Results Presentation
Fiscal 2020 Second Quarter

Kyowa Kirin Co., Ltd.

Agenda

Q2 Financial Review & FY2020 Revised Plan
R&D Review
Business Topics

President and Chief Executive Officer Masashi Miyamoto, Ph.D

Q&A

President and Chief Executive Officer Masashi Miyamoto, Ph.D
Executive Officer, Head of Finance Motohiko Kawaguchi
Executive Officer, Head of R&D Yoshifumi Torii, Ph.D
Executive Officer, Head of Strategic Product Planning 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.

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Q2 Financial Review
& FY2020 Revised Plan

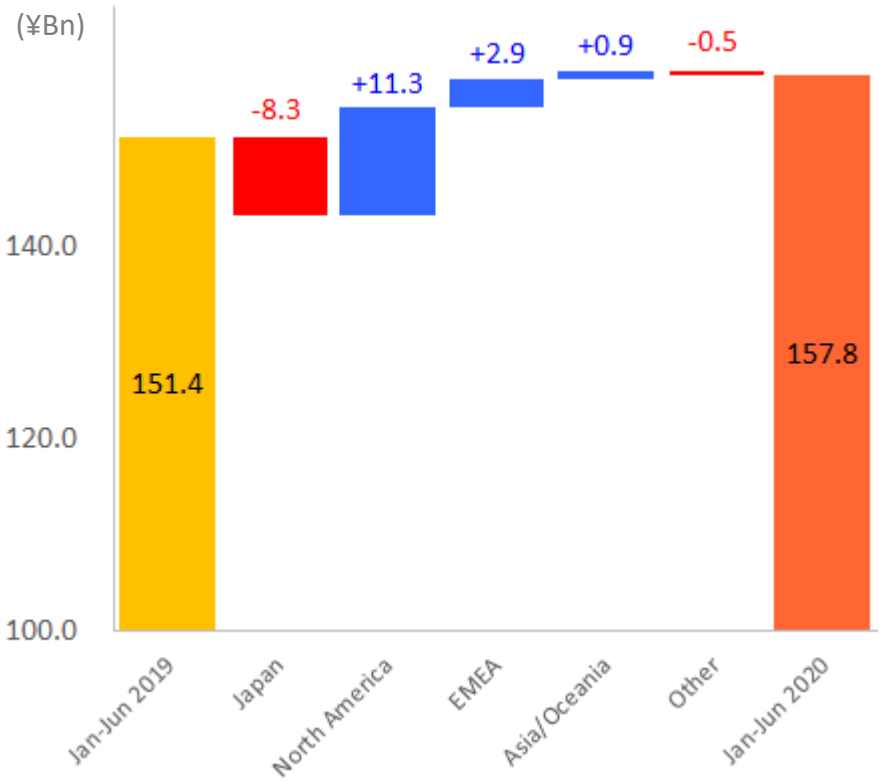
Summary of Q2 Results

(Billion Yen / Rounded)

	2019Q2 Results	2020Q2 Results	Changes	2020 New Plan	Progress
Revenue *[Overseas Ratio]*	151.4 *[37%]*	157.8 *[46%]*	+6.4 (+4%)	327.0→ 313.0 *[53%]*	48%→ 50%
Gross Profit *[Gross Profit margin]*	112.8 *[74%]*	116.9 *[74%]*	+4.1 (+4%)	250.0→ 236.0 *[75%]*	47%→ 50%
Core OP *[Core OP margin]*	32.2 *[21%]*	34.5 *[22%]*	+2.3 (+7%)	65.0→ 60.0 *[19%]*	53%→ 57%
Profit from continued operation	18.7	27.8	+9.1 (+49%)	49.0→ 44.0	57%→ 63%
Profit from discontinued operation	29.4	—	-29.4 (-100%)	—	—
Profit	48.1	27.8	-20.3 (-42%)	49.0→ 44.0	57%→ 63%

YoY Analysis -Revenue-

+6.4 billion yen
(incl. forex effect -1.8)



- **Japan -8.3**
  - Romiplate (Additional indication), G-Lasta and Rituximab BS made strong growth, however they could not cover the impact such as the shrink of Nesp (Shift to Nesp-AG) and Patanol/Allelock (Lower pollen and COVID-19*) and NHI price-cut applied on Oct 2019 and Apr 2020.

  *Voluntary ban on doctor's visit/outing or wearing a mask etc.

- **North America +11.3 (incl. forex effect -0.1)**
  - Sales of Crysvita and Nourianz increased. Poteligeo was at the same level as 2019 due to the COVID-19 pandemic.

- **EMEA +2.9 (incl. forex effect -1.1)**
  - Crysvita went up, and Moventig & Pecfent slightly increased as well.

- **Asia/Oceania +0.9 (incl. forex effect -0.6)**
  - Regpara maintained favorable sales in China.

- **Other -0.5 (incl. forex effect -0.1)**

Revenue of Major Items (Japan)

(Billion yen / Rounded)

Item	2019Q2 Results	2020Q2 Results	Changes	Reason	2020 New Plan	Progress
Nesp + Nesp-AG*	25.6	14.6	-11.0 (-43%)	Nesp-AG launched in Aug 2019 and Biosimilars' penetration	34.7→ 28.9	50%
Nesp	25.6	2.2	-23.4 (-91%)		4.0→ 4.6	48%
Nesp-AG	–	12.4	+12.4		30.7→ 24.4	51%
Regpara	3.8	2.0	-1.8 (-48%)	Switch to Orkedia	3.2→ 3.3	60%
Orkedia	3.0	4.3	+1.2 (+41%)	Steady market penetration	10.1→ 9.5	45%
G-Lasta	11.5	12.8	+1.3 (+11%)	Market expansion	28.1→ 27.6	46%
Poteligeo	1.0	1.0	+0.0 (+2%)		2.0→ 2.0	49%
Rituximab BS	4.2	5.4	+1.2 (+30%)	Steady market penetration	10.1→ 11.4	47%
Romiplate	1.6	4.1	+2.4 (+148%)	Indication added in Jun 2019	7.2→ 7.4	55%
Allelock	6.4	4.9	-1.6 (-25%)	Lower pollen in the air and COVID-19 (Voluntary ban on doctor's visit/outing or wearing a mask etc.)	8.3→ 8.3	59%
Patanol	9.9	7.1	-2.8 (-28%)		9.5→ 9.8	73%
Nouriast	4.8	4.6	-0.2 (-3%)	COVID-19 (Voluntary ban on doctor's visit)	10.5→ 9.9	47%
Haruropi	–	0.2	+0.2	Launched in Dec 2019	1.9→ 1.1	22%
Crysvita	–	1.3	+1.3	Launched in Dec 2019	3.5→ 3.5	37%
Technology licensing	1.7	1.0	-0.6 (-37%)		3.7→ 3.5	30%

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

Revenue of Major Items (Overseas)

(Billion yen / Rounded)

Item	2019Q2 Results	2020Q2 Results	Changes	Reason	2020 New Plan	Progress
Crysvita*1	13.4	24.7	+11.4 (+85%)	Steady market penetration	56.6→ 51.1	48%
North America	9.9	19.1	+9.3 (+95%)			
EMEA	3.5	5.6	+2.0 (+58%)			
Poteligeo	5.4	5.4	-0.0 (-1%)	COVID-19 (Treatment Interval Extension)	14.3→ 10.0	54%
Nourianz	–	1.0	+1.0	Launched in Oct 2019	2.8→ 2.6	37%
Abstral	5.8	5.8	-0.0 (-1%)		9.0→ 9.7	60%
Regpara	2.4	3.9	+1.5 (+60%)	Listed on Chinese NEDL*2 in Oct 2018	7.3→ 7.4	52%
Technology licensing	5.2	7.0	+1.8 (+34%)		18.8→ 18.3	38%
Benralizumab Royalty*3	3.8	5.2	+1.4 (+38%)	Launched in 2018		

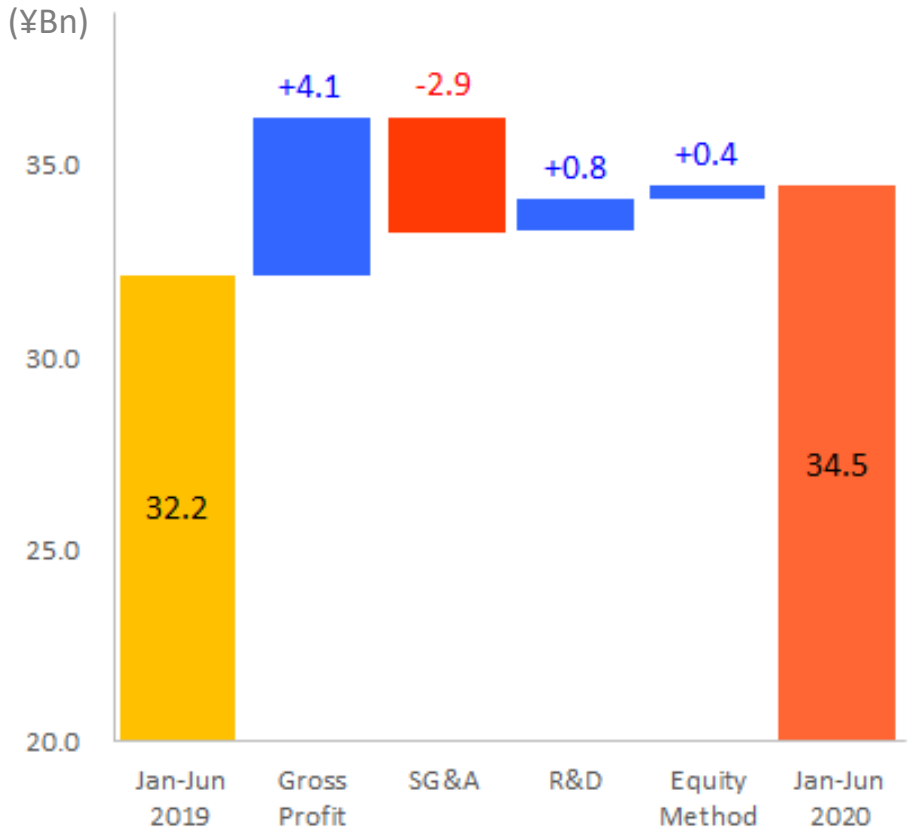
*1 Launched countries as of June 30, 2020 (excluding South America):
USA, Canada, Germany, Netherland, Luxembourg, England, Wales, North Ireland, Slovakia, Sweden, Israel, UAE, Czech, Denmark, Italy, Japan, Norway, Bahrain Scotland, Oman

*2 National Essential Drug List

*3 Sales royalties of Fasenra, marketed by AstraZeneca. Includes our own estimation.

YoY Analysis -Core OP-

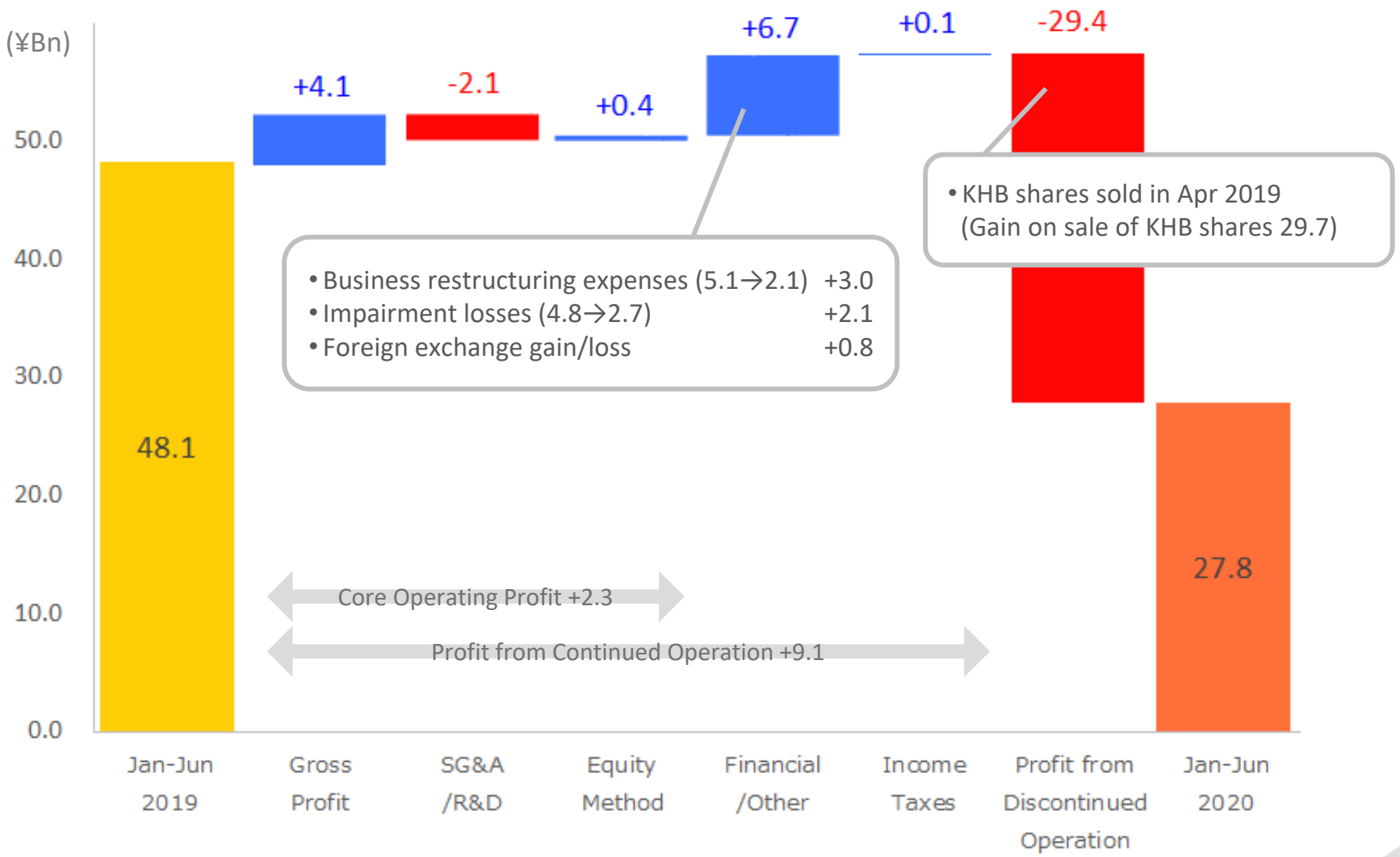
+2.3 billion yen (incl. forex effect -0.8)



- **Gross Profit +4.1 (incl. forex effect -1.5)**
  - Increased in conjunction with the rise in revenue.
- **SG&A -2.9 (incl. forex effect +0.7)**
  - Increased selling and launch readiness expenses in the EU/US, including Crysvita's profit sharing expenses in North America.
- **R&D +0.8 (incl. forex effect +0.1)**
- **Gain/Loss on Equity Method +0.4**
  - Sales of Hulio increased.

YoY Analysis -Profit-

Profit (Jan-Jun) -20.3 billion yen



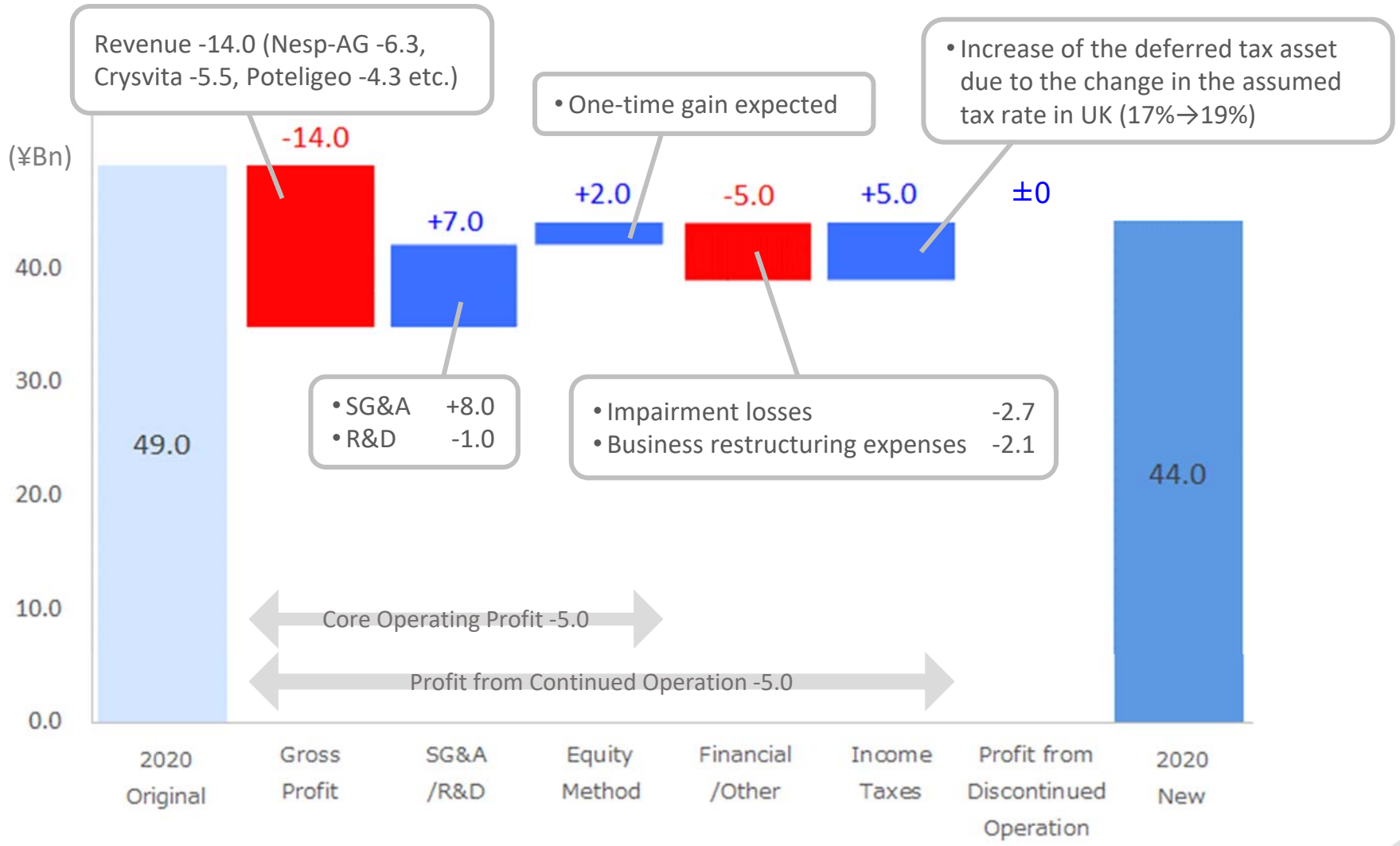
Summary of FY2020 Revised Plan

(Billion Yen / Rounded)

	2019 Results	2020 Original Plan	2020 New Plan	Changes
Revenue *[Overseas Ratio]*	305.8 *[39%]*	327.0 *[47%]*	313.0 *[47%]*	-14.0 (-4%)
Gross Profit *[Gross Profit margin]*	226.2 *[74%]*	250.0 *[76%]*	236.0 *[75%]*	-14.0 (-6%)
Core OP *[Core OP margin]*	59.4 *[19%]*	65.0 *[20%]*	60.0 *[19%]*	-5.0 (-8%)
Profit from continued operation	37.7	49.0	44.0	-5.0 (-10%)
Profit from discontinued operation	29.4	–	–	–
Profit	67.1	49.0	44.0	-5.0 (-10%)

FY2020 Original Plan vs Revised Plan

Profit (Jan-Dec) -5.0 billion yen



R&D Review

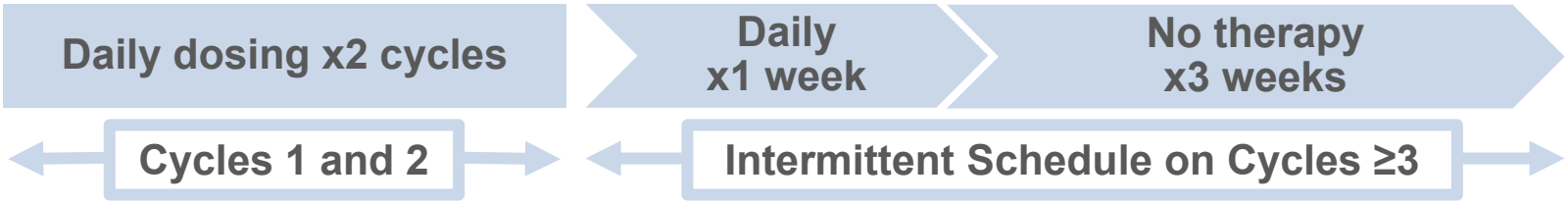
Key Development Updates in 20Q2

- Approval of KRN23 for the treatment of XLH in Hong Kong (April)
- Presentation of updated phase 1b study data on ME-401 for the treatment of follicular lymphoma and other B-cell malignancies at ASCO (May)
- Presentation of phase 2 study data on KHK7791 for the treatment of hemodialysis patients with hyperphosphatemia in Japan at ERA-EDTA (June)
- Approval of KRN321 for the treatment of renal anemia on dialysis in China (June)
- Approval of KHK4827 for the treatment of psoriasis in China (June)
- Approval of supplemental biologics license application of KRN23 for tumor-induced osteomalacia in the U.S. (June)

ME-401: Results of a Phase 1b study in B-cell malignancies

ASCO 2020
ClinicalTrials.gov Identifier: NCT02914938

Treatment: ME-401 monotherapy or in combination with rituximab on IS



Overall Response Rate and Adverse Events of Special Interest

Diagnosis	No. Evaluable Patients	ORR n (%)
FL	**36**	**30 (83%)**
By treatment group		
monotherapy	17	13 (76%)
Combo with rituximab	19	17 (89%)
CLL/SLL	**9**	**8 (89%)**
By treatment group		
ME-401 monotherapy	3	3 (100%)
ME-401 + rituximab	6	5 (83%)

Adverse Event of Special Interest (AESI)	Grade ≥ 3
Diarrhea or colitis	
Diarrhea	2 (3.5%)
Colitis	2 (3.5%)
Rash, all types	1 (1.8%)
ALT/AST elevation	1 (1.8%)
Stomatitis	0
Pneumonia/Infectious pneumonitis	0*
Non-Infectious pneumonitis	1 (1.8%)

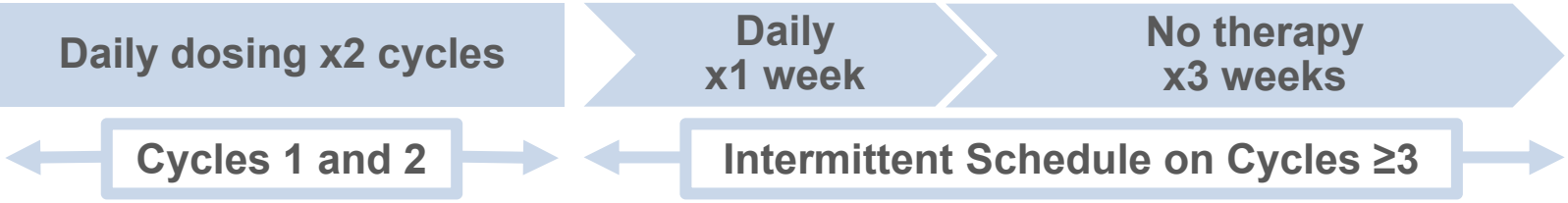
ME-401 on intermittent schedule showed a high response rate and was generally well tolerated in r/r FL and CLL/SLL

*1 patient with grade 5 COVID-19 pneumonia in Cycle 15.
IS: Intermittent schedule, 1 cycle: 28 days, ORR: Overall response rate, FL: Follicular Lymphoma, CLL/SLL: Chronic Lymphocytic Leukemia/Small Lymphocytic Lymphoma, r/r: Relapsed/Refractory

ME-401: Phase 2 TIDAL study in r/r FL patients

ClinicalTrials.gov Identifier: NCT03768505

- **Subjects: Patients with r/r FL after failure of at least two prior systemic therapies including chemotherapy and an anti-CD20 antibody**
- **Treatment: ME-401 on intermittent schedule as monotherapy**



- **Target number of cases: 120**
- **Study location: US, Europe, Oceania, South Korea, Taiwan**
- **Primary endpoint: Overall response rate**
- **Phase 2 clinical trial which may support an accelerated approval marketing application with the U.S. Food and Drug Administration**

TIDAL (**Tri**als of PI3K **D**elt**A** in Non-Hodgkin's **L**ymphoma)

KHK7791: Phase 2 (PB switching) study design

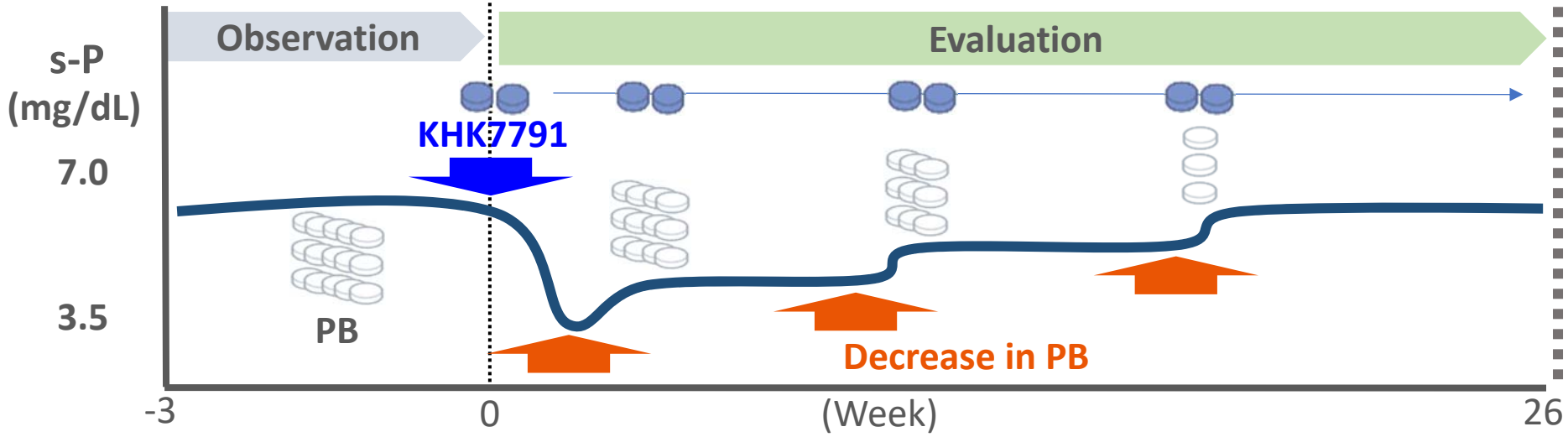
ClinicalTrials.gov Identifier: NCT03831607

- Open-label, single-arm, phosphate binder switch study for subjects with hyperphosphatemia undergoing hemodialysis

Screening 92 subjects → Pre-enrollment 83 subjects → Enrollment 67 subjects → Phosphate binder (PB) + KHK7791

Enrollment criteria
serum phosphorus levels (s-P)
: 3.5-7.0 mg/dL

Observation period (3 weeks) → Evaluation period (26 weeks)



- Primary endpoint: Achievement of 30% decrease in the mean of the total tablets number (PB+KHK7791) compared to baseline PB

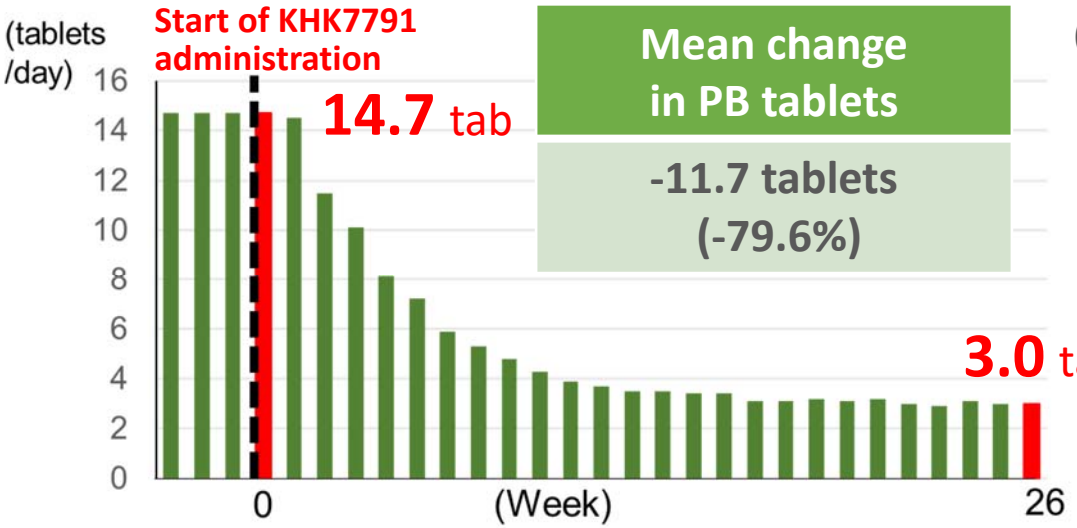
KHK7791: Phase 2 (PB switching) study results

Primary endpoint

	Achievement ratio	*P* value [95%CI]
30% decrease	71.6% (48/67)	<0.001 [59.3, 82.0]

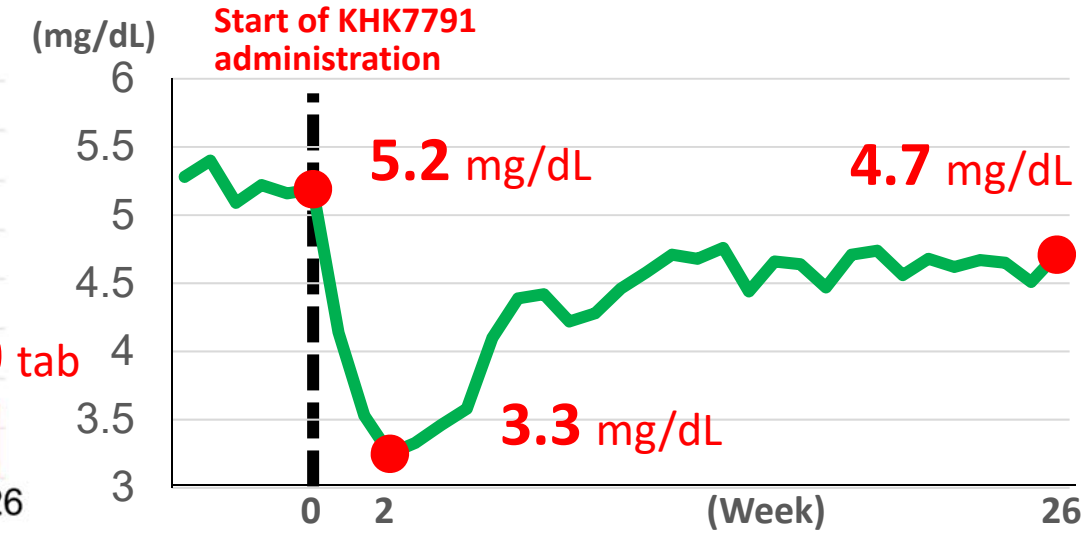
CI confidence interval.

Mean change in total PB tablets over time



Mean change in PB tablets
-11.7 tablets (-79.6%)

Mean change in s-P over time



More than 70% patients achieved a 30% decrease of the total number of PB and KHK7791.

Key Development Updates after June

- **Receipt of a positive opinion by the Committee for Medicinal Products for Human Use on the marketing authorization application for KRN23 for adult XLH in Europe (July)**

Business Topics

Impact of COVID-19 and Countermeasures

Supply Chains

- March to April: Disrupted logistics mainly in the EU, but no problems now
- No delays in the import and supply of raw materials
- Logistics costs are rising

Region Status

- **Japan (sales)**
  - In principle, working from home
  - Product recall, PV activities, and quality information: Responding to the requirements of medical institutions
  - Information dissemination: Telephone, e-mail, and web conferencing as needed
- **NA**
  - Major office locations closed or remote working and other flexible working practices encouraged until the end of July
- **EMEA**
  - Encourages flexible work styles, including telecommuting
  - Online training to prepare for the resumption of office and field activities
- **AP**
  - Depending on the infection situation by region or country, each business has its attendance system and other arrangements
  - China: Back to business as usual, but the transition to digital promotion is accelerated

Basic policy for working in Japan: Working from home. If the current situation continues, a 30% maximum attendance limit until the end of the year

Laboratories: Research work is conducted with adequate measures to prevent infection (Restrictions on the percentage of employees coming to work are lifted. Employees may work from home)

Production: Manufacturing operations are carried out with adequate infection prevention measures in place

Development: Cautiously following the authorities' guidance of each country

Our Supporting Activities Related to COVID-19

China	Zhong Nanshan Medical Foundation	Donation	March
Japan	National Institution of Infectious Diseases	Provided API	March - April
EMEA Counties	Supported local medical systems and patients (e.g. The Red Cross in Spain)	Donation	April
EMEA Countries	Supported healthcare professionals who are volunteering in front line services		
US	AmeriCares*	Donation	April
Singapore	Donated Singapore Association of Pharmaceutical Industry for providing personal PPE	Donation	April
Singapore	Agency for Integrated Care （Sector Emergency Fund）	Donation	
Japan	International Medical Volunteers Japan Heart	Donation	May

＊Supports globally including the US

Business topics

June Commercial launch of “Poteligeo” in Europe

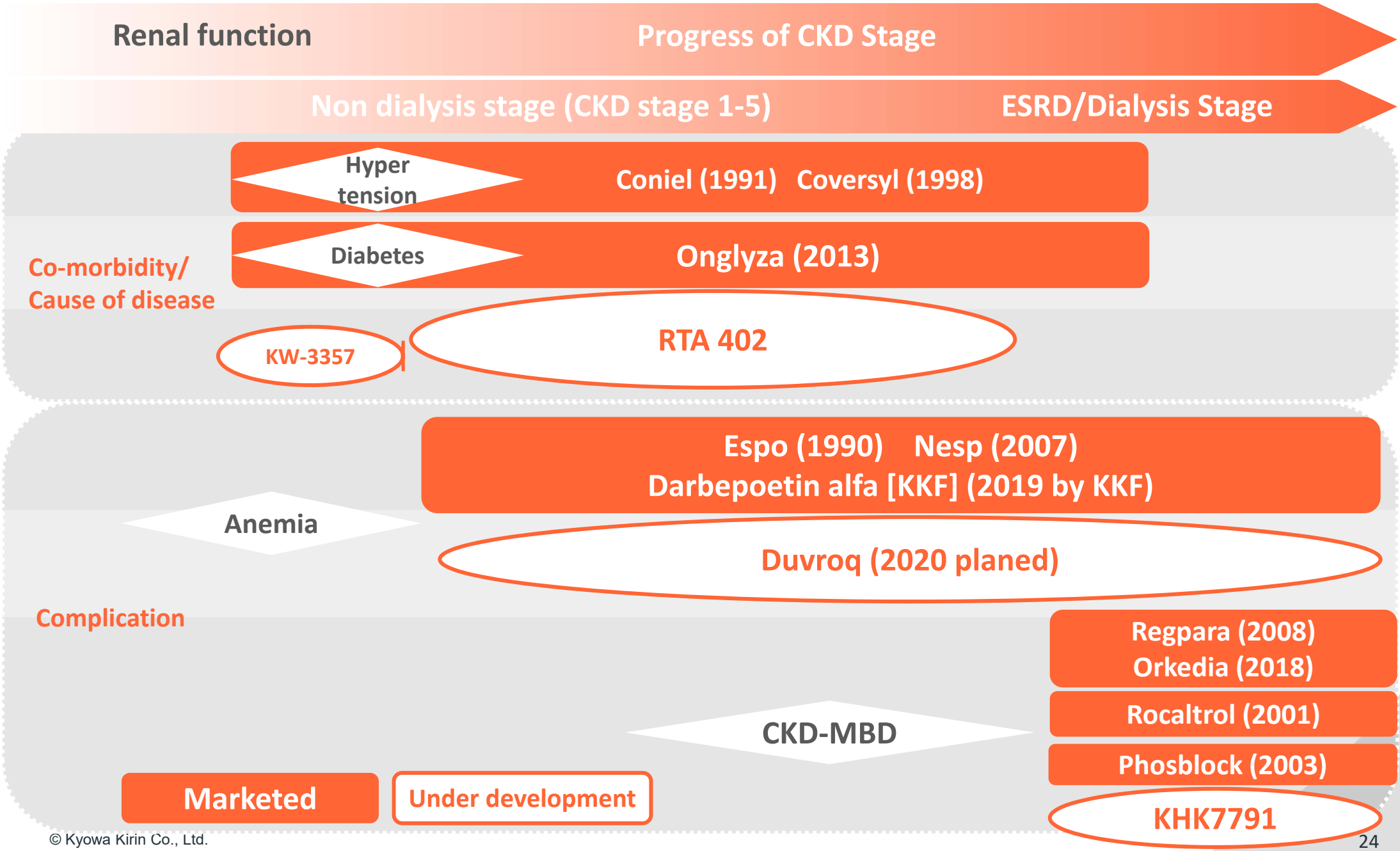
June Approval of “Duvroq” in Japan for Patients with Renal Anemia

June Approval of “Adalimumab (Genetical Recombination) [Adalimumab Biosimilar 1]” in Japan

July U.S. FDA Approval of “Hulio” (adalimumab-fkjp)

July Positive CHMP Opinion for “Equidacent” (FKB238), Biosimilar Bevacizumab

Product portfolio in Nephrology area (as of July 2020)



Appendix

FOREX Information

Average FOREX Rate

(Yen)

Currency	2019Q2 Results	2020Q2 Results	Changes	2020 New Plan
USD/JPY	110	109	-1	108
GBP/JPY	143	138	-5	135

2020Q2 FOREX Impact (YoY)

(Billion yen)

Currency	Revenue	Core OP
USD/JPY	-0.2	-0.1
GBP/JPY	-1.1	-0.4

FY2020 Currency Fluctuation Sensitivity (New)

(Billion yen)

Currency	Changes	Revenue	Core OP
USD/JPY	+1 yen	-0.8	-0.4
GBP/JPY	+1 yen	-0.3	-0.0

Key progress in development (2020 Q2)

Note: Listed events were completed between January 1st, 2020 and June 30th, 2020.

Month	Generic name Code	Indication	Country/region	Event
Jan.	Istradefylline KW-6002	Parkinson's disease	EU	Accepted MAA
Feb.	Pegfilgrastim KRN125	Chemotherapy induced febrile neutropenia	JP	Initiated phase 1 study
Feb.	Burosumab KRN23	Tumor-induced osteomalacia	US	Accepted sBLA submission & priority review designation
May	ME-401	follicular lymphoma and other B-cell malignancies	US	Presented phase 1b data at ASCO
June	Tenapanor KHK7791	Hemodialysis patients with hyperphosphatemia	JP	Presented phase 2 data at ERA-EDTA
June	Burosumab KRN23	Tumor Induced osteomalacia	US	Approved sBLA

[1] Filed indication is an adjunctive treatment to levodopa-based regimens in adult patients with Parkinson's disease experiencing "wearing-off" time

Submission plan of major pipeline

As of June 30th, 2020

Generic name Code	Indication	Country/region	2020 H1	2020 H2	2021
Brodalumab KHK4827	Psoriasis	CN	+		
Brodalumab KHK4827	Psoriasis	AS		+	
Brodalumab KHK4827	Axial spondyloarthritis	JP		+	
Brodalumab KHK4827	Axial spondyloarthritis Psoriatic arthritis	TW	Filed	+	
Burosumab[1] KRN23	XLH (adult)	EU		+	
Burosumab KRN23	XLH[2]	AS	+[4]	+	
Burosumab[1] KRN23	Tumor-induced osteomalacia	US	+		
Istradefylline KW-6002	Parkinson's disease	EU			+
Romiplostim AMG531	Aplastic anemia[3]	TW		+	
Romiplostim AMG531	ITP	CN			+

[1] Jointly developed with Ultragenyx
[2] Filed/approved indications are FGF23-related hypophosphatemic rickets and osteomalacia in KR
[3] Aplastic anemia in patients who have had an inadequate response to conventional therapy
[4] Hong Kong
AS: Asia, CN: China, EU: Europe, JP: Japan, US: United States, TW: Taiwan

+ Estimated time of regulatory decisions
Completed
Planned

Development plan of major pipeline

As of June 30th, 2020

Generic name Code	Indication	Country/region	2020	2021	2022~
Bardoxolone methyl RTA 402	Diabetic Kidney disease	JP	Phase 3		
Bleselumab[1] ASKP1240	Recurrence of FSGS in *de novo* kidney transplant	US	Phase 2		Phase 3
Entinostat KHK2375	Breast cancer	JP	Phase 2		
Evocalcet KHK7580	SHPT	AS	Phase 3		
KHK4083	Atopic dermatitis	JP US EU	Phase 2		Phase 3
KW-6356	Parkinson's disease	JP	Phase 2		Phase 3
Tenapanor KHK7791	Hyperphosphatemia under maintenance dialysis	JP	Phase 2		Phase 3
Antithrombin gamma KW-3357	Preeclampsia	JP	Phase 3		
Brodalumab KHK4827	Systemic sclerosis	JP	Phase 3		
Brodalumab KHK4827	Palmoplantar Pustulosis	JP	Phase 3		
Mogamulizumab KW-0761	HAM	JP	Phase 3		
Pegfilgrastim KRN125	Mobilization of HSCs into peripheral blood	JP	Phase 2		
Romiplostim AMG531	Aplastic anemia[2]	JP AS	Phase 2/3		

[1] Jointly developed with Astellas

[2] Aplastic anemia who were previously untreated with immunosuppressive therapy

AS: Asia, EU: Europe, JP: Japan, US: United States

On going
Planned

Estimated annual incidence/prevalence

Disease	Country /Region	Estimated # of Incidence (i) or Prevalence (p)	Source
ATL	Japan	i: 1,150 per year p: 2,000	i: Survey and countermeasures to HTLV-1 infection and related diseases in Japan. 2009 summary research report (Yamaguchi, 2010) p: Ministry of Health, Labour and Welfare: 2017 Patient survey (illness classification)
ATL	U.S.	i: 180 per year	US Lymphoid Malignancy Statistics by World Health Organization Subtypes (Lauren R et al., CA Cancer J Clin., 2016)
PTCL	Japan	p: 1,000	Ministry of Health, Labour and Welfare: 2017 Patient survey (illness classification)
CTCL	Japan	p: 2,000	Ministry of Health, Labour and Welfare: 2017 Patient survey (illness classification)
CTCL	U.S.	i: 1,500 per year	SEER Data (2001-2007)
XLH	Japan	p: 5,000 (adult), 1,000 (ped.)	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	Europe	p: 12,000 (adult), 3,000 (ped.)	Estimate based on reported prevalence of 1 in 20,000 people
XLH	U.S.	p: 12,000 (adult), 3,000 (ped.)	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/ENS	Japan	p: 30 (TIO)	2010 Ministry of Health, Labour and Welfare Epidemiological Research on abnormalities in Hormone Receptor Mechanisms
TIO/ENS	U.S.	p: 500 - 1,000	Survey by Ultragenyx Pharmaceutical
PD	Japan	p: 162,000	Ministry of Health, Labour and Welfare: 2017 Patient survey (illness classification)
PD	U.S.	p: more than 570,000	Study by Decision Resources
AD	U.S.	p: 9,000,000 - 17,000,000	Studies by Decision Resources and Global Data
CKD	Japan	p: 13,300,000	Clinical Practice Guidebook for Diagnosis and Treatment of Chronic Kidney Disease (2012)
AA	Japan	i: 1,000 per year	Cited from the website of Japan Intractable Diseases Information Center http://www.nanbyou.or.jp/entry/106 Accessed July 29, 2020.
ITP	Japan	i: 3,000 per year	Cited from the website of Japan Intractable Diseases Information Center http://www.nanbyou.or.jp/entry/157 Accessed July 29, 2020.
HAM	Japan	i: 30 per year p: 3,000 - 3,600	HTLV-1 associated myelopathy (HAM) practice guideline 2019
FL	U.S.	i: 15,000 per year	Cited from Cancer.net https://www.cancer.net/ Accessed July 29, 2020.

Crysvita - Collaboration with Ultragenyx -

	Kyowa Kirin Group	Ultragenyx
U.S.A /Canada	● Books sales ● For first 5 years, splits profits in half ● After 5 years, pays mid to high 20% range sales royalty	● For first 5 years, splits profits in half ● After 5 years, receives mid to high 20% range sales royalty
Europe	● Books sales ● Pay up to 10% sales royalty to Ultragenyx	● Receives up to 10% sales royalty
Latin America	● Receives low single-digit sales royalty from Ultragenyx	● Books sales ● Pays low single-digit sales royalty
Turkey	● Receives up to 20% sales royalty from Ultragenyx ● Retains an option to take over commercialization rights after a certain period	● Books sales ● Pays up to 20% sales royalty
Japan/Asia /ROW	● Books sales	

* Kyowa Kirin supplies commercial products in all regions.

List of acronyms

AA	Aplastic Anemia
AD	Atopic Dermatitis
ATL	Adult T-Cell Leukemia/Lymphoma
BS	Biosimilar
CKD	Chronic Kidney Disease
CTCL	Cutaneous T-Cell Lymphoma
DKD	Diabetic Kidney Disease
ENS	Epidermal Nevus Syndrome
FL	Follicular Lymphoma
FSGS	Focal Segmental Glomerulosclerosis
HAM	HTLV-1 Associated Myelopathy
HSC	Hematopoietic Stem Cell
ITP	Idiopathic Thrombocytopenic Purpura
PD	Parkinson’s Disease
PHPT	Primary Hyperparathyroidism
PTCL	Peripheral T-Cell Lymphoma
SHPT	Secondary Hyperparathyroidism
TIO	Tumor Induced Osteomalacia
XLH	X-linked Hypophosphatemia

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