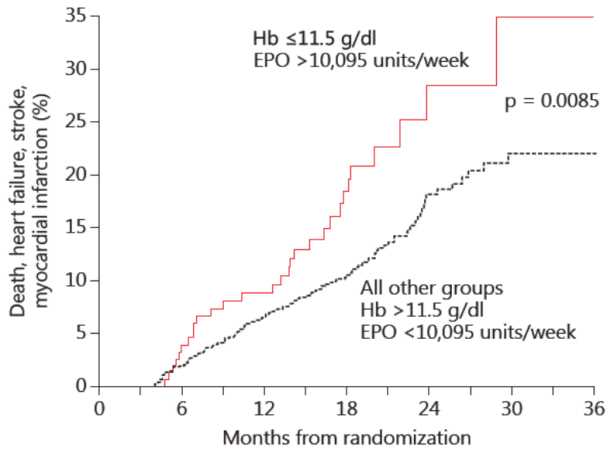
Vadadustat's ongoing Phase 3 cardiovascular outcomes trials for anemia due to CKD in non-dialysis and dialysis patients are now fully enrolled.
Vadadustat is an investigational HIF PH inhibitor that is not approved by the FDA or any other regulatory authority.

Vadadustat is an oral HIF PH inhibitor designed to stimulate endogenous EPO production, with the potential to increase hemoglobin while avoiding supra-physiological EPO levels

HIGH EPO LEVEL IS ASSOCIATED WITH INCREASED CV RISK

Kaplan-Meier Survival Curves[1]

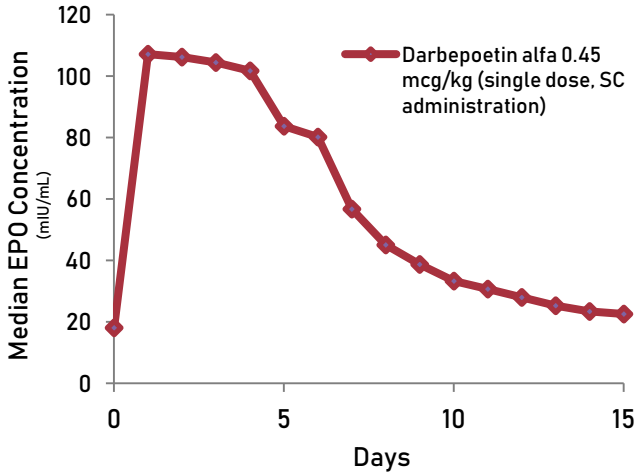
Death, Heart Failure, Stroke, Myocardial Infarction



SoC WITH INJECTABLE ESAs RESULT IN SUPRA-PHYSIOLOGICAL EPO LEVELS

EPO vs Time with Darbepoetin Alfa (SC)

PK-PD Model in CKD Subjects[2]

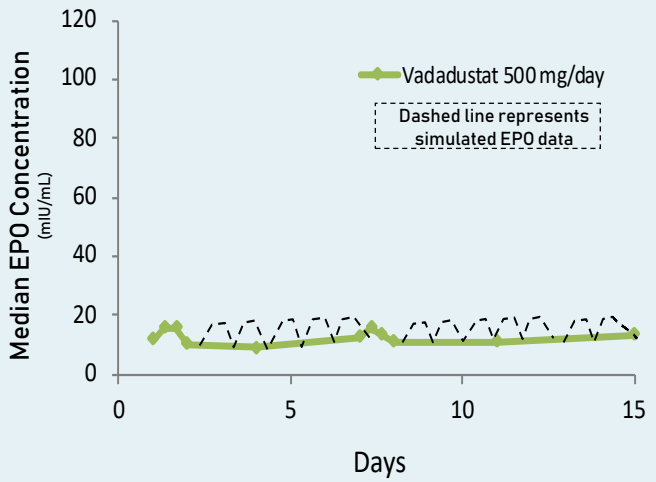


NOT A HEAD-TO-HEAD COMPARISON

VADADUSTAT, ORAL HIF PH INHIBITOR, AVOIDED SUPRA-PHYSIOLOGICAL EPO LEVELS

EPO vs Time with vadadustat (Oral)

Phase 1 Study in Healthy Volunteers[3]



ESA is erythropoietin stimulating agent. SOURCES: 1 McCullough P.A., et al. Am J Nephrol 2013;37:549-558 (DOI:10.1159/000351175); Permission granted by S. Karger AG, Basel.; 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 mU/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.; 3 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.

Non-Dialysis

Vadadustat for the treatment of anemia due to CKD in non-dialysis dependent adult patient population

Otsuka

Mitsubishi Tanabe Pharma

Dialysis

Vadadustat for the treatment of anemia due to CKD in dialysis dependent adult patient population

Otsuka

Mitsubishi Tanabe Pharma

VIFOR PHARMA

Vadadustat's ongoing Phase 3 cardiovascular outcomes trials for anemia due to CKD in non-dialysis and dialysis patients are now fully enrolled. Vadadustat is an investigational HIF PH inhibitor that is not approved by the FDA or any other regulatory authority.

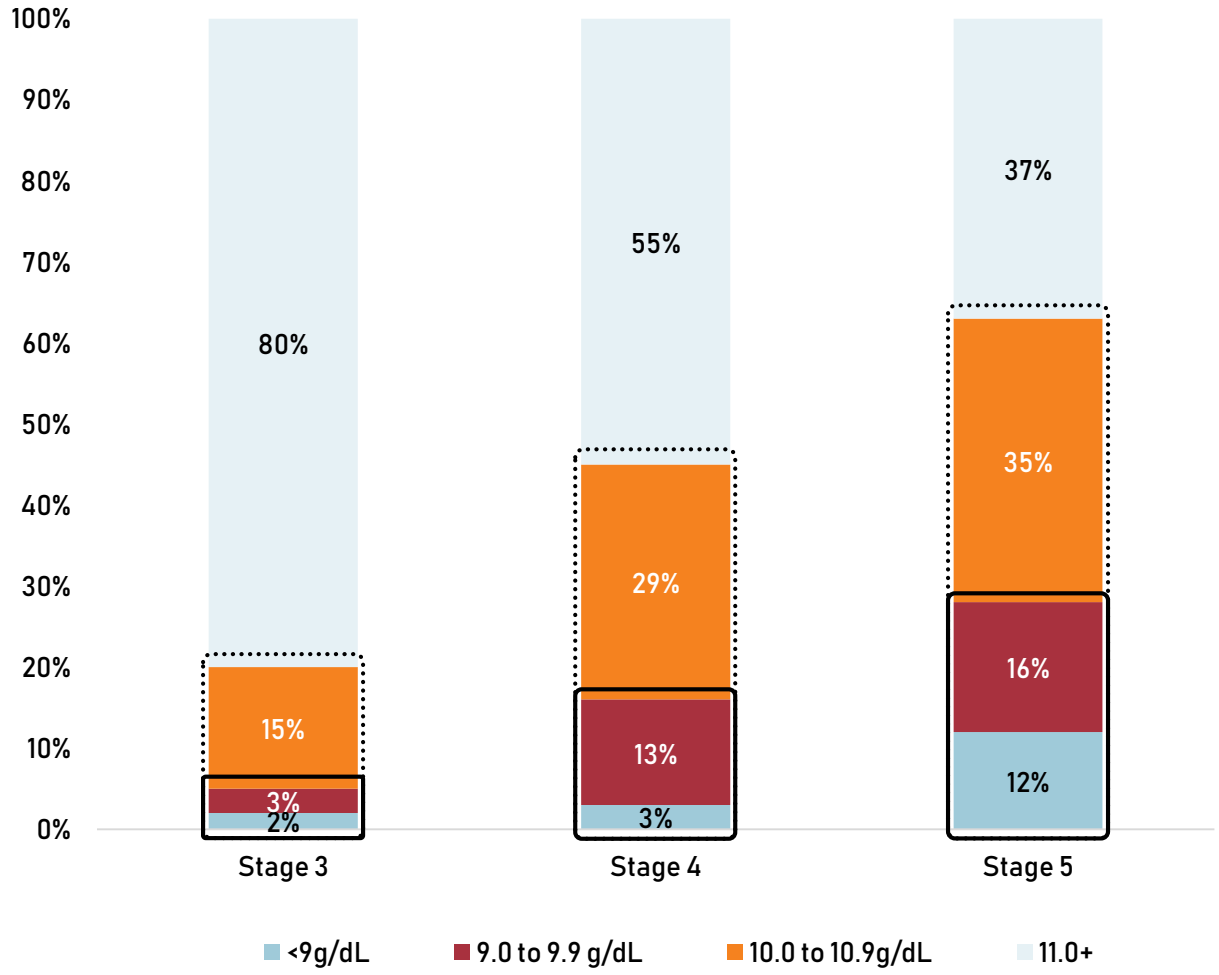
Non-Dialysis

Limitations of Current Standard of Care/ Treatment Option (ESAs)

- Risk Of CV Events
- Variability In Hemoglobin Levels
- Risk Of Hypertension
- Inconvenient Dosing

High Proportion Of Patients With Low Hb Are Not Treated

Hemoglobin Levels in patients NOT treated with ESA by CKD Stage[1]



SOURCE: 1 Spherix RealWorld Dynamix Renal Anemia market research survey and chart review, Feb 2018 (172 nephrologists, 1059 patient records).

Vadadustat

SIGNIFICANT OPPORTUNITY TO ADDRESS UNMET NEEDS OF NON-DIALYSIS PATIENT POPULATION WITH ANEMIA DUE TO CKD

(US MARKET)

37 Million

People with CKD[1]

5.7 Million

People with anemia due to CKD[2]

250K

Size of non-dialysis dependent patient population currently treated for anemia due to CKD[3]

400-500K

Estimated size of addressable non-dialysis dependent patient population with anemia due to CKD[4]

$2-3 Billion

Estimated **US** Market Opportunity within Non-Dialysis Patient Population[4]

Vadadustat's ongoing Phase 3 cardiovascular outcomes trials for anemia due to CKD in non-dialysis and dialysis patients are now fully enrolled. Vadadustat is an investigational HIF PH inhibitor that is not approved by the FDA or any other regulatory authority.

Source: 1. 2018 USRDS Annual Data Report: https://www.usrds.org/2018/download/v2_c01_IncPrev_18_web.xlsx. 2. Stauffer et al, PLOSONE, 2014. 3. Spherix RealWorld Dynamix Renal Anemia market research survey and chart review, Feb 2018. 4. Based on internal estimates for pts with Hb<11 and not treated with ESA and industry reports estimating ESA pricing.

Non-Dialysis

GLOBAL PHASE 3 PROGRAM FOR VADADUSTAT FOR NON-DIALYSIS

NDA/MAA Core Package

Cardiovascular Outcomes PRO$_2$TECT Phase 3 Studies
Anemia due to CKD in Non-Dialysis Dependent Patients

FULLY ENROLLED

PRO$_2$TECT CORRECTION

Not ESA Treated
Vadadustat vs
Darbepoetin Alfa

N ≈ 3,513

PRO$_2$TECT 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 accrual of MACE.

Vadadustat

SIGNIFICANT OPPORTUNITY TO ADDRESS UNMET NEEDS OF DIALYSIS DEPENDENT PATIENT POPULATION WITH ANEMIA DUE TO CKD

(US MARKET)

37 Million

People with CKD[1]

5.7 Million

People with anemia due to CKD[2]

510K

Dialysis patient population[1]

90%

Of dialysis patient population is treated for anemia due to CKD[3]

$2 Billion

Estimated **US** Market Opportunity within Dialysis Patient Population[4]

Vadadustat's ongoing Phase 3 cardiovascular outcomes trials for anemia due to CKD in non-dialysis and dialysis patients are now fully enrolled. Vadadustat is an investigational HIF PH inhibitor that is not approved by the FDA or any other regulatory authority.

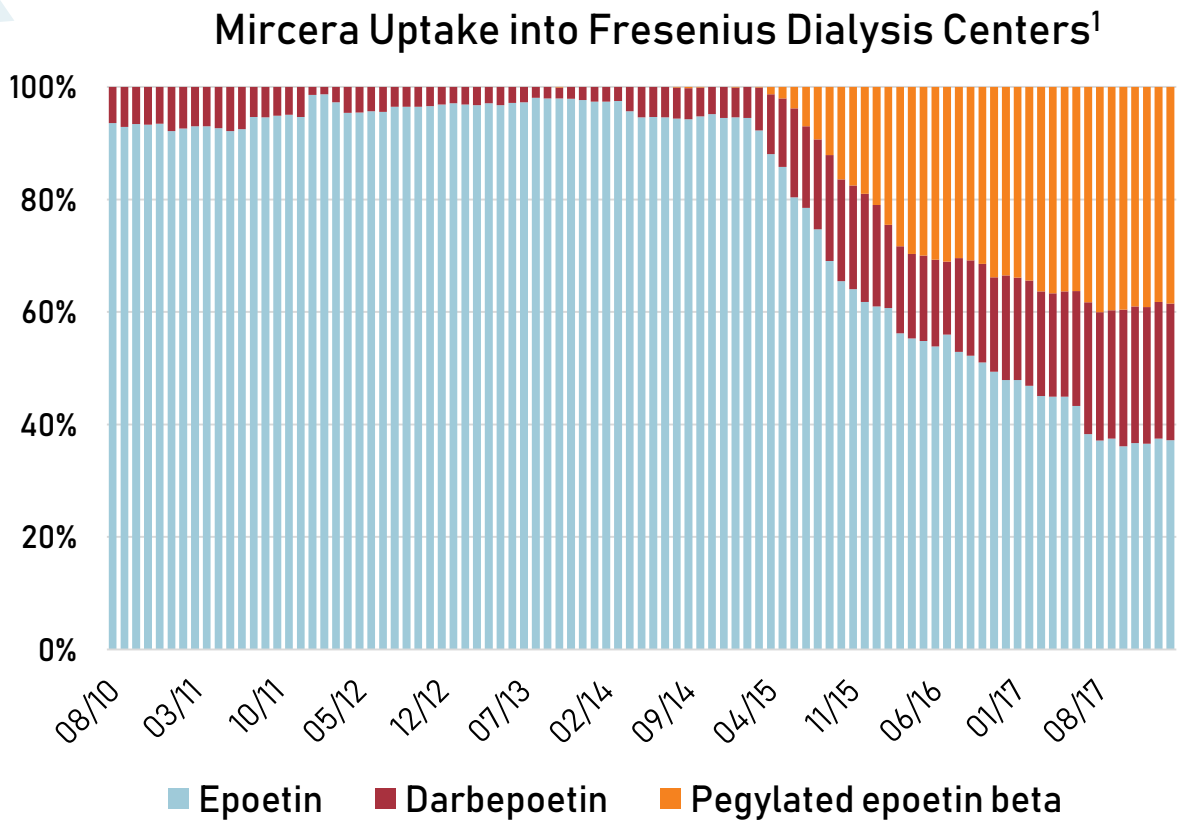
Source: 1. 2018 USRDS Annual Data Report: https://www.usrds.org/2018/download/v2_c01_IncPrev_18_web.xlsx. 2. Stauffer et al, PLOSONE, 2014. 3. Based on internal estimates and industry reports. 4. Based on internal estimates and industry reports estimating ESA pricing.

Dialysis

STRONG DISTRIBUTION CHANNEL TO U.S. DIALYSIS NETWORK

VIFOR PHARMA

- Unique market dynamics with treatment adoption driven by protocols
- Agreement with Vifor (International) Ltd. positions vadadustat for rapid uptake in Fresenius LDO subject to FDA approval*
- 2019 amendment expands agreement, facilitating access to up to 60% of U.S dialysis patients
- TDAPA[2] creates major additional opportunity for value creation for Akebia



*Subject to the earlier of reimbursement under TDAPA (defined below) or inclusion in the ESRD bundle. SOURCES: 1 ESA Use, by type. DOPPS Practice Monitor. https://www.dopps.org/dpm/DPMSlideBrowser.aspx. Accessed 12/19/2018.; 2 TDAPA: Transitional drug add-on payment adjustment, CMS Ruling CMS-1691-F. Medicare Program; End-Stage Renal Disease Prospective Payment System, Payment for Renal Dialysis Services Furnished to Individuals with Acute Kidney Injury, End-Stage Renal Disease Quality Incentive Program, Durable Medical Equipment, Prosthetics, Orthotics and Supplies (DMEPOS) Competitive Bidding Program (CBP) and Fee Schedule Amounts, and Technical Amendments to Correct Existing Regulations Related to the CBP for Certain DMEPOS.

Dialysis

GLOBAL PHASE 3 PROGRAM FOR VADADUSTAT FOR DIALYSIS

NDA/MAA Core Package

Cardiovascular Outcomes INNO$_2$VATE Phase 3 Studies
Anemia due to CKD in Dialysis Patients

FULLY ENROLLED

INNO$_2$VATE CORRECTION CONVERSION

New-Onset Dialysis
Vadadustat vs
Darbepoetin Alfa

N ≈ 3,923

INNO$_2$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 Q2 2020*

*Subject to accrual of MACE.

GLOBAL PHASE 3 PROGRAM FOR VADADUSTAT

Additional Non-Primary Endpoints

- INDIVIDUAL MACE COMPONENTS
- THROMBOEMBOLIC EVENTS
- HOSPITALIZATION FOR HEART FAILURE
- HB EXCURSIONS
- TIME IN RANGE (HB)
- IV IRON USE
- CKD PROGRESSION

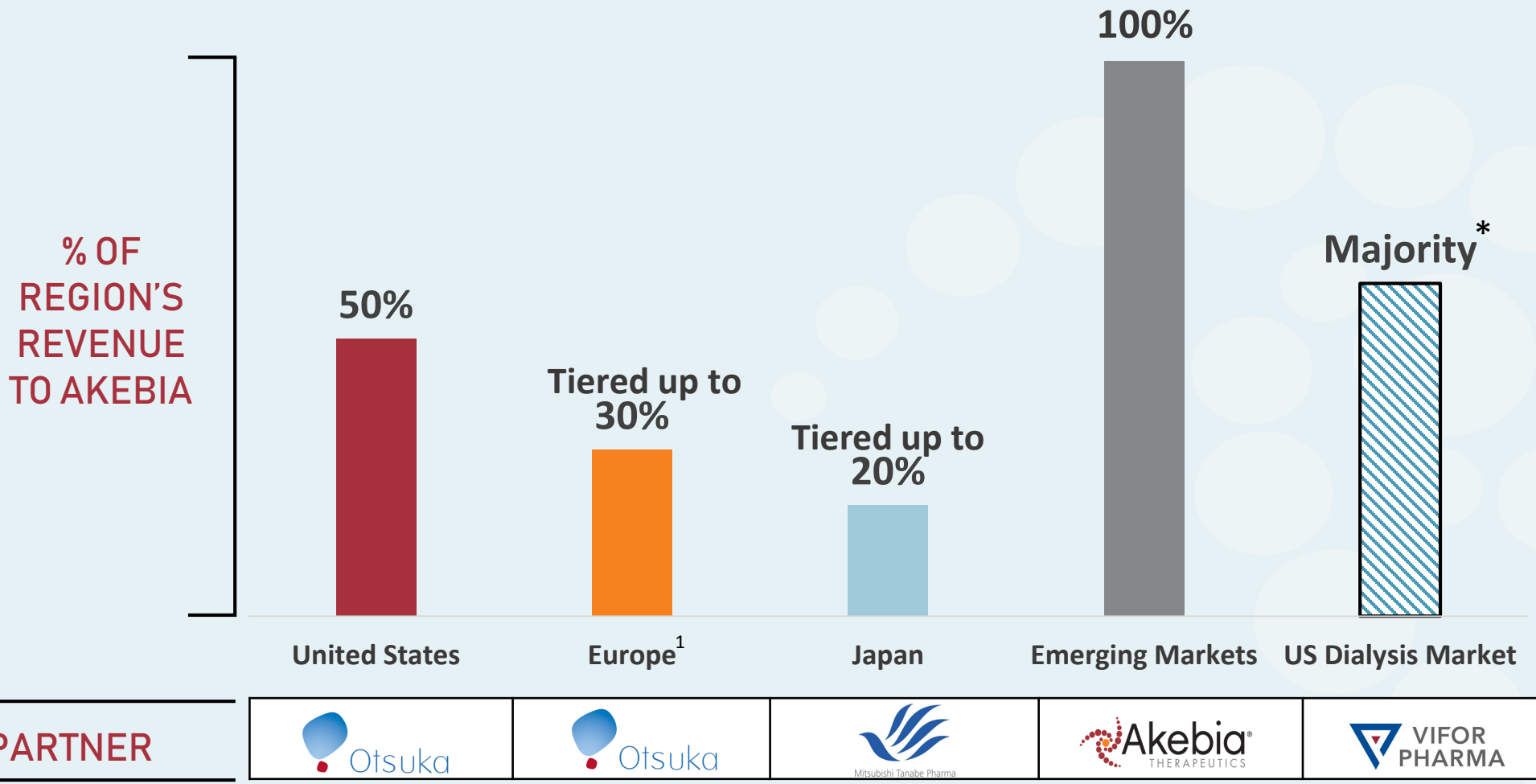
Expected Data from Additional Ongoing/Planned Studies to Support Value Proposition

Comparative data of vadadustat versus ESA in dialysis-dependent CKD subjects who are ESA hyporesponders

Modified QD dosing of vadadustat compared to ESA for additional dosing flexibility

TIW dosing of vadadustat compared to ESA for additional dosing flexibility

Global Partnerships for Vadadustat



Vadadustat's ongoing Phase 3 cardiovascular outcomes trials for anemia due to CKD in non-dialysis and dialysis patients are now fully enrolled. Vadadustat is an investigational HIF PH inhibitor that is not approved by the FDA or any other regulatory authority.

1. Includes Russia, China, Canada, Australia and Middle East

*Pursuant to the Vifor Amended Agreement, Akebia granted Vifor Pharma an exclusive license to sell vadadustat to Fresenius Kidney Care and to certain other third party dialysis organizations in the U.S., upon approval of vadadustat by the FDA and other conditions discussed in Akebia's SEC filings. Akebia will share revenue from the profit share with Vifor Pharma with Otsuka pursuant to the Otsuka U.S. Agreement. Akebia currently retains rights to commercialize vadadustat for use in other dialysis organizations in the U.S., which will be done in collaboration with Otsuka following FDA approval

Well Positioned for Success at Vadadustat Launch

EXPERIENCED AND ESTABLISHED NEPHROLOGY FOCUSED SALES TEAM

GROWING RENAL PRODUCT PORTFOLIO WITH AURYXIA

FOCUSED PRESCRIBER BASE

KEY STRATEGIC PARTNERS WITH ROBUST COMMERCIAL FOOTPRINTS

STRATEGIC PARTNERSHIP WITH VIFOR PHARMA FOR DIALYSIS MARKET

>140

Akebia Commercial Team Members

95

Akebia Sales Representatives

10 yrs.

Average Years of Sales Experience including Nephrology/Renal

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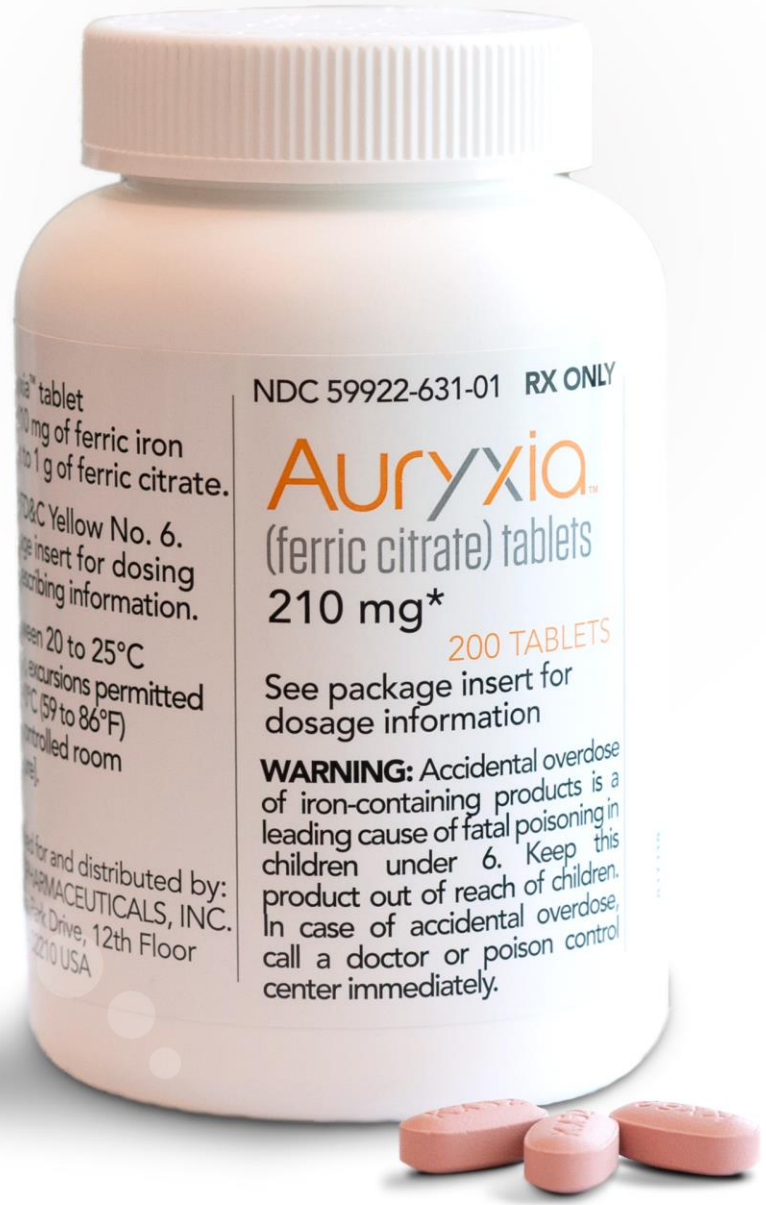


Auryxia

Akebia THERAPEUTICS

TWO APPROVED INDICATIONS

HYPERPHOSPHATEMIA

Dialysis

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

IRON DEFICIENCY ANEMIA

Non-Dialysis

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

COMMERCIAL PROGRESS

Total Pro-forma Unaudited Full-Year Auryxia Revenue Increased 72% To

$96M

IN 2018 COMPARED TO 2017

Total Prescriptions Increased

85%

IN 2018 COMPARED TO 2017

6.6% Market Share In 2018

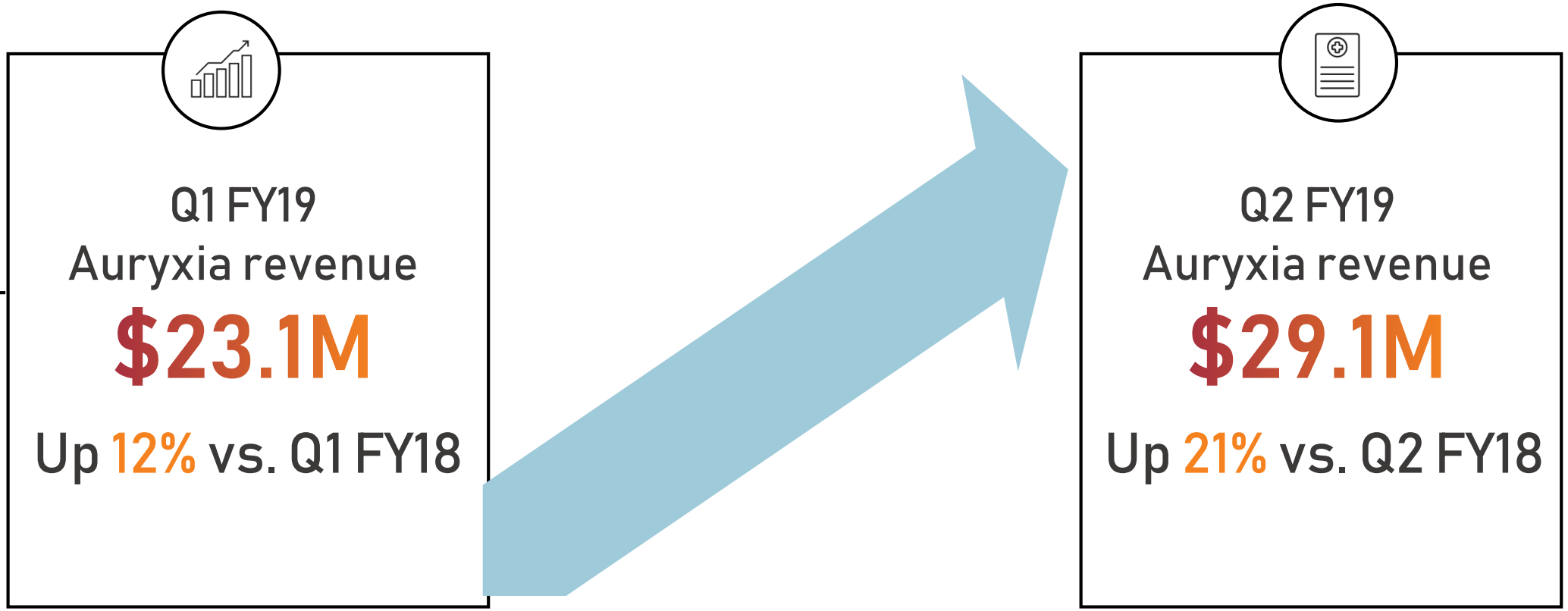
▲

4.2% Market Share In 2017

HIGHEST SHARE GAIN OF ALL PHOSPHATE BINDERS

Source: IQVIA NPA, Dialysis SP TRx Data *2018 Annual Auryxia revenue is pro-forma unaudited combined full-year Auryxia sales including sales recorded by Keryx prior to the merger on December 12, 2018.

QUARTERLY PERFORMANCE



Well Positioned to Drive Growth in 2019

AURYXIA LONG-TERM GROWTH POTENTIAL

AN INVESTIGATOR-SPONSORED STUDY OF FERRIC CITRATE PUBLISHED IN THE JOURNAL OF THE AMERICAN SOCIETY OF NEPHROLOGY (JASN), AUGUST 2019

- Single-center, open-label, randomized trial comparing ferric citrate (FC) and standard of care in subjects with advanced non-dialysis CKD (Stage 4/5)
- Assessed hypothesis that "provision of fixed dose FC to subjects with advanced CKD, independent of serum phosphate or degree of anemia, would improve multiple biochemical aspects simultaneously and reduce the need for exogenous ESA or intravenous (IV) iron"[1]
- Baseline characteristics were comparable with the exception of diabetes (FC 47%, SOC 77%, p=0.001)[1]

A Pilot Randomized Trial of Ferric Citrate Coordination Complex for the Treatment of Advanced Chronic Kidney Disease

Geoffrey A Block, Martha S. Block, Gerard Smits, Rupal Mehta, Tamara Isakova, Myles Wolf and Glenn Chertow

"The data from this study suggest that administering ferric citrate to late-stage pre-dialysis patients not only improves biochemical parameters associated with chronic kidney disease, but also has the potential to delay the need for dialysis," said Geoffrey Block, M.D,, Director of Clinical Research at Denver Nephrology.

"With the impact of ferric citrate across multiple aspects of CKD, it is worth further investigation to determine which of these many factors is contributing to the reduced risk of renal replacement therapy observed in this study."

*Keryx Biopharmaceuticals, Inc. (now a wholly owned subsidiary of Akebia) provided funding and study medication for this investigator initiated study.; SOURCE 1 Geoffrey Block, Martha Block, Gerard Smits, Laura Kooienga, Rupal Mehta, Tamara Isakova, Myles Wolf, Glenn Chertow. "A pilot randomized trial of the effects of ferric citrate in patients with advanced chronic kidney disease." JASN August 2019, 30 (8) 1495-1504; DOI: https://doi.org/10.1681/ASN.2018101016


Akebia®
THERAPEUTICS
THANK YOU | QUESTIONS?