Results Presentation
Fiscal 2019 Second Quarter

Kyowa Kirin Co., Ltd.

Agenda

Q2 Summary & Financial Review
R&D Review
Business Topics

President & CEO Masashi Miyamoto, Ph.D

Q&A

President & CEO Masashi Miyamoto, Ph.D
Executive Officer, Head of R&D Division Mitsuo Satoh, Ph.D
Executive Officer, Head of Finance Dept Motohiko Kawaguchi

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Q2 Summary & Financial Review

Summary of Q2 Results

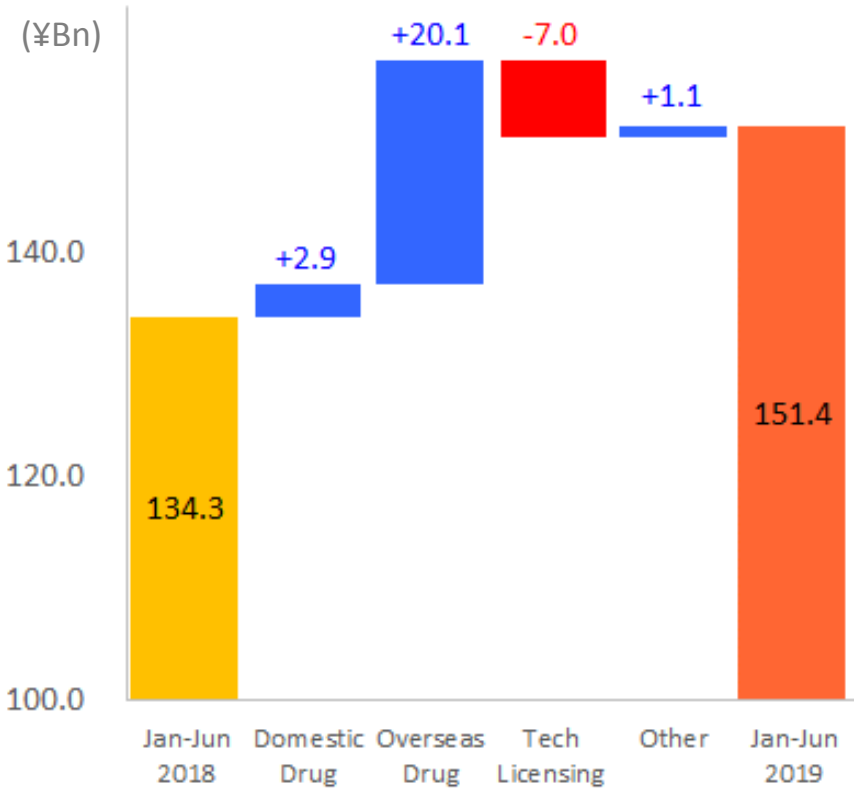
(Billion Yen / Rounded)

	2018Q2 Results [Cumulative]	2019Q2 Results [Cumulative]	Changes	2019Q4 Plan* [Cumulative]	Progress
Revenue	134.3	151.4	+17.1 (+13%)	305.0	50%
Gross Profit *[Gross Profit margin]*	97.4 *[72%]*	112.8 *[74%]*	+15.4 (+16%)	224.0 *[73%]*	50%
Core OP *[Core OP margin]*	28.4 *[21%]*	32.2 *[21%]*	+3.8 (+13%)	53.0 *[17%]*	61%
Profit from continued operation	31.5	18.7	-12.9 (-41%)	37.0	50%
Profit from discontinued operation	2.7	29.4	+26.7 (+970%)	31.0	95%
Profit	34.3	48.1	+13.8 (+40%)	68.0	71%

*Announced on February 5, 2019.

YoY Analysis -Revenue-

+17.1 billion yen
(incl. forex effect -1.4)



● Domestic Drug +2.9

- **Positive:** New product line including Rituximab BS (+3.0), Orkedia (+2.7), G-Lasta (+2.0), Dovobet (+0.6), Nouriast (+0.4) and Lumicef (+0.3) maintained steady growth.
- **Negative:** There are negative impacts by the drug price revision in April 2018. In addition, Regpara (-4.0) dropped due to the presence of a competing product and switch to Orkedia. Long-listed products such as Allelock (-1.0), Coniel (-0.5) and Depakene (-0.4) decreased mainly due to the penetration of generic drugs.

● Overseas Drug +20.1 (incl. forex effect -1.2)

- **EU/US:** Crysvita (+12.5) and Poteligeo (+5.4), launched last year, strongly penetrated into the market.
- **Asia:** Regpara (+0.9) recorded favorable sales mainly in China . Neulasta/Peglasta (+0.9) also increased due to the launch in Middle Eastern countries.

● Tech Licensing -7.0 (incl. forex effect -0.1)

- **Benralizumab:** Increased sales royalties were not able to offset the absence of milestone revenue booked last year.
- **Other:** There was gain on sale of Priority Review Voucher (US$80.6M × 50%) last year.

● Other -1.1

- Increase in the sales of FKB327 (Hulio)'s API.

Revenue of Major Items (Japan)

(Billion yen / Rounded)

Item	2018Q2 Results [Cumulative]	2019Q2 Results [Cumulative]	Changes	Reason	2019Q4 Plan*2 [Cumulative]	Progress
Nesp+Authorized ver.*1	25.6	25.6	+0.1 (+0%)		48.4	53%
Regpara	7.8	3.8	-4.0 (-51%)	A competitor's penetration & switch to Orkedia	5.1	74%
Orkedia	0.4	3.0	+2.7 (+717%)	Launched on May 2018	9.5	32%
G-Lasta	9.5	11.5	+2.0 (+21%)	Steady market penetration	22.8	50%
Rituximab BS	1.1	4.2	+3.0 (+262%)	Launched on Jan 2018	8.4	50%
Allelock	7.5	6.4	-1.0 (-14%)	Generic drugs' market penetration	9.3	69%
Patanol	9.7	9.9	+0.2 (+2%)		11.3	88%
Nouriast	4.4	4.8	+0.4 (+8%)	Steady market penetration	10.0	48%
Technology licensing	1.5	1.7	+0.2 (+12%)		4.4	37%

*1 Authorized version of Nesp "Darbepoetin Alfa Injection Syringe [KKF]" is to be released since August 5, 2019.
*2 Announced on February 5, 2019.

Revenue of Major Items (Overseas)

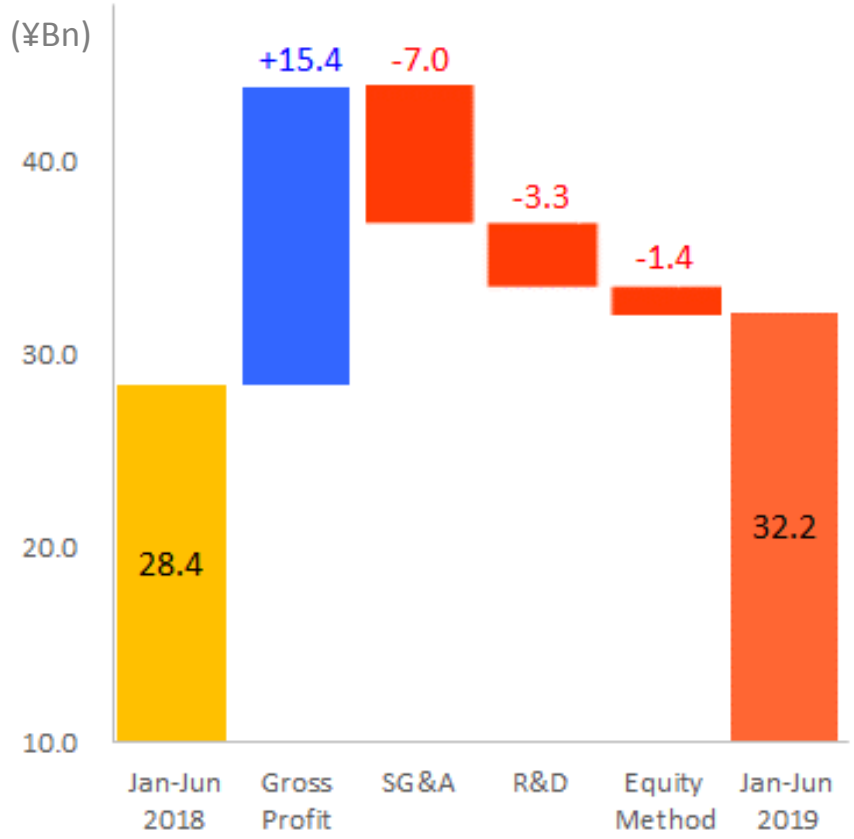
(Billion yen / Rounded)

Item	2018Q2 Results [Cumulative]	2019Q2 Results [Cumulative]	Changes	Reason	2019Q4 Plan*4 [Cumulative]	Progress
Crysvita*1	0.8	13.4	+12.5 (+1474%)	Launched in Apr 2018	Undisclosed	—
North America		9.9				
Europe & other		3.5				
Poteligeo	—	5.4	+5.4	Launched in Oct 2018	10.0	54%
Abstral	6.5	5.8	-0.6 (-10%)		12.3	47%
Technology licensing	12.4	5.2	-7.2 (-58%)	Benralizumab milestone & Crysvita PRV*3 in 2018	12.9	40%
Benralizumab Royalty*2	0.5	3.8	+3.3 (+639%)	Launched in 2018	Undisclosed	—

*1 In January, sales started in UK (England etc) at the list price of GBP2,992 per 10mg vial.
Since May, the List price in Germany has been revised to €2,550 per 10mg vial (from €3,388).
Launched countries excluding South America as of June 30, 2019:
USA, Canada, Germany, Netherland, Luxembourg, England, Wales, North Ireland, Slovakia, Sweden, Israel, UAE
*2 Sales royalties from Fasenra marketed by AstraZeneca. Includes our own estimation.
*3 PRV = Priority Review Voucher
*4 Announced on February 5, 2019.

YoY Analysis -Core OP-

+3.8 billion yen (incl. forex effect -0.2)



- **Gross Profit +15.4 (incl. forex effect -1.0)**
  - Increased in conjunction with the rise in the revenue. Gross profit margin up by 2 points, from 72% to 74%.

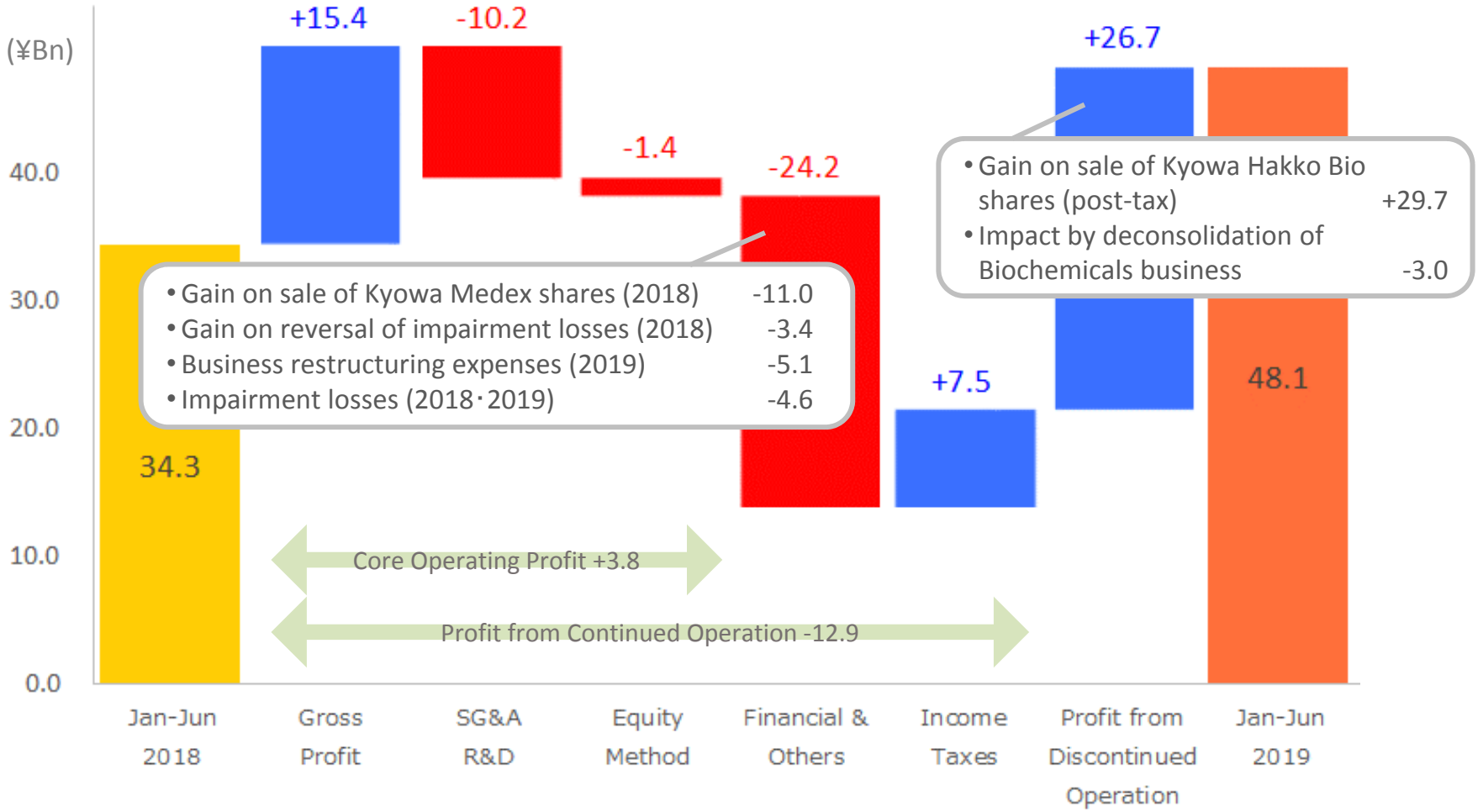
- **SG&A -7.0 (incl. forex effect +0.7)**
  - Increased selling and launch readiness expenses in the EU/US. *Including Crysvita's profit sharing expenses in North America.

- **R&D -3.3 (incl. forex effect +0.1)**
  - **Negative:** KHK4083 (P2 initiated in Oct 2018), RTA402 (P3 initiated in May 2018), KW-6356 (P2 initiated in Nov 2018), etc.

- **Gain/Loss on Equity Method -1.4**
  - Decreased due to the absence of FKB327-related revenue from the license-out of EU commercial rights and the achieved development milestone booked last year.

YoY Analysis -Profit-

Profit (Jan-Jun) +13.8 billion yen



R&D Review

Key development updates in 19Q2

- Application for approval of KHK4827 for the treatment of psoriasis in China (April)
- Application for approval of KRN23 for the treatment of XLH in Korea[1] (May) and China (June)
- Completion of the patient enrollment in the phase 3 AYAME study of RTA 402 for the treatment of DKD in Japan (June)
- Approval of AMG531 for the treatment of aplastic anemia in Japan (June)

[1]Filed indications are FGF23-related hypophosphatemic rickets and osteomalacia

Business Topics

Business Topics

- Kyowa Kirin Frontier to sell Darbepoetin Alfa Injection Syringe [KKF] (August)

- Kyowa Kirin buys back Tivozanib non-oncology rights from Aveo Oncology (August)

Appendix

FOREX Information

Average FOREX Rate

(Yen)

Currency	2018Q2 Results	2019Q2 Results	Changes	2019Q4 Plan
USD/JPY	109	110	+1	110
GBP/JPY	151	143	-8	145

YoY FOREX Impact

(Billion yen)

Currency	On Revenue	On Core OP
USD	+0.1	+0.0
GBP	-1.2	+0.0

Key progress in development (2019 H1)

Note: Listed events were completed between January 1st ,2019 and June 30th, 2019.

Month	Generic name Code	Indication	Country/region	Event
Jan.	Burosumab KRN23	XLH[1]	**JP** US EU RoW	Filed
Feb.	Tenapanor KHK7791	Hyperphosphatemia under maintenance dialysis	**JP** US EU RoW	Initiated phase 2 study
Apr.	Istradefylline KW-6002	Parkinson's disease	JP **US** EU RoW	Accepted resubmission
Apr.	Evocalcet KHK7580	Hypercalcemia in patients with parathyroid carcinoma or primary hyperparathyroidism	**JP** US EU RoW	Filed additional indications
Jun.	Brodalumab KHK4827	Systemic sclerosis	**JP** US EU RoW	Initiated phase 3 study
Jun.	Romiplostim AMG531	Aplastic anemia in patients with inadequate response to conventional therapy	**JP** US EU RoW	Approved additional indication

[1]Filed indications are FGF23-related hypophosphatemic rickets and osteomalacia

Submission plan of major pipeline

As of June 30th, 2019

Generic name Code	Indication	Country/region	2019	2020	2021~
Burosumab[1] KRN23	XLH (adult)	EU	Submission	+	
Burosumab KRN23	XLH[2]	JP	Filed	+	
Burosumab KRN23	XLH	AS	Filed[3]	+	
Evocalcet KHK7580	PHPT	JP	Filed	+	
Istradefylline KW-6002	Parkinson's disease	US	Filed	+	
Romiplostim AMG531	Aplastic anemia	JP	+		
Romiplostim AMG531	Aplastic anemia	KR	Submission / +		
Romiplostim AMG531	ITP	CN		Submission	+
Brodalumab KHK4827	Psoriasis	AS	Filed[3]	+	
Brodalumab KHK4827	Axial spondyloarthritis	JP, AS	Phase 3	Submission / +	
Mogamulizumab KW-0761	HAM	JP	Phase 3	Submission / +	

[1] Jointly developed with Ultragenyx
[2] Filed indications are FGF23-related hypophosphatemic rickets and osteomalacia in JP and KR
[3] Korea and China
AS: Asia, CN: China, EU: Europe, JP: Japan, KR: Korea, US: United States

+ Estimated time of regulatory decisions
Completed
On going
Planned

Development plan of major pipeline

As of June 30th, 2019

Generic name Code	Indication	Country/region	2019	2020	2021~
Bardoxolone methyl RTA 402	Diabetic Kidney disease	JP	Phase 3		
Bleselumab[1] ASKP1240	Recurrence of FSGS in *de novo* kidney transplant	US	Phase 2		
Entinostat KHK2375	Breast cancer	JP	Phase 2		
Evocalcet KHK7580	SHPT	AS	FPI	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	FPI	Phase 2	Phase 3
KHK4827 Brodalumab	Systemic sclerosis	JP	FPI	Phase 3	
Romiplostim AMG531	Aplastic anemia[2]	JP	FPI	Phase 3	
Pegfilgrastim KRN125	Mobilization of HSCs into peripheral blood	JP	FPI	Phase 2	

[1] Jointly developed with Astellas

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

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

Completed
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 (as of July, 2019) http://www.nanbyou.or.jp/entry/106
HAM	Japan	i: 30 per year p: 3,000 - 3,600	HTLV-1 associated myelopathy (HAM) practice guideline 2019

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 from Kyowa Kirin International (KKI)
Europe	● Books sales ● Pay up to 10% sales royalty to Ultragenyx	● Receives up to 10% sales royalty from KKI
Latin America	● Receives low single-digit sales royalty from Ultragenyx	● Books sales ● Pays low single-digit sales royalty to Kyowa Kirin
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 to KKI
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
FSGS	Focal Segmental Glomerulosclerosis
HAM	HTLV-1 Associated Myelopathy
HSC	Hematopoietic Stem Cell
ITP	Idiopathic (immune) 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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