

Delivering Life-changing Value as a Global Specialty Pharma

J.P. Morgan Healthcare Conference
January 10, 2022

Masashi Miyamoto, Ph.D.

Representative Director of the Board,
President and Chief Executive Officer



This document contains certain forward-looking statements relating to such items as the Company's forecasts, targets and plans (including those of its domestic and overseas subsidiaries). These forward-looking statements are based upon information available to the Company at the present time and upon reasonable assumptions made by the Company in making its forecasts, but actual results may differ substantially due to uncertain factors.

These uncertain factors include, but are not limited to, inherent risks in the business activities of the pharmaceutical industry in Japan and overseas, intellectual property risks, risk of product side effects, legal regulation risks, product defect risks, risks of changes to prices for raw materials, risks of changes to market prices, as well as risks of changes to foreign exchange rates and financial markets.

This document is provided to investors for informational purposes only. It contains information on pharmaceutical products (including products under development), but the contents should not be construed as promotion, advertising or as a medical recommendation.

The registered trademarks contained herein are the property of Kyowa Kirin Co., Ltd. or respective owners.

Agenda

-
- 4** Who We Are
 - 12** Global Strategic Products Driving Our Growth
 - 18** Next-generation Strategic Products
 - 30** Outlook for the Future
-

Who We Are

Vision

Our New Vision toward 2030

Kyowa Kirin will realize the successful creation and delivery of **life-changing value** that ultimately makes people smile, as a Japan-based Global Specialty Pharmaceutical company built on the diverse team of experts with shared passion for innovation.

Provide
pharmaceuticals for
unmet medical needs

Address
patient-centric
healthcare needs

Retain the trust
of society

Overview

Kyowa Kirin



5,423

Valuable employees worldwide



¥318.4 billion

Annual consolidated revenue in 2020

Global



39 countries/
regions

Extensive market reach, with a presence worldwide



48 %

Overseas revenue ratio in 2020



152 %

Sales growth for the three global strategic products (vs. 2019)

Specialty Pharma



over 50

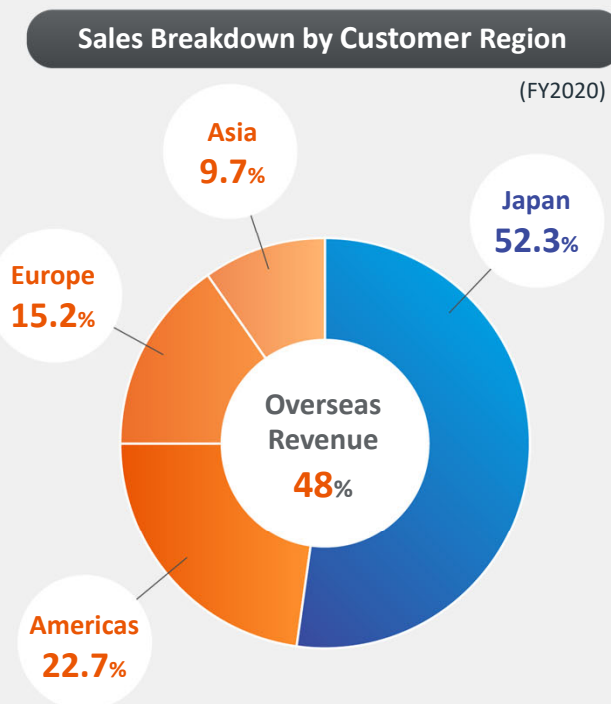
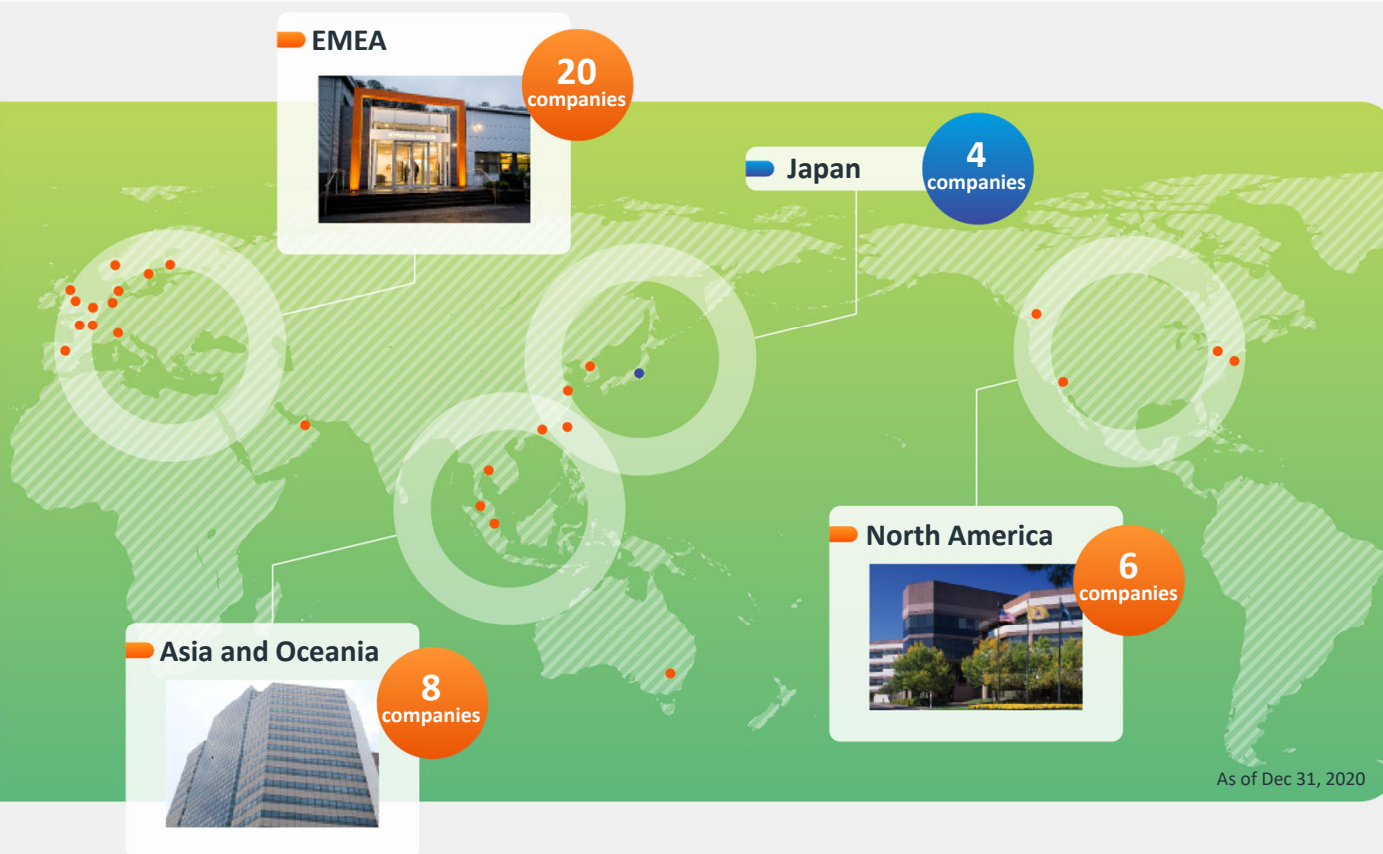
Pharmaceutical products on the market including 4 monoclonal antibodies



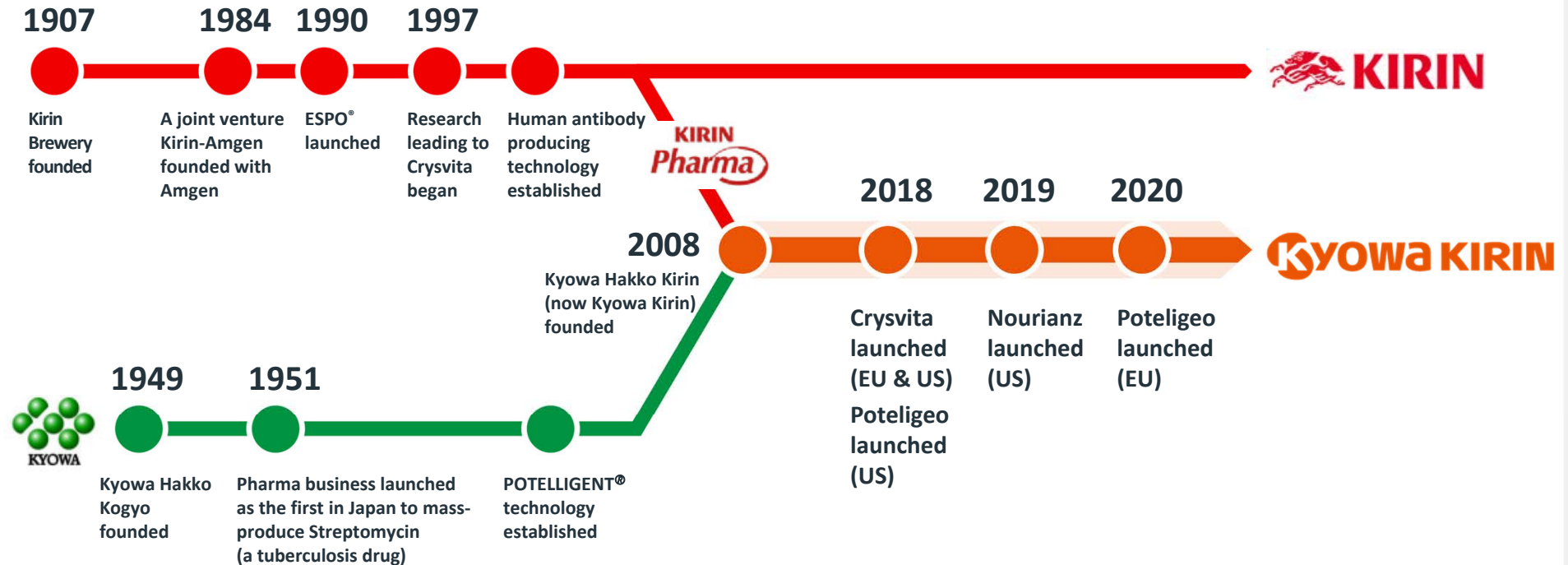
Strengths in

4 Therapeutic fields

Global Network



History



Key Marketed Products



Nephrology



Oncology



Immunology/Allergy



Central Nervous System



Others

NESP®

Erythropoiesis Stimulating Agent
Japan and Asia

REGPARA®

Calcium receptor agonist
Japan and Asia

オルケディア®

Calcium receptor agonist
Japan

オングリザ®

DPP-4 inhibitor
Japan

POTELIGEO®
(mogamulizumab)

Anti-CCR4 mAb
Japan, US and EU

Abstral®
SUBLINGUAL FENTANYL CITRATE TABLET

Sublingual fentanyl
Japan and EU

ジーラスト® GRAN™

Long-lasting G-CSF/G-CSF
Japan and Asia*1

リツキシマブ®BS

Rituximab biosimilar
Japan

Allelock®

Anti-allergic agent
Japan and Asia

パタノール®

Ophthalmic Anti-allergic agent
Japan

ルミセフ®

Anti-IL17R mAb
Japan

ドボベット®

Vitamin D3/Corticosteroid
Japan



NOURIANZ®
(istradefylline) tablets

Adenosine A_{2A}R antagonist
Japan and US

デパケン®

Antiepileptic agent
Japan

トピナ®

Antiepileptic agent
Japan

CRYSVITA®
curosumab

Anti-FGF23 mAb
Japan, US*2 and EU

Romiplate®

Romiplostim
Thrombopoietin R agonist
Japan and Asia

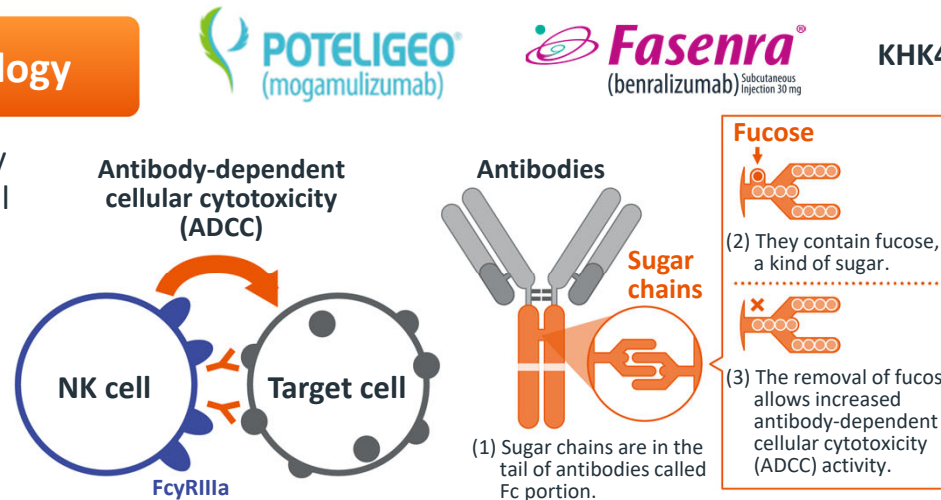
Coniel®

Calcium channel blocker
Japan and Asia

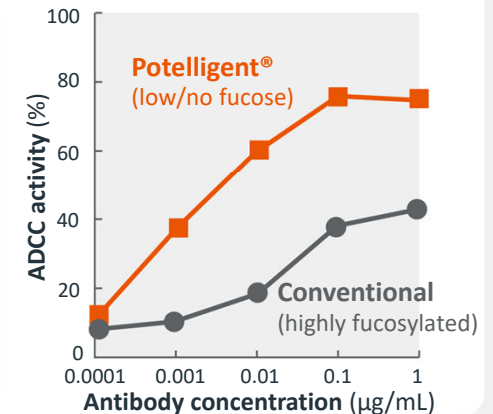
Proprietary Technologies for Life-changing Value Creation

POTELLIGENT® Technology

An ADCC-enhancing technology that realizes effective target cell elimination by antibody afucosylation



KHK4083/AMG 451



Human Antibody Producing Technology

A technology that enables to generate fully human antibodies with the same diversity as natural antibodies using chromosome engineering

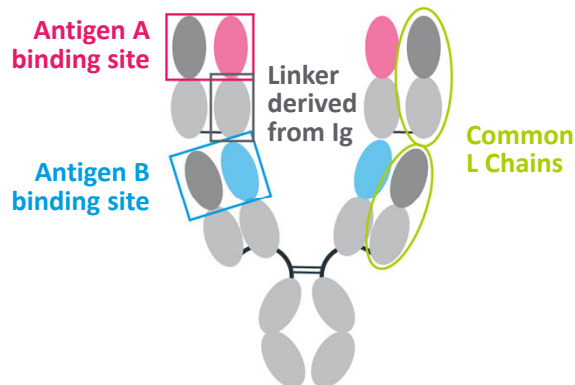


KHK4083/AMG 451

Next-Generation Technologies for Life-Changing Value Creation

Bispecific Antibody Technology

- Selection of linkers derived from Immunoglobulin (Ig) and the common sequence of L Chains
- Versatility equivalent to wild type IgG
- Unique biology based on bivalent x bivalent binding



Drug Discovery Collaborations

- Revolutionary small molecule drug discovery (with Axcelead)
- Data-driven drug discovery (with InveniAI)
- RNA structure-targeted drug discovery (with xFOREST and Axcelead)



Global Strategic Products Driving Our Growth

Strong Growth of Global 3 Brands (G3B)



Crysvita

(Burosumab, an anti-FGF23 fully human monoclonal antibody)

X-linked hypophosphatemia (XLH)
Tumor-induced osteomalacia (TIO)



Poteligeo

(Mogamulizumab, an anti-CCR4 humanized antibody)

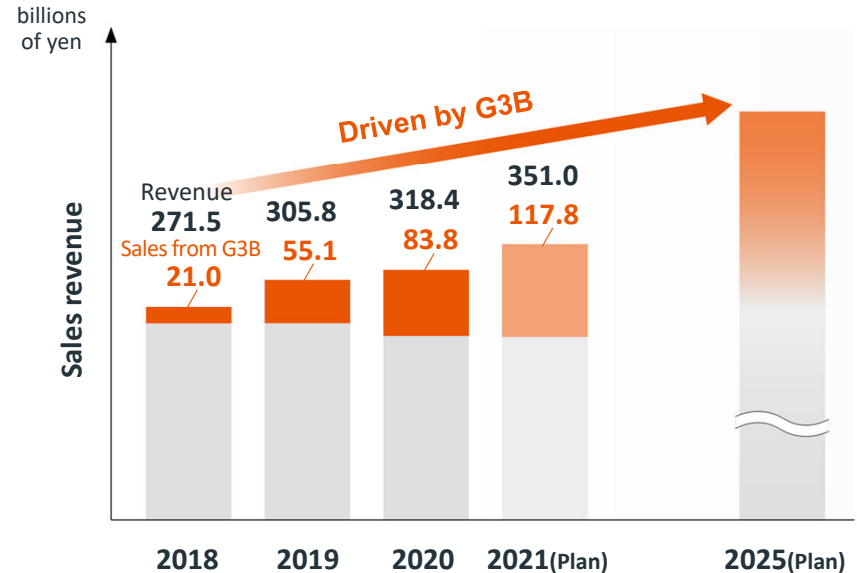
Mycosis fungoides (MF)
Sézary syndrome (SS)



Nourist/Nourianz

(Istradefylline, an Adenosine A_{2A} receptor antagonist)

Parkinson's disease (PD)
experiencing "off" episodes



Crysvita



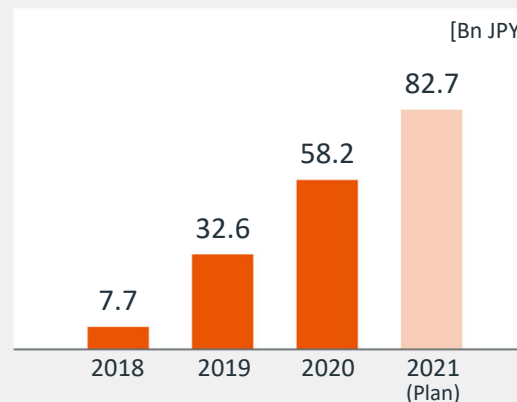
Burosumab, an anti-FGF23 fully human monoclonal antibody



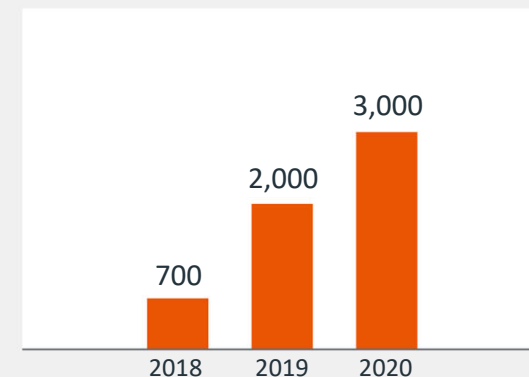
• Indications

US	EU	JP
XLH TIO	XLH (TIO)	FGF23-related hypophosphatemic rickets and osteomalacia

Annual sales growth^{*2} (NA+EMEA+JP)



Number of patients^{*2} (global total)

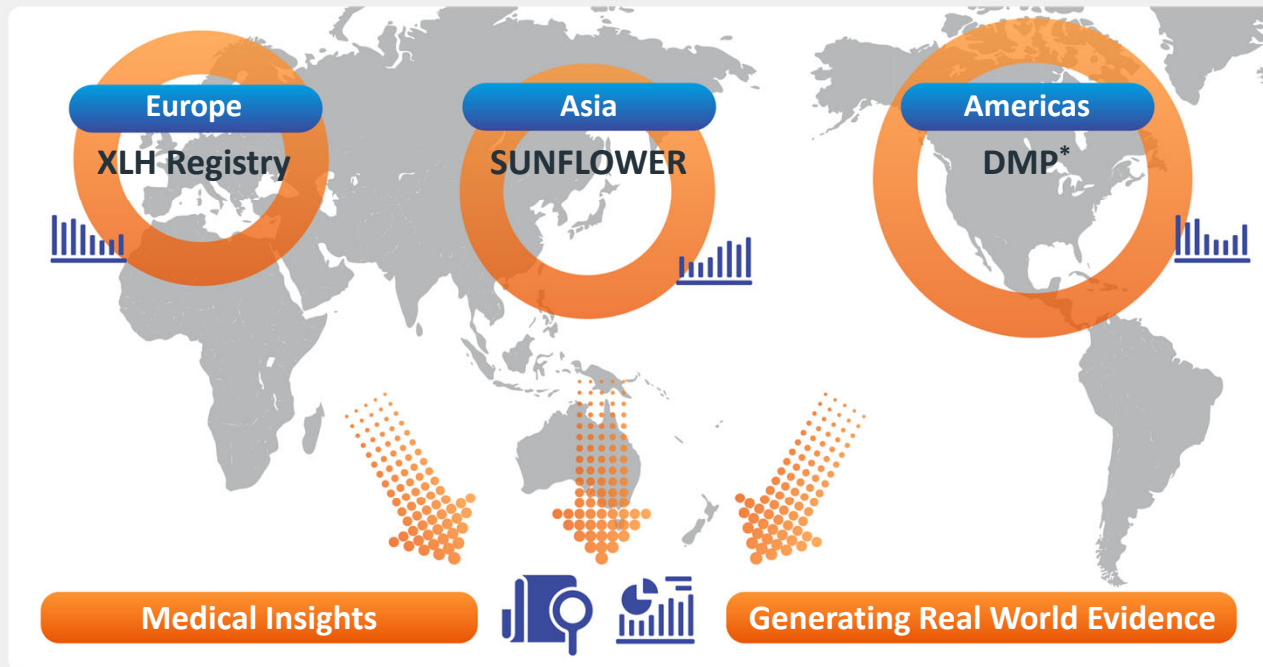


Crysvita

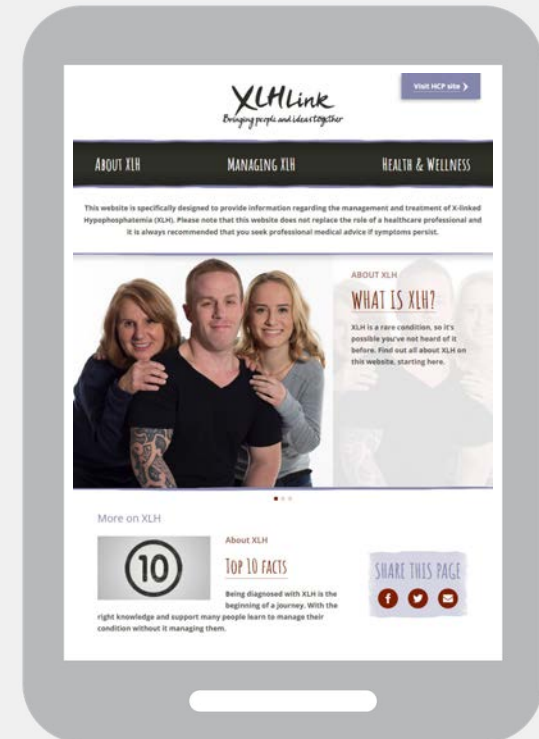


Burosumab, an anti-FGF23 fully human monoclonal antibody

Global Evidence Generation Project



* Disease Monitoring Program

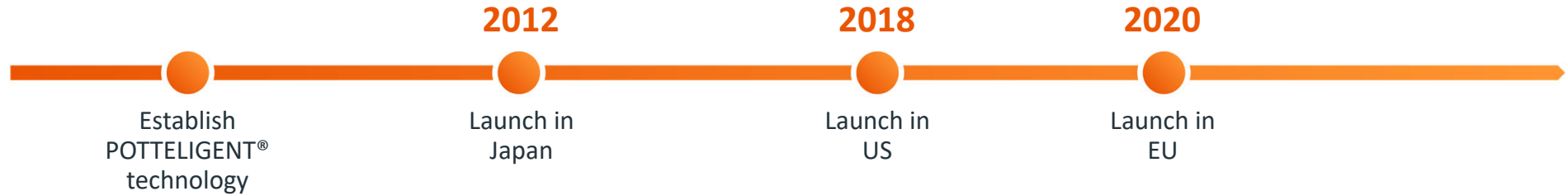


<https://xlhlink.eu/>

Poteligeo



Mogamulizumab, an anti-CCR4 humanized antibody

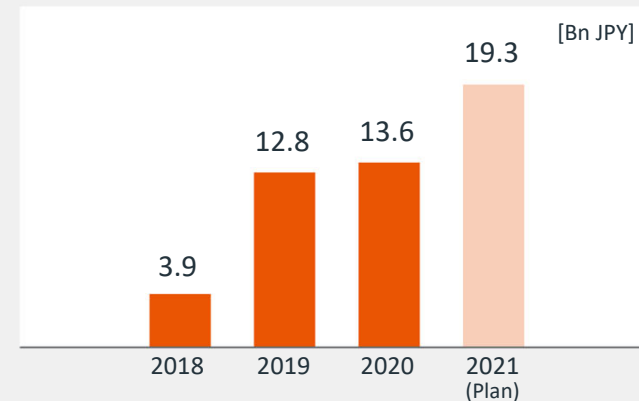


- The first glyco-engineered antibody to reach the market
- Indications

US	EU	JP
MF SS	MF SS	ATL PTCL CTCL

ATL: adult T-cell leukemia/lymphoma; PTCL: peripheral T-cell lymphoma; CTCL: cutaneous T-cell lymphoma

Annual sales growth* (NA+EMEA+JP)



* Excl. patients under Early Access Program and patients who have not started the reimbursement process

Nourianz/Nourias



Istradefylline, an Adenosine A_{2A} receptor antagonist



• MOA (Simple illustration)

Levodopa replaces lost dopamine, like pressing down on the gas



Istradefylline blocks A_{2A} receptors, like lifting the brake

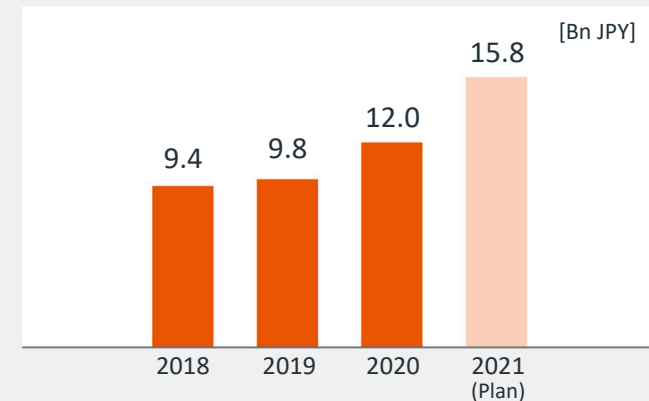


• Indications

US / JP

PD
experiencing
"off"
episodes

Annual sales growth (NA+JP)



Next-generation Strategic Products

Next-generation Strategic Products

	Country / region ^{*1}	Indication ^{*2}	Approval year ^{*3}	Total addressable market ^{*4}	No. of patients ^{*5}
KHK4083/ AMG 451	NA/EU/JP	Atopic dermatitis	2025/2026	★★★	16,000K
KW-6356	NA/EU/JP	Parkinson's disease	2025	★★★	3,500K
ME-401 Zandelisib	NA/EU/JP	Follicular lymphoma Marginal zone lymphoma	2023	★★★	~800K
RTA 402 Bardoxolone methyl	JP/Asia	Alport syndrome Diabetic kidney disease (DKD) Autosomal dominant polycystic kidney disease (ADPKD)	2022 2023 2025	★★★	2,500K ~
KHK7791 Tenapanor	JP	Hyperphosphatemia under maintenance dialysis	2023	★★☆	250K

As of Feb 4, 2021

^{*1} Countries or regions where Kyowa Kirin currently has marketing rights and will launch products (or will conduct marketing activities); products may not be launched in all countries or regions shown in the table

^{*2} Expected indications as of the date of this document; indications may ultimately differ to expectations due status of approvals from regulatory authorities

^{*3} Expected year of first approval

^{*4} Expected total addressable market, which is the sum of all products for the indications shown in ^{*2}, in all countries or regions defined in ^{*1}, not projected sales or the Company's targets;

★ = less than ¥50bn, ★★ = ¥50-100bn, ★★★ = Over ¥100bn

^{*5} Total number of estimated patients in all countries or regions defined in ^{*1}.

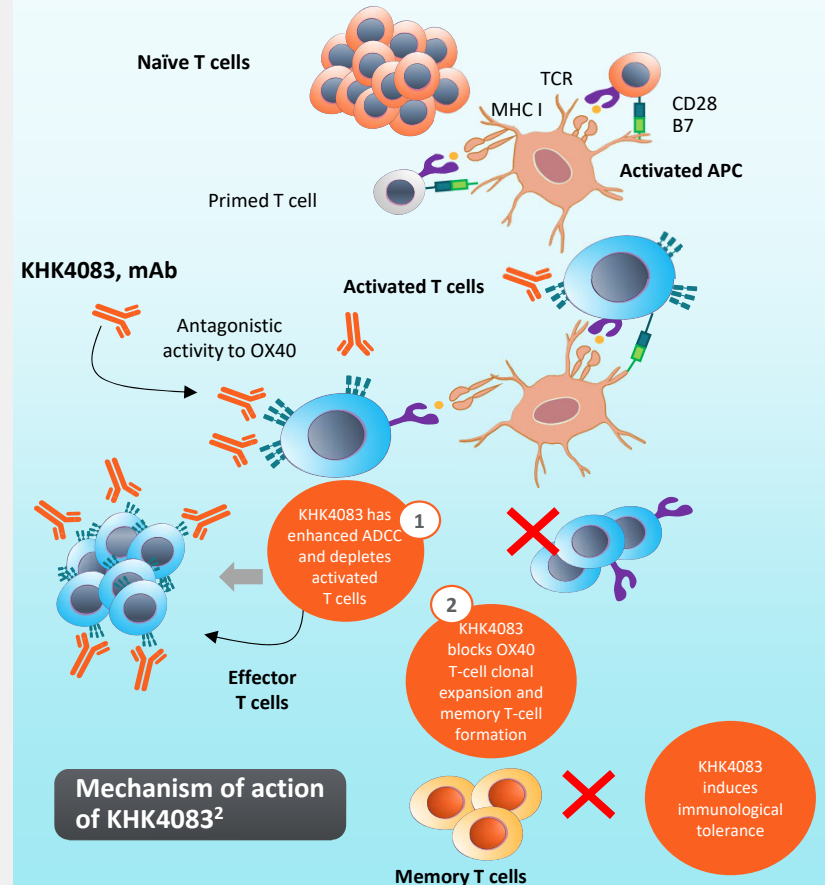
^{*6} The size of the total addressable market and patient numbers are based on our estimates

KHK4083/AMG 451 targets OX40 as a potential novel target for AD treatment

- Activation of Th2 and other T-cell subsets is central in atopic dermatitis (AD)
- The OX40–OX40L axis plays a critical role in long-lasting T-cell responses
 - OX40 is expressed by activated T cells after antigen recognition and binds OX40L on APCs, facilitating the effector function of T cells
- KHK4083/AMG 451 is a fully human, anti-OX40, non-fucosylated IgG1 mAb with enhanced ADCC¹ that acts by
 - Partially depleting activated T cells² ①
 - Blocking T-cell clonal expansion and memory T-cell formation² ②

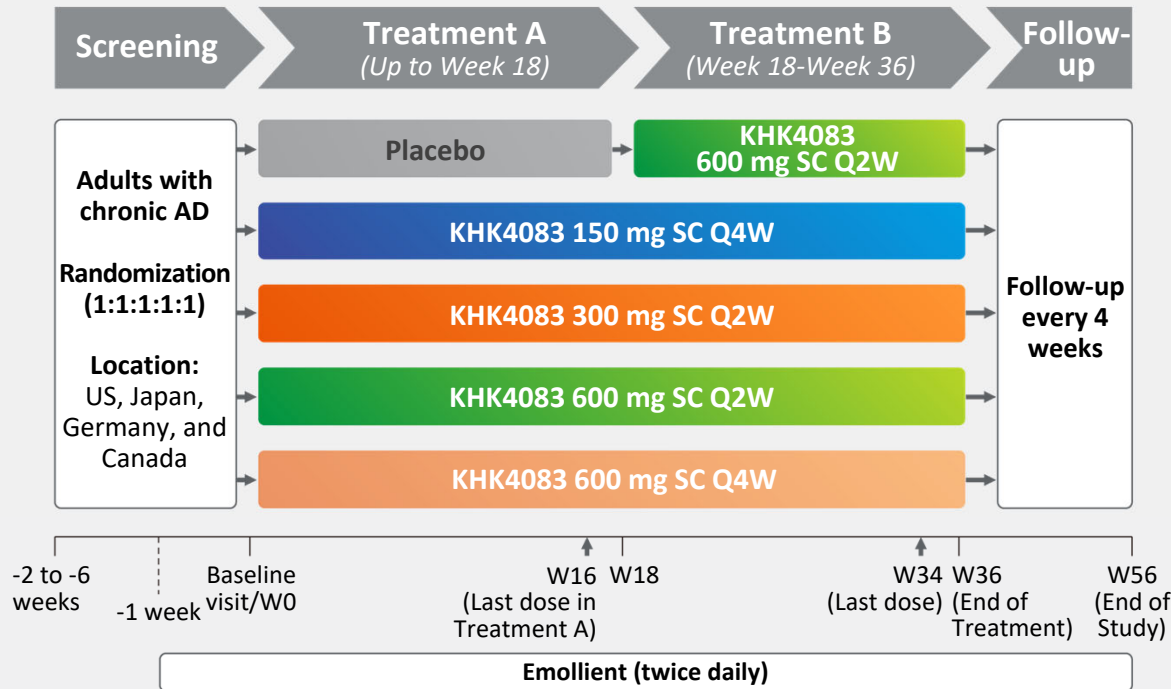
AD, atopic dermatitis; ADCC, antibody-dependent cellular cytotoxicity; APC, antigen-presenting cell; CD28, cluster of differentiation 28; IgG, immunoglobulin G; MHC, major histocompatibility complex; mAb, monoclonal antibody; TCR, T-cell receptor; Th2, T-helper 2

¹Nakagawa H et al. J Dermatol Sci. 2020; 99(2):82–89; ²Papp KA et al. J Eur Acad Dermatol Venereol. 2017; 31(8):1324–1332.

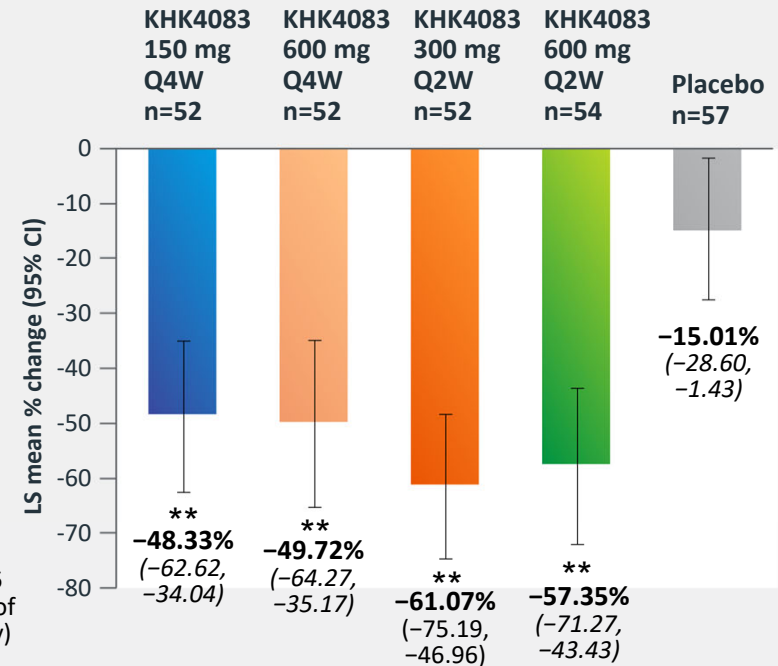


KHK4083/AMG 451: Achieved Primary Endpoint in Phase 2 Study

Phase 2 Study Design



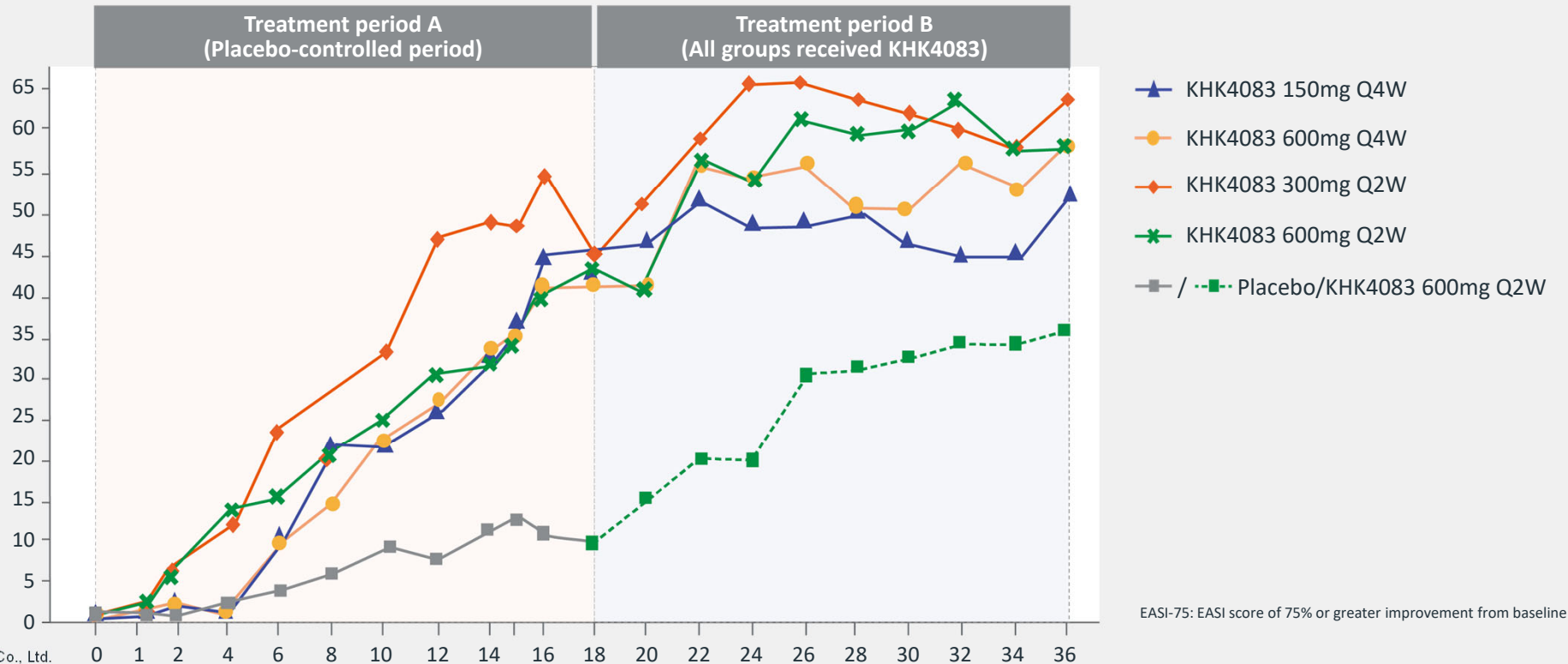
Primary Endpoint: % change from baseline in EASI score at W16 (LOCF, FAS)



EASI, Eczema Area and Severity Index; LS, least square; Q2W, every 2 weeks; Q4W, every 4 weeks
 **p<0.001 for difference versus placebo

KHK4083/AMG 451: Durable EASI-75 Response Confirmed

Secondary Endpoint: EASI-75 Responder Proportion



KHK4083/AMG 451: Collaboration with Amgen

	US	Europe and Asia (ex. JP)	JP
Development	<ul style="list-style-type: none"> Amgen leads development Share development cost 	<ul style="list-style-type: none"> Amgen leads development Share development cost 	<ul style="list-style-type: none"> Kyowa Kirin leads development
Commercialization	<ul style="list-style-type: none"> Amgen commercializes and books sales Kyowa Kirin co-promotes and shares promotion cost 	<ul style="list-style-type: none"> Amgen commercializes and books sales Kyowa Kirin has opt-in rights for co-promotion 	<ul style="list-style-type: none"> Kyowa Kirin commercializes and books sales
Sales royalties	<ul style="list-style-type: none"> Double-digit royalty to Kyowa Kirin 	<ul style="list-style-type: none"> Double-digit royalty to Kyowa Kirin 	
Commercial supply	<ul style="list-style-type: none"> Amgen supplies 	<ul style="list-style-type: none"> Amgen supplies 	<ul style="list-style-type: none"> Kyowa Kirin supplies

Amgen makes a \$400 million up-front payment to Kyowa Kirin and future contingent milestone payments potentially worth up to an additional \$850 million, as well as royalty payments on future global sales.

Zandelisib (ME-401): Clinical and Commercial Opportunity

>8,000 U.S. Patients
With Relapsed/Refractory
Follicular Lymphoma

\$1B
Addressable
Market

Market Opportunity

- PI3K δ inhibitors deliver potent efficacy, but utility limited by the extensive T-reg mediated toxicity
- R/R FL has several treatment options but no standard of care
- PI3K δ inhibitors limited to modest $\geq 3^{\text{rd}}$ Line FL use due to risk/benefit of current therapies

Zandelisib Opportunity

- Product attributes and novel treatment schedule could reset expectations of PI3K δ inhibitors
- Compelling emerging profile supports best-in-class opportunity in 3L+ FL
- Unique zandelisib properties and combinability could expand utility to earlier lines of FL and into other BCMs

MEI Pharma

KYOWA KIRIN

Global License, Development and Commercialization Agreement to Optimize Zandelisib Value (April 2020)

US: cost-sharing, co-promotion, MEI Pharma books sales

Ex-US: Kyowa Kirin has exclusive rights, escalating tiered sales royalty payments to MEI starting in teens

BCMs: B-cell malignancies

Zandelisib: Emerging Profile

Phase 2 study (TIDAL) in patients with r/r FL

Overall Response Rate (ORR)

95% CI (59.8, 79.5)

70.3%

Complete Response Rate (CR)

95% CI (25.4, 45.9)

35.2%

Duration of Response:

Insufficiently mature to estimate final DOR: with median follow-up time for response of 8.4 months, median DOR had not been reached

N=91 in the primary efficacy population for the evaluation of ORR and DOR.

Note: ORR assessed by IRC after a minimum follow-up of 6 months and represents the primary endpoint of the TIDAL study. Safety and duration of response data are as of the data cutoff date; the data cutoff date is approximately 6 months after the last patient in the primary efficacy population received their first dose of zandelisib. With exception of the ORR and CR data reported in the primary follicular lymphoma efficacy population of 91 patients, the data reported today provides an initial look at the data as of the data cutoff date and is interim and subject to change as more patient data become available. Because the data reported today is from an ongoing study, the final data may differ materially from the data reported in this presentation.



Discontinuation Rate Due to Any Drug Related Adverse Event

9.9%

Adverse Events of Special Interest (Grade ≥3)

- 1.7% ALT/AST Elevation
- 1.7% Colitis
- 5.0% Diarrhea
- 2.5% Mucositis
- 0.8% Pneumonitis
- 3.3% Rash

≤ 5% each

Median Follow-up of 9.4 Months (0.8-24)

N=121 in the total study population for the evaluation of safety.

Zandelisib: Exploring Full Potential as Backbone Therapy

Zandelisib Single Agent

- Ph 2 Study TIDAL in 3L+ FL and MZL
- Ph 2 Study K02 in 3L+ in iNHL (Japan)

Zandelisib + Rituximab

- Ph 3 Study COASTAL in 2L+ FL and MZL

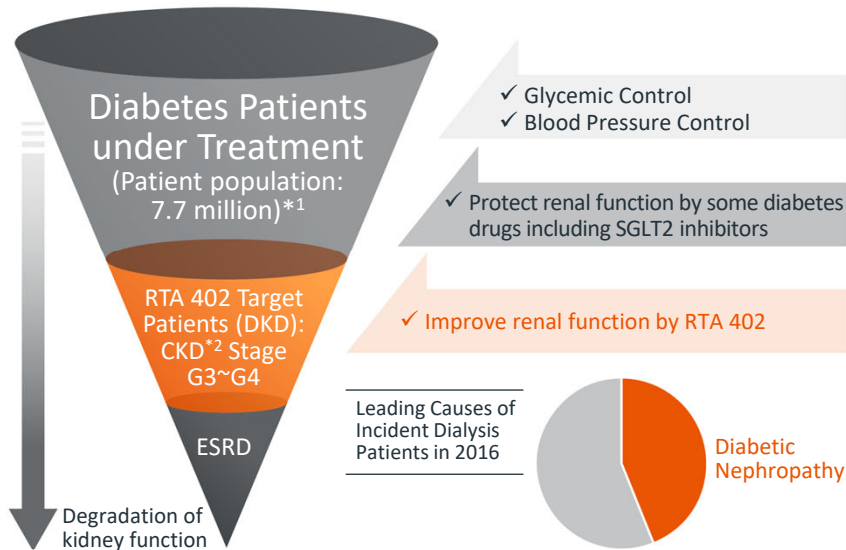
Other Zandelisib Combinations

- + Zanubrutinib in FL and MCL in 2L+
- + R-CHOP in DLBCL in 1L
- + Ven-R in CLL

Additional Studies in Active Planning and More Under Consideration

CLL: chronic lymphocytic lymphoma; DLBCL: diffuse large B-cell lymphoma; FL: follicular lymphoma; iNHL: indolent B-cell non-Hodgkin lymphoma; MCL: mantle cell lymphoma; MZL: marginal zone lymphoma; R-CHOP: rituximab-cyclophosphamide/doxorubicin/prednisone/vincristine; Ven-R: venetoclax-rituximab

Bardoxolone Methyl (RTA 402): Clinical and Commercial Opportunity



Market Opportunity

- Increasing diabetes patients in Japan*¹
- DKD - the leading cause of incident dialysis (around 40%)
- Innovative drug is highly anticipated (No drug can improve renal function)

RTA 402 Opportunity

- Novel MOA - Nrf2 activation
- Priority Review Designation under Japanese SAKIGAKE system
- Potential to be the first drug to improve renal function

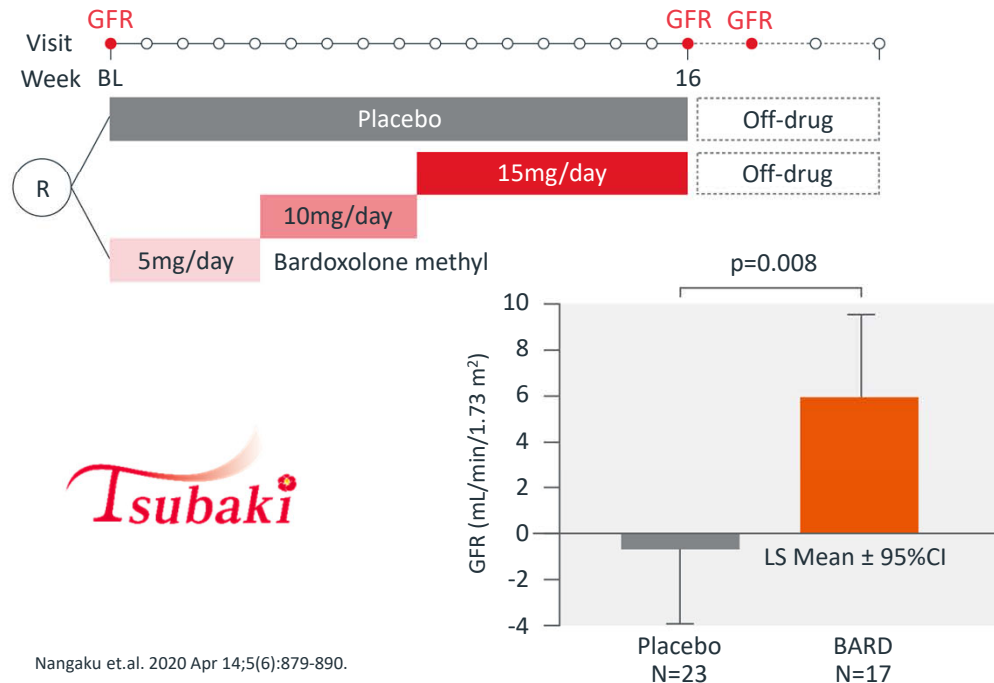


License Agreement on Bardoxolone Methyl for Japan and Certain Asian Markets (January 2010)
Kyowa Kirin has exclusive rights, sales royalty payments to Reata ranging from the low teens to the low 20%

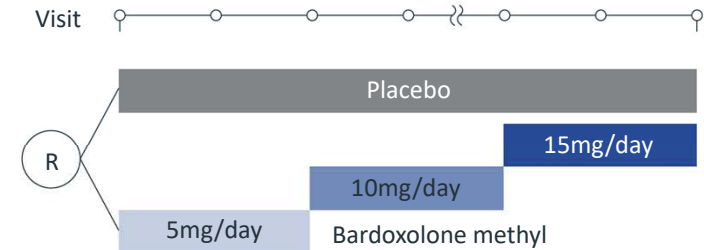
*¹ National Health and Nutrition Survey (2016) , MHLW/Ministry of Health, Labour and Welfare; *² CKD: chronic kidney disease

Bardoxolone Methyl: Increase GFR in CKD Patients with Type 2 Diabetes

Phase 2 study (TSUBAKI) in CKD patients with Type 2 Diabetes



Phase 3 study (AYAME) in DKD patients



Expected News Flow in FY2022

As of Nov 1, 2021

Code generic name	Target disease	2022
KHK4083/AMG 451	Atopic dermatitis	P3 FPI
KW-6356	Parkinson's disease	P2b detailed data P3 FPI
ME-401 Zandelisib	FL/MZL (2L, R combo)	Enrollment ongoing
	iNHL (3L, mono)	P2 Topline data
	CLL (Ven-R combo)	P2 FPI
RTA 402 Bardoxolone methyl	Alport syndrome	Regulatory decision (JP)
	Diabetic kidney disease	P3 LPO
KHK7791 Tenapanor	Hyperphosphatemia under maintenance dialysis	Marketing application (JP)

FPI: first patient in; FL: follicular lymphoma; MZL: marginal zone lymphoma; iNHL: indolent B-cell non-Hodgkin lymphoma; CLL: chronic lymphocytic leukemia; Ven-R: venetoclax-rituximab; LPO: last patient out

Outlook for the Future

- Delivering Life-changing Value as a GSP -

Outlook toward 2030



FY2021-2025 Medium Term Business Plan

Delivering **Life-changing** Value as a GSP



Appendix

Stock Information (As of December 31, 2020)

Stock Listing

Tokyo

Securities Code

4151

Transfer Agent of Common Stock

Sumitomo Mitsui Trust Bank, Limited
1-4-1, Marunouchi, Chiyoda-ku, Tokyo
100-8233, Japan
<http://www.smtb.jp/personal/agency/index.html>

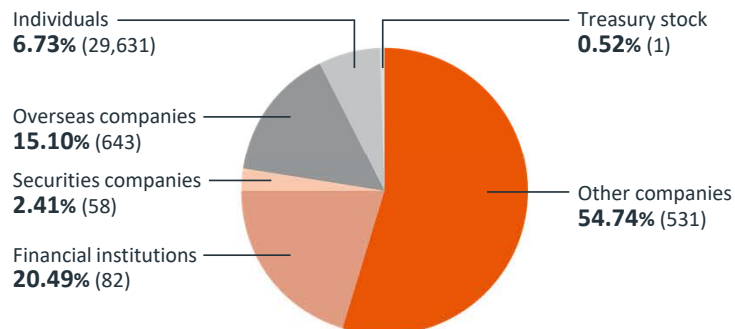
Number of Shares of Common Stock

Authorized: 987,900,000
Issued: 540,000,000

Number of Shareholders

30,946

Shareholding by Type of Investor (Number)



Principal Shareholders

	Number of Shares Held(Thousands)	Percentage of Total Shares Issued (%)
Kirin Holdings Company, Limited	288,819	53.77
The Master Trust Bank of Japan, Ltd. (Trust account)	43,422	8.08
Custody Bank of Japan, Ltd. (Trust account)	23,827	4.44
State Street Bank & Trust Company 505223	7,839	1.46
Custody Bank of Japan, Ltd. (Trust account 7)	5,527	1.03
Mizuho Trust & Banking Co., Ltd. (Retirement Benefit Trust for Mizuho Bank, Ltd.)	4,539	0.85
State Street Bank West Client-Treaty 505234	4,337	0.81
SMBC Nikko Securities Inc.	3,812	0.71
JPMorgan Chase Bank 385781	3,651	0.68
State Street Bank & Trust Company 505103	3,416	0.64

*1 The 4,539 thousand shares held by Mizuho Trust & Banking Co., Ltd. (Retirement Benefit Trust for Mizuho Bank, Ltd.) are the trust assets entrusted by Mizuho Bank for its retirement benefit trust, and voting rights for the shares are retained by Mizuho Bank.

*2 The 2,823 thousand shares held by the Company as treasury stock are excluded from the above because treasury stock has no voting rights.

Stock Price and Trading Volume



Total Shareholder Return (TSR)

	Past 4 years	Past 3 years	Past 2 years	Past 1 year	Current year
Kyowa Kirin Co., Ltd.	85.7%	116.5%	113.0%	141.1%	156.0%
TOPIX Total Return Index	100.3%	122.6%	103.0%	121.7%	130.7%

