Results Presentation
Fiscal 2024 First Quarter

協和キリン株式会社

Agenda

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

Commercial Update
Managing Executive Officer, Chief Strategy Officer (CSO) Yasuo Fujii

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

News Flow in 2024
Managing Executive Officer, Chief Strategy Officer (CSO) Yasuo Fujii

Q&A
Senior Managing Executive officer, Chief Medical Officer (CMO) Takeyoshi Yamashita, Ph.D.
Managing Executive Officer, Chief Financial Officer (CFO) Motohiko Kawaguchi
Managing Executive Officer, Chief Strategy Officer (CSO) Yasuo Fujii

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

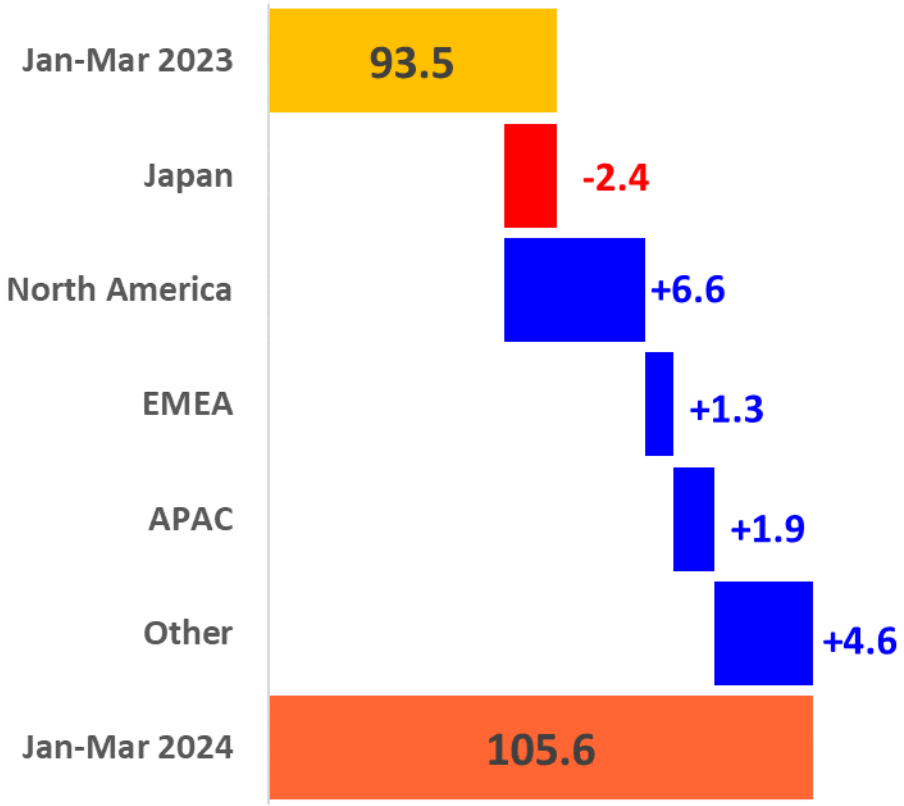
Summary of Q1 Results

(Billion Yen / Rounded)

	2023Q1 Results	2024Q1 Results	Changes	FY2024 Plans	Progresses
Revenue *[Overseas Ratio]*	93.5 *[63%]*	105.6 *[68%]*	+12.0 (+13%)	473.0 *[70%]*	22%
Gross Profit *[Gross Profit Margin]*	74.6 *[80%]*	80.0 *[76%]*	+5.4 (+7%)	348.0 *[74%]*	23%
SG&A *[SG&A Ratio]*	41.8 *[45%]*	40.2 *[38%]*	-1.6 (-4%)	166.0 *[35%]*	24%
R&D *[R&D Ratio]*	*16.6* *[18%]*	23.3 *[22%]*	+6.7 (+40%)	100.0 *[21%]*	23%
Gain/Loss on Equity Method	0.8	0.9	+0.1 (+13%)	3.0	30%
Core Operating Profit *[Core OP Margin]*	17.0 *[18%]*	17.4 *[16%]*	+0.4 (+2%)	85.0 *[18%]*	20%
Profit	12.8	14.6	1.9 (+15%)	63.0	23%

YoY Analysis -Revenue-

+12.0 billion yen
(incl. forex effect +7.3)



- **Japan -2.4**
  Although Duvroq, Phozevel, and Crysvita increased, revenue in Japan region decreased by 7% due mainly to negative impact by annual NHI price-cut and shrink in G-Lasta affected by competitive products.

- **North America +6.6 (incl. forex effect +3.4)**
  Revenue in North America region increased by 26% with the growth of Crysvita(+21%) and Poteligeo(+44%).

- **EMEA +1.3 (incl. forex effect +2.0)**
  Revenue in EMEA region increased by 8% with the growth of Crysvita(+49%) 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 +1.9 (incl. forex effect +0.7)**
  APAC revenue increased by 26% with the growth of Crysvita, and Nesp.

- **Other +4.6 (incl. forex effect +1.2)**
  42% 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	2023Q1 Results	2024Q1 Results	Changes	Reasons	2024 Plans	Progresses
Crysvita	2.3	2.5	+0.2 (+7%)	Market penetration (Launched in Dec 2019)	12.9	19%
Poteligeo	0.4	0.4	+0.0 (+1%)		1.9	23%
Nesp + Nesp-AG[1]	4.2	3.5	-0.8 (-18%)	NHI price-cut & Biosimilars' penetration	14.4	24%
Nesp	0.8	0.7	-0.1 (-9%)		2.8	25%
Nesp-AG	3.5	2.8	-0.7 (-20%)		11.7	24%
Duvroq	1.8	2.5	+0.7 (+37%)	Market penetration (Launched in Aug 2020)	12.2	20%
Phozevel	-	0.6	+0.6 (- %)	Launched in Feb 2024	3.3	19%
Orkedia	2.2	2.2	-0.1 (-2%)		11.7	19%
G-Lasta	7.0	5.8	-1.3 (-18%)	Biosimilars' penetration	20.5	28%
Rituximab BS	2.2	1.9	-0.3 (-13%)	NHI price-cut	7.9	24%
Romiplate	2.7	3.0	+0.3 (+12%)	Market penetration (New indication in Jun 2019)	13.2	23%
Nouriast	1.7	1.5	-0.2 (-9%)		7.1	21%
Haruropi	0.9	1.0	+0.0 (+4%)		5.2	19%

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.

Revenue of Major Items (ex-Japan)

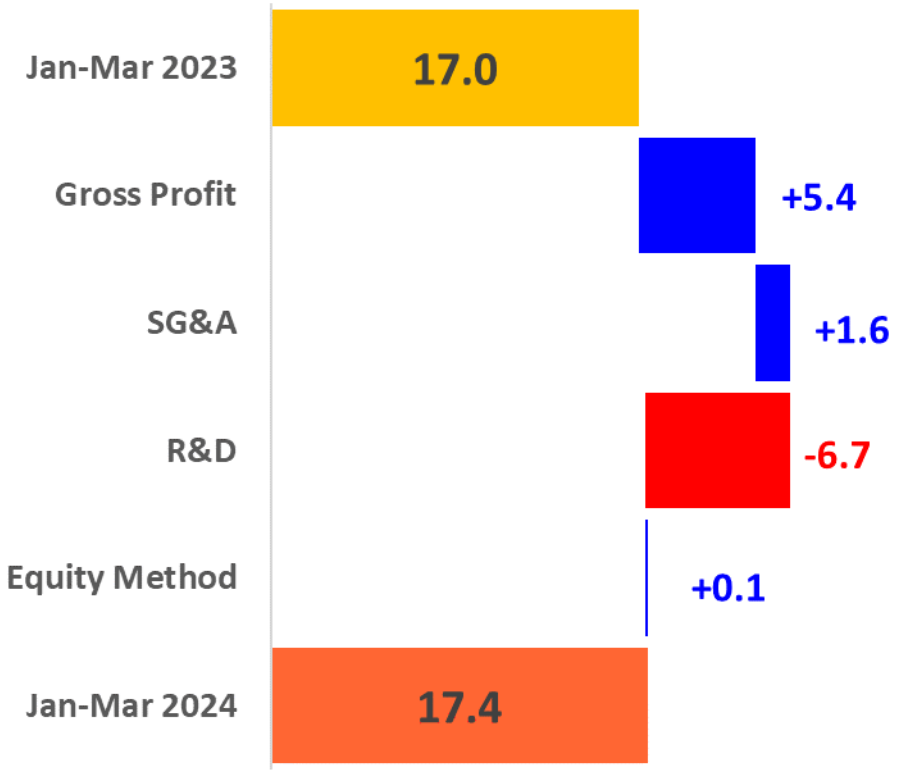
(Billion Yen / Rounded)

Item	2023Q1 Results	2024Q1 Results	Changes	Reasons	2024 Plans	Progresses
Crysvita	27.1	35.4	+8.2 (+30%)		175.9	20%
North America	18.8	22.8	+4.0 (+21%)	[North America] Market penetration [EMEA] Geographical expansion & Additional indication (Adult/TIO) [APAC] Geographical expansion		
EMEA	8.0	11.9	+3.9 (+49%)			
APAC	0.3	0.6	+0.4 (+144%)			
Poteligeo	5.8	8.2	+2.4 (+41%)		32.5	25%
North America	4.3	6.3	+1.9 (+44%)	[North America] Market penetration [EMEA] Geographical expansion & Market penetration	23.3	27%
EMEA	1.5	1.9	+0.4 (+29%)		8.8	22%
APAC	-	0.0	+0.0 (- %)		0.5	4%
Libmeldy / Lenmeldy	-	1.1	+1.1(- %)	New consolidation of Orchard (FDA approval in Mar 2024)	4.5	25%
Nourianz	1.7	1.6	-0.1 (-7%)	Market penetration	8.5	18%
Nesp	2.2	2.9	+0.7 (+32%)		10.7	27%
Gran	1.4	1.8	+0.3 (+24%)		7.2	25%
Tech-licensing	8.9	11.7	+2.8 (+31%)	Upfront revenue from Boehringer Ingelheim and growth of Fasenra	45.0	26%
Benralizumab Royalty[1]	5.7	6.4	+0.7 (+12%)			

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

YoY Analysis -Core OP-

+0.4 billion yen (incl. forex effect +2.1)



- **Gross Profit +5.4 (incl. forex effect +6.4)**
  Increased in conjunction with JPY12.0B rise in revenue. COGs have increased due to the North America Crysvita Sales royalty after Apr 27, 2023. Hence, gross profit % declined YoY. (80% →76%)

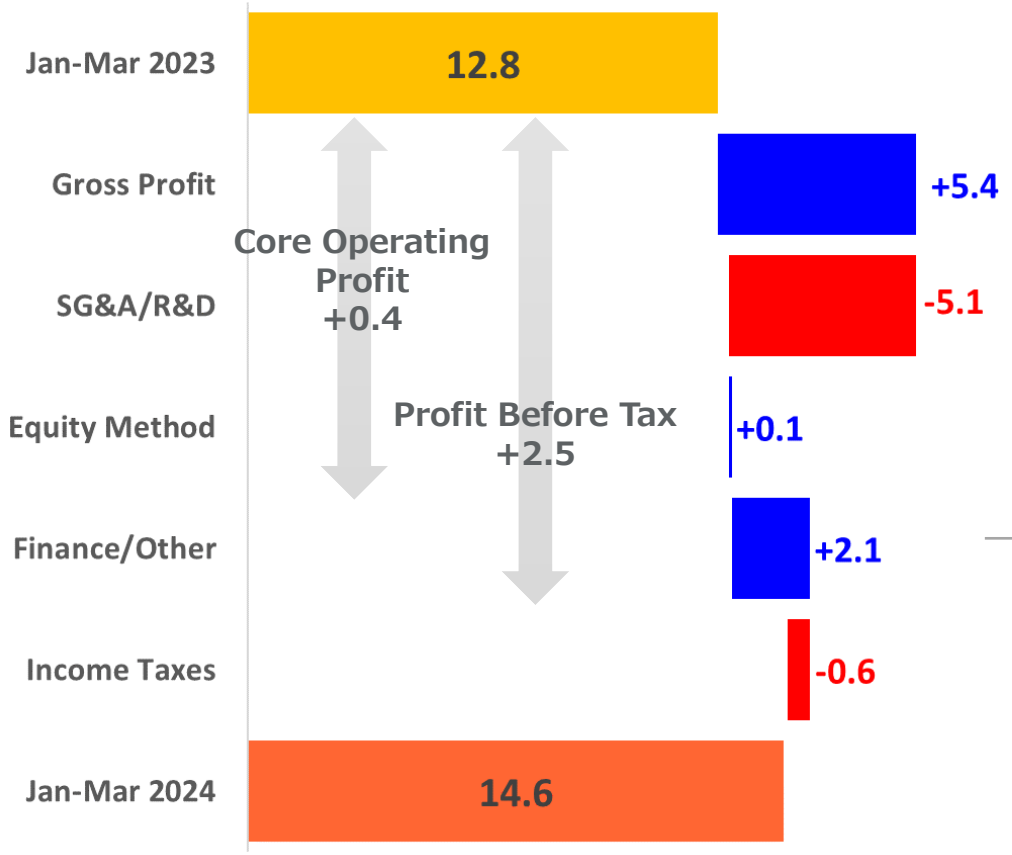
- **SG&A +1.6 (incl. forex effect -2.5)**
  While HR expenses and FX impact increased, SG&A decreased due to the North America Crysvita-related scheme change after Apr 27, 2023.
  [HR exp -3.0 / Sales promotion +7.0 (incl. Crysvita profit sharing expenses +7.6)]

- **R&D -6.7 (incl. forex effect -1.8)**
  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 +0.1**
  Despite the reversal of deferred tax assets, sales of Hulio (FKB327/Adalimumab biosimilar) continue to grow.
  FKB; Fujifilm Kyowa Kirin Biologics Co., Ltd.

YoY Analysis -Profit-

Profit (Jan-Mar) +1.9 billion yen



● **Finance/Other +2.1**

- Gain on disposal of fixed assets +2.4 (0 → 2.4)
- Impairment losses +0.9 (-1.1 → -0.2)
- Provision for loss on contracts +0.6 (-0.6 → -)
- Business restructuring expenses -1.8 (-0.5 → -2.3)

Commercial Update

2024 Key Actions & Q1 Topics

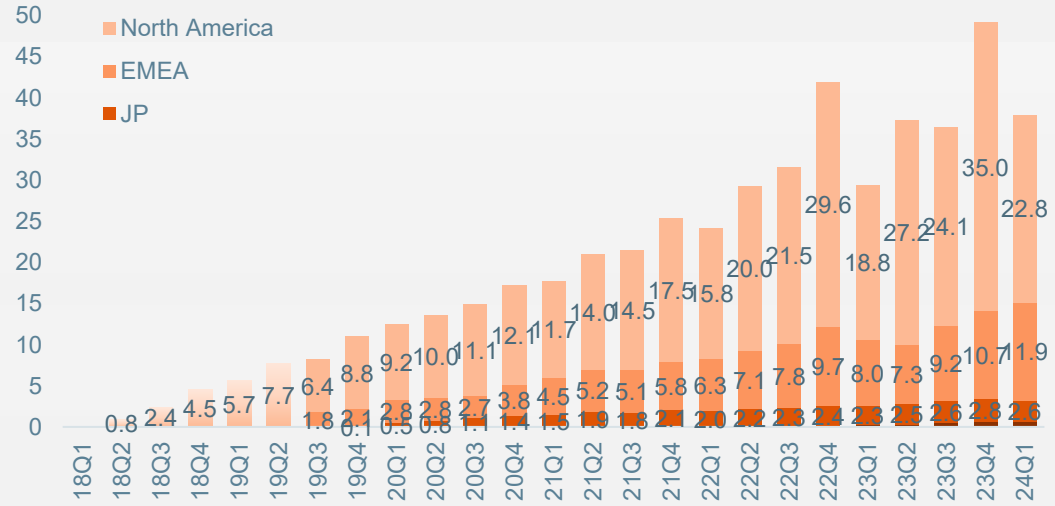
2024 Key Actions

- 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.

Q1 Topics

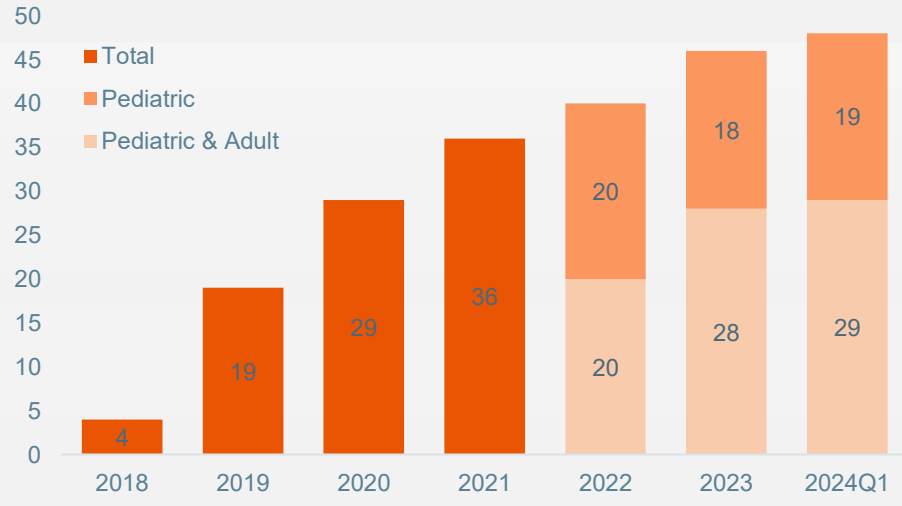
- Strengthen evidence-based marketing activities.
- North America
  - The number of patient enrollments in the treatment preparation stage and treatment patients continued to increase steadily.
  - Continued to strengthen patient support programs from diagnosis to treatment initiation.
  - Despite seasonal factors, revenue increased 21% YoY and were generally in line with plans.
- EMEA:
  - Revenue increased 49% YoY. Growth due to geographic expansion and increased patient penetration with the launch of sales for adult XLH compared to the same period last year.
- Japan:
  - Continued to strengthen promotional activities by the dedicated personnel.

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 (XLH)



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

2024 Key Actions & Q1 Topics

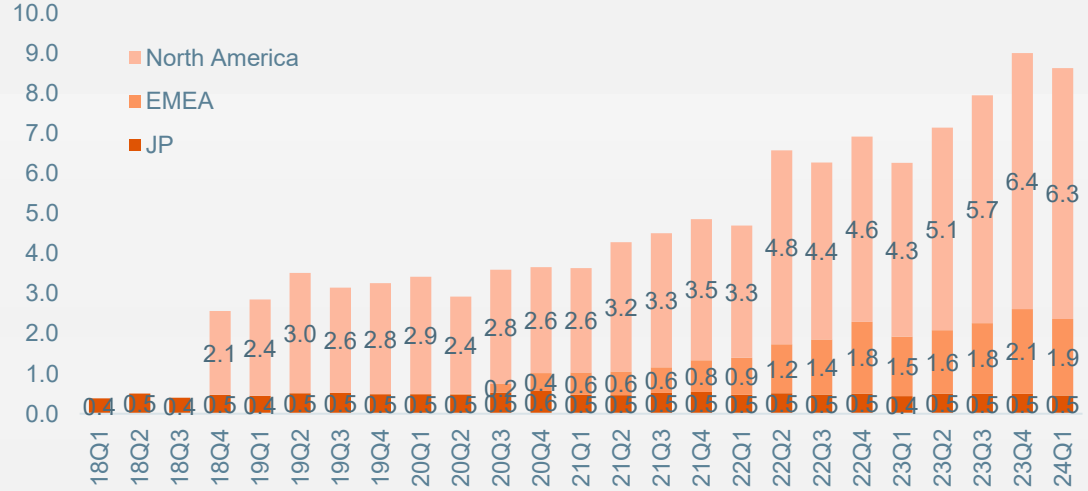
2024 Key Action

- 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 MF patients with visible skin symptoms (2nd half)
  - ◆ Geographic Expansion

Q1 Topics

- NA : Sales revenue increased by 44% YoY. Patient penetration has increased through enhanced customer & account focus.
- EMEA : Sales revenue increased by 29% YoY and generally in-line with plan.

Sales Revenue (Billion Yen)



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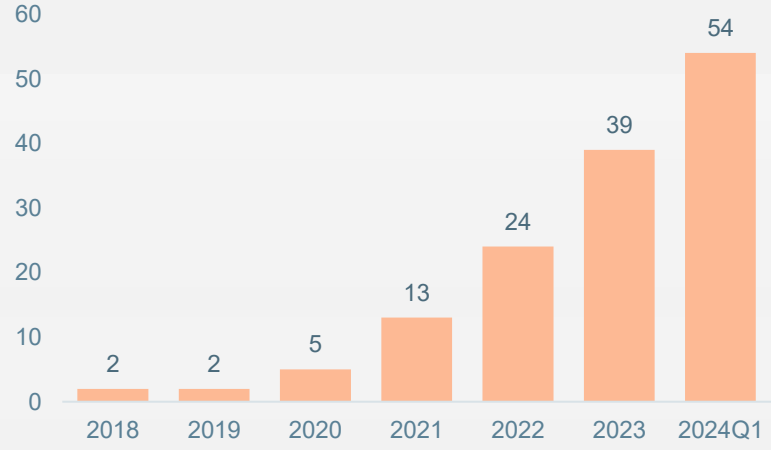
Launched Countries / Regions



R&D Update

News Flow of Main Development Pipeline Products

As of May 7, 2024

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[1]	Q2 2024
	Prurigo nodularis	P3 initiation	H2 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	Q2 2024
KK8123	XLH	P1 initiation	Q2-Q3 2024
Atidarsagene autotemcel (formerly OTL-200)	**MLD**	**US approval**	Mar. 2024
OTL-203	**MPS-IH (Hurler syndrome)**	**Registrational study[2]**	In progress
OTL-201	**MPS-IIIA (Sanfilippo syndrome type A)**	**PoC study[3] Data - Conference presentation**	Feb. 2024

1. Achieved First Site Activated; 2. Equivalent to P3 study; 3. Equivalent to P1/2 study.

News Flow in 2024

Year-to-date Key News Flow

As of May 7, 2024

Category	Date	Headline
PP	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
PP	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)
		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

Appendix

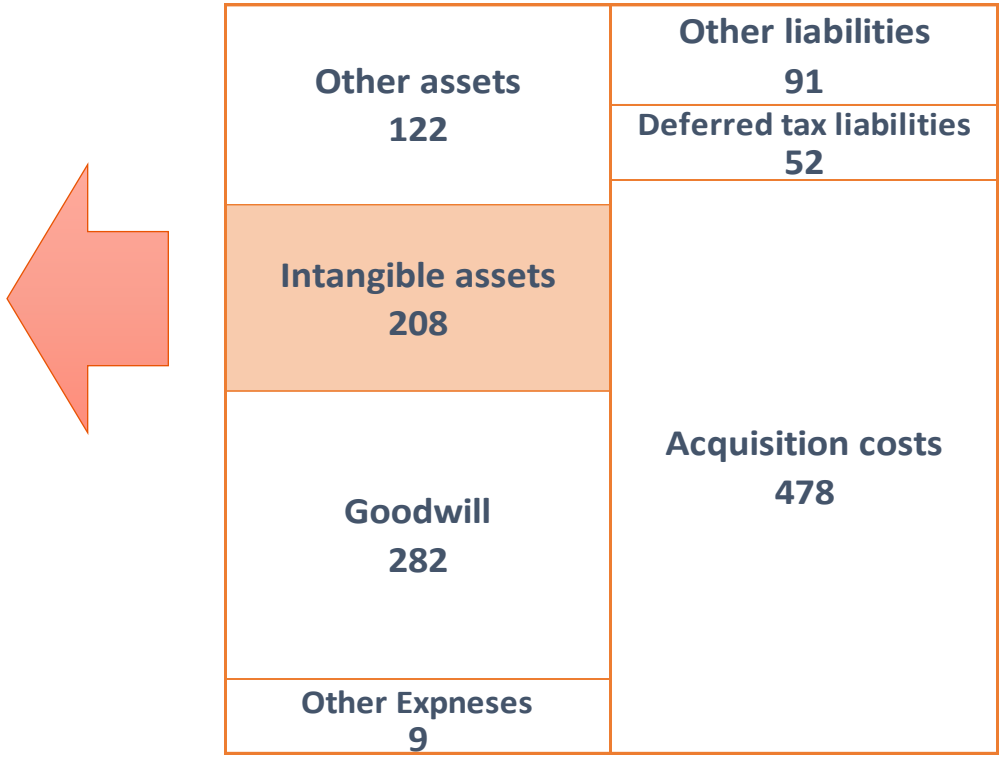
Accounting treatment of share acquisition of Orchard Therapeutics (Tentative)

- ✓ Completed the share acquisition on January 24, 2024, and started consolidation from the February 2024
- ✓ Recognized intangible assets of $208M and goodwill of $282M
- ✓ Intangible assets will be amortized over 20 years (19 years for Libmeldy/Lenmeldy)

【Breakdown of Intangible $208M】
- Libmeldy/Lenmeldy $118M
- OTL-203 $90M

【Annual amortization amount】
- Libmeldy/Lenmeldy $6M /year
  ⇒Amortization started from Feb 2024
- OTL-203 $4M /year
  ⇒To be amortized after market launch

(Unit: Million USD)



- ✓ The above is a tentative calculation while the purchase price allocation has not been completed as of the end of Q1 2024.
- ✓ The acquisition costs above ($478 million) include amounts for options, Restricted Stock Units and other instruments which are paid by Orchard. The acquisition costs under business combination accounting is $386 million (approximately 57.1 billion yen)

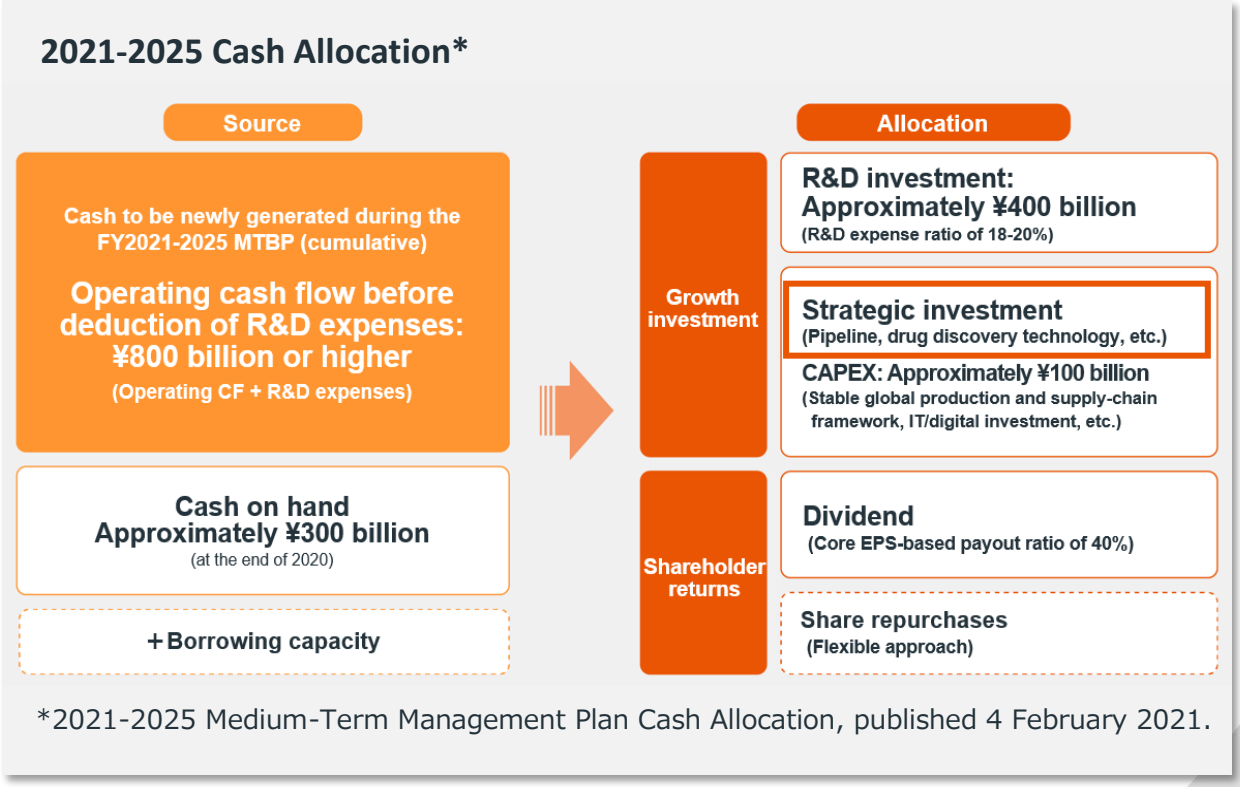
Strategic Investment ～For successful creation and delivery of life-changing value

Licensing-in and M&A investments to strengthen the portfolio

- Priority will be given to the focus disease areas
  - bone & mineral
  - intractable hematological diseases/hemato oncology
  - rare diseases

Investment in science and technology to create new strengths

- Investments aimed at acquiring new drug discovery technologies and early pipelines and accelerating cooperation and collaborations
- VC investment and CVC activities for exploring and accessing information.



*2021-2025 Medium-Term Management Plan Cash Allocation, published 4 February 2021.

Main Development Pipeline Products (After Ph2)

As of May 7, 2024

	Diseases under development[*1]	Planned Approval Year[*2]	Development status	Total addressable market[*3]	No. of Patients[*4]
KHK4083/AMG 451 rocatinlimab	Moderate and severe Atopic Dermatitis	2026/2027	P3 (Global)	★★★★★	16M
	Moderate and severe Asthma	TBD	Preparation underway for P2 (Global)	★★★★★	13.5M
	Prurigo nodularis	TBD	Preparation underway for 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	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 May 7, 2024

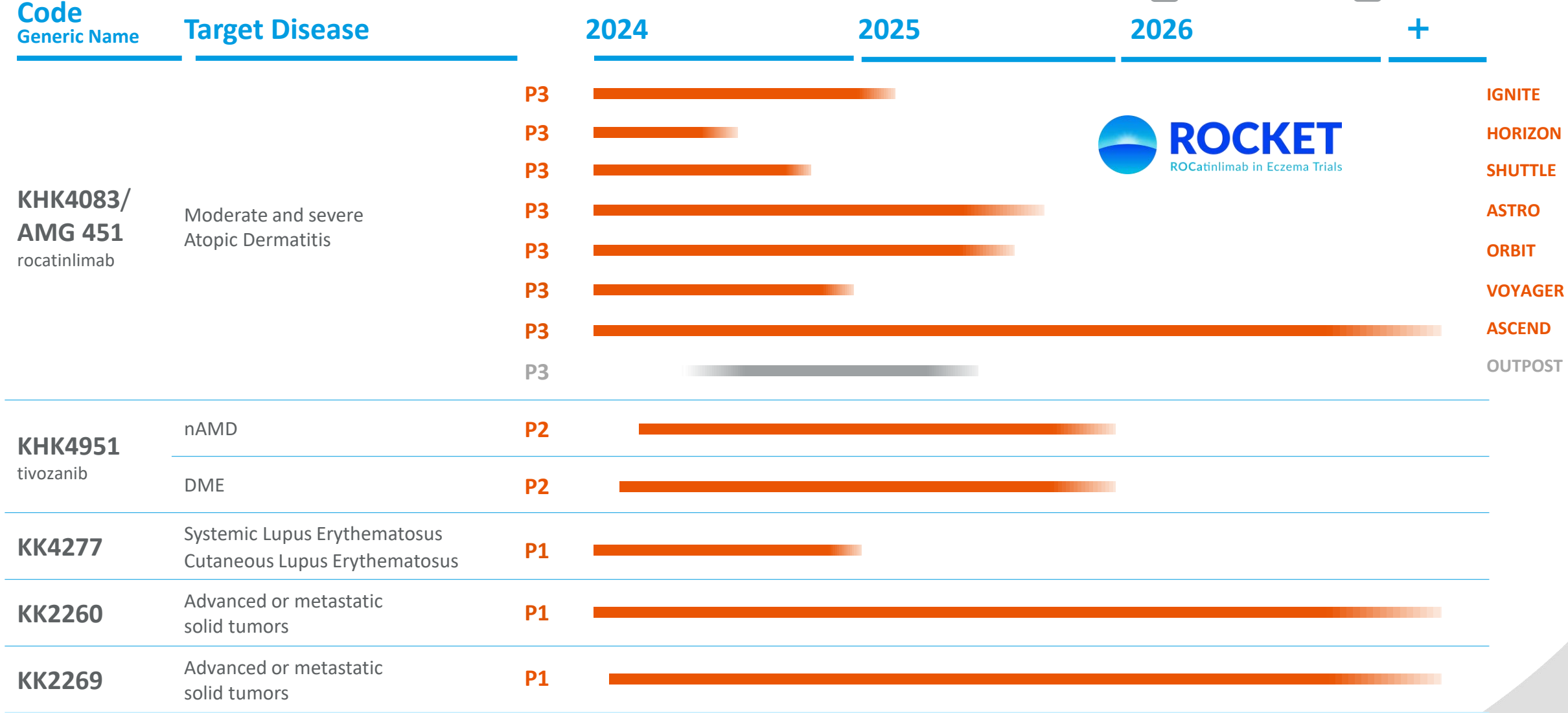
	Diseases under development*1	Development status	Modality, technology
KK4277	SLE, CLE	P1 (JP, Asia)	Antibody, POTELLIGENT®
KK2260	Advanced or metastatic solid tumors	P1 (JP: in progress, US: in preparation)	Antibody, REGULGENT™
KK2269	Advanced or metastatic solid tumors	P1 (JP, US)	Antibody, REGULGENT™
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 May 7, 2024

T : Topline data　D : Detailed data



FOREX Information

Average FOREX Rates (yen)

	2023Q1	2024Q1	Changes	2024 Plans
USD	132	147	+15	140
GBP	161	187	+26	180
EUR	141	160	+19	155

Q1 YoY FOREX Impacts (billion yen)

	Revenue	Core OP
USD	+4.2	+1.0
GBP	+0.4	-0.5
EUR	+1.6	+1.0

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

Crysvita - Collaboration with Ultragenyx -

	Economic Terms
US & Canada	• Kyowa Kirin books sales • 50/50 profit share for 5 years from the U.S. launch - Supply price: 35% of net sales through 2022, 30% thereafter (No impact on the sales royalties stated below) • After 5 years (April 27, 2023-), Kyowa Kirin pays tiered sales royalties in mid-high 20% range to Ultragenyx *Ultragenyx has sold 30% of its royalty interest, subject to a 1.45x cap, to OMERS Capital Markets
Europe	• Kyowa Kirin books sales • Kyowa Kirin pays sales royalties in up to 10% range to Ultragenyx *Ultragenyx has sold its royalty interest, subject to a 1.9x or 2.5x cap depending on when the cap is achieved, to Royalty Pharma
Latin America	• Ultragenyx books sales • Kyowa Kirin receives low single-digit sales royalties from Ultragenyx • Supply price: 35% of net sales through 2022, 30% thereafter
Turkey	• Ultragenyx books sales • Kyowa Kirin receives sales royalties in up to 20% range from Ultragenyx
Asia & Others	• Kyowa Kirin books sales

* Kyowa Kirin supplies commercial products in all territories.

KHK4083/AMG 451 - Collaboration with Amgen -

	US	Europe & Asia (ex. JP)	JP
Development	• Amgen leads development • Share development cost	• Amgen leads development • Share development cost	• Kyowa Kirin leads development
Commercialization	• Amgen commercializes and books sales • Kyowa Kirin co-promotes and shares promotion cost	• Amgen commercializes and books sales • Kyowa Kirin has opt-in rights for co-promotion	• Kyowa Kirin commercializes and books sales
Sales Royalties	• Double-digit royalty to Kyowa Kirin	• Double-digit royalty to Kyowa Kirin	
Commercial supply	• Amgen supplies	• Amgen supplies	• 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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