Results Presentation
Fiscal 2021 Second Quarter

Kyowa Kirin Co., Ltd.

Agenda

Financial Review
Commercial Update
R&D Update

President and CEO Masashi Miyamoto, Ph.D.

Q&A

President and CEO Masashi Miyamoto, Ph.D.
Executive Officer, Head of Finance Motohiko Kawaguchi
Executive Officer, Vice President, Head of R&D Yoshifumi Torii, Ph.D.
Executive Officer, Head of Global Product Strategy Tomohiro Sudo

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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.

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Financial Review

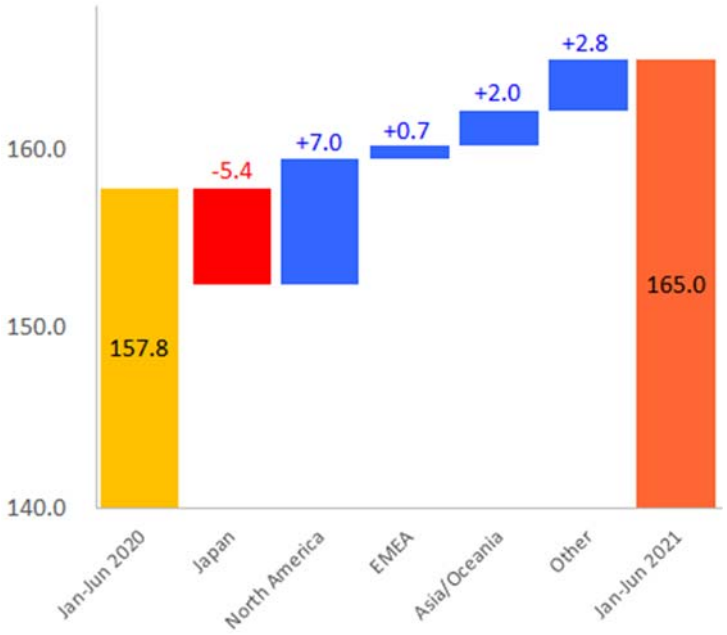
Summary of Q2 Results

(Billion Yen / Rounded)

	2020Q2 Results	2021Q2 Results	Changes	2021 Plans	Progresses
Revenue *[Overseas Ratio]*	157.8 *[46%]*	165.0 *[52%]*	+7.2 (+5%)	351.0 *[54%]*	47%
Gross Profit *[Gross Profit Margin]*	116.9 *[74%]*	123.8 *[75%]*	+6.9 (+6%)	270.0 *[77%]*	46%
SG&A *[SG&A Ratio]*	58.2 *[37%]*	67.2 *[41%]*	+9.0 (+15%)	141.0 *[40%]*	48%
R&D *[R&D Ratio]*	24.1 *[15%]*	26.5 *[16%]*	+2.4 (+10%)	65.0 *[19%]*	41%
Gain/Loss on Equity Method	-0.1	0.9	+1.0 (－)	1.0	92%
Core Operating Profit *[Core OP Margin]*	34.5 *[22%]*	30.9 *[19%]*	-3.5 (-10%)	65.0 *[19%]*	48%
Profit	27.8	25.1	-2.7 (-10%)	50.0	50%

YoY Analysis -Revenue-

+7.2 billion yen
(incl. forex effect +1.5)



- **Japan -5.4**
  Crysvita, Haruropi, Duvroq (New products), G-Lasta (Market's expansion & recovery) and Patanol (Higher pollen) and increased. However, Nesp-AG (Competitors) and Romiplate (Supply constraints) were down. In addition, there were negative impacts by NHI price-cut applied in April 2020 & April 2021 and co-promotion termination of Asacol, Minirinmelt and Desmopressin.

- **North America +7.0 (incl. forex effect -0.6)**
  Crysvita and Nourianz made a steady growth. Poteligeo that has been affected by the pandemic saw signs of market recovery.

- **EMEA +0.7 (incl. forex effect +1.7)**
  Crysvita maintained steady growth supported by farther launches and label expansion to adult. Poteligeo increased after its first launch in Germany in June 2020. On the other hand, Abstral decreased due to emergence of generics and supply constraints.

- **Asia/Oceania +2.0 (incl. forex effect +0.7)**
  China business centered on Regpara was favorable.

- **Other +2.8 (incl. forex effect -0.3)**
  Sales royalties of Fasenra (Benralizumab) made steady growth. In addition, we received US$10M upfront payment from Aevi Genomic Medicine regarding license-out of anti-LIGHT antibody.

Revenue of Major Items (Japan)

(Billion Yen / Rounded)

Item	2020Q2 Results	2021Q2 Results	Changes	Reasons	2021 Plans	Progresses
Nesp + Nesp-AG[*1]	14.6	12.9	-1.7 (-11%)	Biosimilars' penetration & NHI price-cut	23.2	56%
Nesp	2.2	1.9	-0.3 (-14%)		3.8	50%
Nesp-AG	12.4	11.0	-1.4 (-11%)		19.4	57%
Duvroq	—	0.5	+0.5 (—)	Launched in Aug 2020	4.0	14%
Regpara	2.0	1.5	-0.5 (-26%)		2.0	73%
Orkedia	4.3	4.6	+0.4 (+9%)		10.4	45%
G-Lasta	12.8	13.8	+1.0 (+8%)	Market's expansion & recovery	29.8	46%
Poteligeo	1.0	0.9	-0.0 (-4%)		2.0	48%
Rituximab BS	5.4	5.3	-0.1 (-3%)		11.5	46%
Romiplate	4.1	2.8	-1.2 (-31%)	Supply constraints (2020.6-2021.3)	8.7	32%
Allelock	4.9	4.8	-0.0 (-0.0%)		6.8	71%
Patanol	7.1	7.8	+0.6 (+8%)	Higher pollen	10.9	71%
Nouriast	4.6	4.2	-0.4 (-9%)	Competitors' penetration	9.1	46%
Haruropi	0.2	1.3	+1.1 (+457%)	Launched in Dec 2019	4.6	29%
Crysvita	1.3	3.3	+2.0 (+153%)	Launched in Dec 2019	5.5	61%
Tech-licensing	1.0	1.3	+0.3 (+27%)		2.5	54%

*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)

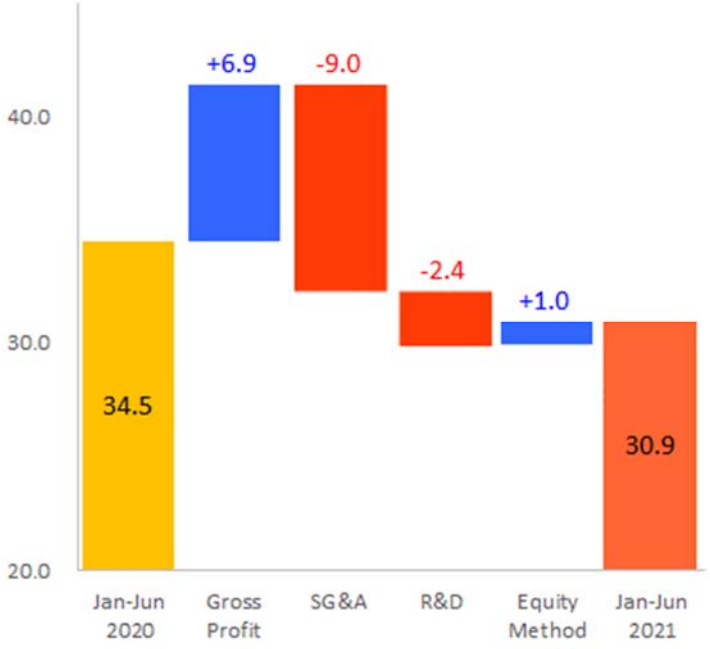
(Billion Yen / Rounded)

Item	2020Q2 Results	2021Q2 Results	Changes	Reasons	2021 Plans	Progresses
Crysvita	24.7	35.4	+10.7 (+43%)	[North America] Market penetration & Expanded indication (TIO) [EMEA] Farther launches & Expanded indication (Adult)	77.2	46%
North America	19.2	25.7	+6.5 (+34%)			
EMEA	5.6	9.7	+4.1 (+75%)			
Poteligeo	5.4	7.0	+1.6 (+30%)	[North America] Signs of market recovery [EMEA] Launched in Germany in Jun 2020 & Farther launches	17.3	40%
North America	5.4	5.8	+0.5 (+9%)			
EMEA	0	1.1	+1.1 (—)			
Nourianz	1.0	1.9	+1.0 (+102%)	Market penetration	6.7	29%
Abstral	5.8	4.0	-1.8 (-31%)	Generic's penetration & Supply constraints	8.1	50%
Regpara	3.9	4.6	+0.7 (+18%)	Listed on Chinese NEDL*1 in Oct 2018	9.3	49%
Tech-licensing	7.0	9.1	+2.1 (+31%)	Growth of Fasenra & Upfront revenue of anti-LIGHT antibody license-out	23.7	38%
Benralizumab Royalty*2	5.2	7.1	+1.9 (+37%)			

*1 National Essential Drug List
*2 Sales royalties of Fasenra, marketed by AstraZeneca (Including our own estimation)
＊ Revenue from Early Access Program (EAP) are not included in the figures above.

YoY Analysis -Core OP-

-3.5 billion yen
(incl. forex effect +0.2)



- **Gross Profit +6.9 (incl. forex effect +1.1)**
  - Increased in conjunction with 7.2B yen rise in revenue. Gross profit margin made 1 point improvement from 74% to 75%.

- **SG&A -9.0 (incl. forex effect -0.9)**
  Increased for the maximization of "Global Strategic 3 Brands" (G3B) and the early consolidation of global business foundation. [Sales promotion -3.4 (incl. Crysvita's profit sharing expenses in North America) / Labor -2.5 / Other -3.1]

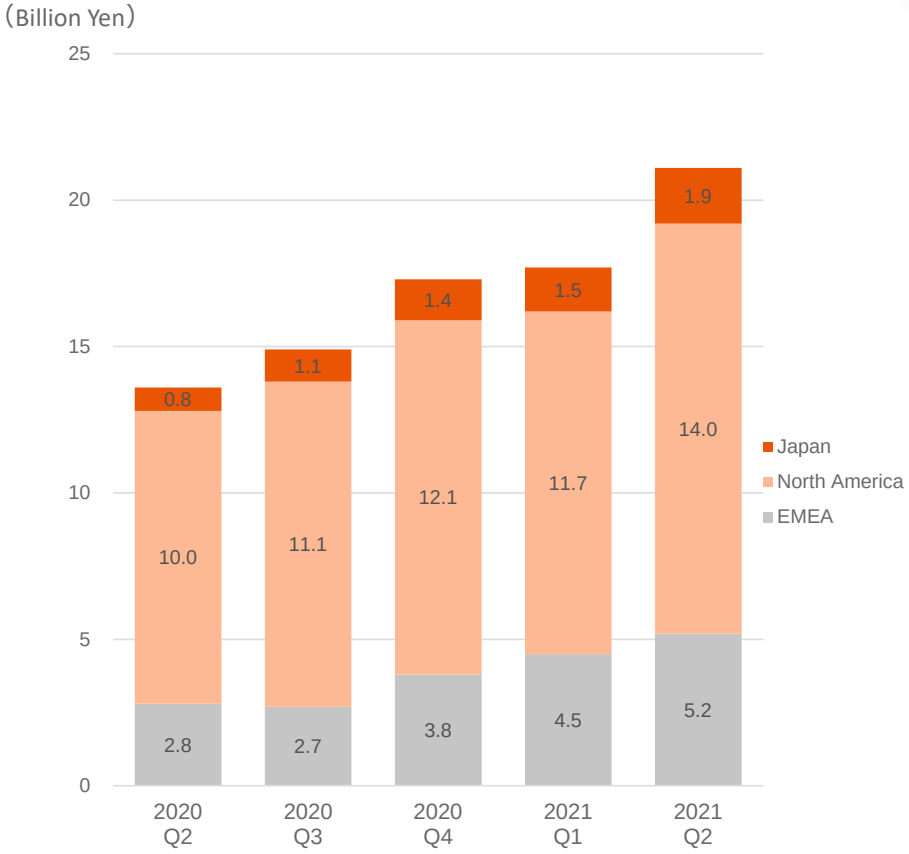
- **R&D -2.4 (incl. forex effect +0.1)**
  Development cost of ME-401 and KHK7791 increased.

- **Gain/Loss on Equity Method +1.0**
  Sales of Hulio (FKB327/Adalimumab BS) increased.

YoY Analysis -Profit-

Profit (Jan-Jun) -2.7 billion yen

Commercial Update



Topics

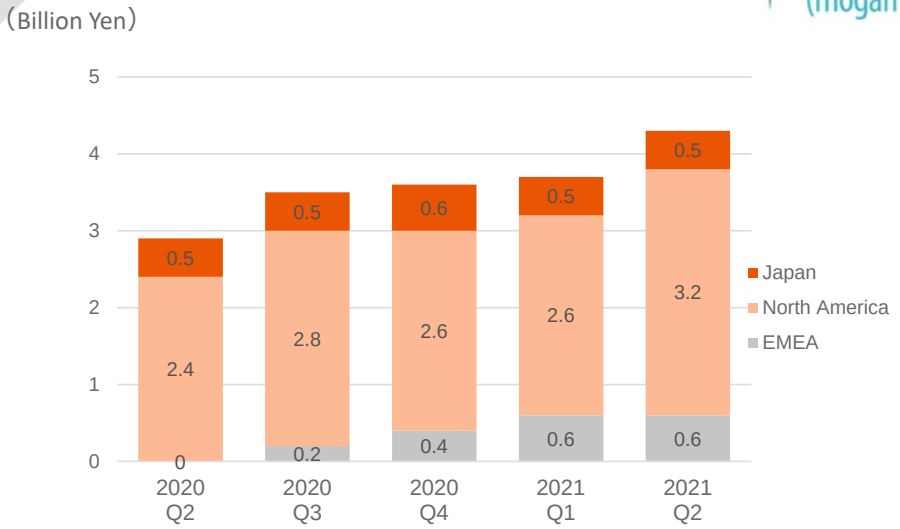
- North America: Steady progress in the number of newly treated patients.
- EMEA: (EU) Self-injection approved. (Germany) Expansion of indication for adult use contributed.
- AP: (China) Preparation for launch on track. (Singapore) XLH approval.

Launched Countries/Regions (XLH) as of June 30,2021

Excluding South America
Underlines: Pediatric and Adult
Bolded types: New launches in Q2 2021

USA , Canada, Germany, Netherland, Luxembourg, England, Wales, North Ireland, Slovakia, Sweden, Israel, UAE, Czech, Denmark, Italy, Japan, Norway, Bahrain, Scotland, Oman, Kuwait, Qatar, Romania, Slovenia, France, Finland, Estonia, Spain, **Ireland, Hungary, Belgium, Saudi Arabia, Hong Kong**

POTELIGEO® (mogamulizumab)



Topics

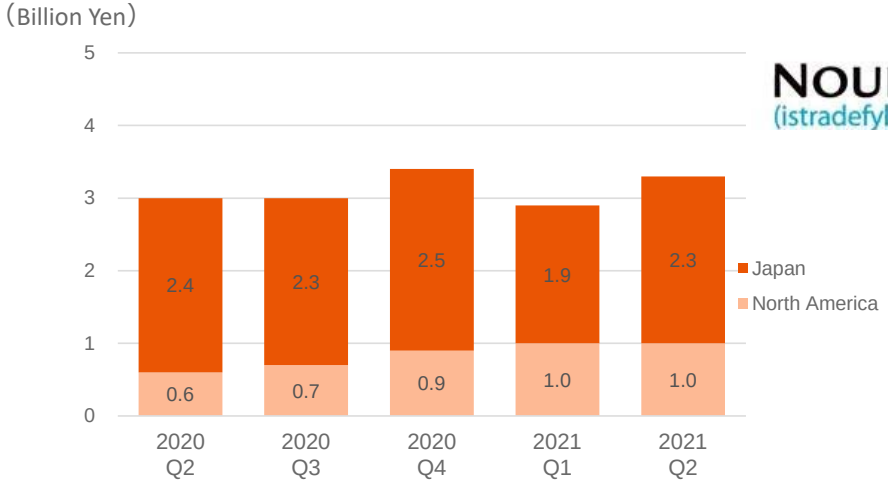
- Therapeutic effect in cases with hematological tumor.
- North America: Sales improved (Signs of recovery).
- EMEA: Reimbursement negotiations are delaying due to Cov19.
- Asia: (China) Filed for regulatory approval in June.

Launched Countries as of June 30, 2021

Bolded types: New launches in Q2 2021

Japan, USA, Germany, Austria, Luxembourg, Italy, **Scotland, Netherlands, Belgium, Slovenia, Denmark**

NOURIANZ® (istradefylline) tablets



Topics

- Japan: Impact of competitive products
- North America: Targeting and other market penetration measures underway
- Europe: CHMP issued a negative opinion

Launched Countries as of June 30, 2021

Japan, USA

Business Topics

Signed an agreement with Amgen for joint development and commercialization of KHK4083, a potent treatment for atopic dermatitis (Disclosed on June 1)

- On June 1, Kyowa Kirin and Amgen entered into an agreement to jointly develop and commercialize KHK4083, an anti-OX40 fully human monoclonal antibody discovered and being developed globally by Kyowa Kirin

  *This agreement became effective as of July 31, 2021, following the expiration of the waiting period under the U.S. antitrust laws.

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

- KHK4083 has a unique mechanism of action that is expected to become a new therapeutic option for treating atopic dermatitis, with potential in other autoimmune diseases.

R&D Update

Upcoming Events: Next-generation Strategic Products

✓ : Completed events from Jan 1 to May 6, 2021
✓ : Completed events from May 7 to Aug 3, 2021

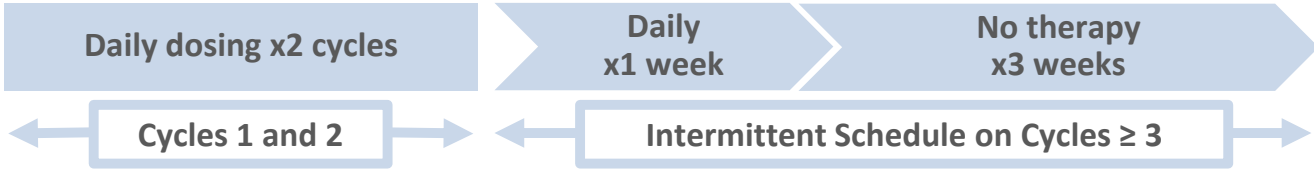
Code generic name	Target disease	H1 2021	H2 2021	H1 2022
KHK4083	Atopic dermatitis	P2b topline data ✓	P2b detailed data	P3 FPI
KW-6356	Parkinson's disease			P2b detailed data P3 FPI
ME-401 Zandelisib	FL (3L, mono)	P2 LPI ✓	P2 topline data	
	MZL (3L, mono)	P2 FPI ✓		
	FL/MZL (2L, combo)		P3 FPI	
RTA 402 Bardoxolone methyl	Alport syndrome		MA (JP) ✓	* (JP)
	DKD			
	ADPKD	P3 FPI (JP) ✓		
KHK7791 Tenapanor	Hyperphosphatemia under maintenance dialysis	P3 FPI (JP) ✓		

* Anticipated timing of regulatory decision; FPI: first patient in; LPI: last patient in; FL: follicular lymphoma; MZL: marginal zone lymphoma; ADPKD: autosomal dominant polycystic kidney disease; DKD: diabetic kidney disease; MA: marketing application

Zandelisib: Results of a Phase 1b Study in B-cell Malignancies Efficacy and Safety in patients with R/R FL with POD24

ASCO 2021
ClinicalTrials.gov Identifier: NCT02914938

- **Treatment: Zandelisib monotherapy or in combination with rituximab administered in IS**



High Response Rate in All FL Patients
Including Both POD24 and Non-POD24 Groups

	POD 24 N = 22	Non-POD24 N = 15	Total N = 37
ORR, N(%)	18 (82%)	14 (93%)	32 (87%)
- Monotherapy	8/11 (73%)	6/7 (86%)	14/18 (78%)
- Combination with rituximab	10/11 (91%)	8/8 (100%)	18/19 (95%)
Prior lines of therapy			
- 1 line of prior therapy	5/7 (71%)	9/9 (100%)	14/16 (88%)
- ≥ 2 lines of treatment	13/15 (87%)	5/6 (83%)	18/21 (86%)
CR rate, N (%)	4 (18%)	6 (40%)	10 (27%)

Treatment Well-tolerated:
8% Discontinuations Due to Adverse Events

Grade ≥ 3 Adverse Events in ≥ 2 Patients, N (%)	N = 37
Neutropenia	6 (16)
ALT/AST increased	3 (8)
Rash	3 (8)
Diarrhea	2 (5)
Colitis	2 (5)
Hypokalemia	2 (5)
Hyponatremia	2 (5)
COVID-19 infection	2 (5)

* POD24: progression of disease within 24 months of first-line chemoimmunotherapy

Zandelisib: Results of a Phase 1b Study in B-cell Malignancies
Zandelisib in combination with Zanubrutinib

ASCO 2021
ClinicalTrials.gov Identifier: NCT02914938

Group	Treatment schedule	
	Zandelisib, 60mg	Zanubrutinib
Group A (n = 7)	Daily dosing x2 cycles → Daily x1 week → No therapy x3 weeks Cycles 1 and 2 / IS on Cycles ≥3	160 mg, oral, bid
Group B (n = 13)	Daily x1 week → No therapy x3 weeks IS from Cycles 1	80 mg, oral, bid

Combination of zandelisib 60 mg on an IS from Cycle 1 and zanubrutinib 80 mg twice daily is well tolerated

Grade 3-4 AESI, N (%)	Group A N = 7 (%)	Group B N = 13 (%)
ALT / AST increased	2 (29%)	2 (15%)
Rash	1 (14%)	0
CMV colitis	1 (14%)	0
Pneumonia	1 (14%)	0
Diarrhea	0	0
Atrial fibrillation	0	0

Disease Response in 100% of Patients with iNHL and CLL

Evaluable N = 18	FL N = 8	CLL/SLL N = 5	MZL N = 2	MCL N = 1	DLBCL/HGBCL N = 2
ORR, N (%)	8 (100%)	5 (100%)	2 (100%)	1 (100%)	0
Group A	1 (100%)	3 (100%)	1 (100%)	1 (100%)	0
Group B	7 (100%)	2 (100%)	1 (100%)	0	0

Currently enrolling in expansion cohorts for FL and MCL with Regimen B

Expanding Zandelisib Development Activities to Explore Full Potential

Zandelisib Single Agent	Zandelisib + Rituximab	Other Zandelisib Combinations
・Ph 2 Study TIDAL in 3L+ FL and MZL	・Ph 3 Study COASTAL in 2L+ FL and MZL	・+ Zanubrutinib in FL and MCL in 2L+
・Ph 2 Study K02 in 3L+ in iNHL (Japan)		・+ R-CHOP in DLBCL in 1L
		・+ Ven-R in CLL

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

Appendix

FOREX Information

Average FOREX Rate

(Yen)

Currency	2020Q2 Results	2021Q2 Results	Change	2021 Plans
USD/JPY	109	107	-2	105
GBP/JPY	138	147	+9	140

2021Q2 FOREX Impacts (YoY)

(Billion yen)

Currency	Revenue	Core OP
USD/JPY	-1.0	-0.5
GBP/JPY	+1.7	+0.3

FY2021 FOREX Sensitivity

(Billion yen)

Currency	Changes	Revenue	Core OP
USD/JPY	+1 yen	-0.9	-0.5
GBP/JPY	+1 yen	-0.4	-0.1

Crysvita - Collaboration with Ultragenyx -

Territories	Economic terms
U.S. & Canada	• Kyowa Kirin books sales • 50/50 profit share for 5 years from the U.S. launch • After 5 years, Kyowa Kirin pays tiered sales royalties in mid-high 20% range to Ultragenyx • Supply price: 35% of net sales through 2022, 30% thereafter
Europe	• Kyowa Kirin books sales • Kyowa Kirin pays sales royalties in up to 10% range to Ultragenyx *Ultragenyx have sold a royalty right on/after 2020 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.

Development Plan of Next-generation Strategic Products

T : Topline data　D : Detailed data　As of Aug 3rd, 2021

Code generic name	Target disease		2021	2022	2023	+
KHK4083	Atopic dermatitis	P2b	T D			
		P3				
KW-6356	Parkinson's disease	P2b		D		
		P3				
ME-401 **Zandelisib**	FL (mono, 3L)	P2	T			
	MZL (mono, 3L)	P2				
	FL/MZL (combo, 2L)	P3				
	iNHL (mono, 3L)	P2				
RTA 402 **Bardoxolone methyl**	Alport syndrome		MA[1]	*[2]		
	Diabetic kidney disease	P3				
	ADPKD	P3				
KHK7791 **Tenapanor**	Hyperphosphatemia under maintenance dialysis	P3				
		P3				
		P3				
		P3				

[1] Japan; [2] Anticipated timing of regulatory decision; MA: marketing application; FL: follicular lymphoma; MZL: marginal zone lymphoma; iB-NHL: indolent B-cell non-Hodgkin's lymphoma; ADPKD: autosomal dominant polycystic kidney disease; 3L: third-line therapy ; 2L: second-line therapy

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)
CTCL	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)
XLH	EU	1:20,000	Adult: 12,000 Ped: 3,000	Estimate based on reported prevalence of 1 in 20,000 people
XLH	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
TIO	US		500-1,000	Survey by Ultragenyx Pharmaceutical
AD	JP, NA, EU5		30,000,000	Study by Decision Resources
PD	JP		162,000	Ministry of Health, Labour and Welfare: 2017 Patient survey (illness classification)
PD	US	60,000 / y	~1,000,000	Cited from Parkinson's Foundation https://www.parkinson.org/Understanding-Parkinsons/Statistics Accessed July 31, 2021.
FL	US	15,000 / y		Cited from Cancer.net https://www.cancer.net/ Accessed July 31, 2021.
FL	JP	6,750 / y		Cited from Cancer Registry and Statistics. Cancer Information Service, National Cancer Center, Japan (Ministry of Health, Labour and Welfare, National Cancer Registry) and Epidemiology of malignant lymphoma and recent progress in research on adult T-cell leukemia/lymphoma in Japan (Miyoshi H et al., Int J Hematol, 2018)
MZL	US	6,000 / y		Cited from Lymphoma.org https://lymphoma.org/ Accessed July 31, 2021.
MZL	JP	1,060 / y		Cited from Cancer Registry and Statistics. Cancer Information Service, National Cancer Center, Japan (Ministry of Health, Labour and Welfare, National Cancer Registry) and Epidemiology of malignant lymphoma and recent progress in research on adult T-cell leukemia/lymphoma in Japan (Miyoshi H et al., Int J Hematol, 2018)
AS	JP		1,200	Cited from the website of Japan Intractable Diseases Information Center https://www.nanbyou.or.jp/entry/4348 Accessed July 31, 2021.
ADPKD	JP		31,000	Cited from the website of Japan Intractable Diseases Information Center https://www.nanbyou.or.jp/entry/295 Accessed July 31, 2021.
CKD	JP		13,300,000	Japanese Society of Nephrology, Clinical Practice Guidebook for Diagnosis and Treatment of Chronic Kidney Disease (2012)
CKD (Dialysis)	JP	40,885 / y	344,640	The Japanese Society for Dialysis Therapy, An Overview of Regular Dialysis Treatment in Japan (As of 31 December 2019)

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

List of Acronyms

AD	Atopic Dermatitis
AG	Authorized Generic
ATL	Adult T-Cell Leukemia/Lymphoma
BS	Biosimilar
CKD	Chronic Kidney Disease
CLL	Chronic Lymphocytic Leukemia
DKD	Diabetic Kidney Disease
DLBCL	Diffuse Large B-Cell Lymphoma
FL	Follicular Lymphoma
iNHL	Indolent B-cell Non-Hodgkin Lymphoma
MCL	Mantle Cell Lymphoma
MZL	Marginal Zone Lymphoma
PD	Parkinson’s Disease
PTCL	Peripheral T-Cell Lymphoma
R-CHOP	Rituximab-Cyclophosphamide/Hydroxydaunorubicin(doxorubicin)/Oncovin(vincristine)/Prednisone
TIO	Tumor Induced Osteomalacia
Ven-R	Venetoclax-Rituximab
XLH	X-linked Hypophosphatemia

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