

Results Presentation

Fiscal 2024 Second Quarter

協和キリン株式会社



Agenda

Financial Review
Commercial Update
R&D Update
News Flow in 2024
Q&A

President and Chief Executive Officer (CEO) **Masashi Miyamoto, Ph.D.**

Senior Managing Executive officer, Chief Medical Officer (CMO) **Takeyoshi Yamashita, Ph.D.**

Managing Executive Officer, Chief Financial Officer (CFO) **Motohiko Kawaguchi**

This document contains certain forward-looking statements relating to such items as the company's (including its domestic and overseas subsidiaries) forecasts, targets and plans. 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 the actual results in practice may differ substantially due to uncertain factors.

These uncertain factors include, but are not limited to, potential risks of the business activities in the pharmaceutical industry in Japan and overseas, intellectual property risks, risk of side effects, regulatory risks, product defect risks, risks of changes to the 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 used only for the purpose of providing the information to investors. Though it may contain the information concerning pharmaceutical products (including products under development), it is not for the purpose of promotion, advertising, or medical advice.

Financial Review

Summary of Q2 Results

Rev. Plan FX Rates (full-year)
 USD 140 → 151 / USD
 GBP 180 → 191 / GBP
 EUR 155 → 163 / EUR

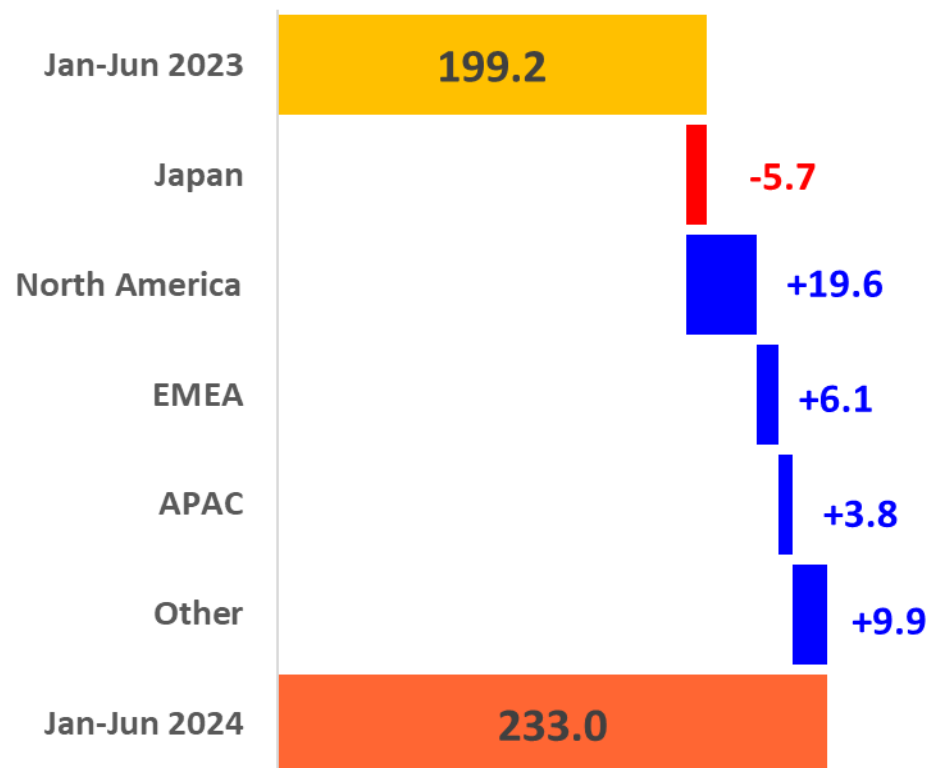


(Billion Yen / Rounded)

	2023Q2 Results	2024Q2 Results	Changes	FY2024 Rev. Plans	Progress to goal
Revenue <i>[Overseas Ratio]</i>	199.2 <i>[63%]</i>	233.0 <i>[71%]</i>	+33.8 (+17%)	473.0→ 492.0 <i>[71%]</i>	47%
Gross Profit <i>[Gross Profit Margin]</i>	152.2 <i>[76%]</i>	173.5 <i>[74%]</i>	+21.3 (+14%)	348.0→ 364.0 <i>[74%]</i>	48%
SG&A <i>[SG&A Ratio]</i>	82.4 <i>[41%]</i>	83.2 <i>[36%]</i>	+0.8 (+1%)	166.0→ 168.0 <i>[34%]</i>	50%
R&D <i>[R&D Ratio]</i>	33.7 <i>[17%]</i>	49.2 <i>[21%]</i>	+15.6 (+46%)	100.0→ 105.0 <i>[21%]</i>	47%
Gain/Loss on Equity Method	1.4	3.1	+1.7 (+124%)	3.0→ 1.0	311%
Core Operating Profit <i>[Core OP Margin]</i>	37.5 <i>[19%]</i>	44.1 <i>[19%]</i>	+6.7 (+18%)	85.0→ 92.0 <i>[19%]</i>	48%
Profit	21.6	37.8	+16.1 (+75%)	63.0→ 68.0	56%

YoY Analysis -Revenue-

+33.8 billion yen
(incl. forex effect +18.6)



● Japan -5.7

Although Phozevel, Duvroq and Crysvita increased, revenue in Japan region decreased by 8% due mainly to negative impact by annual NHI price-cut and shrink in G-Lasta affected by competitive products.

● North America +19.6 (incl. forex effect +9.4)

Revenue in North America region increased by 32% with the growth of Crysvita(+27%) and Poteligeo(+50%).

● EMEA +6.1 (incl. forex effect +4.6)

Revenue in EMEA region increased by 20% with the growth of Crysvita(+66%) and Poteligeo(+29%) although the shift from product sales to sales royalties/license fees for 13 established medicines portfolio, such as Abstral, by entered into the Joint Venture Collaboration with Grünenthal on Aug 1, 2023

● APAC +3.8 (incl. forex effect +1.7)

APAC revenue increased by 24% with the growth of Crysvita, and Nesp.

● Other +9.9 (incl. forex effect +2.9)

47% growth in the other revenue was due to the royalties of growing Fasenra (Benralizumab), upfront revenue from Boehringer Ingelheim, and new consolidation of Orchard.

Revenue of Major Items (Japan)

(Billion Yen / Rounded)

Item	2023Q2 Results	2024Q2 Results	Changes	Reasons	2024 Rev. Plans*	Progress to goal
Crysvita	4.8	5.4	+0.5 (+11)	Market penetration (Launched in Dec 2019)	12.9	42%
Poteligeo	0.9	1.0	+0.0 (+4%)		1.9	50%
Nesp + Nesp-AG ¹	8.4	6.9	-1.4 (-17%)		14.4	48%
Nesp	1.5	1.4	-0.2 (-10%)	NHI price-cut & Biosimilars' penetration	2.8	49%
Nesp-AG	6.9	5.6	-1.3 (-19%)		11.7	48%
Duvroq	4.2	5.7	+1.4 (+34%)	Market penetration (Launched in Aug 2020)	12.2	46%
Phozevel	-	1.7	+1.7 (- %)	Launched in Feb 2024	3.3	51%
Orkedia	5.0	4.9	-0.1 (-1%)		11.7	42%
G-Lasta	15.0	10.5	-4.5 (-30%)	NHI price-cut & Biosimilars' penetration	20.5	51%
Rituximab BS	4.4	3.8	-0.6 (-15%)	NHI price-cut	7.9	48%
Romiplate	5.7	6.5	+0.7 (+13%)	Market penetration (New indication in Jun 2019)	13.2	49%
Nouriaast	3.7	3.4	-0.3 (-9%)		7.1	47%
Haruopi	2.1	2.2	+0.1 (+4%)		5.2	42%

1 AG stands for Authorized Generic. Official product name is Darbepoetin Alfa [KKF]. Kyowa Kirin Frontier is a marketing authorization holder; Kyowa Kirin is a distributor.

* 2024 Revised Plan announced on August 1, 2024, there is no changes to the "Revenue of Major Items (Japan)"

Revenue of Major Items (ex-Japan)

(Billion Yen / Rounded)

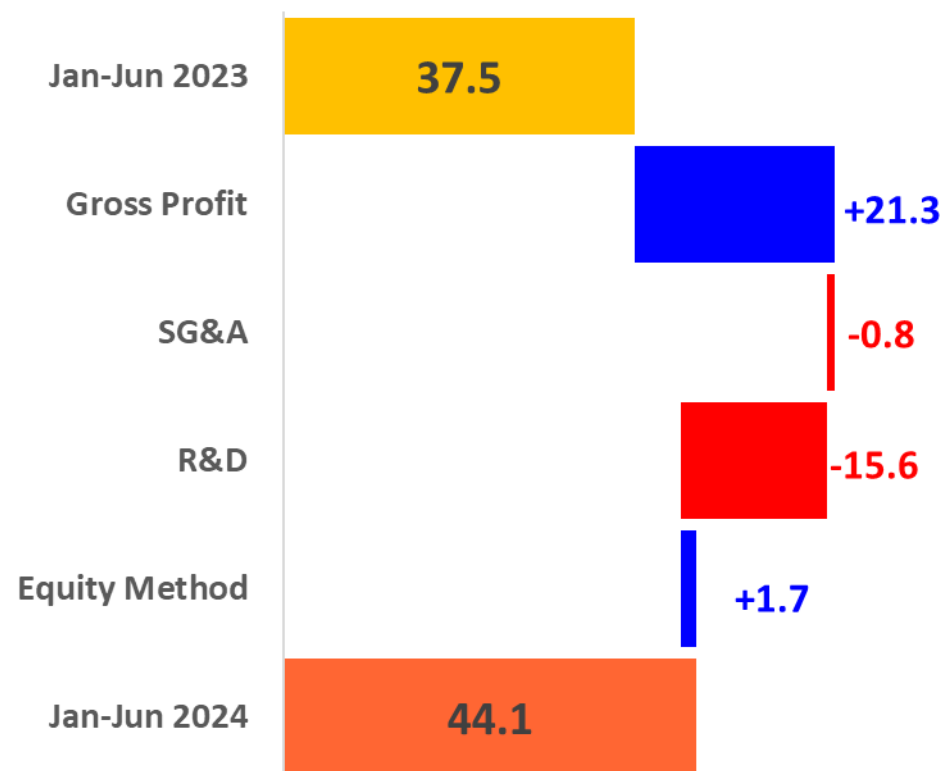
Item	2023Q2 Results	2024Q2 Results	Changes	Reasons
Crysvita	61.9	85.5	+23.6 (+38%)	[North America] Market penetration [EMEA] Geographical expansion & Additional indication (Adult/TIO) [APAC] Market penetration
North America	46.0	58.7	+12.7 (+27%)	
EMEA	15.3	25.4	+10.1 (+66%)	
APAC	0.6	1.3	+0.8 (+141%)	
Poteligeo	12.5	18.1	+5.6 (+45%)	[North America] Market penetration [EMEA] Geographical expansion & Market penetration
North America	9.4	14.1	+4.7 (+50%)	
EMEA	3.1	3.9	+0.9 (+29%)	
APAC	-	0.1	+0.1 (- %)	
Libmeldy / Lenmeldy	-	1.4	+1.4(- %)	New consolidation of Orchard (FDA approval in Mar 2024)
Nourianz	3.5	3.5	+0.0 (+0%)	
Nesp	4.4	5.7	+1.2 (+28%)	
Gran	3.2	3.7	+0.5 (+14%)	
Tech-licensing	17.8	23.3	+5.5 (+31%)	Upfront revenue from Boehringer Ingelheim and growth of Fasenra
Benralizumab Royalty ¹	11.6	14.4	+2.8 (+24%)	

2024 Rev. Plans	Progress to goal
175.9→ 187.8	46%
32.5→ 34.8	52%
23.3→ 25.1	56%
8.8→ 9.3	42%
0.5→ 0.5	18%
4.5→ 4.9	29%
8.5→ 9.1	39%
10.7→ 10.7	53%
7.2→ 7.2	51%
45.0→ 47.8	49%

1 Sales royalties of Fasenra which has been marketed by AstraZeneca, including our own estimation.

YoY Analysis -Core OP-

**+6.7 billion yen
(incl. forex effect +6.3)**



● Gross Profit +21.3 (incl. forex effect +16.2)

Increased in conjunction with JPY 33.8B rise in revenue. COGs have increased due to the North America Crysvita Sales royalty after Apr 27, 2023. Hence, gross profit % declined YoY. (76% → 74%)

● SG&A -0.8 (incl. forex effect -5.5)

SG&A slightly increased due to increase in HR expenses and FX impact, despite the decrease in Crysvita profit-sharing expenses because of the North America Crysvita-related scheme change after Apr 27, 2023.

[HR exp -5.4 / Sales promotion +9.7 (incl. Crysvita profit sharing expenses +10.8)]

● R&D -15.6 (incl. forex effect -4.3)

Increased in clinical study costs of KHK4083 which is undergoing joint global Phase III clinical study and new consolidation of Orchard

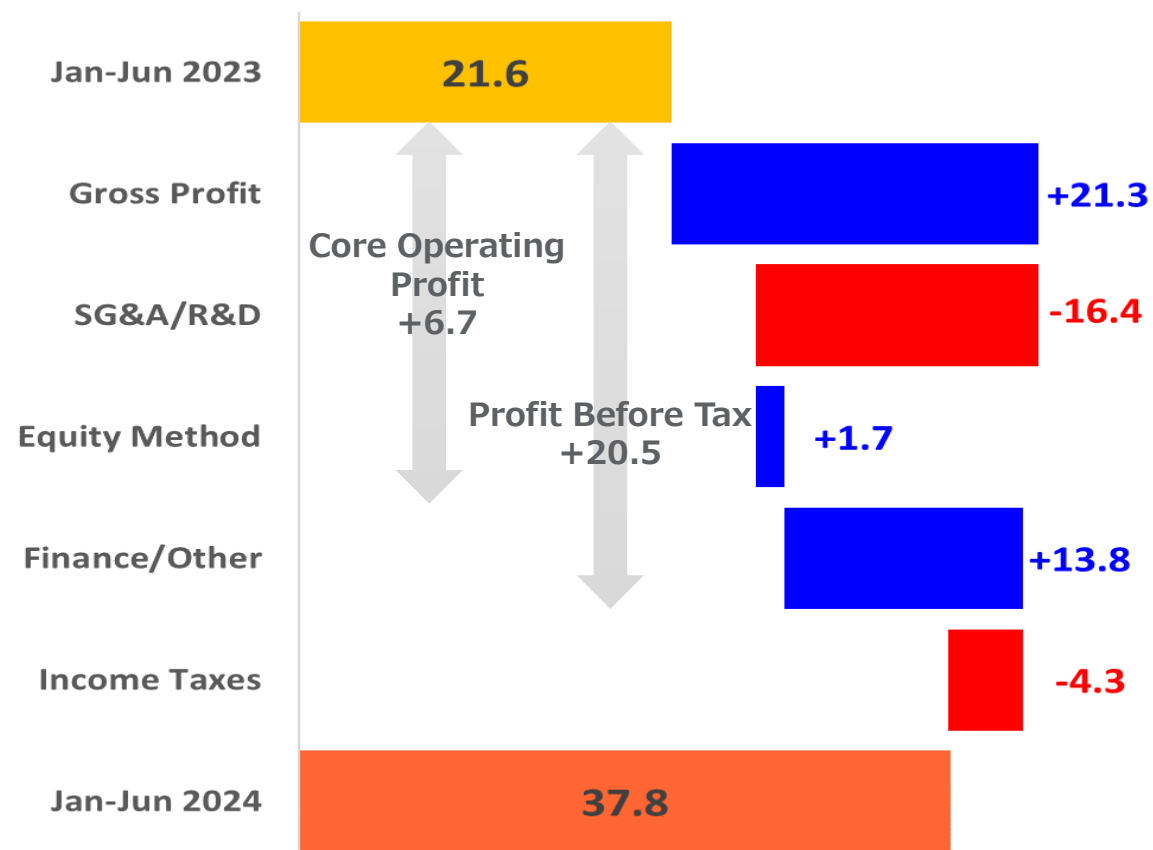
● Gain/Loss on Equity Method +1.7 (incl. forex effect +0.1)

FKB's Hulio (FKB327/Adalimumab biosimilar) continued to grow in Europe.

FKB; Fujifilm Kyowa Kirin Biologics Co., Ltd.

YoY Analysis -Profit-

Profit (Jan-Jun)
+16.1 billion yen



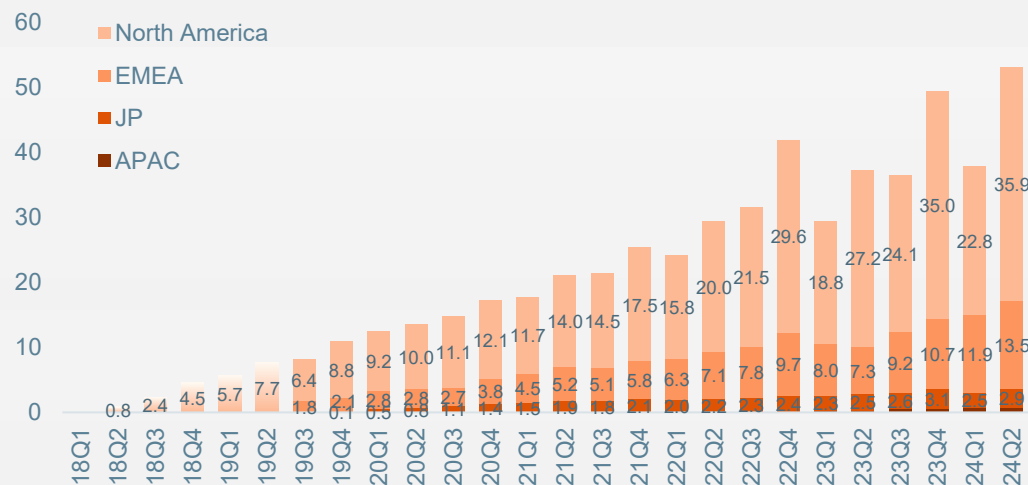
Commercial Update

2024 Key Actions & Q2 Topics

2024 Key Actions

- Strengthen evidence-based marketing activities.
- North America:
Enhance disease awareness activities. Strengthen further the foundation of the own sales structure.
- EMEA:
Continue to focus on geographical & indication expansion. Increase market penetration in adult XLH.
- Japan:
Further strengthen promotional activities by the dedicated personnel to accelerate growth.

Sales Revenue (Billion Yen)

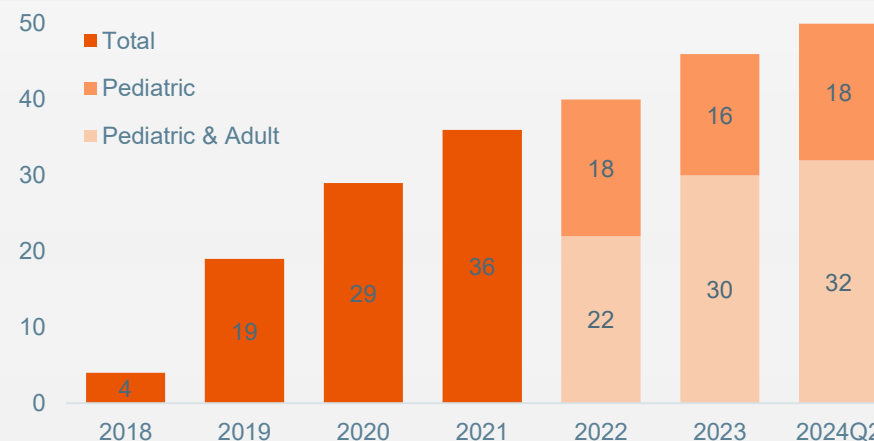


*Revenue from EAP (Early Access Program) is not included in sales until FY2022, and is included in sales from FY2023 onwards as it is insignificant in monetary terms.

Q2 Topics

- Strengthen evidence-based marketing activities.
- North America
• Seasonal factors have dissipated, and solid growth continues.(generally in line with plans)
- EMEA:
• In addition to growth from market expansion and patient penetration due to adult insurance reimbursement, some sales were shipped ahead of schedule.
Sales increased significantly YoY, partly due to price adjustments payment in the last year.
- NICE (National Institute for Health and Care Excellence) recommended this drug for the treatment of adult patients with XLH.
- Japan:
• Continued to strengthen promotional activities by the dedicated personnel.

Launched Countries / Regions (XLH)



*Excludes Latin America and Turkey, where Ultragenyx records sales.

2024 Key Actions & Q2 Topics

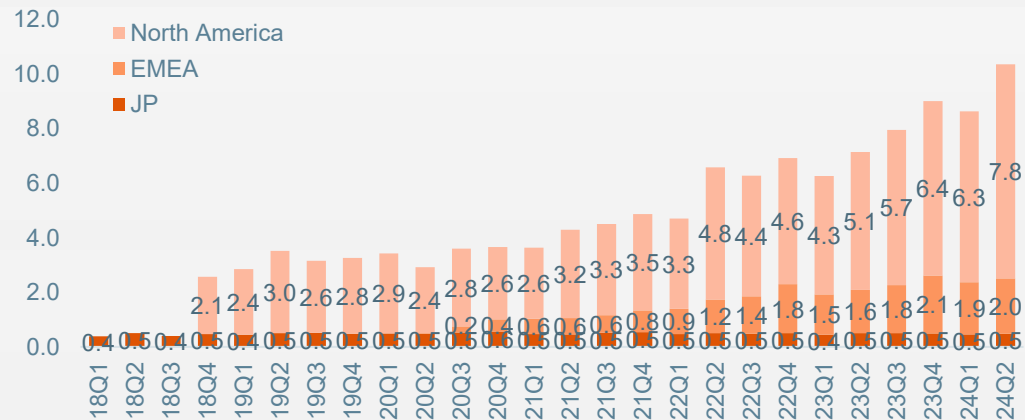
2024 Key Action

- Deeper penetration into the existing markets as well as expansion of targets through further progression of evidence-based promotional activities.
 - ◆ Continue to raise awareness of importance of blood testing to accurately stage disease.
 - ◆ Start promotional activities focusing on progressing CTCL patients with visible skin symptoms.
 - ◆ Geographic Expansion

Q2 Topics

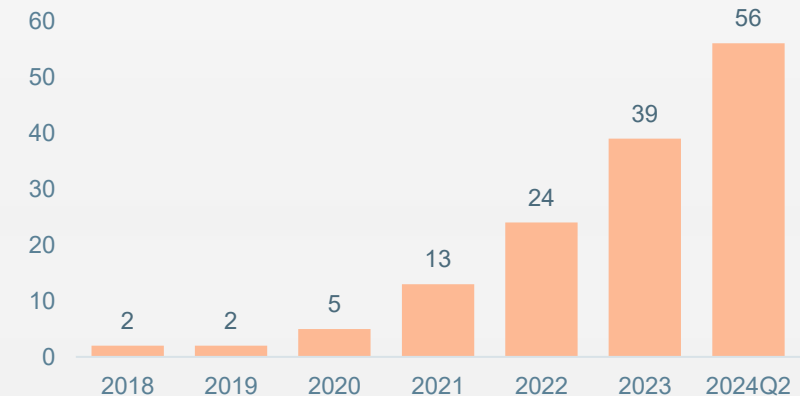
- NA : Sales revenue increased 50% YoY due to:
 - Expand evidence-based promotional activities to focus not only on cases with predominantly blood involvement, but also on early-stage cases with predominantly skin involvement.
 - Promotional activities focused on medical facilities with high potential for use based on data analysis.
- EMEA : Sales revenue increased by 29% YoY due to:
 - Geographic expansion
 - Deeper penetration into the existing markets

Sales Revenue (Billion Yen)



*Revenue from EAP (Early Access Program) is not included in sales until FY2022, and is included in sales from FY2023 onwards as it is insignificant in monetary terms.

Launched Countries / Regions



R&D Update

News Flow of Main Development Pipeline Products

Code
Generic Name

Events (Completed are in bold)

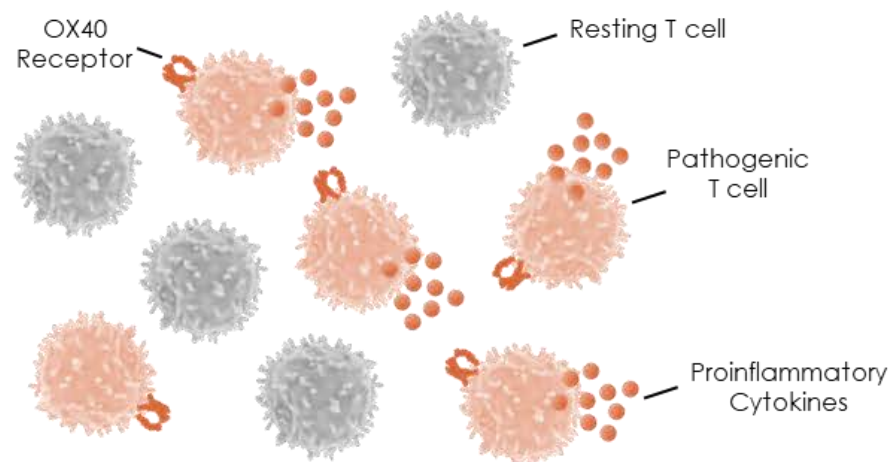
Timeline
(Completed are in orange)

KHK4083/AMG 451 rocatinlimab	Atopic Dermatitis	P3 (ROCKET Program)	In progress
	Asthma	P2 initiation	May 2024
	Prurigo nodularis	P3 initiation	July 2024
KHK4951 tivozanib	nAMD	P2	In progress
	DME	P2	In progress
KK4277	SLE, CLE	P1	In progress
KK2260	Advanced or metastatic solid tumors	P1	In progress
KK2269	Advanced or metastatic solid tumors	P1	In progress
KK2845	AML	P1 initiation	Q3 2024
KK8123	XLH	P1 initiation	Q3 2024
OTL-203	MPS-IH (Hurler syndrome)	Registrational study ¹	In progress
OTL-201	MPS-IIIA (Sanfilippo syndrome type A)	Proof-of-concept study ²	In progress

Rocatinlimab is a Potential T-cell Rebalancing Therapy

T-cell Imbalance

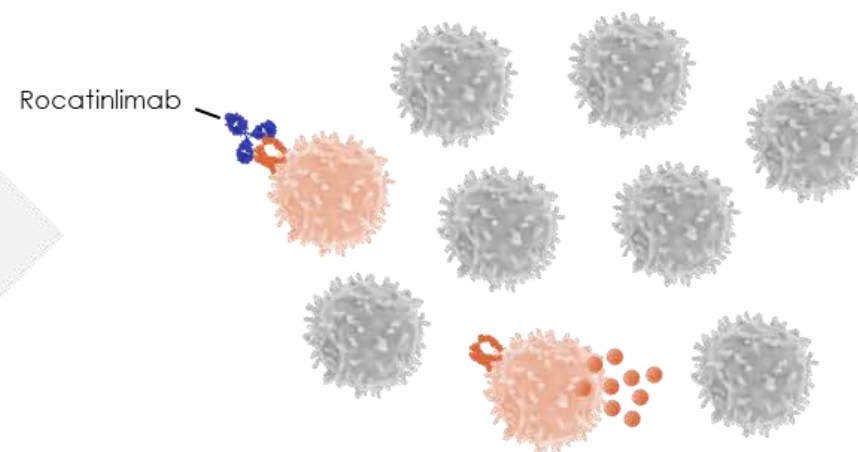
Driven by increased activity and number of pathogenic T cells



Rocatinlimab
rebalances
pathogenic T cells

T-cell Rebalance

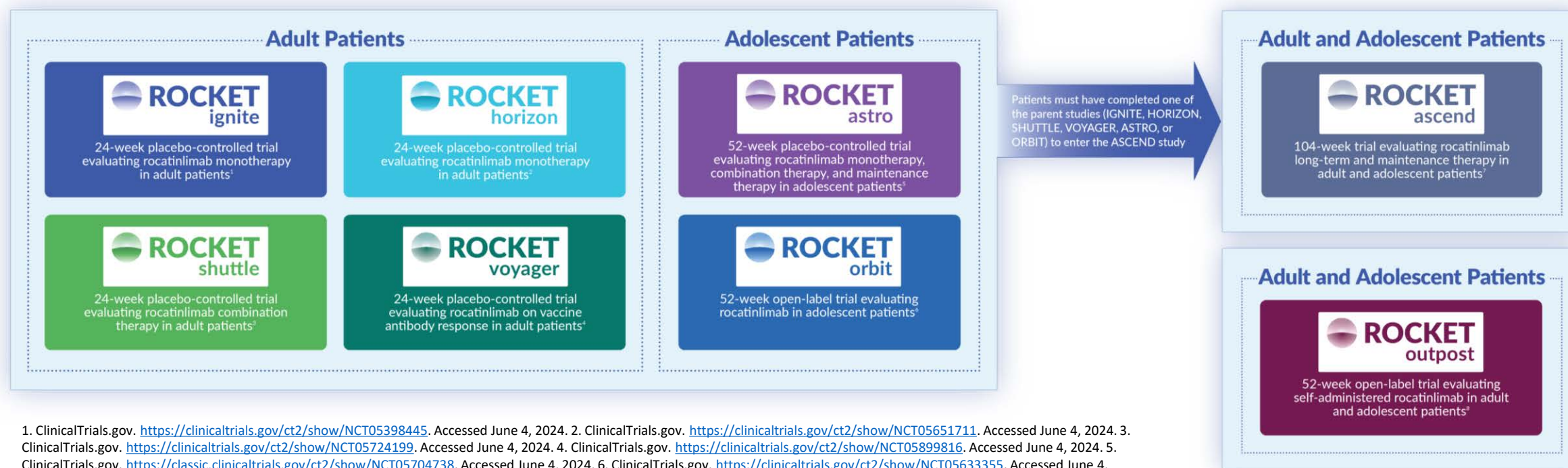
Achieved by inhibition and reduction of pathogenic T cells



- T-cell imbalance is a root cause of inflammatory disease
- Rocatinlimab is the potential first and only T-cell rebalancing therapy that inhibits and reduces pathogenic T cells by targeting OX40 receptor
- Rocatinlimab is a T-cell rebalancing therapy designed to relieve inflammatory diseases across heterogeneous patient types

Rocatinlimab – Progress of the ROCKET Program

- Composed of eight studies enrolling adult and adolescent patients
- To date, over 3,100 patients have been enrolled in the ROCKET Program with five studies having completed enrollment



1. ClinicalTrials.gov. <https://clinicaltrials.gov/ct2/show/NCT05398445>. Accessed June 4, 2024. 2. ClinicalTrials.gov. <https://clinicaltrials.gov/ct2/show/NCT05651711>. Accessed June 4, 2024. 3. ClinicalTrials.gov. <https://clinicaltrials.gov/ct2/show/NCT05724199>. Accessed June 4, 2024. 4. ClinicalTrials.gov. <https://clinicaltrials.gov/ct2/show/NCT05899816>. Accessed June 4, 2024. 5. ClinicalTrials.gov. <https://classic.clinicaltrials.gov/ct2/show/NCT05704738>. Accessed June 4, 2024. 6. ClinicalTrials.gov. <https://clinicaltrials.gov/ct2/show/NCT05633355>. Accessed June 4, 2024. 7. ClinicalTrials.gov. <https://clinicaltrials.gov/ct2/show/NCT05882877>. Accessed June 4, 2024. 8. ClinicalTrials.gov. <https://clinicaltrials.gov/study/NCT06224192>. Accessed June 4, 2024.

Currently preparing for the disclosure of the topline data of the ROCKET-horizon study
Data readout is anticipated in Q3 2024

News Flow in 2024

Year-to-date Key News Flow

As of Aug 1, 2024

Category	Date	Headline
SP	Jan 5	Out-licensed the exclusive and worldwide rights to Boehringer Ingelheim of developing first-in-class treatment for fibro-inflammatory diseases.
SI	Jan 24	Completion of share acquisition of Orchard Therapeutics plc, UK biopharmaceutical company
R&D	Feb 6	First Patient Randomized in Registrational Trial of OTL-203 for MPS-I Hurler Syndrome
R&D	Feb 6	First Patient Enrolled in the Phase2 Clinical Trial Evaluating Tivozanib Eye Drop for Diabetic Macular Edema
SI	Feb 7	Conclusion of Agreement with BridgeBio Pharma for an Exclusive License on Infigratinib in Skeletal Dysplasias in Japan
Finance	Feb 7	Acquisition of Own Shares and Cancellation of Treasury Shares
MKT	Feb 19	Launch of PHOZEVEL® Tablets for Improvement of Hyperphosphatemia in Chronic Kidney Disease Patients on Dialysis (Japan)
R&D	Mar 11	Presented the post-hoc analysis data from the Phase 2b study of rocatinlimab (AMG 451/KHK4083) at American Academy of Dermatology (AAD) 2024 Annual Meeting
R&D	Mar 19	Receives FDA Approval of OTL-200 (Lenmeldy) for the treatment of children with early-onset—metachromatic leukodystrophy (MLD)

ESG: environmental, social, and governance; LCM: lifecycle management; R&D: research and development; SCM: supply chain management; SI: strategic investment; SP; strategic partnering MKT; marketing

Year-to-date Key News Flow

As of Aug 1, 2024

Category	Date	Headline
ESG	May 14	Announced the Publication of a Patient-focused Global Consensus Statement for Improving Diagnosis and Care in Cutaneous T-Cell Lymphoma (Kyowa Kirin, Inc.)
LCM	May 17	Approval for Partial Change of Approved Indication of G-Lasta® for the Mobilization of Hematopoietic Stem Cells into Peripheral Blood for Autologous Blood Stem Cell Transplantation in Japan
SCM	Jun 10	Announced Establishing New Biologics Manufacturing Plant in North Carolina, in the United States
LCM	Jun 28	Application for Additional Formulation of “LUMICEF® Subcutaneous Injection 210 mg Pen” in Japan
MKT	Jul 1	Announced Global Progress toward Advancing Newborn Screening for MLD (Orchard Therapeutics)
ESG	Jul 29	Joined the Pharmaceutical Supply Chain Initiative (PSCI)
SCM	Aug 1	Restructuring of APAC Region Business and Change in Kyowa Kirin China Pharmaceutical Co., LTD
R&D	Aug 1	Transition to a Research Organization to Realize Our Vision toward 2030, and Introduction of a Voluntary Retirement Program
Updates after the previous earnings announcement		

ESG: environmental, social, and governance; LCM: lifecycle management; R&D: research and development; SCM: supply chain management; SI: strategic investment; SP; strategic partnering MKT; marketing

Efforts toward the expansion of newborn screening (NBS) for MLD

<https://ir.orchard-tx.com/news-releases/news-release-details/orchard-therapeutics-celebrates-global-progress-toward-advancing>

Nomination to add MLD to the RUSP¹ submitted by multi-disciplinary expert working group

- Submitted on June 27 to ACHDNC².
- The submission initiates the review process for the benefit of NBS for MLD.
- The committee will analyze:
 - The effectiveness and precision of the screening test to detect newborns with MLD
 - Treatment guidelines for diagnosed children
 - The clinical benefit of pre-symptomatic diagnosis and treatment
- Currently, 12 states have legislation to expedite adding new conditions to state NBS panels once added to RUSP.

Making progress toward the implementation of national MLD NBS in U.S.

Activities toward the implementation of MLD NBS are steadily progressing on a global scale

Norway has adopted MLD into its national NBS

- The Ministry of Health and Care Services in Norway has added MLD to its expanded national NBS panel on June 25.

Norway becomes the first country in the world to add MLD to its national NBS program

Multiple papers on NBS for MLD published in 1H 2024

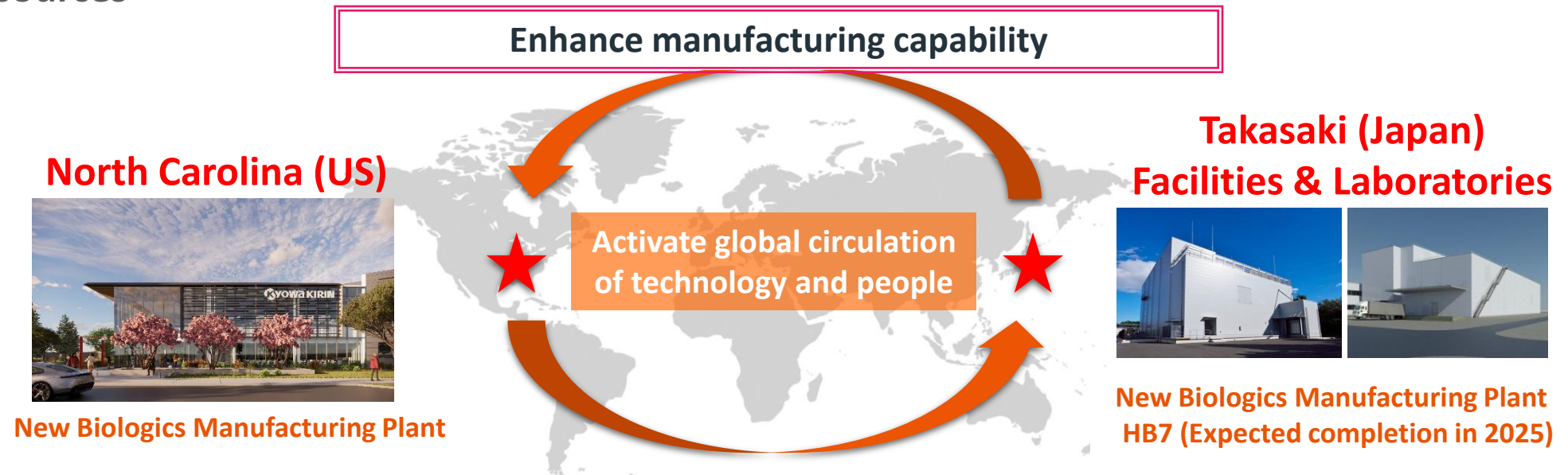
- Consensus guidelines for monitoring and managing MLD (US)
- European consensus-based recommendations for clinical management of NBS-identified MLD
- Details of the preliminary NBS test results and the proposed optimal screening algorithm (UK)
- Details of a high-specificity screening assay for detecting MLD
- A health economic analysis demonstrating the cost-effectiveness of NBS for MLD (UK)

Evidence generation toward universal NBS for MLD is progressing

1. The U.S. Recommended Uniform Screening Panel; 2. Advisory Committee on Heritable Disorders in Newborns and Children

Establishing New Biologics Manufacturing Plant in North Carolina

- ✓ Investing up to \$530M in construction with two-bioreactor facility
- ✓ Scheduled to commence construction in H2 2024 and complete in 2027
- ✓ Planning to manufacture innovative biologic therapies, including next-generation antibodies, for our planned clinical trials and future commercial use
- ✓ Aiming to enhance manufacturing capability by activating global circulation of technology and human resources



Restructuring of APAC business

- ✓ APAC business will be restructured in accordance with Story For Vision 2030
- ✓ China business: Divest all the equity to WinHealth
- ✓ Established medicines portfolio (excl. China): Grant commercial license to DKSH
- ✓ Global products (Crysvita and Poteligeo): Grant commercial license to partners in certain countries / regions

	Established medicines portfolio	Global Products
China	Divest to WinHealth	Partner with WinHealth
Korea / Taiwan	Partner with DKSH	Kyowa Kirin
Australia	N/A	Kyowa Kirin
Other Asia *	Partner with DKSH	Partner with DKSH

* Hong Kong / Macau / Thailand / Malaysia / Singapore

P/L Impact on Restructuring of APAC business

		Country / region	Until September 2024	October 2024 onwards
Revenue	Divest (Established medicines portfolio)	CN	Sales to market	Sales to Partner
	Partnering (Established medicines portfolio & Global products)	CN/HK/MO/ MY/SG/TH /KR/TW		Sales to Partner
	Continuation of in-house (Global products)	KR/TW/AU		Sales to market
COGs		ALL	COGs	COGs
SG&A	Divest / Partnering	CN/HK/MO/ MY/SG/TH /KR/TW	SG&A	
	Continuation of in-house (Global products)	KR/TW/AU		SG&A
Other income / expenses			Gain on sales of shares Business restructuring expenses	Business restructuring expenses

Transition of the Research Organization aiming to realize Vision toward 2030

- ✓ Implement the transition to research functions in line with the 'Story for Vision 2030' and aim to further strengthen our drug discovery capabilities.
 - 1) Shifting focus disease areas: bone & mineral, intractable hematological diseases/hemato oncology, and rare diseases
 - 2) Strengthening Innovative Modalities (advanced antibody technologies and hematopoietic stem cell gene therapy (HSC-GT) etc..)
 - 3) Globalization and restructuring of Research Organization
- ✓ During the transition, we are clarifying our focus area, and planning a significant reduction in our in-house small molecule drug discovery research activities

As part of the restructuring, a temporary voluntary retirement program will be introduced for the Research Division, the Production Division's CMC R&D Center, and certain groups in the Quality Division's Global CMC Quality Unit

Appendix

Our Vision toward 2030

Our 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

We are focused on developing medicines for diseases where there is a clear patient need for new options. We make full use of multiple therapeutic modalities, including biotechnology such as antibody technology, and beyond, building on our Kyowa Kirin established strengths.

Address patient-centric healthcare needs

We will meet the needs of patients and society by providing value across the entire patient care pathway, delivering cutting-edge science and technology, grounded in our in-depth pharmaceutical knowledge and expertise.

Retain the trust of society

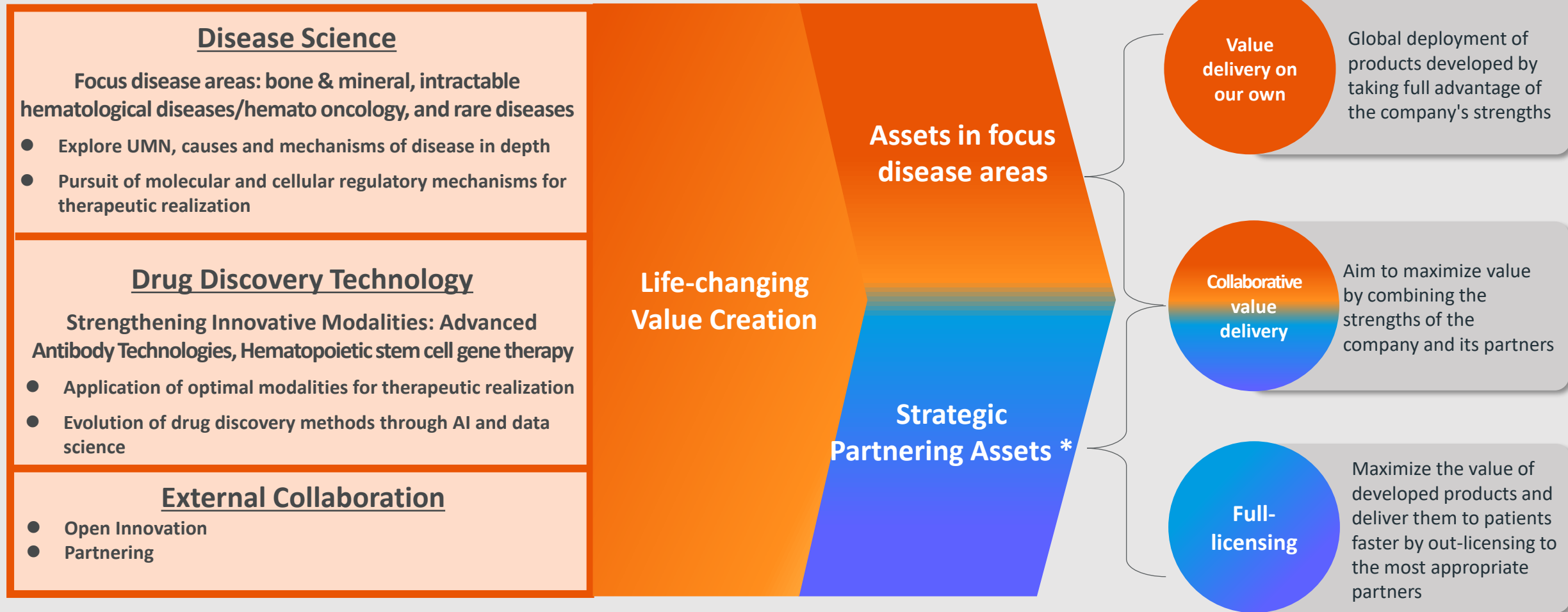
We pursue world-class product quality and operational excellence to grow our business in ways which build long-term trust with our stakeholders.

* Make patients smile through dramatic improvements in quality of life by identifying the unmet medical needs of people battling with medical conditions and by creating and supplying new drugs or services that help them overcome those challenges.



Story for Vision 2030

Strategies for creating and delivering life-changing value

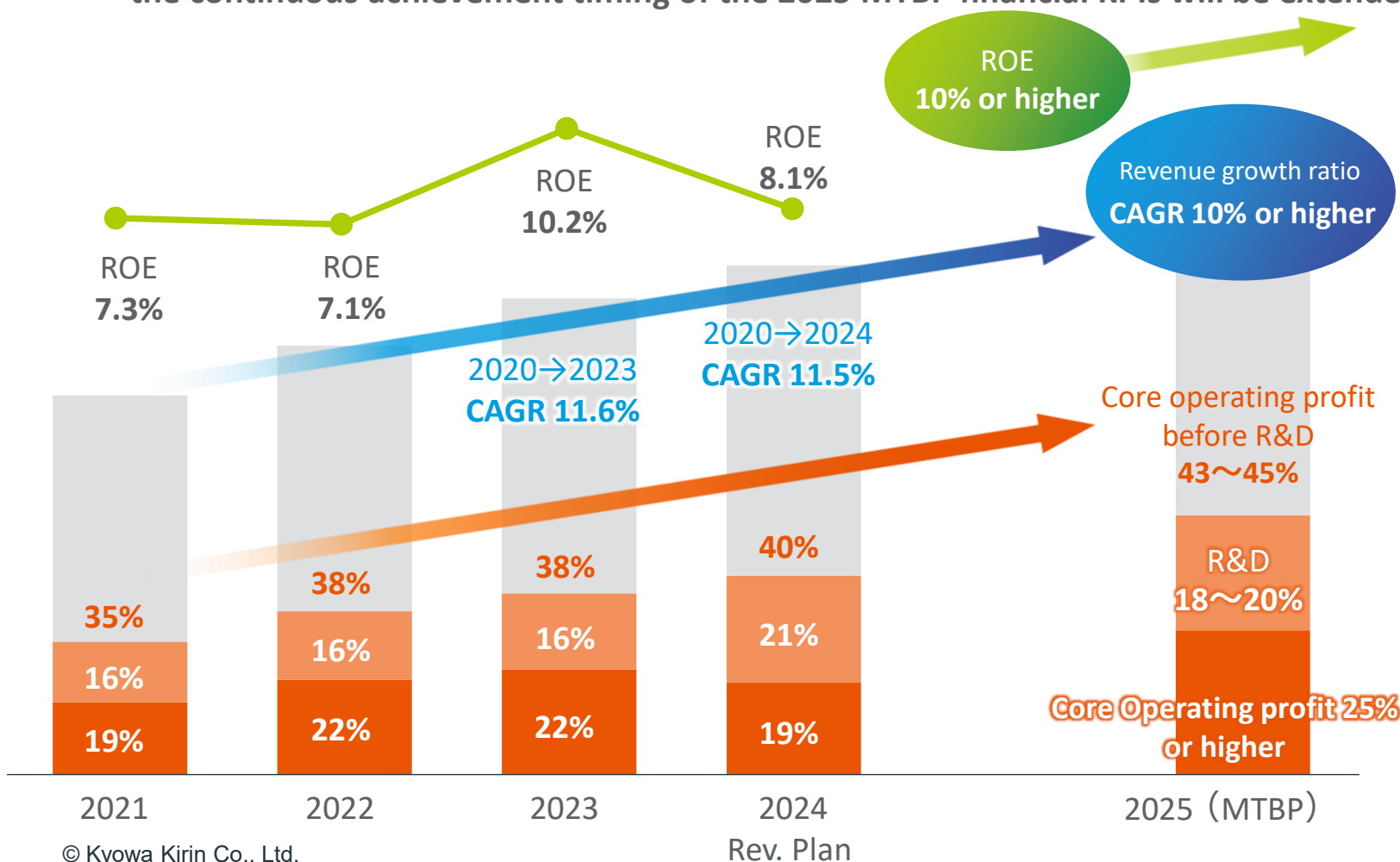


*Assets outside of the disease areas of focus are designated as strategic partnering assets, and value maximization is achieved through collaboration with partners.

2021-2025 Medium Term Business Plan

- Revision of Financial KPI -

- Record high in Core operating profit for FY2023. Achieved KPIs such as “ROE of 10%” and “CAGR of 10% or higher”
- Due to the restructuring of our business model to adapt to environmental changes, the continuous achievement timing of the 2025 MTBP financial KPIs will be extended to 2026 or beyond.



2025 MTBP financial KPIs
Achievement timing will be 2026 or beyond



- Steady growth in Crysvita sales
- Collaboration with Amgen on KHK4083
- Depreciation of Yen



- Short term financial impact on Orchard acquisition
- Increasing investment in KHK4083 development
- Depreciation of Drug price environment (Japan, Europe, and China)
- Unlaunched new products (discontinued pipelines, Nourianz in Europe)

Main Development Pipeline Products (After Ph2)

As of Aug 1, 2024

	Diseases under development* ¹	Planned Approval Year* ²	Development status	Total addressable market* ³	No. of Patients* ⁴
KHK4083/AMG 451 rocatinlimab	Moderate to severe Atopic Dermatitis	2026/2027	P3 (Global)	★★★★★	16M
	Moderate to severe Asthma	TBD	P2 (Global)	★★★★★	13.5M
	Prurigo nodularis	TBD	P3 (Global)	★★★★★	1M
KHK4951 tivozanib	nAMD	TBD	P2 (JP, US)	★★★★★	2,600K
	DME	TBD	P2 (JP, US)	★★★★★	3,400K
OTL-203	MPS-IH (Hurler syndrome)	2029/2030	Registrational study* ⁵ (US, EU)	★	(1 in 100K live birth)* ⁶
OTL-201	MPS-IIIA (Sanfilippo syndrome type A)	TBD	Proof-of-concept* ⁷	★	(1 in 100K)

*1 Expected indications as of the date of this document; indications may ultimately differ to expectations due status of approvals from regulatory authorities. *2 Expected year of first approval. *3 Expected total addressable market estimated by Kyowa Kirin, which is the sum of all products for the indications shown in *1, not projected sales or the Company's targets. **Colored areas represent estimates for global, and the rest are for Japan.** ★: less than ¥50Bn、★★: ¥50Bn-¥100Bn、★★★: Over ¥100Bn-¥500Bn、★★★★: Over ¥500Bn-¥1Tn、★★★★★: Over ¥1Tn. *4 Total number of estimated patients by Kyowa Kirin. **Colored areas represent in-house estimates for global, and the rest are in-house estimates for Japan.** *5 Equivalent to P3 study. *6 "1 in 100k live birth" is estimated incidence for all of MPS-I, of which approximately 70 percent are cases of Hurler syndrome. *7 Equivalent to P1/2 study.

Main Development Pipeline Products (nonclinical ~ Ph1)

As of Aug 1, 2024

	Diseases under development*1	Development status	Modality, technology
KK4277	SLE, CLE	P1 (JP, Asia)	Antibody, POTELLIGENT®
KK2260	Advanced or metastatic solid tumors	P1 (JP: in progress, US: in preparation)	Antibody, REGULGENT™
KK2269	Advanced or metastatic solid tumors	P1 (JP, US)	Antibody, REGULGENT™
KK2845	AML	Preparation underway for P1 (JP)	Antibody-Drug Conjugate
KK8123	XLH	Preparation underway for P1 (US, EU)	Antibody

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

Main Development Pipeline Products: Future plans

As of Aug 1, 2024

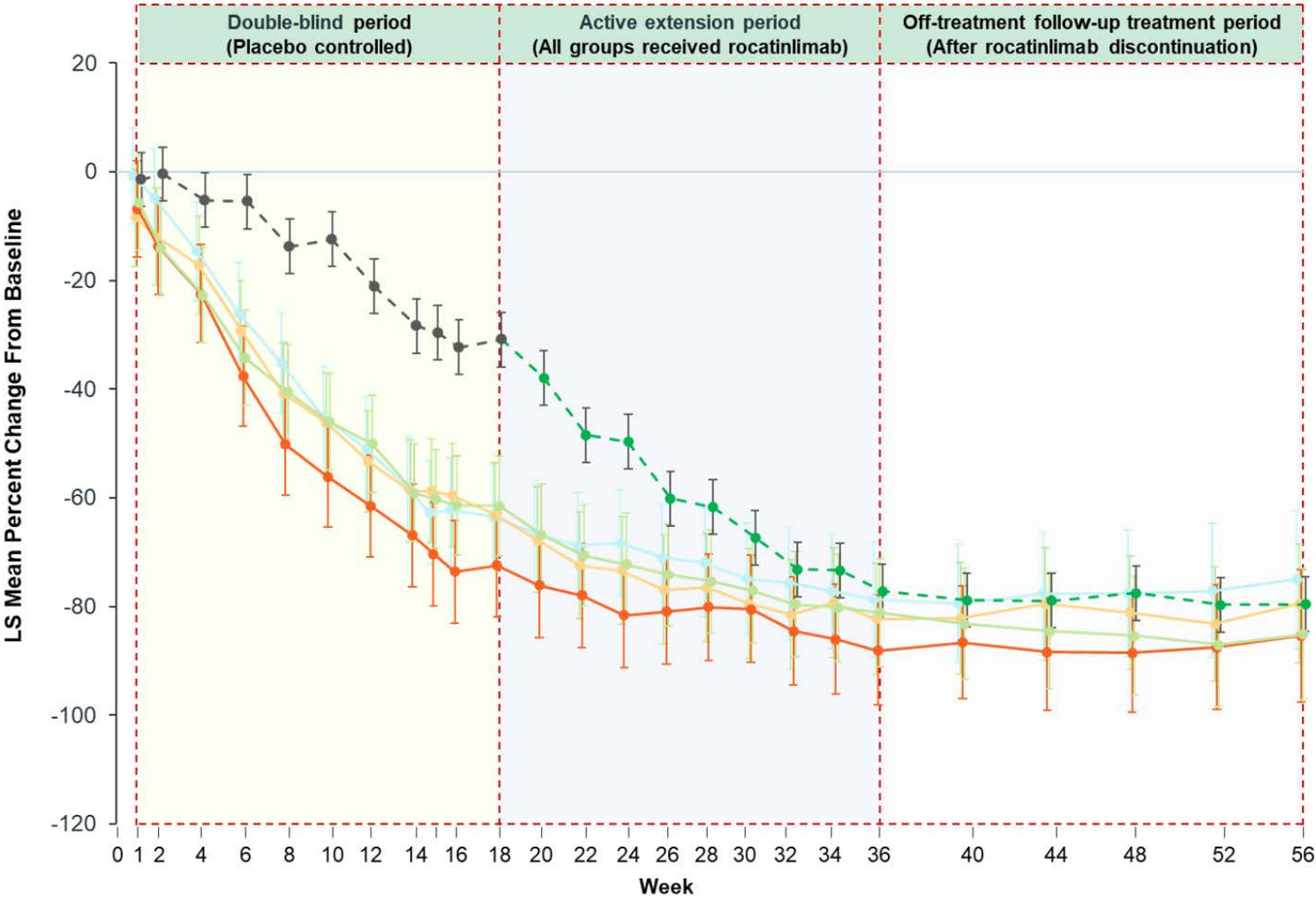
T : Topline data

D : Detailed data

Code Generic Name	Target Disease		2024	2025	2026	+
KHK4083/ AMG 451 rocatinlimab	Moderate to severe atopic dermatitis	P3				IGNITE
		P3				HORIZON
		P3				SHUTTLE
		P3				ASTRO
		P3				ORBIT
		P3				VOYAGER
		P3				ASCEND
		P3				OUTPOST
KHK4951 tivozanib	Moderate to severe asthma	P2				
	nAMD	P2				
	DME	P2				
KK4277	Systemic lupus erythematosus Cutaneous lupus erythematosus	P1				
KK2260	Advanced or metastatic solid tumors	P1				
KK2269	Advanced or metastatic solid tumors	P1				



Rocatinlimab: Phase 2b data¹



The Least-squares (LS) mean percent change in Eczema Area and Severity Index (EASI) score

	Week 16	Week 24	Week 36	Week 56
Rocatinlimab 150 mg Q4W, %	-62.2	-68.3	-78.7	-75.0
Rocatinlimab 600 mg Q4W, %	-59.5	-73.4	-82.3	-79.5
Rocatinlimab 300 mg Q2W, %	-73.6	-81.6	-88.1	-85.4
Rocatinlimab 600 mg Q2W, %	-61.4	-72.2	-81.1	-85.1
Placebo/rocatinlimab 600 mg Q2W, %	-32.3	-49.7	-77.2	-79.6

Sustained improvement in EASI after treatment discontinuation (Week 36)

Libmeldy® (OTL-200, atidarsagene autotemcel)

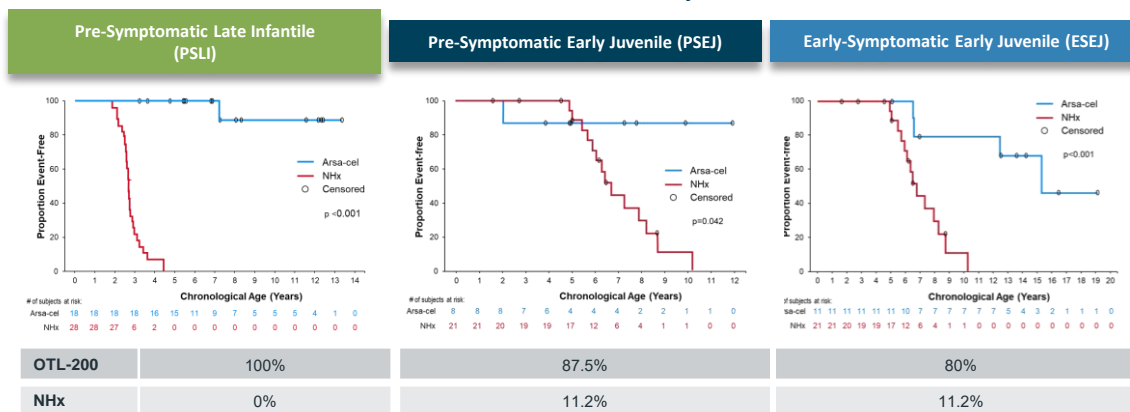
■ MLD (Metachromatic Leukodystrophy)

- Fatal genetic CNS disorder
- Rapid and irreversible loss of motor and cognitive function
- In its most severe form, most children pass away within five years of symptom onset¹

Severe Motor Impairment Free Survival (sMFS)

Interval from birth to first occurrence GMFC-MLD ≥ 5
(no locomotion and unable to sit) or death

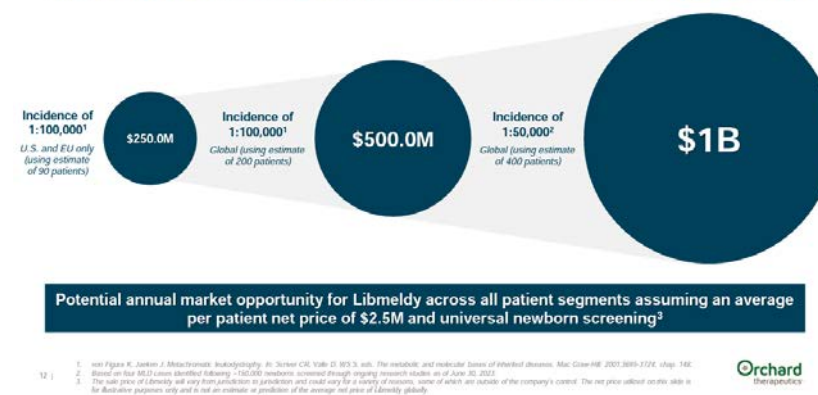
Treatment with OTL-200 resulted in statistically significant and clinically meaningful improvement in sMFS in the PSLI ($p < 0.001$), PSEJ ($p = 0.042$) and ESEJ ($p < 0.001$) MLD subgroups compared to disease natural history.



Data presented at the at the Society for the Study of Inborn Errors of Metabolism (SSIEM) Annual Symposium 2023, August 2023

Ref.) Orchard Therapeutics plc, Q2 2023 Financial Results and Webcast
<https://ir.orchard-tx.com/static-files/9fed8b65-2fd9-491a-97c0-69bf6595c0c3>

MLD Represents a Significant Annual Global Market Opportunity



Potential annual market opportunity for Libmeldy® across all patient segments assuming an average per patient net price of \$2.5M and universal newborn screening²

1. van Rappard DF, Boelens JJ, Wolf NI. Metachromatic leukodystrophy: disease spectrum and approaches for treatment. Best Pract Res Clin Endocrinol Metab 2015; 29: 261–73.
2. The sale price of Libmeldy® will vary from jurisdiction to jurisdiction and could vary for a variety of reasons, some of which are outside of the company's control. The net price utilized on this slide is for illustrative purposes only and is not an estimate or prediction of the average net price of Libmeldy® globally.

FOREX Information

Average FOREX Rates (yen)

	2023Q2	2024Q2	Changes	2024 Rev. Plans
USD	134	151	+17	151
GBP	164	191	+27	191
EUR	144	163	+19	163

Q1 YoY FOREX Impacts (billion yen)

	Revenue	Core OP
USD	+11.8	+3.9
GBP	+1.1	-0.6
EUR	+3.2	+1.8

FY2024 FOREX Sensitivities (based on 2024 Plans, billion yen)

	Changes	Revenue	Core OP
USD	+1 yen	+1.4	+0.4
GBP	+1 yen	+0.2	-0.0
EUR	+1 yen	+0.3	+0.2

KHK4083/AMG 451 - Collaboration with Amgen -

	US	Europe & 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 (done) and future contingent milestone payments potentially worth up to an additional \$850 million, as well as royalty payments on future global sales, to Kyowa Kirin.

Estimated Patient Numbers

Disease	Country/ Region	Incidence	Prevalence*	Reference
ATL	JP	1,150 / y		Survey and countermeasures to HTLV-1 infection and related diseases in Japan. 2009 summary research report (Yamaguchi, 2010)
PTCL	JP		2,000	Ministry of Health, Labour and Welfare: 2017 Patient survey (illness classification)
CTCL	JP		2,000	Ministry of Health, Labour and Welfare: 2017 Patient survey (illness classification)
	US	1,500 / y		SEER Data (2001-2007)
XLH	JP	1:20,000	Adult: 5,000 Ped: 1,000	Estimate based on reported prevalence of 1 in 20,000 people; Nationwide survey of fibroblast growth factor 23 (FGF23)-related hypophosphatemic diseases in Japan: prevalence, biochemical data and treatment. (Endo I et al., Endocr J., 2015)
	EU	1:20,000	Adult: 12,000 Ped: 3,000	Estimate based on reported prevalence of 1 in 20,000 people
	US	1:20,000	Adult: 12,000 Ped: 3,000	Estimate based on reported prevalence of 1 in 20,000 people; New perspectives on the biology and treatment of X-linked hypophosphatemic rickets. (Carpenter TO, Pediatr Clin North Am., 1997)
TIO	JP		30	2010 Ministry of Health, Labour and Welfare Epidemiological Research on abnormalities in Hormone Receptor Mechanisms
	US		500-1,000	Survey by Ultragenyx Pharmaceutical
AD	JP, NA, EU		30,000,000	Study by Decision Resources
nAMD	JP, US		2,300,000	Study by Decision Resources
PE	JP		15,000	Estimate based on the Demographic Survey by the Ministry of Health, Labour and Welfare and the estimated incidence of this disease

*Prevalence represents the estimated patient number per the entire population of each country or region.

List of Acronyms

AD	Atopic Dermatitis
AG	Authorized Generic
APAC	Asia-Pacific
AML	Acute Myeloid Leukemia
BS	Biosimilar
CTCL	cutaneous T cell lymphoma
DME	Diabetic Macular Edema
EMEA	Europe, the Middle East and Africa
JP	Japan
LCM	Lifecycle Management
MDS	Myelodysplastic syndromes
MF	Mycosis fungoides
MLD	Metachromatic Leukodystrophy
MPS-IH	Mucopolysaccharidosis type I, Hurler syndrome
MPS-IIIA	Mucopolysaccharidosis type IIIA
NA	North America
nAMD	neovascular Age-related Macular Degeneration
PTCL	peripheral T-cell lymphoma
SS	Sézary syndrome
TIO	Tumor Induced Osteomalacia
XLH	X-linked Hypophosphatemia



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