

Results Presentation

Fiscal 2023 Second Quarter

協和キリン株式会社



Agenda

Financial Review
Commercial Update
R&D Update
News Flow in 2023

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

Q&A

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

Managing Executive Officer, Head of Finance **Motohiko Kawaguchi**

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

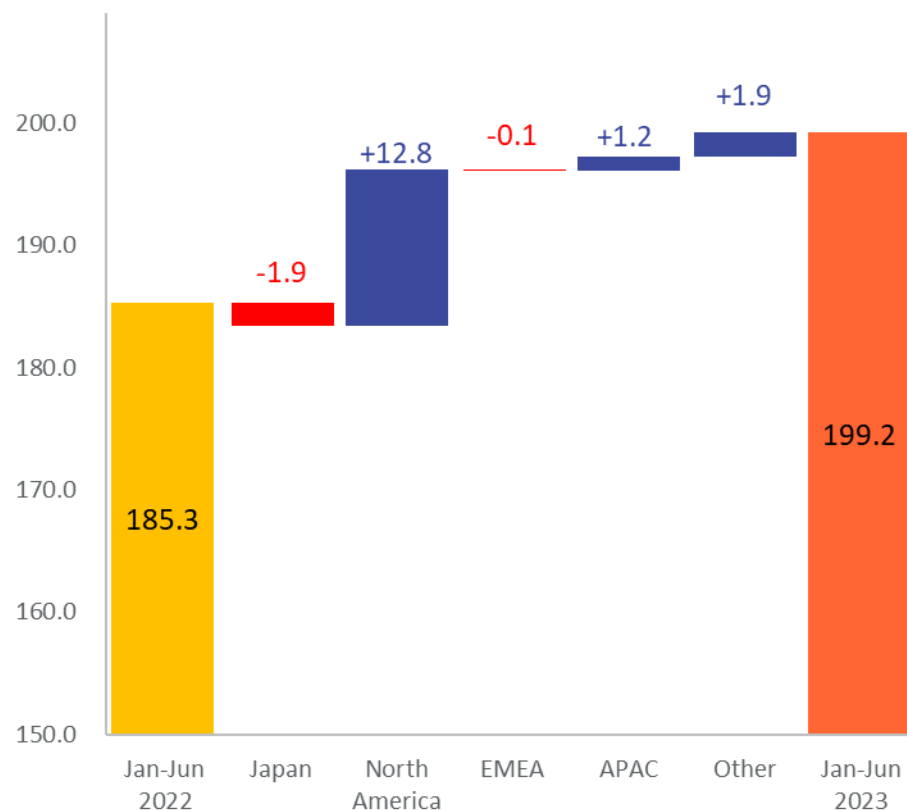
Summary of Q2 Results

(Billion Yen / Rounded)

| | 2022Q2 Results | 2023Q2 Results | Changes | 2023 Revised Plans | Progresses |
|--|-----------------------|-----------------------|--------------|-----------------------|------------|
| Revenue <i>[Overseas Ratio]</i> | 185.3 <i>[59%]</i> | 199.2 <i>[63%]</i> | +13.9 (+8%) | 426.0 <i>[64%]</i> | 47% |
| Gross Profit <i>[Gross Profit Margin]</i> | 141.9 <i>[77%]</i> | 152.2 <i>[76%]</i> | +10.3 (+7%) | 326.0 <i>[77%]</i> | 47% |
| SG&A <i>[SG&A Ratio]</i> | 76.4 <i>[41%]</i> | 82.4 <i>[41%]</i> | +6.0 (+8%) | 162.0 <i>[38%]</i> | 51% |
| R&D <i>[R&D Ratio]</i> | 27.9 <i>[15%]</i> | 33.7 <i>[17%]</i> | +5.7 (+21%) | 79.0 <i>[19%]</i> | 43% |
| Gain/Loss on Equity Method | 2.4 | 1.4 | -1.0 (-41%) | 3.0 | 46% |
| Core Operating Profit <i>[Core OP Margin]</i> | 39.9 <i>[22%]</i> | 37.5 <i>[19%]</i> | -2.4 (-6%) | 88.0 <i>[21%]</i> | 43% |
| Profit | 35.0 | 21.6 | -13.4 (-38%) | 76.0→70.0 | 31% |

YoY Analysis -Revenue-

+13.9 billion yen
(incl. forex effect +9.8)



● Japan -1.9

Although Duvroq, Romiplate, and Crysvita increased, revenue in Japan region decreased by 3% due mainly to negative impact by annual NHI price-cut and shrink in Nesp-AG and Allelock affected by competitive products.

● North America +12.8 (incl. forex effect +5.8)

Revenue in North America region increased by 27% with the growth of Crysvita(+28%), Poteligeo(+15%), and Nourianz(+37%).

● EMEA -0.1 (incl. forex effect +2.0)

Revenue in EMEA region decreased by JPY0.1B due to the impact of generic penetration of Abstral despite the continued growth of Crysvita(+13%), and Poteligeo(+42%).

● APAC +1.2 (incl. forex effect +0.7)

APAC revenue increased by 8% with the growth of Crysvita, and Nesp, while Gran was down due to the Chinese national tender system.

● Other +1.9 (incl. forex effect +1.3)

10% growth in the other revenue was due to the royalties of growing Fasentra (Benralizumab).

Revenue of Major Items (Japan)

(Billion Yen / Rounded)

| Item | 2022Q2 Results | 2023Q2 Results | Changes | Reasons | 2023 Plans | Progresses |
|-----------------------------|----------------|----------------|-------------|---|------------|------------|
| Nesp + Nesp-AG ¹ | 10.5 | 8.4 | -2.1 (-20%) | NHI price-cut & Biosimilars' penetration | 16.6 | 50% |
| Nesp | 1.6 | 1.5 | -1.5 (-8%) | | 2.8 | 54% |
| Nesp-AG | 8.8 | 6.9 | -2.2 (-22%) | | 13.8 | 50% |
| Duvroq | 2.7 | 4.2 | +1.5 (+57%) | Market penetration (Launched in Aug 2020) | 7.8 | 54% |
| Orkedia | 4.9 | 5.0 | +0.1 (+1%) | | 11.2 | 44% |
| G-Lasta | 14.8 | 15.0 | +0.2 (+1%) | | 33.5 | 45% |
| Poteligeo | 1.0 | 0.9 | -0.0 (-5%) | | 2.0 | 47% |
| Rituximab BS | 5.0 | 4.4 | -0.6 (-12%) | NHI price-cut | 8.7 | 51% |
| Romiplate | 4.8 | 5.7 | +1.0 (+20%) | Market penetration (New indication in 2019) | 11.2 | 51% |
| Allelock | 3.8 | 3.1 | - 0.7(-19%) | NHI price-cut | 4.7 | 66% |
| Nourias | 3.9 | 3.7 | -0.2 (-5%) | | 7.5 | 49% |
| Haruopi | 1.8 | 2.1 | +0.3 (+17%) | Market penetration (Launched in Dec 2019) | 4.7 | 44% |
| Crysvita | 4.1 | 4.8 | +0.7 (+17%) | Market penetration (Launched in Dec 2019) | 11.1 | 44% |

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.

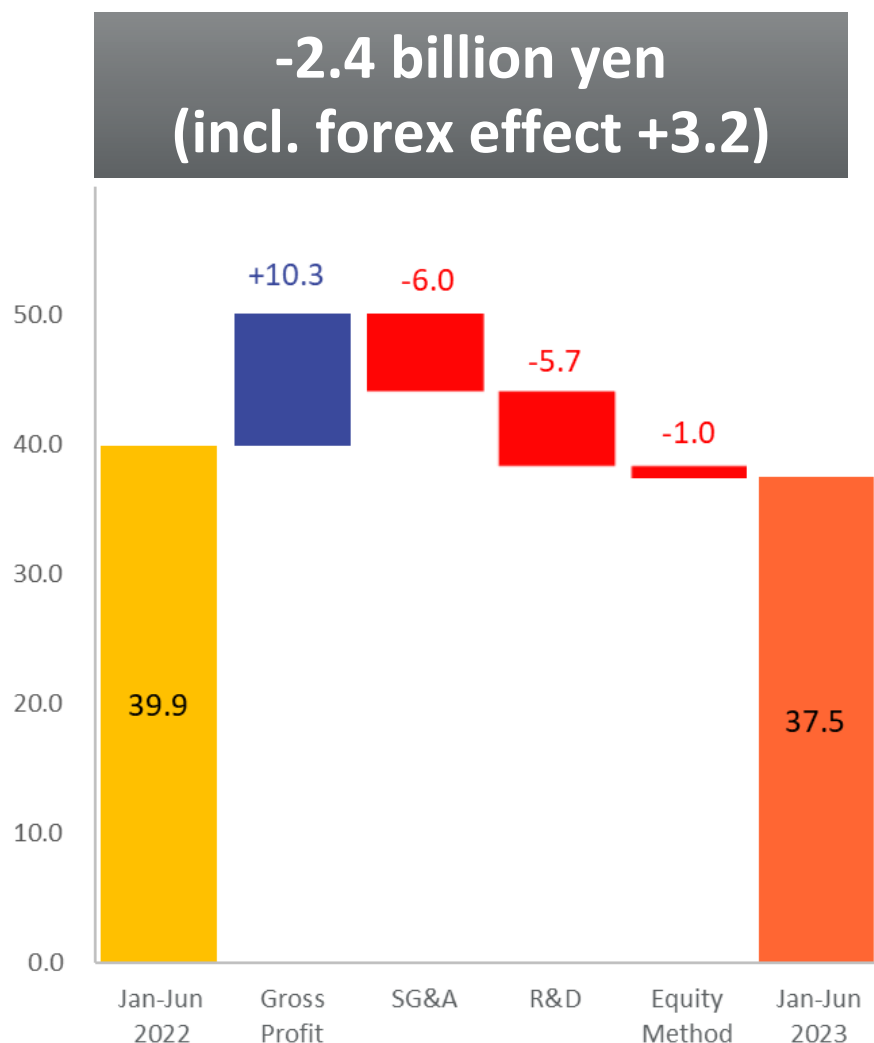
Revenue of Major Items (ex-Japan)

(Billion Yen / Rounded)

| Item | 2022Q2 Results | 2023Q2 Results | Changes | Reasons | 2023 Plans | Progresses |
|-----------------------------------|----------------|----------------|--------------|---|------------|------------|
| Crysvita | 49.4 | 61.9 | +12.5 (+25%) | [North America] Market penetration [EMEA] Geographical expansion & Additional indication (Adult/TIO) [APAC] Geographical expansion | 138.0 | 45% |
| North America | 35.9 | 46.0 | +10.2 (+28%) | | | |
| EMEA | 13.5 | 15.3 | +1.8 (+13%) | | | |
| APAC | 0.1 | 0.6 | +0.5 (+600%) | | | |
| Poteligeo | 10.3 | 12.5 | +2.2 (+21%) | [North America] Market penetration [EMEA] Geographical expansion & Market penetration | 27.5 | 45% |
| North America | 8.1 | 9.4 | +1.3 (+15%) | | 19.4 | 49% |
| EMEA | 2.1 | 3.1 | +0.9 (+42%) | | 8.0 | 38% |
| APAC | - | - | - | | 0.2 | - |
| Nourianz | 2.6 | 3.5 | +0.9 (+37%) | Market penetration | 7.5 | 47% |
| Nesp | 3.9 | 4.2 | +0.3 (+9%) | | 8.0 | 53% |
| Gran | 3.8 | 2.8 | -0.6 (-17%) | Listed on Chinese tender list | 8.2 | 39% |
| Neulasta | 3.0 | 2.8 | -0.2(-7%) | | 5.7 | 49% |
| Tech-licensing | 15.3 | 17.8 | +2.5 (+16%) | Growth of Fasenra | 39.0 | 46% |
| Benralizumab Royalty ¹ | 9.3 | 11.6 | +2.3 (+25%) | | | |

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

YoY Analysis -Core OP-



● Gross Profit +10.3 (incl. forex effect +9.0)

Increased in conjunction with JPY13.9B rise in revenue. Margin % remains at the same level (76.6% → 76.4%) thanks to increased proportion of profitable “Global 3 brands” and tech-licensing revenue, despite the COGs have increased due to the North America Crysvita-related scheme change from ‘Profit-sharing (SG&A)’ to ‘Sales royalty (COGs)’ after Apr 27, 2023.

● SG&A -6.0 (incl. forex effect -4.1)

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 and investment in IT/Digital infrastructure and human resources for global business foundation.

[HR exp -5.1 / Sales promotion +2.1 (incl. Crysvita profit sharing expenses +3.2)]

● R&D -5.7 (incl. forex effect -1.6)

Increased in clinical study costs of KHK4083.

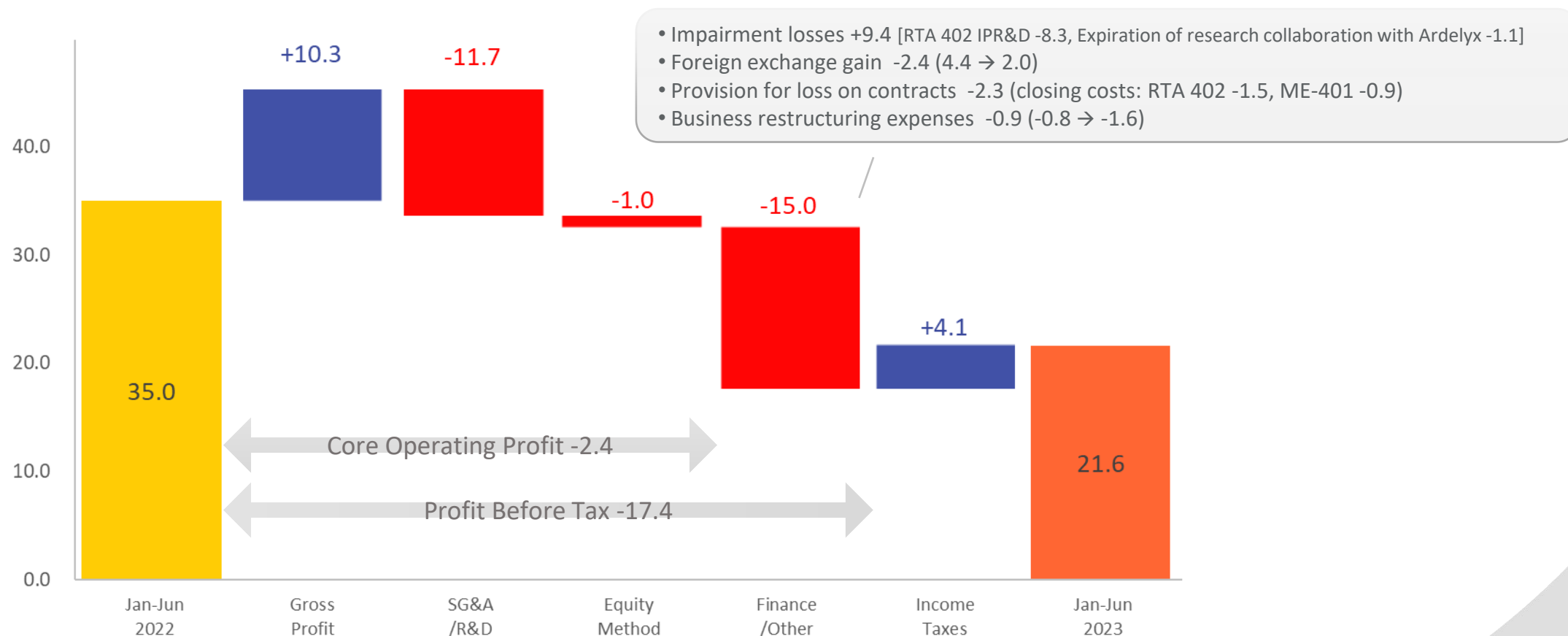
● Gain/Loss on Equity Method -1.0

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-Jun) -13.4 billion yen



Commercial Update

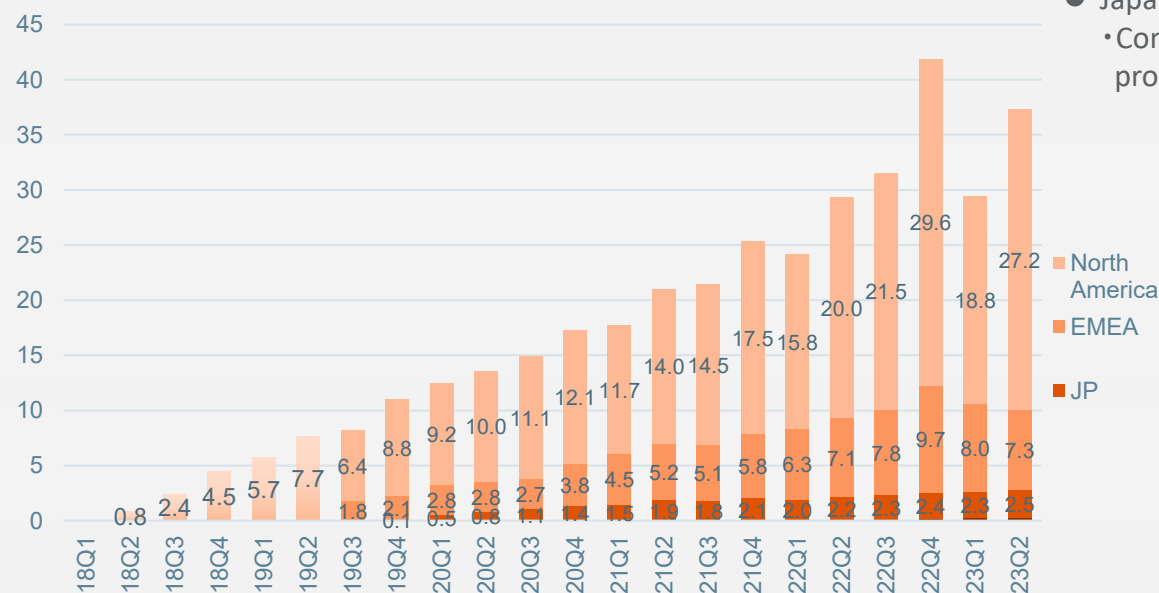
Coordinated Actions to Maximize the Patient Access to G3B

2023 Key Actions & Q2 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.

Sales Revenue (Billion Yen)

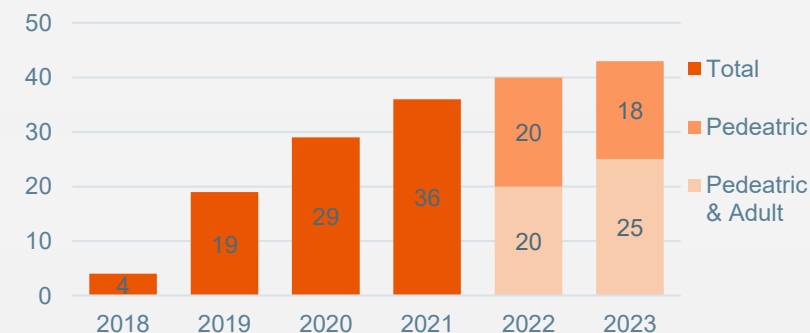


*Revenue from EAP (Early Access Program) is not included in sales until FY2022, and is included in sales from FY2023 onwards as it is insignificant in monetary terms.

Q2 Topics

- North America:
 - Own sales operation has made smooth start with the establishment of solid base for it.
 - The seasonal factor has been resolved, and sales grew as expected.
 - The number of patient enrollments in the treatment preparation stage and treatment patients continued to increase steadily.
- EMEA:
 - Patient penetration in each country has been steadily increased.
 - Revenues increased only slightly YoY due to price settlement payments including those for previous years for renegotiation of insurance reimbursement prices in connection with the expansion of sales scale in Germany.
- Japan:
 - Continued to strengthen promotional activities, such as holding webinars for medical professionals.

Launched Countries / Regions (XLH)



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

2023 Key Actions & Q2 Topics

2023 Key Actions

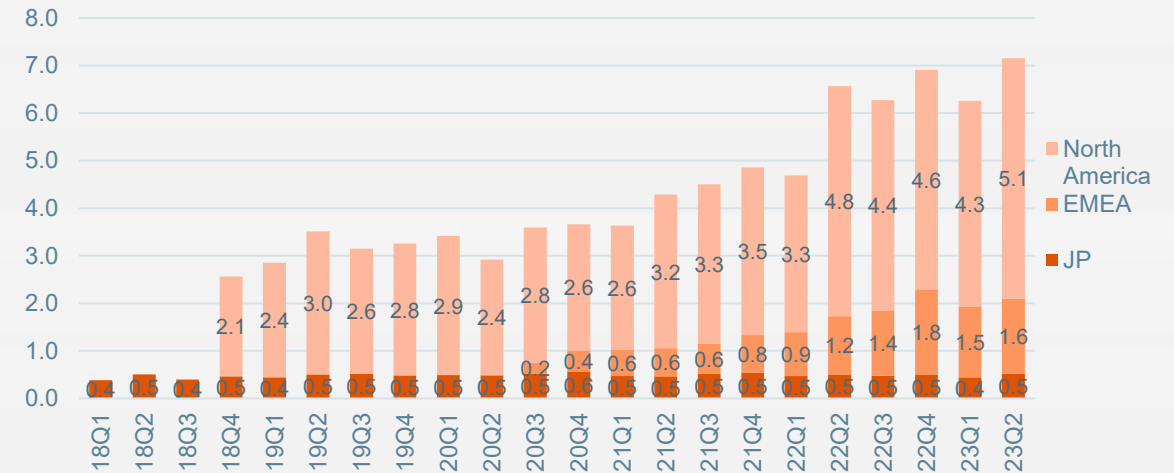
- Strengthen promotional activities utilizing evidences including blood tumor data.
- Raise awareness of importance of blood testing among early-stage patients

Q2 Topics

- North America: Steady growth by virtue of promotional activities utilizing evidences including blood tumor data or raising awareness of importance of earlier consultation with specialists and blood testing.
- EMEA: Sales grew steadily y-o-y, despite an increase in Mandatory Discount to public health insurance organizations in Germany and the U.K.

*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.

Sales Revenue (Billion Yen)



2023 Key Actions & Q2 Topics

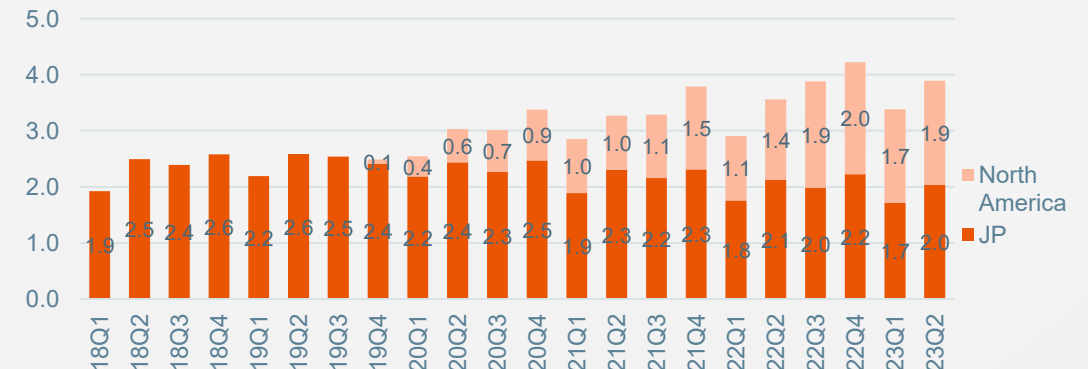
2023 Key Actions

- 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.

Q2 Topics

- US: Sales increased YoY. Introduced a new interactive case-based speaker program and continued strong field activity to enhance the messaging of importance of A2A pathway in treating OFF episodes. Lower coverage gap utilization had a positive cause for this quarter sales.

Sales Revenue (Billion Yen)



R&D Update








Upcoming Events: Main Development Pipeline Products

As of August 3, 2023

| Code Generic Name | Events (Completed are in bold) | Timeline (Completed are in orange) |
|---|---|---------------------------------------|
| KHK4083/AMG 451 rocatinlimab | Atopic Dermatitis P3 (ROCKET PROGRAM) Asthma P2 | In progress TBD |
| KHK4951 tivozanib | nAMD P1 data presentation P2 initiation | Q4 2023 Q4 2023 |
| KHK7791 tenapanor | Hyperphosphatemia under maintenance dialysis: Regulatory decision to be announced (JP) | Sep. 2023 |
| KW-3357 antithrombin gamma (genetical recombination) | Preeclampsia P3 LPO Topline data | Jun. 2023 Q3 2023 |

LPO: Last Patient Out; nAMD: Neovascular age-related macular degeneration

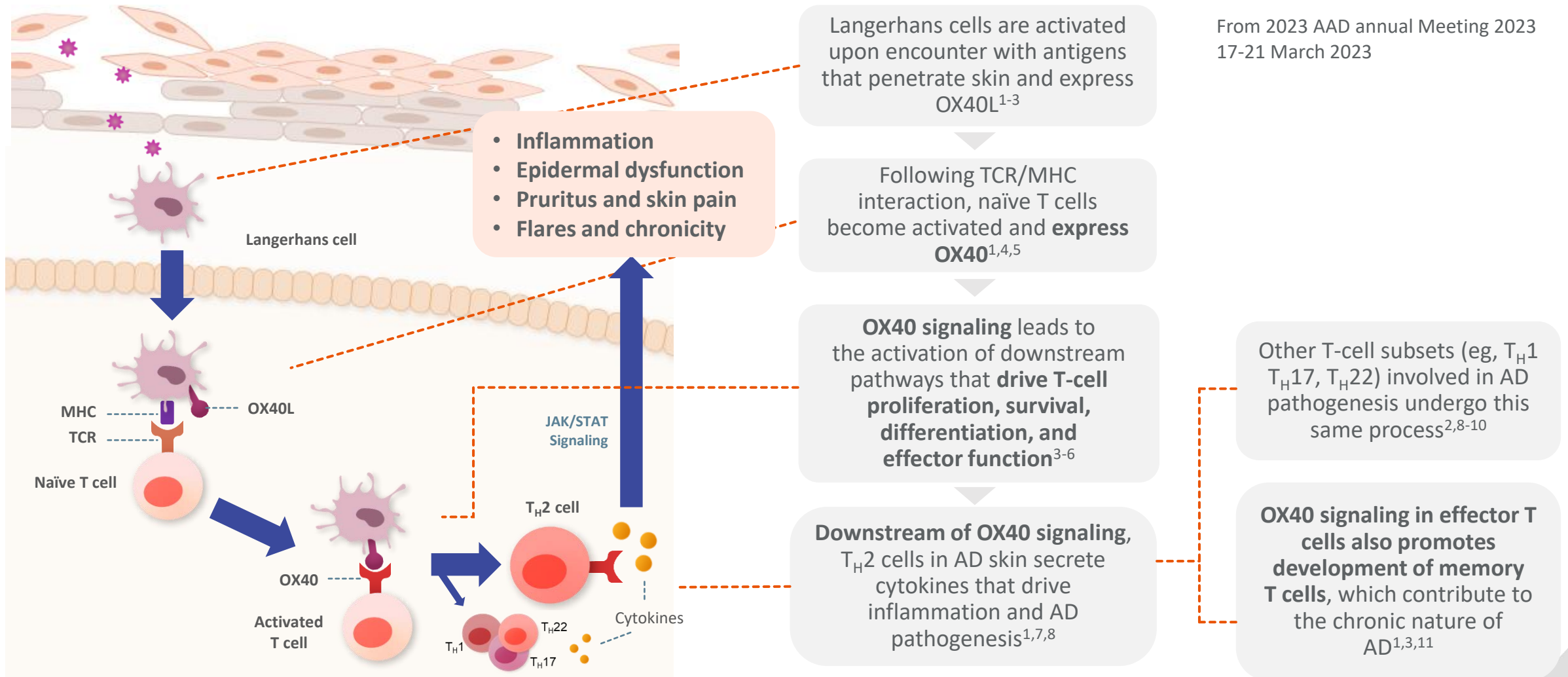
ROCKET is a comprehensive phase 3 development program designed to evaluate the efficacy, safety, and tolerability of rocatinlimab (monotherapy or combination therapy) in adult and adolescent patients with moderate-to-severe AD

| |  ROCKET ignite |  ROCKET horizon |  ROCKET shuttle |  ROCKET voyager |  ROCKET astro |  ROCKET orbit |
|--|---|--|--|--|--|--|
| Trial Duration | 24 weeks | 24 weeks | 24 weeks | 24 weeks | 52 weeks | 52 weeks |
| Trial Design | Placebo-controlled | Placebo-controlled | Placebo-controlled | Placebo-controlled | Placebo-controlled with re-randomized maintenance | Open-label |
| Rocatinlimab Dose | Dose 1 or Dose 2 Q4W* | Dose 1 Q4W* | Dose 1 or Dose 2 Q4W* + TCS/TCI | Dose 1 Q4W* | Initial Period: Dose 1 or Dose 2 Q4W* ±TCS/TCI Maintenance Period: Dose 1 or Dose 2 Q4W or Q8W ±TCS/TCI | Dose 1 Q4W* |
| Population | Adults with msAD [†] | Adults with msAD [†] | Adults with msAD [†] | Adults with msAD [†] | Adolescents with msAD [†] | Adolescents with msAD [†] |
| <div>  ROCKET ascend </div> <p>Adults and adolescents who complete a rocatinlimab parent study are eligible to enter ROCKET-Ascend, where they will be randomized to receive rocatinlimab dose 1 or dose 2 Q4W or Q8W or placebo Q4W[‡]</p> | | | | | | |

- *Loading dose at week 2. [†]All trials will enroll patients with moderate-to-severe AD with inadequate response, contraindication, or intolerance to topical medications (biologic-naïve and biologic-experienced patients). [‡]Available doses for subjects entering ROCKET-Ascend will vary based on the parent study they participated in.
- AD, atopic dermatitis; msAD, moderate-to-severe atopic dermatitis; Q4W, every 4 weeks; Q8W, every 8 weeks; TCI, topical calcineurin inhibitor; TCS, topical corticosteroid.

Critical Role of OX40 Signaling in Orchestrating T-Cell Driven Inflammation and AD Pathogenesis

From 2023 AAD annual Meeting 2023
17-21 March 2023



AD=atopic dermatitis; MHC=major histocompatibility complex; OX40=OX40 receptor; OX40L=OX40 ligand; TCR=T-cell receptor; Th=T helper cell.

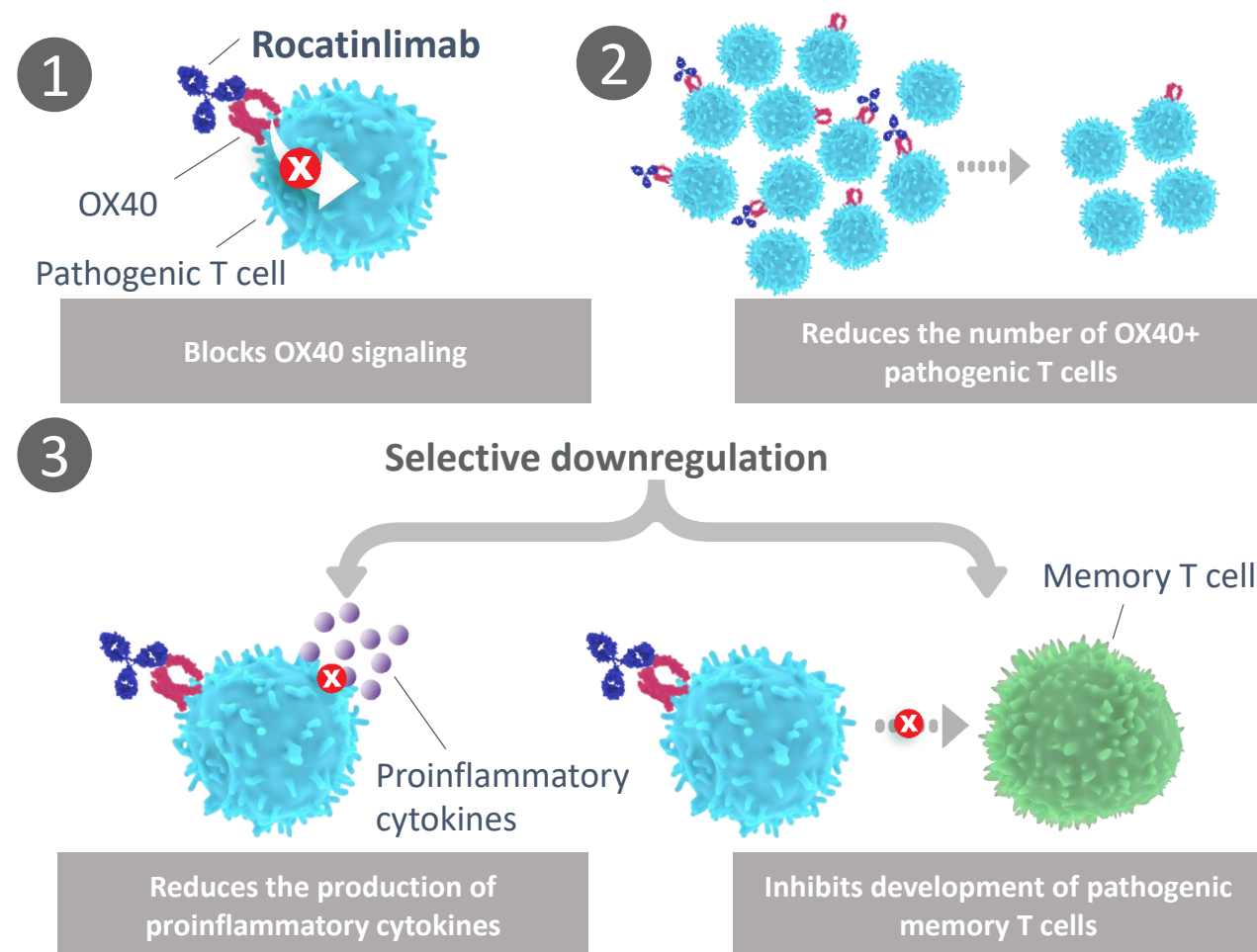
1. Furue M, et al. *J Clin Med*. 2021;10:2578. 2. Guttman-Yassky E, et al. *Semin Cutan Med Surg*. 2017;36:100-103. 3. Croft M, et al. *Immunol Rev*. 2009;229:173-191. 4. Magee CN, et al. *Am J Transplant*. 2012;12:2588-2600. 5. Goronzy JJ, et al. *Arthritis Res Ther*. 2008;10(suppl 1):S3. 6. Mascarelli DE, et al. *Front Cell Dev Biol*. 2021;9:692982. 7. Krohn IK, et al. *Allergy*. 2022;77:827-842. 8. De Bruyn Carlier T, et al. *J Autoimmun*. 2021;120:1026345. 9. Kumar S, et al. *Int J Mol Sci*. 2019;20:2159. 10. Fu Y, et al. *Acta Pharm Sin B*. 2020;10:414-433. 11. Chen L, et al. *Cell Mol Immunol*. 2020;17:64-75.

Rocatinlimab is an investigational anti-OX40 monoclonal antibody that targets the underlying AD pathogenesis¹⁻³

From 25th WCD Congress 3-8 July 2023

OX40 is a co-stimulatory molecule expressed on **Th2 cells**, as well as other T-cell subsets (**Th1, Th17, Th22**) that are **upregulated** in patients with AD⁴

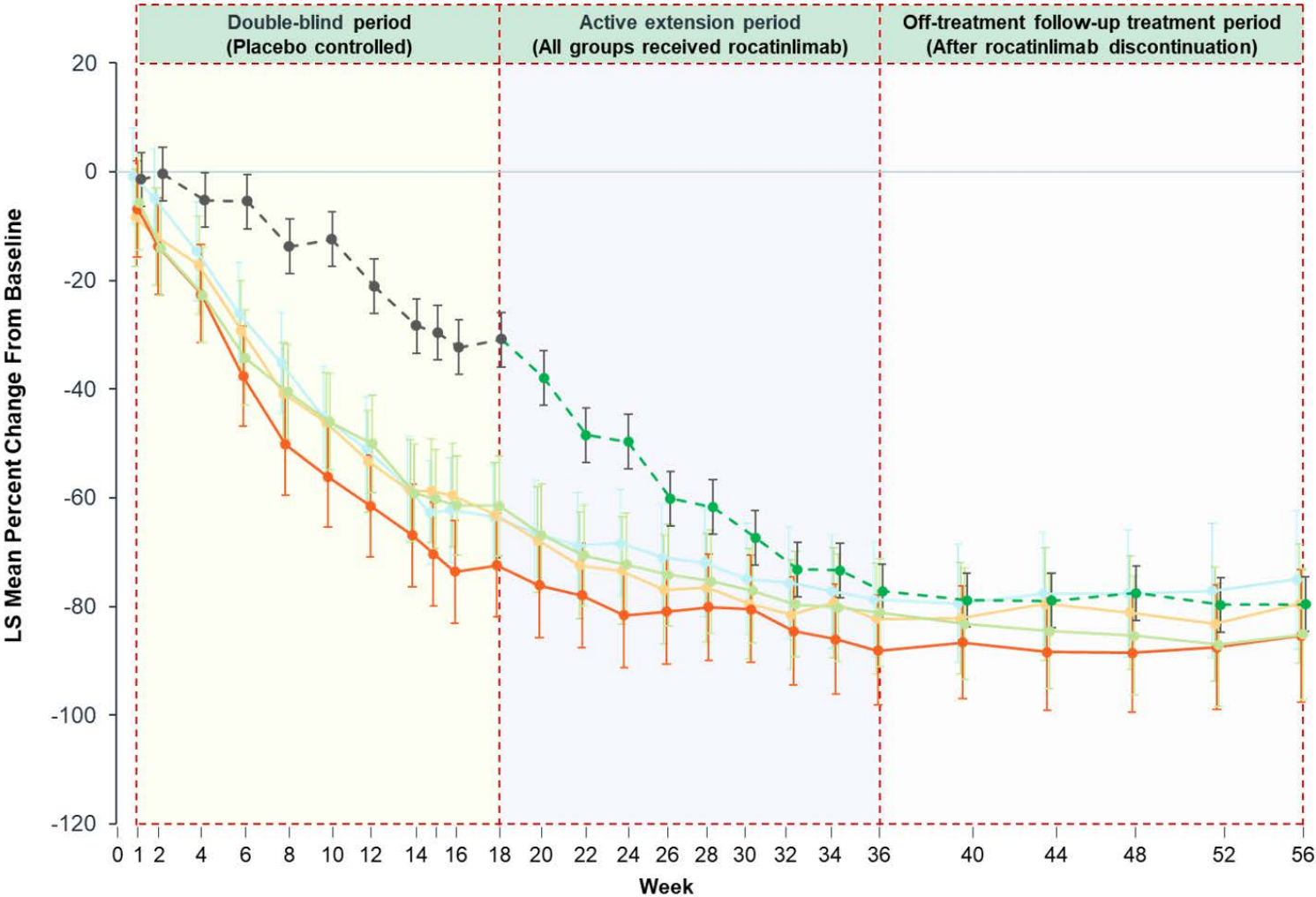
Rocatinlimab inhibits and reduces the number of OX40-expressing pathogenic T cells, responsible for driving systemic and local inflammatory responses¹⁻³



AD, atopic dermatitis; Th, T helper

¹. Nakagawa et al. *J Dermatol Sci.* 2020;99(2):82–89; ². Papp KA, et al. *J Eur Acad Dermatol Venereol.* 2017;31(8):1324–1332; ³. Guttman-Yassky et al. *Lancet* 2023;401(10372):204–214; ⁴. Furue & Furue. *J Clin Med* 2021;10(12):2578.

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)

News Flow in 2023

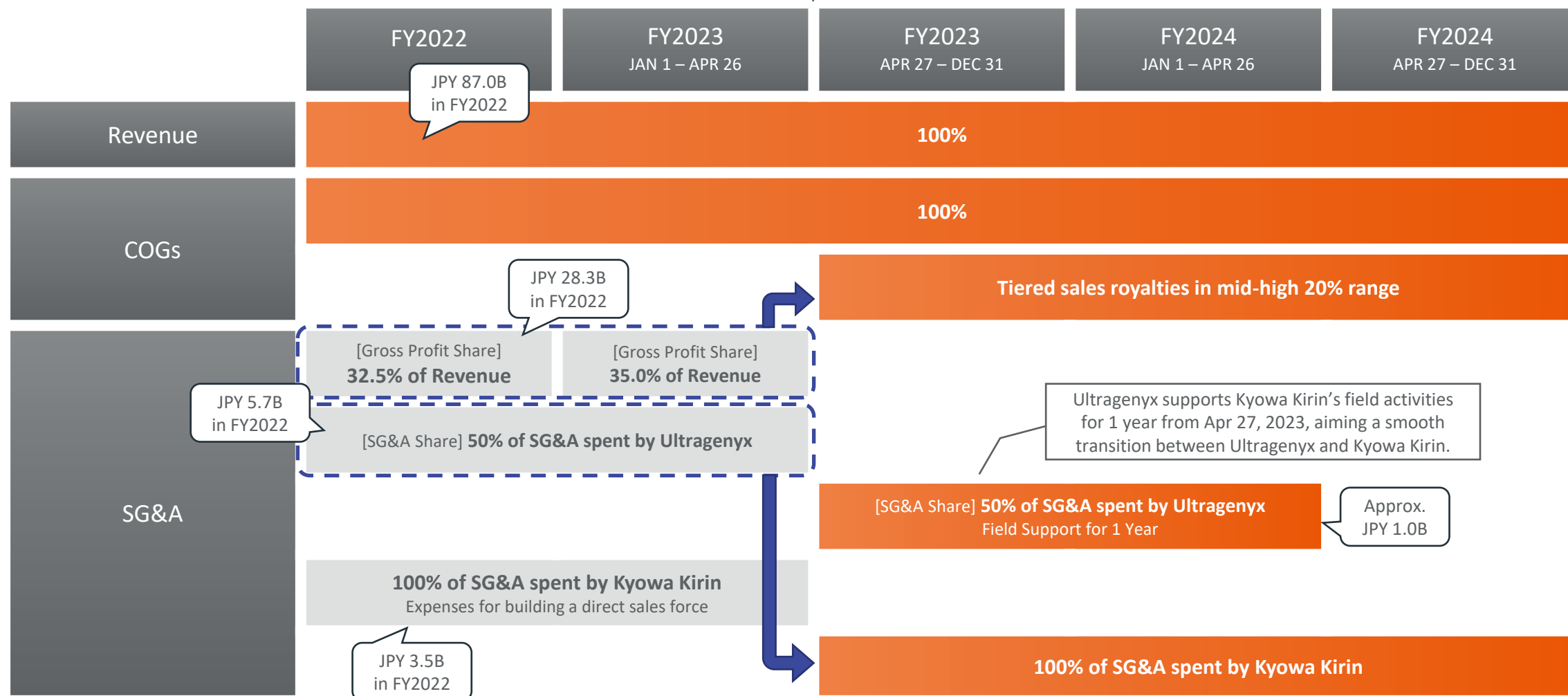
Year-to-date Key News Flow

| Category | Date | Headline | As of August 3, 2023 |
|----------|--------|--|----------------------|
| 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) | |

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

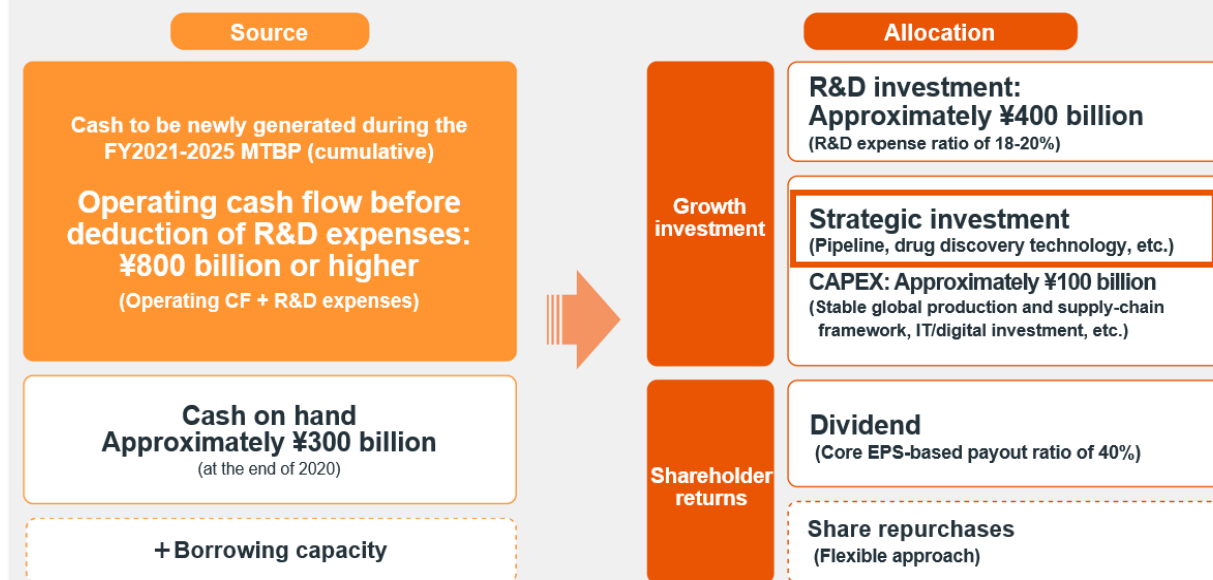
Licensing-in and M&A investments to strengthen the portfolio

- Development pipeline with synergies with Crysvisa 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 Cash Allocation*



*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 August 3, 2023

| Code Generic Name | Target Disease | | 2023 | 2024 | 2025 | + |
|--|---|----|------|------|------|------------|
| KHK4083/ AMG 451 rocatinlimab | Atopic dermatitis | P3 | | | | IGNITE |
| | | P3 | | | | HORIZON |
| | | P3 | | | | SHUTTLE |
| | | P3 | | | | ASTRO |
| | | P3 | | | | ORBIT |
| | | P3 | | | | VOYAGER |
| | | P3 | | | | ASCEND |
| KHK4951 tivozanib | Neovascular age-related macular degeneration | P1 | | D | | |
| | | P2 | | | | |
| KHK7791 tenapanor | Hyperphosphatemia under maintenance dialysis | | | ★ | | |
| KW-3357PE antithrombin gamma (genetical recombination) | Preeclampsia | P3 | | T | | KOUNO-TORI |



★: Anticipated timing of regulatory decision

Main Development Pipeline Products

| | 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 |
| KHK4951 tivozanib | Neovascular (wet) age-related macular degeneration | TBD | Preparing for Ph2 (US and JP) | ★★★★ | 2,300K~ |
| KHK7791 tenapanor | Hyperphosphatemia under maintenance dialysis | 2023 | Filed (JP) | ★ | 250K |
| KW-3357 antithrombin gamma (genetical recombination) | Preeclampsia | 2024 | Ph3 (JP) | ★ | 15K |

^{*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.**

FOREX Information

Average FOREX Rates (yen)

| | 2022Q2 | 2023Q2 | Changes | 2023 Plans |
|-----|--------|--------|---------|------------|
| USD | 120 | 134 | +14 | 130 |
| GBP | 158 | 164 | +6 | 160 |
| EUR | 133 | 144 | +11 | 135 |

Q2 YoY FOREX Impacts (billion yen)

| | Revenue | Core OP |
|-----|---------|---------|
| USD | +7.0 | +2.3 |
| GBP | +0.2 | -0.1 |
| EUR | +1.6 | +0.7 |

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 | <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 |
| ADPKD | Autosomal Dominant Polycystic Kidney Disease |
| AG | Authorized Generic |
| AP, APAC | Asia-Pacific |
| AS | Alport Syndrome |
| ATL | Adult T-Cell Leukemia/Lymphoma |
| BS | Biosimilar |
| CKD | Chronic Kidney Disease |
| CLL | Chronic Lymphocytic Leukemia |
| DKD | Diabetic Kidney Disease |
| EMEA | Europe, the Middle East and Africa |
| FL | Follicular Lymphoma |
| iB-NHL | Indolent B-cell Non-Hodgkin Lymphoma |
| JP | Japan |
| LCM | Lifecycle Management |
| MZL | Marginal Zone Lymphoma |
| NA | North America |
| nAMD | neovascular Age-related Macular Degeneration |
| PD | Parkinson's Disease |
| PE | Preeclampsia |
| PTCL | Peripheral T-Cell Lymphoma |
| TIO | Tumor Induced Osteomalacia |
| XLH | X-linked Hypophosphatemia |



Kyowa Kirin Co., Ltd.
Corporate Communications Dept., IR Group
+81-3-5205-7206 / ir@kyowakirin.com