Results Presentation
Fiscal 2023 Third Quarter

協和キリン株式会社

Agenda

Financial Review	Managing Executive Officer, CFO Head of Finance Motohiko Kawaguchi
Commercial Update	Executive Officer, Head of Global Product Strategy Tomohiro Sudo
R&D Update	Executive Officer, Head of R&D Yoshifumi Torii, Ph.D.
News Flow in 2023	Managing Executive Officer, CSO Head of Strategy Yasuo Fujii

Q&A	Managing Executive Officer, CFO Head of Finance Motohiko Kawaguchi
	Managing Executive Officer, CSO Head of Strategy Yasuo Fujii
	Executive Officer, Head of R&D Yoshifumi Torii, Ph.D.
	Executive Officer, Head of Global Product Strategy Tomohiro Sudo

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

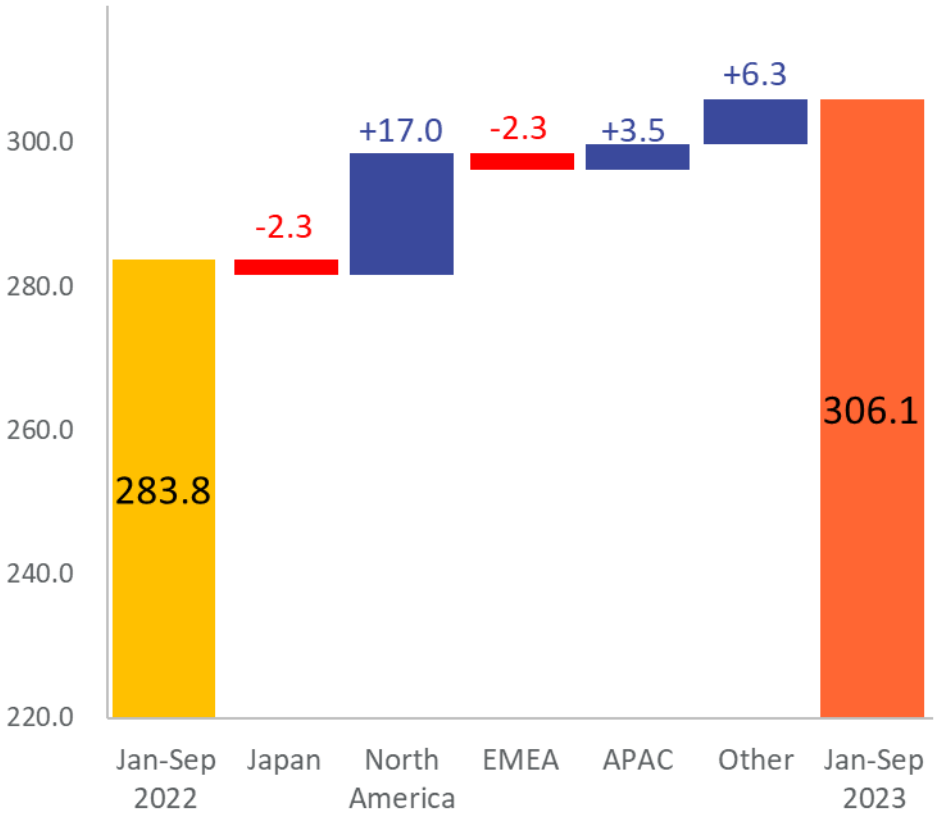
Summary of Q3 Results

(Billion Yen / Rounded)

	2022Q3 Results	2023Q3 Results	Changes	2023 Revised Plans	Progresses
Revenue *[Overseas Ratio]*	283.8 *[61%]*	306.1 *[64%]*	+22.3 (+8%)	426.0 *[64%]*	72%
Gross Profit *[Gross Profit Margin]*	219.6 *[77%]*	229.1 *[75%]*	+9.4 (+4%)	326.0 *[77%]*	70%
SG&A *[SG&A Ratio]*	117.3 *[41%]*	119.3 *[39%]*	+2.0 (+2%)	162.0 *[38%]*	74%
R&D *[R&D Ratio]*	44.1 *[16%]*	51.2 *[17%]*	+7.0 (+16%)	79.0 *[19%]*	65%
Gain/Loss on Equity Method	2.6	2.3	-0.4 (-14%)	3.0	76%
Core Operating Profit *[Core OP Margin]*	60.9 *[21%]*	60.9 *[20%]*	+0.0 (+0%)	88.0 *[21%]*	69%
Profit	49.2	53.6	4.3 (+9%)	76.0→70.0	77%

YoY Analysis -Revenue-

+22.3 billion yen
(incl. forex effect +13.9)



- **Japan -2.3**
  Although Duvroq, Romiplate, and Crysvita increased, revenue in Japan region decreased by 2% due mainly to negative impact by annual NHI price-cut and shrink in Nesp-AG affected by competitive products.

- **North America +17.0 (incl. forex effect +7.6)**
  Revenue in North America region increased by 22% with the growth of Crysvita(+22%), Poteligeo(+20%), and Nourianz(+23%).

- **EMEA -2.3 (incl. forex effect +3.6)**
  Revenue in EMEA region decreased by JPY2.3B due to 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, despite the continued growth of Crysvita(+15%), and Poteligeo(+37%).

- **APAC +3.5 (incl. forex effect +1.0)**
  APAC revenue increased by 15% with the growth of Crysvita, and Nesp, while Gran was down due to the Chinese national tender system.

- **Other +6.3 (incl. forex effect +1.8)**
  23% growth in the other revenue was due to the royalties of growing Fasenra (Benralizumab).

Revenue of Major Items (Japan)

(Billion Yen / Rounded)

Item	2022Q3 Results	2023Q3 Results	Changes	Reasons	2023 Plans	Progresses
Nesp + Nesp-AG[1]	15.7	12.7	-3.1 (-20%)	NHI price-cut & Biosimilars' penetration	16.6	76%
Nesp	2.5	2.3	-0.2 (-8%)		2.8	82%
Nesp-AG	13.2	10.3	-2.9 (-22%)		13.8	75%
Duvroq	4.4	6.9	+2.4 (+54%)	Market penetration (Launched in Aug 2020)	7.8	88%
Orkedia	7.5	7.6	+0.1 (+1%)		11.2	68%
G-Lasta	22.7	23.2	+0.5(+2%)		33.5	69%
Poteligeo	1.5	1.4	-0.0 (-1%)		2.0	73%
Rituximab BS	7.6	6.7	-0.9 (-12%)	NHI price-cut	8.7	77%
Romiplate	7.5	8.7	+1.2 (+16%)	Market penetration (New indication in Jun 2019)	11.2	77%
Allelock	4.8	4.1	-0.6 (-13%)	NHI price-cut	4.7	88%
Nouriast	5.9	5.5	-0.3 (-6%)		7.5	73%
Haruropi	2.8	3.2	+0.4 (+15%)	Market penetration (Launched in Dec 2019)	4.7	68%
Crysvita	6.4	7.4	+1.0 (+16%)	Market penetration (Launched in Dec 2019)	11.1	67%

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)

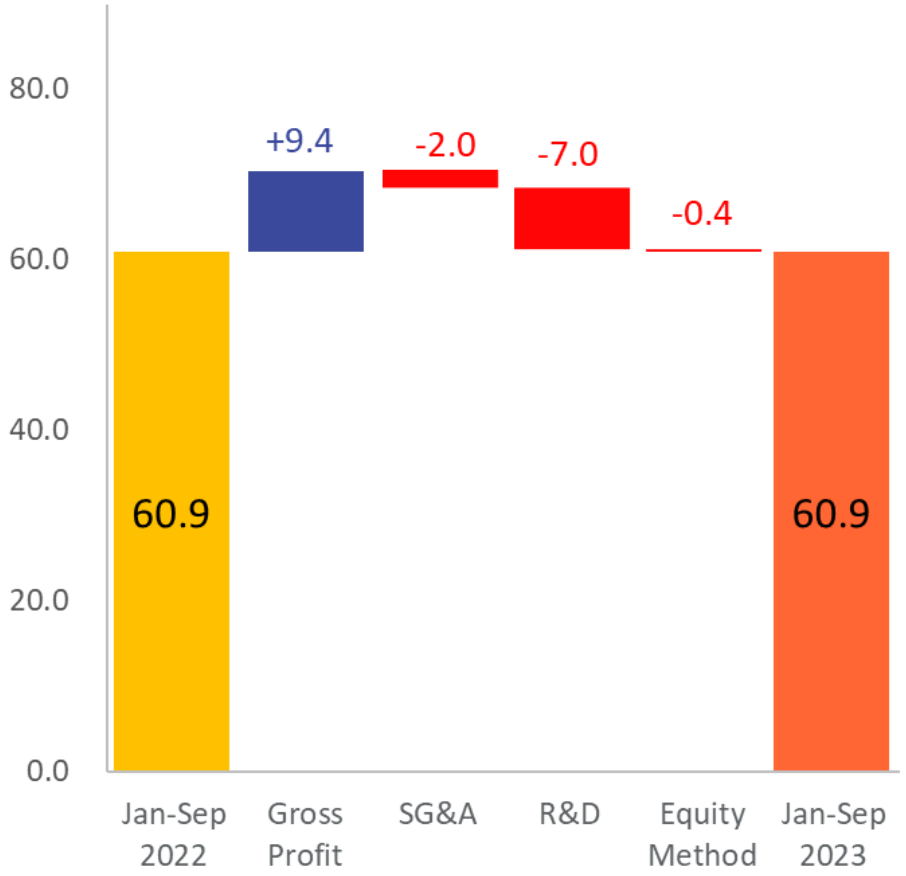
(Billion Yen / Rounded)

Item	2022Q3 Results	2023Q3 Results	Changes	Reasons	2023 Plans	Progresses
Crysvita	78.7	95.7	+17.0 (+22%)		138.0	69%
North America	57.4	70.2	+12.8 (+22%)	[North America] Market penetration [EMEA] Geographical expansion & Additional indication (Adult/TIO) [APAC] Geographical expansion		
EMEA	21.2	24.5	+3.2 (+15%)			
APAC	0.1	1.1	+0.9 (+709%)			
Poteligeo	16.1	19.9	+3.8 (+24%)		27.5	72%
North America	12.6	15.1	+2.5 (+20%)	[North America] Market penetration [EMEA] Geographical expansion & Market penetration	19.4	78%
EMEA	3.5	4.8	+1.3 (+37%)		8.0	61%
APAC	-	0.0	+0.0 (- %)		0.2	3%
Nourianz	4.5	5.5	+1.0 (+23%)	Market penetration	7.5	73%
Nesp	5.9	7.0	+1.2 (+20%)		8.0	89%
Gran	6.4	5.2	-1.2 (-19%)	Listed on Chinese tender list	8.2	63%
Neulasta	4.4	4.5	+0.1 (+2%)		5.7	79%
Tech-licensing	23.3	29.3	+6.0 (+26%)	Growth of Fasenra	39.0	75%
Benralizumab Royalty[1]	15.4	19.1	+3.8 (+25%)			

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

YoY Analysis -Core OP-

+0.0 billion yen
(incl. forex effect +4.2)



- **Gross Profit +9.4 (incl. forex effect +12.0)**
  Increased in conjunction with JPY22.3B rise in revenue. COGs have increased due to the North America Crysvita Sales royalty after Apr 27, 2023. Hence, gross profit % declined YoY. (77% →75%)

- **SG&A -2.0 (incl. forex effect -6.2)**
  Although decreased in Crysvita profit sharing expenses due to the North America Crysvita-related scheme change after Apr 27, 2023, increased in HR exp, etc by the Crysvita commercial operation. In addition to that, increased due to FX impact.
  [HR exp -7.1 / Sales promotion +9.1 (incl. Crysvita profit sharing expenses +11.3)]

- **R&D -7.0 (incl. forex effect -1.8)**
  Increased in clinical study costs of KHK4083 which is undergoing joint global Phase III clinical study.

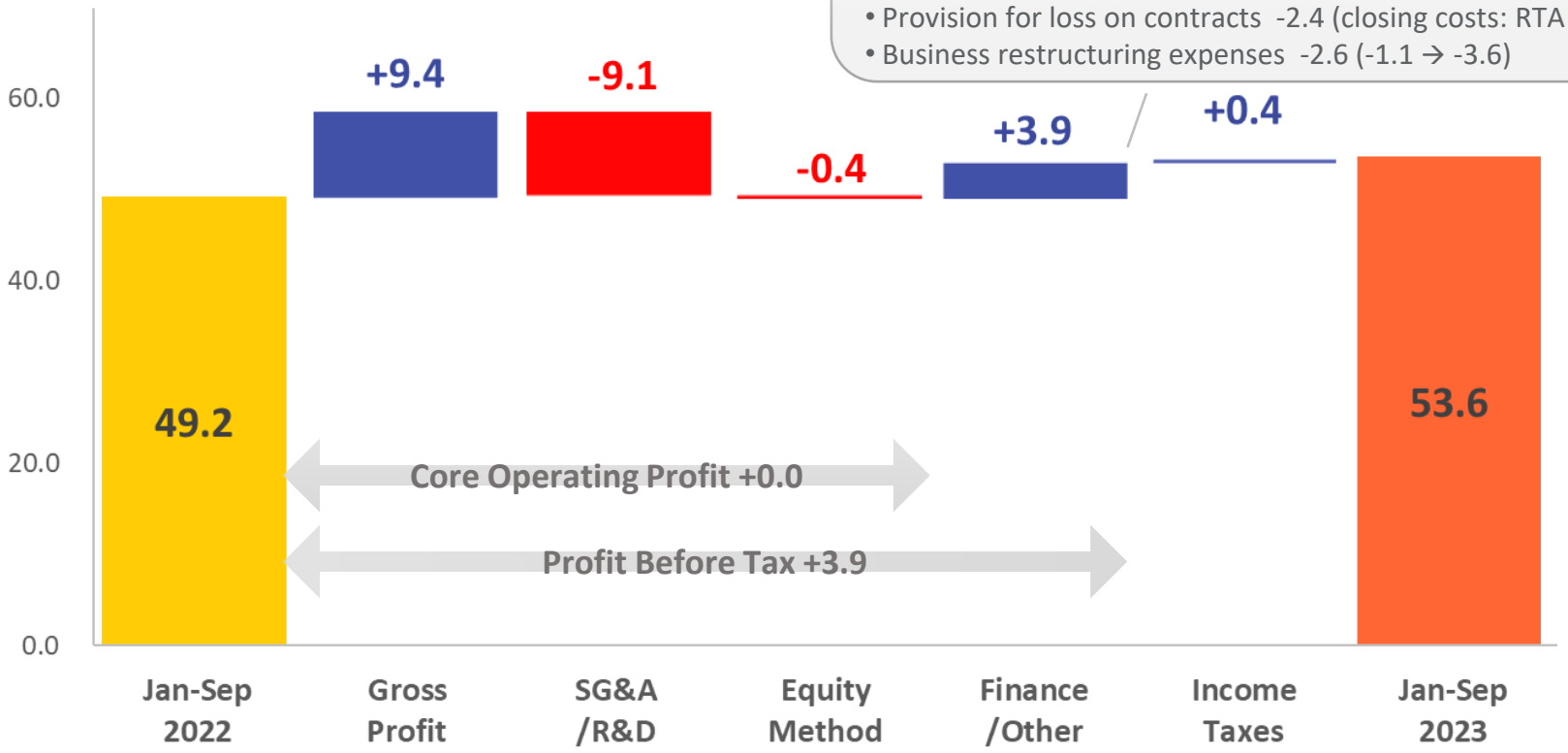
- **Gain/Loss on Equity Method -0.4**
  While revenue of Hulio (FKB327/Adalimumab biosimilar) increased, FKB's profit declined due to decrease in tax-accounting effect.

FKB; Fujifilm Kyowa Kirin Biologics Co., Ltd.

YoY Analysis -Profit-

Profit (Jan-Sep) +4.3 billion yen

- EMEA established medicines portfolio:
  Gain on sales of 51% shares plus gain on valuation of remaining 49% shares +14.8
- Impairment losses -6.2
  [FY22: irofotase alfa IPR&D -3.2]
  [FY23: RTA 402 IPR&D -8.3, Expiration of research collaboration with Ardelyx -1.1]
- Provision for loss on contracts -2.4 (closing costs: RTA 402 -1.5, ME-401 -0.8)
- Business restructuring expenses -2.6 (-1.1 → -3.6)



Commercial Update

Coordinated Actions to Maximize the Patient Access to G3B

2023 Key Actions & Q3 Topics

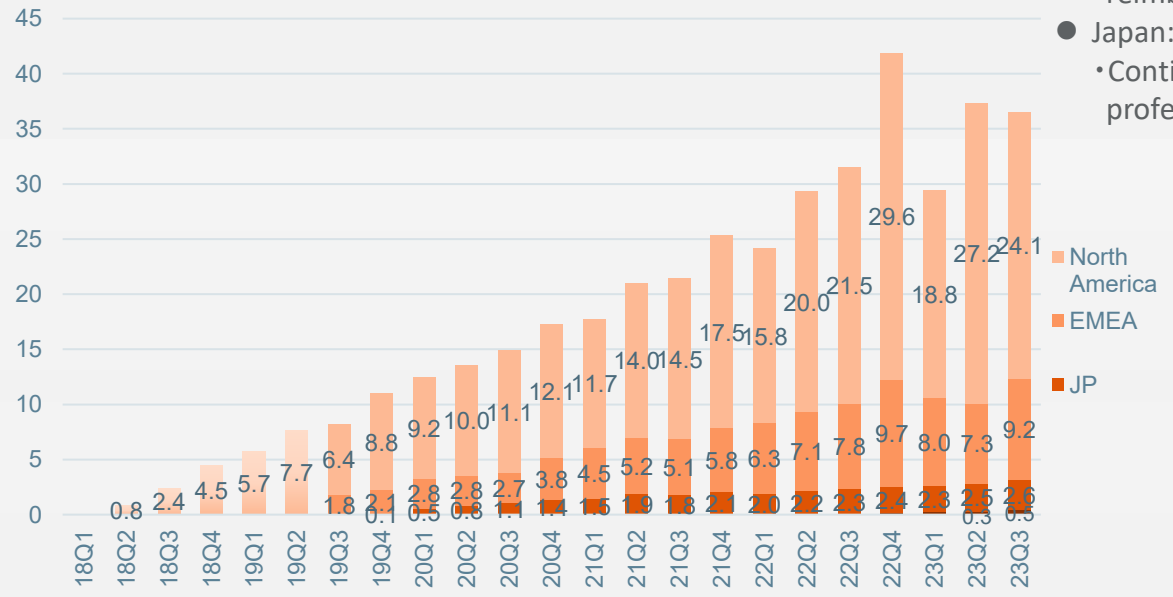
2023 Key Actions

- North America:
  Start own sales (Establish and start own operation of the direct sales force).
- EMEA:
  Continue to focus on geographical & indication expansion.
- Japan:
  Strengthen promotional activities centered by the dedicated personnel.

Q3 Topics

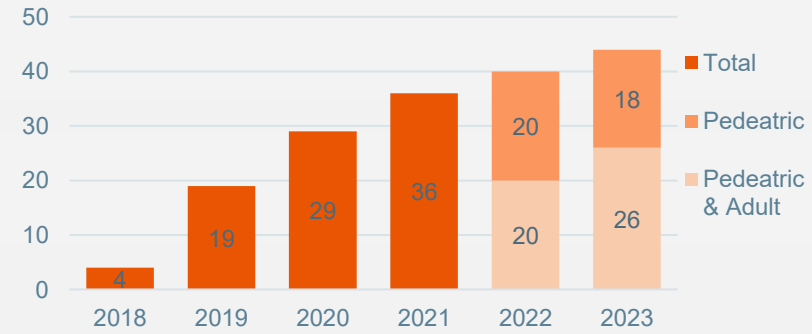
- North America:
  - The number of patient enrollments in the treatment preparation stage and treatment patients continued to increase steadily, through addressing the individual issues in the sales transfer.
  - As for sales revenue, estimated allowances for returns corresponding to the inventory of Ultragenyx-labeled products as of end of September was recorded due to the switch from Ultragenyx-labeled products to Kyowa Kirin-labeled products.
- EMEA:
  - Patient penetration in each country has been steadily increased.
  - Revenues increased 15% YoY, despite the impact of the reduction of insurance reimbursement prices in Germany.
- Japan:
  - Continued to strengthen promotional activities, such as holding webinars for medical professionals.

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.

POTELIGEO® (mogamulizumab)

2023 Key Actions & Q3 Topics

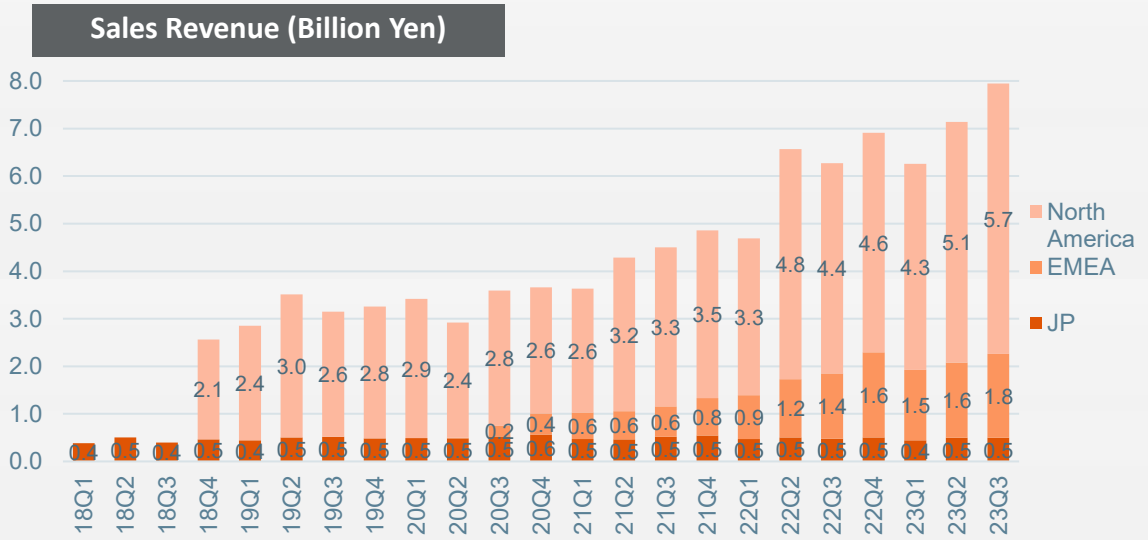
2023 Key Actions

- Strengthen promotional activities utilizing evidences including efficacy in blood involvement.
- Raise awareness of importance of blood testing among early-stage patients

Q3 Topics

- North America: Sales revenue has been growing steadily by virtue of promotional activities for raising awareness of importance of earlier consultation with specialists and blood testing.
- EMEA: Shipping quantity grew steadily, and sales revenue increased 37% YoY. On the other hand, due to specific issues in each countries, such as sales price or duration of medication, it is slightly below the annual plan. Began reorganization, etc., to make the organization suitable more efficient marketing activity.

*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.



NOURIANZ® (istradefylline) tablets

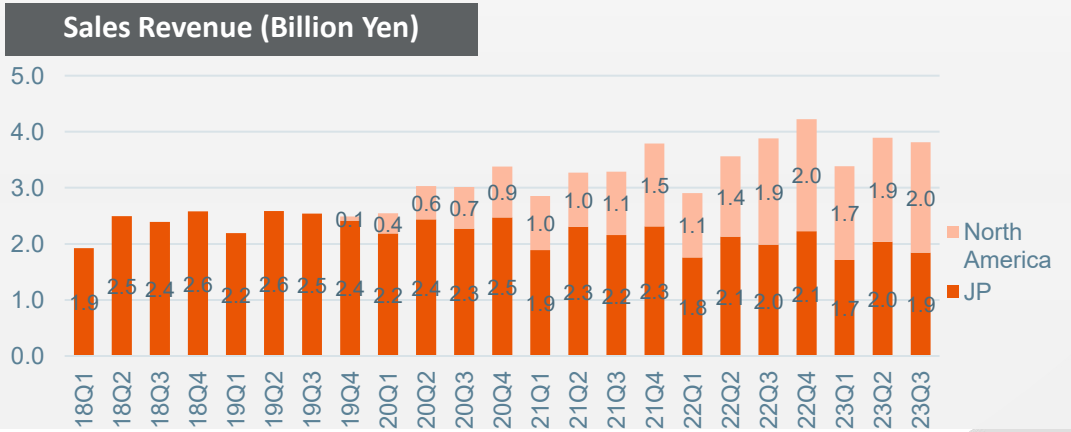
2023 Key Actions & Q3 Topics

2023 Key Actions

- Further instill importance of adenosine A_{2A} receptor antagonism in treating wearing-off.
- Strengthen field level activity through further collaboration and knowledge sharing between Japan and the US, and through maximizing available resources by utilizing effective approaches including digital.

Q3 Topics

- US: Sales increased YoY. Strengthen cross-functional execution on earlier use in adjunctive setting supported by our unique MOA. Continue effort on reducing patient Rx abandonment rate in specialty pharmacies.



R&D Update

Upcoming Events: Main Development Pipeline Products

As of November 1, 2023

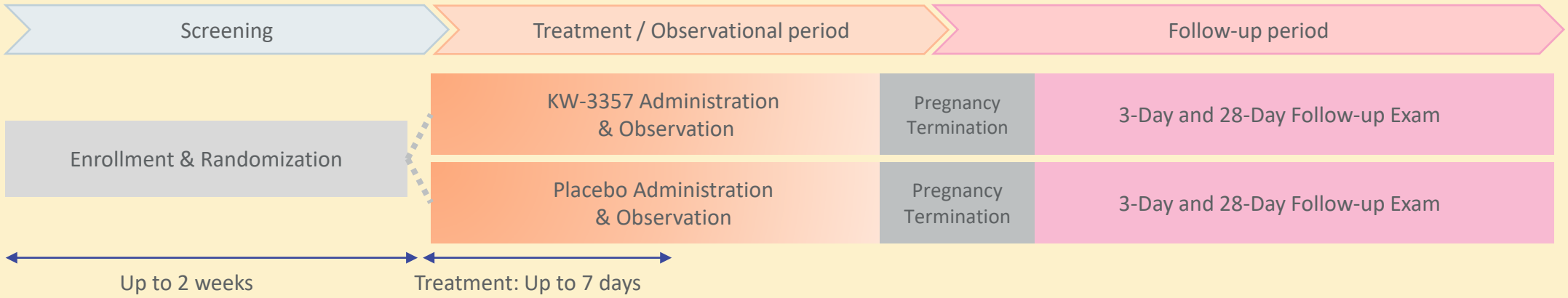
Code Generic Name	Events (Completed are in bold)		Timeline (Completed are in orange)
KHK4083/AMG 451 rocatinlimab	**Atopic Dermatitis**	**P3 (ROCKET PROGRAM)**	In Progress
	Asthma	P2	To be initiated
KHK4951 tivozanib	**nAMD**	**P1 data presentation**	Oct. 2023
	nAMD	P2 initiation	Q4 2023
	DME	P2 initiation	Q4 2023
KK2260	Advanced or metastatic solid tumors	P1 initiation	Q4 2023
KK2269	Advanced or metastatic solid tumors	P1 initiation	Q1 2024
KW-3357 antithrombin gamma (genetical recombination)	**Discontinuation of development for Preeclampsia**		Nov. 2023

Cf.) Orchard Therapeutics' development pipelines

OTL-200	MLD[1]	FDA decision to be announced	Mar. 2024[2]
OTL-203	MPS-IH[3]	Ph3 Study Initiation	Q4 2023
OTL-201	MPS-IIIA[4]	Ph1/2 Data readout	Q1 2024

1. Metachromatic Leukodystrophy; 2. PDUFA date: Mar. 18, 2024; 3. Mucopolysaccharidosis type I, Hurler syndrome; 4. Mucopolysaccharidosis type IIIA (Sanfilippo Syndrome type A)

KW-3357 Preeclampsia Ph3 data



■ Primary Endpoint：Days of maintaining pregnancy (Days from the start date of investigational drug to the date of pregnancy termination)		The trend toward prolongation was observed compared with the placebo group, but no statistically significant improvement was observed
■ Safety	Amount of blood lost during delivery	No significant difference was observed compared with placebo
	Bleeding-related events and anemia	Occurred 3 times more frequently than in the placebo group*

* Bleeding-related events: KW-3357 28%, Placebo 9%　Anemia: KW-3357 27%, Placebo 10%

Due to the failure to meet the primary endpoint and the higher incidence of bleeding-related events and anemia in KW-3357 group, we have decided to discontinue the development of this product for preeclampsia.

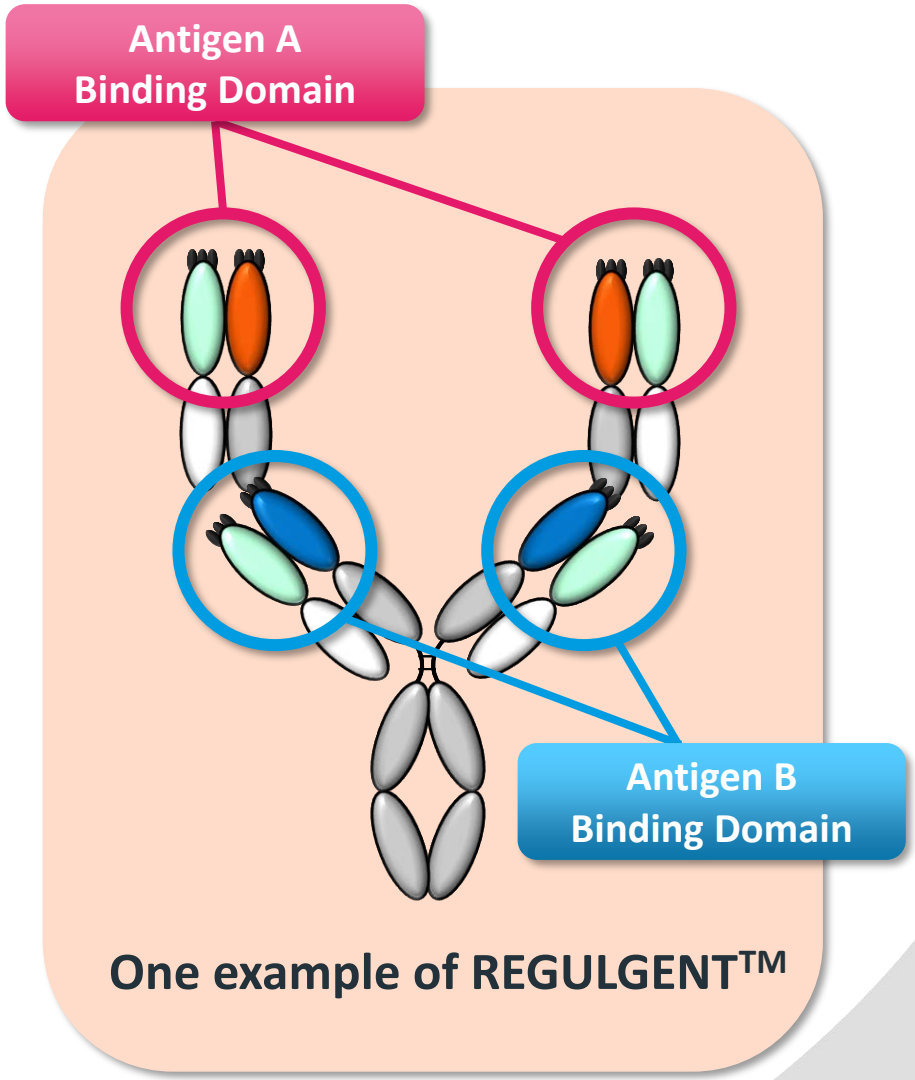
Overview of KK2260 and KK2269

- Both are Kyowa Kirin's proprietary bispecific antibodies with REGULGENT™ technology
- The phase 1 studies information for both products have been disclosed on jRCT*

* Trial ID KK2260: jRCT2031230372, KK2269: jRCT2031230419

	KK2260	KK2269
Countries/ Regions	Global (Japan and US for Phase 1)	
Indications	Advanced or metastatic solid tumors (esophageal cancer, head and neck cancer, etc.)	Advanced or metastatic solid tumors (gastric cancer, lung cancer, etc.)
Method	Single	Single or combination

Further details will be presented in our R&D Meeting (2023 December)



One example of REGULGENT™

News Flow in 2023

Year-to-date Key News Flow

As of November 1, 2023

Category	Date	Headline
ESG	Mar 8	Kyowa Kirin Selected for a “Health & Productivity Stock” and awarded as a “Certified Health & Productivity Management Outstanding Organization”(White 500) (Japan)
R&D	Mar 17	Presented New data from Phase 2b clinical study of Rocatinlimab in Atopic Dermatitis at the American Academy of Dermatology Annual Meeting 2023
SCM	Mar 28	Completed construction of a new building at Ube Plant (Japan)
ESG	Apr 6	Introduced RE100 renewable electricity to all purchased electricity at its two plants and three laboratories (Japan)
SCM	Apr 7	Completed construction of a multipurpose facility relating to Quality Assurance (Q-Tower) at Takasaki Plant (Japan)
R&D	Apr 27	Started collaboration in drug discovery technology with School of Life Science and Technology, Tokyo Institute of Technology (Japan)
R&D	May 10	Announced Phase III Study Results of bardoxolone methyl (RTA 402) in Japan and Discontinuation of Development (Japan)
LCM	Jun 23	Approval for partial change of Antineoplastic Mitomycin C Agent. (Japan)
LCM	Jul 18	Launched of Topical Ophthalmic Mitomycin C Agent and resumed the supply of Antineoplastic Mitomycin C Agent. (Japan)
LCM	Jul 24	Application 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. (Japan)

ESG: environmental, social, and governance; LCM: lifecycle management; R&D: research and development; SCM: supply chain management; BS: business strategy

Year-to-date Key News Flow

As of November 1, 2023

Category	Date	Headline
LCM	Aug 23	Approval for partial change of LUMICEF® for Palmoplantar Pustulosis (Japan)
LCM	Aug 30	Approval for Calcimimetics Agent ORKEDIA® TABLETS 4mg (Japan)
LCM	Sep25	Approval for Partial Change of Romiplate® for Aplastic Anemia (Japan)
R&D	Sep 25	Approval of PHOZEVEL® for Improvement of Hyperphosphatemia in Chronic Kidney Disease Patients on Dialysis (Japan)
R&D	Sep 28	Disclosing Top-Line Results of Phase 3 Clinical Study of KW-3357 for the Treatment of Preeclampsia (Japan)
LCM	Sep 29	Approval for Partial Change of Rituximab Biosimilar Received by Sandoz (Japan)
SI	Oct 5	Conclusion of Agreement to acquire Shares of UK biopharmaceutical company Orchard Therapeutics plc
R&D	Oct 11	Presented the posthoc analysis data from the Phase 2b study of rocatinlimab (AMG 451/KHK4083) at European Academy of Dermatology and Venereology (EADV) Congress 2023
R&D	Oct 17	Presented the Results of Phase 3 Studies of PHOZEVEL® at the American Society of Nephrology Meeting (ASN Kidney Week 2023)

Kyowa Kirin and Grünenthal have entered into a Joint Venture Collaboration for Kyowa Kirin International's 13 established medicines portfolio on Aug 1.

The transfer of global rights for Tostran, an established medicine, from Kyowa Kirin International to ADVANZ PHARMA has been completed on October 13.

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

Acquire shares of Orchard Therapeutics ~Strategic Rationale~

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.

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

Our purpose of the acquisition:

- To enrich our pipeline to address the UMNs** for which there is still no cure
- To obtain capabilities in Cell Gene Therapy R&D to address the future UMNs

** Unmet medical needs

- Strengthen the business following Crysvita and Poteligeo as a Global Specialty Pharmaceutical company
- Enhance ability to address UMNs in the future by combining with our strength in biologics
- Commitment to life by providing not only pharmaceuticals but also treatments

Acquire shares of Orchard Therapeutics ~Transaction Summary~

Items	Summary
Target	■ Orchard Therapeutics plc (London) — Listed on NASDAQ
Purchase Price*	■ $16.00 per ADS / approx. $387.4 million (approx. JPY 57.3 billion) — Orchard shareholders will hold additional contingent value rights (CVR) of $1.00 per ADS. — Additional $1.00 CVR will be paid for a total of $17.00 per ADS, or approximately $477.6 million (approx. JPY 70.7 billion) if OTL-200 is approved by the U.S. Food and Drug Administration for the commercial marketing and sale in the U.S.
Funding Method	■ Cash on balance sheet
Financial Impact	■ To be announced once allocation of goodwill and intangible assets are determined
Transaction Structure and Process	■ Scheme of Arrangement (SoA) — Requires the approval by Orchard's shareholder meeting, UK court, and regulatory authorities — Closing is expected in 2024Q1 through implementation of SoA

*Refers to the amount required to make payments related to all outstanding Orchard common shares, ADS purchases, options, Restricted Stock Units, and other instruments.

Acquire shares of Orchard Therapeutics ~Expected Synergy~

Further Development as a Global Specialty Pharmaceutical Company

- To be a Japan-based Global Specialty Pharmaceutical Company providing life-changing value to high unmet medical needs in concert with our existing business through Crysvita and Poteligeo

Reinforcement of New Drug Discovery & Development

Kyowa Kirin

- Experience and expertise of R&D and commercialization in Biologics and Antibody Drugs
- Proprietary next-generation antibody technology
- Continuous efforts to new modalities

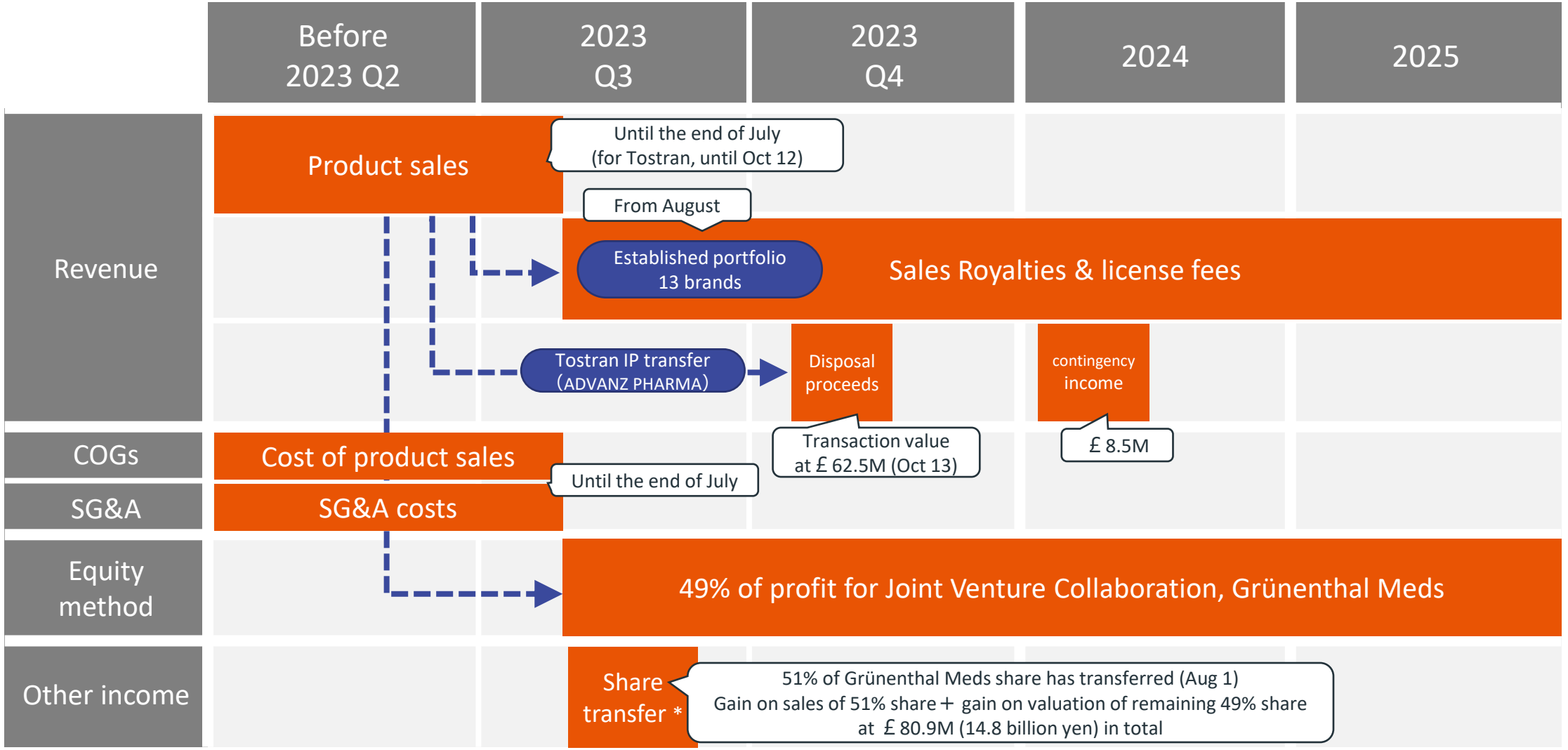
Orchard Therapeutics

- Experience and know-how in the marketing HSC-GT* in EU
- High technology in HSC-GT[1] (Research, CMC, SCM, etc.)
- Patient access and collaboration with medical institutions
- Efforts and experience to new modalities

* Hematopoietic Stem Cell Gene Therapy

- A key step toward delivering advanced value to patients
  - Pursuing the potential for “One-time treatment in life”
  - Challenge to correct the underlying cause of a genetic disease
  - Personalized medicine / Precision medicine
  - Providing treatment beyond the existing drugs
- Address a broader range of UMNs

P/L Impact on EMEA established medicines portfolio

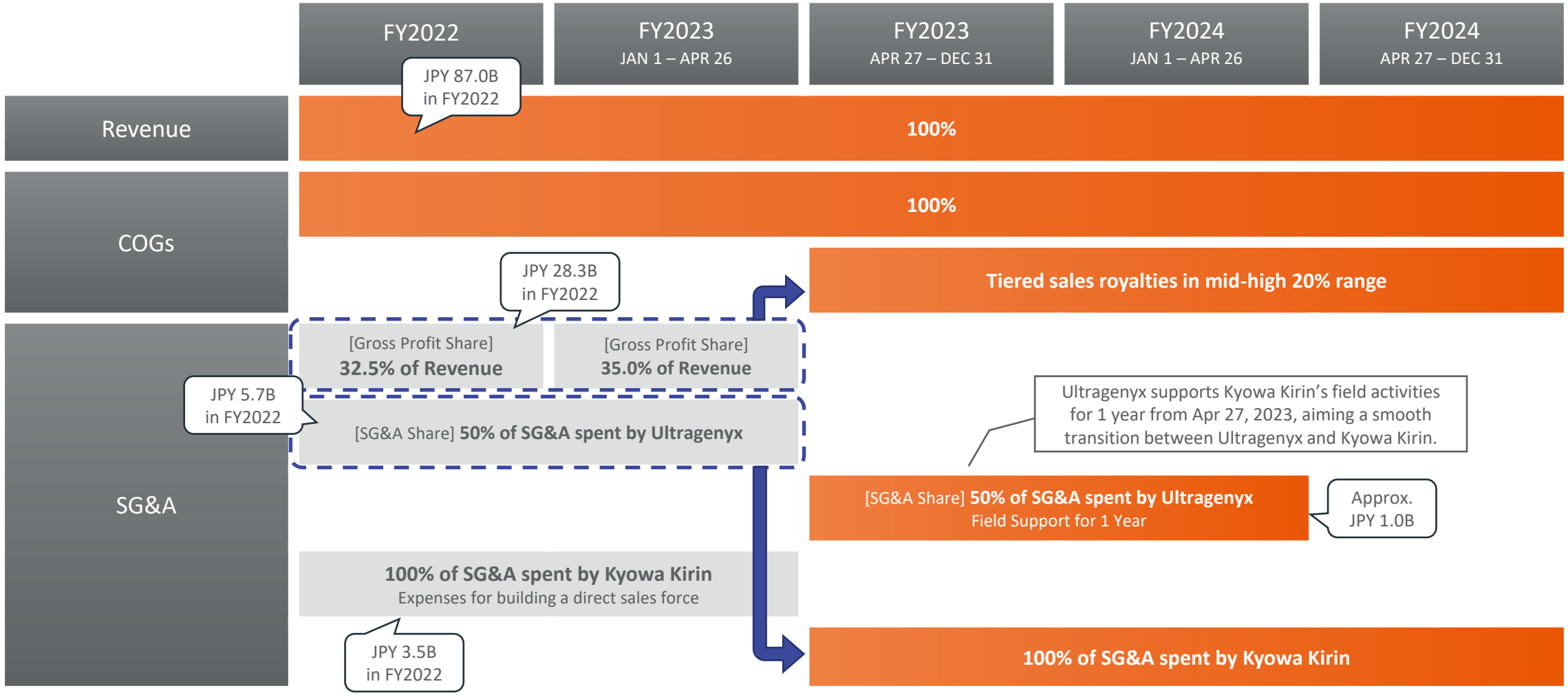


* Grünenthal owns a 51 percent majority share in the Joint Venture Collaboration, while Kyowa Kirin International plc owns a 49 percent share. Grünenthal will have the option to fully acquire the remaining 49 percent share, including intellectual property (IP) of 13 brands, via exercising an option in Q1, 2026

Appendix

P/L Impact on North American Crysvita Business

Based on the Collaboration and License Agreement in 2013, Kyowa Kirin takes over the field activities in North America from Ultragenyx, starting from April 27, 2023 (6th year from launch).



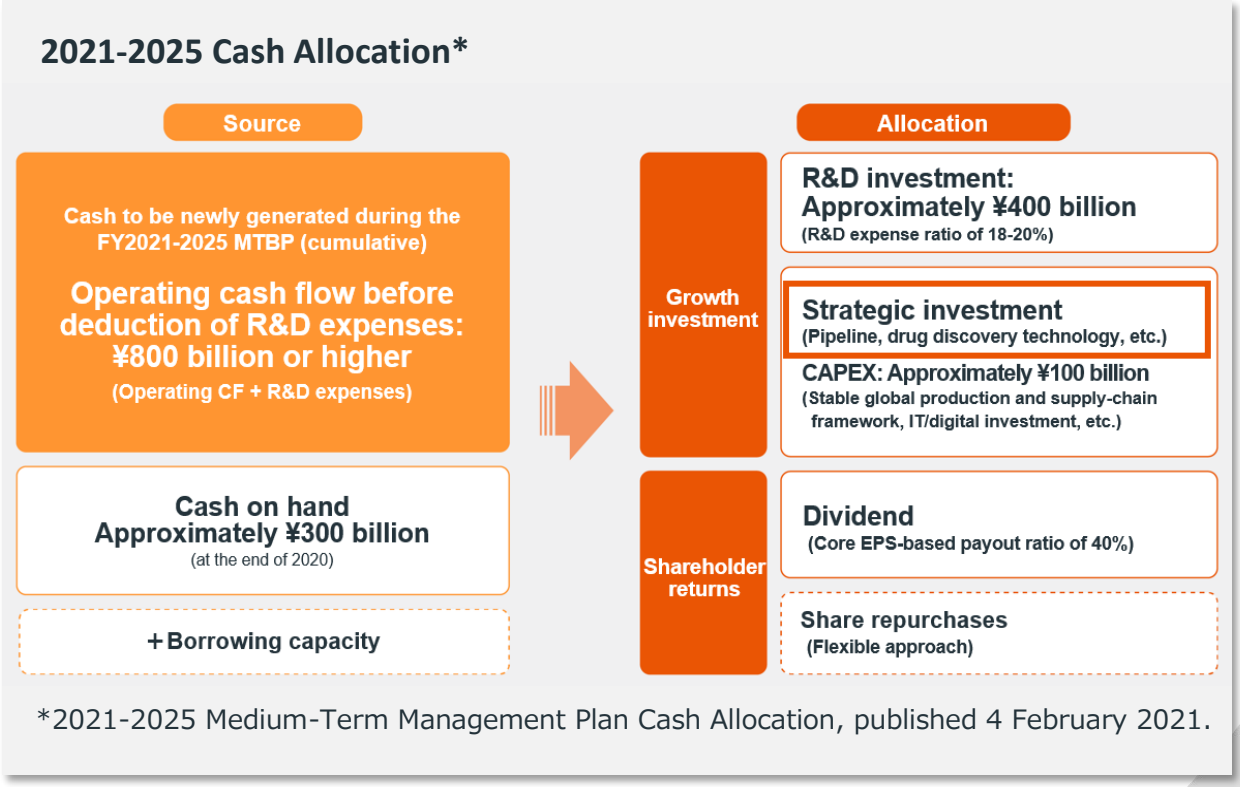
Strategic Investment ～For successful creation and delivery of life-changing value

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

- Development pipeline with synergies with Crysvita and Poteligeo
  - ◆Bone, Mineral ◆Hematologic oncology
- Implementing the strengths of each region
  - ◆Nephrology ◆Hematology / Oncology
  - ◆Immunology

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.

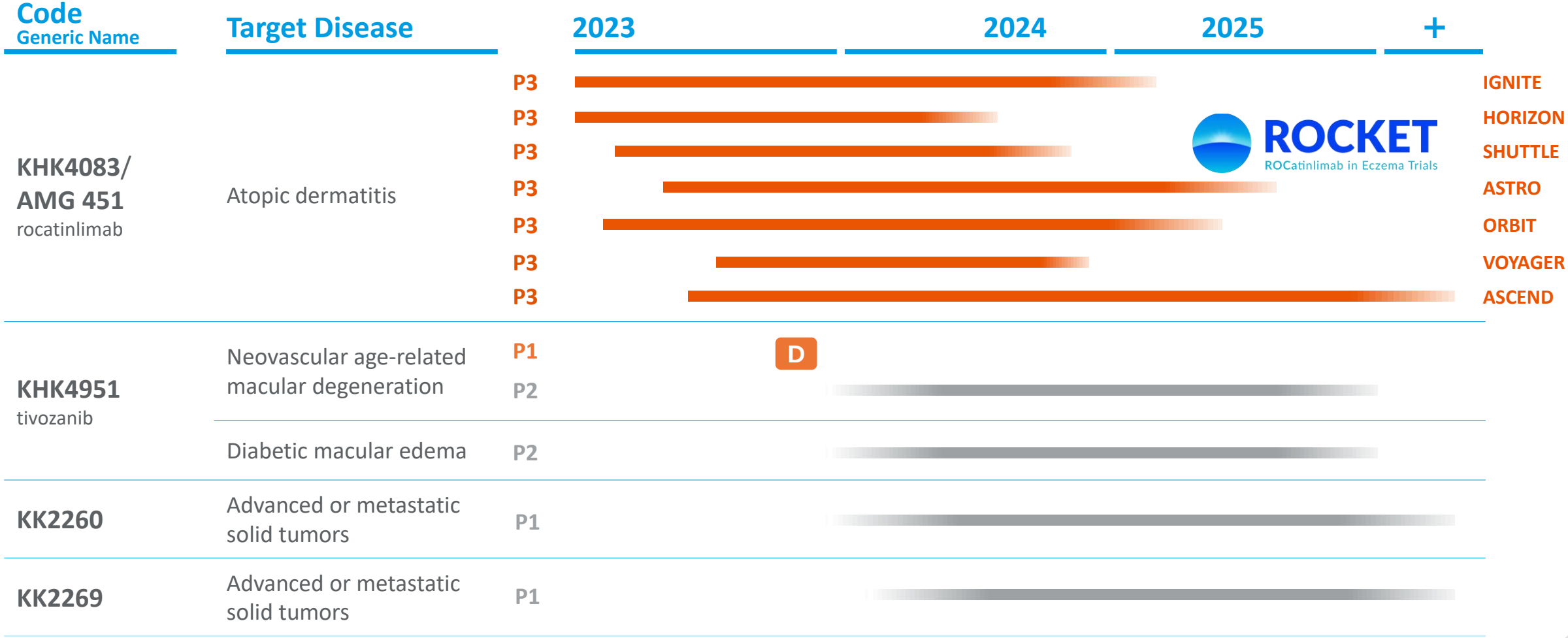


*2021-2025 Medium-Term Management Plan Cash Allocation, published 4 February 2021.

Main Development Pipeline Products: Future plans

T : Topline data　D : Detailed data

As of November 1, 2023



Main Development Pipeline Products

As of November 1, 2023

	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	Ph3 (Global)	★★★★★	16M
KHK4083/ AMG 451 rocatinlimab	Moderate and severe Asthma*5	TBD	Preparation for Ph2 (Global)	★★★★★	13.5M
KHK4951 tivozanib	Neovascular (wet) age-related macular degeneration	TBD	Preparation for Ph2 (US and JP)	★★★★	2,600K
KHK4951 tivozanib	Diabetic macular edema*5	TBD	Preparation for Ph2 (US and JP)	★★★★	3,400K
Pipeline products prior to the initiation of clinical studies					
KK2260	Advanced or metastatic solid tumors (esophageal cancer, head and neck cancer, etc.)	TBD	Preparation for Ph1 (US and JP)	TBD	TBD
KK2269	Advanced or metastatic solid tumors (gastric cancer, lung cancer, etc.)	TBD	Preparation for Ph1 (US and JP)	TBD	TBD

*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、★★★: ¥100Bn-¥500Bn、★★★★: ¥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 These are not yet on our pipeline list because these will start with Ph2 study.

FOREX Information

Average FOREX Rates (yen)

	2022Q3	2023Q3	Changes	2023 Plans
USD	126	137	+11	130
GBP	160	170	+10	160
EUR	135	148	+13	135

Q3 YoY FOREX Impacts (billion yen)

	Revenue	Core OP
USD	+9.2	+2.8
GBP	+0.6	-0.3
EUR	+2.7	+1.3

FY2023 FOREX Sensitivities (based on 2023 Plans, billion yen)

	Changes	Revenue	Core OP
USD	+1 yen	+1.2	+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
ADPKD	Autosomal Dominant Polycystic Kidney Disease
AG	Authorized Generic
AP, APAC	Asia-Pacific
ATL	Adult T-Cell Leukemia/Lymphoma
BS	Biosimilar
DME	Diabetic Macular Edema
EMEA	Europe, the Middle East and Africa
JP	Japan
LCM	Lifecycle Management
NA	North America
nAMD	neovascular Age-related Macular Degeneration
PD	Parkinson’s Disease
TIO	Tumor Induced Osteomalacia
XLH	X-linked Hypophosphatemia

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