Event Summary

[Event Name] 40th Annual J.P. Morgan Healthcare Conference

[Date] January 10, 2022 (EST)

[Speaker] Masashi Miyamoto, Ph.D.
Representative Director of the Board, President and Chief Executive Officer
Delivering Life-changing Value as a Global Specialty Pharma

J.P. Morgan Healthcare Conference
January 10, 2022

Masashi Miyamoto, Ph.D.
Representative Director of the Board,
President and Chief Executive Officer

This document contains certain forward-looking statements relating to such items as the Company’s forecasts, targets and plans (including those of its domestic and overseas subsidiaries). 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 actual results may differ substantially due to uncertain factors.

These uncertain factors include, but are not limited to, inherent risks in the business activities of the pharmaceutical industry in Japan and overseas, intellectual property risks, risk of product side effects, legal regulation risks, product defect risks, risks of changes to 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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In order to make this vision a reality by 2030, we focus on: Provide pharmaceuticals for unmet medical needs, Address patient-centric healthcare needs, and Retain the trust of a society.
Kyowa Kirin was created from the merger of Kyowa Hakko and Kirin Pharma in 2008. At that time, both companies had this long legacy of fermentation technology applied to pharmaceutical manufacturing and also enjoyed long-lasting relationship with research institutions and biotech companies.

For example, Kirin Pharma had a 30-year joint venture with Amgen. Both companies established antibody technologies such as POTELLIGENT and fully human antibody producing technologies, which have spurred innovations in our portfolio.

After the merger, the company began to seek to expand its operation globally. Over the past 13 years, we have followed that plan expanding through organic growth and targeted acquisitions.

Since 2018, we have received FDA approval on three medicines discovered and developed in-house, CrysVita, Poteligeo, and Nourianz.
As you may know, ADCC is a critical function of the immune system that enhances the ability of antibodies to kill target cell. This POTELLIGENT technology has been used for our products and rug candidates. In addition,
it has been licensed out and utilized by numerous major pharmaceutical and biotech companies in their pipelines.

The marketed products in which we have applied this technology include Poteligeo, our drug for cutaneous T-cell lymphoma; Fasenra, an asthma drug licensed out to AstraZeneca; and GSK's Blenrep, a drug for multiple myeloma.

We also have our proprietary human antibody producing technologies applied for several drugs and drug candidates. The products launched using this technology include Crysvita, a treatment for X-linked hypophosphatemia.

In addition, KHK4083, as known as AMG 451, is being developed for the treatment of atopic dermatitis by applying both technologies.

The first is our bispecific antibody technology that would create a rich drug pipeline. We are working hard to have this technology to enter clinical trials. Also, we hope that open innovation will lead to the discovery of innovative new drugs and the creation of fundamental technologies to support them.

We value our partnership with outside collaborators as they are essential to reinforce the strength of our technologies into life-changing value.
Strong Growth of Global 3 Brands (G3B)

**Crysvita**
(Burosumab, an anti-FGF23 fully human monoclonal antibody)
- X-linked hypophosphatemia (XLH)
- Tumor-induced osteomalacia (TIO)

**Poteligeo**
(Mogamulizumab, an anti-CCR4 humanized antibody)
- Mycosis fungoides (MF)
- Sézary syndrome (SS)

**Nouriast/Nourianz**
(Istrilestine, an approved α-synuclein agonist)
- Parkinson’s disease (PD) experiencing “off” episodes

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**Crysvita**
(Burosumab, an anti-FGF23 fully human monoclonal antibody)

- **2000**: Cloning of FGF23 cDNA
- **2013**: Identify Function
- **2018**: Collaborate with Ultragenyx
- **2019**: Launch in EU (pediatrics & adults) 1
- **2020**: Launch in Japan
- **2023**: To initiate own sales in US

**Indications**
- **US**: XLH, TIO
- **EU**: XLH, TIO
- **JP**: FGF23-related hypophosphatemic rickets and osteomalacia

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**Annual sales growth** 1 (NA = MEA HP)

<table>
<thead>
<tr>
<th>Year</th>
<th>2018</th>
<th>2019</th>
<th>2020</th>
<th>2021 (Plan)</th>
<th>[BE: JPY]</th>
</tr>
</thead>
<tbody>
<tr>
<td>US</td>
<td>7.7</td>
<td>32.6</td>
<td>58.2</td>
<td>82.7</td>
<td>710</td>
</tr>
<tr>
<td>EU</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>2,000</td>
</tr>
<tr>
<td>JP</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>3,000</td>
</tr>
</tbody>
</table>

**Number of patients** 2 (global total)

<table>
<thead>
<tr>
<th>Year</th>
<th>2018</th>
<th>2019</th>
<th>2020</th>
</tr>
</thead>
<tbody>
<tr>
<td>US</td>
<td>710</td>
<td>2,000</td>
<td>3,000</td>
</tr>
</tbody>
</table>

1. Marketed by Ultragenyx (Pharmaceutical Inc.)
2. Excl. patients under Early Access Program and patients who have not started the reimbursement process
**Crysvita**

Global Evidence Generation Project

- **Europe**
  - XLH Registry

- **Asia**
  - SUNFLOWER

- **Americas**
  - DMP*

* Disease Monitoring Program

https://whink.eu/

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**Poteligeo**

**POTELIGEO**
Mogamulizumab, an anti-CC chemokine receptor type 4 (C-C chemokine receptor type 4) (CCR4) humanized antibody

- **2012**
  - Establish POTELIGENT® technology
- **2018**
  - Launch in Japan
- **2020**
  - Launch in US
  - Launch in EU

**Annual sales growth** (NA+EMA+JP)

<table>
<thead>
<tr>
<th>Year</th>
<th>US</th>
<th>EU</th>
<th>JP</th>
</tr>
</thead>
<tbody>
<tr>
<td>2018</td>
<td>3.9</td>
<td>12.8</td>
<td>13.6</td>
</tr>
<tr>
<td>2019</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2020</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2021</td>
<td></td>
<td></td>
<td>19.3</td>
</tr>
</tbody>
</table>

* Excl. patients under Early Access Program and patients who have not started the reimbursement process

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Nourianz is the first new mechanism of action drug for Parkinson’s disease treatment in decades and is an innovative adenosine A2a receptor antagonist that we discovered.

Next-generation Strategic Products

<table>
<thead>
<tr>
<th>Country / region*2</th>
<th>Indication*2</th>
<th>Approval year*2</th>
<th>Total addressable market*4</th>
<th>No. of patients*5</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>KHK4083/ AMG 451</strong></td>
<td>Atopic dermatitis</td>
<td>2025/2026</td>
<td>★★★</td>
<td>16,000K</td>
</tr>
<tr>
<td><strong>KW-6356</strong></td>
<td>Parkinson’s disease</td>
<td>2025</td>
<td>★★★</td>
<td>3,500K</td>
</tr>
<tr>
<td><strong>ME-401 Zandelki</strong></td>
<td>Follicular lymphoma Marginal zone lymphoma</td>
<td>2023</td>
<td>★★★</td>
<td>~800K</td>
</tr>
<tr>
<td><strong>RTA 402 Ramiprilac methyl</strong></td>
<td>Alport syndrome Diabetic kidney disease (DKD) Autosomal dominant polycystic kidney disease (ADPKD)</td>
<td>2022 2025</td>
<td>★★★</td>
<td>2,500K+</td>
</tr>
<tr>
<td><strong>KHK7791 Tenaparit</strong></td>
<td>Hyperphosphatemia under maintenance dialysis</td>
<td>2023</td>
<td>★☆☆</td>
<td>250K</td>
</tr>
</tbody>
</table>

*1 Countries or regions where Kyowa Kirin currently has marketing rights and will launch products (will conduct marketing activities; products may not be launched in all countries or regions shown in the table)
*2 Depicted indications on the date of this document; indications may ultimately differ to associations due to status of approvals from regulatory authorities
*3 Approved total addressable market, which is the sum of all products for the indications shown in *1, in all countries or regions defined in *1, not projected sales or the Company’s targets
*4 A total number of estimated patients in all countries or regions defined in *1
*5 The size of the total addressable market and patient numbers are based on our estimates

As of Feb 4, 2021
KHK4083/AMG 451 targets OX40 as a potential novel target for AD treatment

- Activation of Th2 and other T-cell subsets is central in atopic dermatitis (AD).
- The OX40-OX40L axis plays a critical role in long-lasting T-cell responses.
  - OX40 is expressed by activated T cells after antigen recognition and binds OX40L on APCs, facilitating the effector function of T cells.
- KHK4083/AMG 451 is a fully human, anti-OX40, non-fucosylated IgG1 mAb with enhanced ADC (antibody-dependent cell-mediated cytotoxicity) that acts by:
  - Partially depleting activated T cells.
  - Blocking T-cell clonal expansion and memory T-cell formation.

As is commonly known, activation of Th2 cells is central in atopic dermatitis. KHK4083 blocks the OX40 signaling pathway, which plays a major role in upstream T-cell activation by OX40 inhibition or OX40 positive cell depletion.

As expected, clinical results based on this mechanism of action have been obtained to date.

KHK4083/AMG 451: Achieved Primary Endpoint in Phase 2 Study

Phase 2 Study Design

<table>
<thead>
<tr>
<th>Screening</th>
<th>Treatment A (Up to Week 18)</th>
<th>Treatment B (Week 18-Week 36)</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adults with chronic AD</td>
<td>Placebo</td>
<td>KHK4083 150 mg SC Q2W</td>
<td>Follow-up every 4 weeks</td>
</tr>
<tr>
<td>Randomization (1:1:1:1)</td>
<td>KHK4083 600 mg SC Q2W</td>
<td>KHK4083 300 mg SC Q2W</td>
<td>KHK4083 600 mg SC Q2W</td>
</tr>
<tr>
<td>Location: US, Japan, Germany, and Canada</td>
<td>KHK4083 150 mg SC Q2W</td>
<td>KHK4083 300 mg SC Q2W</td>
<td>KHK4083 600 mg SC Q2W</td>
</tr>
</tbody>
</table>

Primary Endpoint: % change from baseline in EASI score at W16 (LOCF, FAS)

- KHK4083 150 mg Q4W: n=52, Placebo: n=57
- KHK4083 600 mg Q2W: n=52, Placebo: n=57

**p<0.01 for difference versus placebo**
To maximize the product value of KHK4083/AMG 451 for atopic dermatitis and its potential in other autoimmune disease treatments, Kyowa Kirin and Amgen have entered into an agreement to develop and commercialize KHK4083/AMG 451 jointly.

Our agreement to co-development and co-commercialize KHK4083 made news in 2021. With Amgen, we are planning to initiate a Phase 3 study of KHK4083 in the first half of 2022. We will also explore the potential use of KHK4083 in indications beyond atopic dermatitis.
Zandelisib has innovative molecular and biologic properties that show the potential to avoid the safety issues that are common among other PI3K treatments.

On November 4th, 2021, FDA granted orphan drug designation to Zandelisib for follicular lymphoma.
Given this, bardoxolone methyl has opportunities as it activates the system that plays an important role in defense responses against oxidative stress as its convincing clinical data and the priority review designation support its potential market opportunity in Japan, and as it could be the first drug addressing the need for renal function protection and improvement.
The Phase 3 AYAME study is scheduled to be complete this year, and then the topline data is available.

There are various catalysts for 2022, so please stay tuned. In particular, I hope you will look forward to the start of several Phase 3 clinical trials including KHK4083 and KW-6356. We will announce in a timely manner when Phase 3 studies actually start.
In order to do so, we will work on open innovation by developing our own technologies as well as successfully incorporating useful technologies from outside, and we will surely produce life-changing value that will make patients smile. We will continue to strive for this goal.

Thank you very much for your time and attention for Kyowa Kirin. [END]