Outline of Consolidated Financial Results for the 1st Quarter Ended June 30, 2021

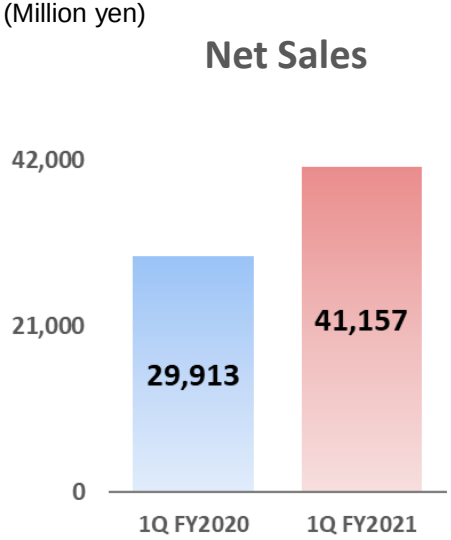
August 10, 2021
NIPPON SHINYAKU CO., LTD.

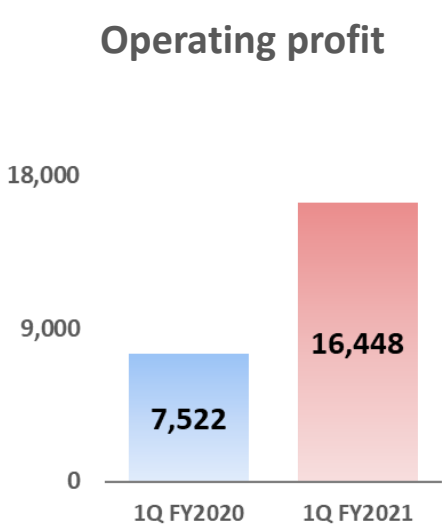
NIPPON SHINYAKU CO., LTD.

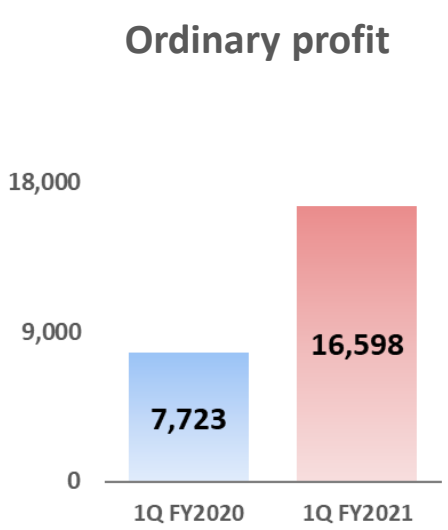
1Q FY2021 Summary

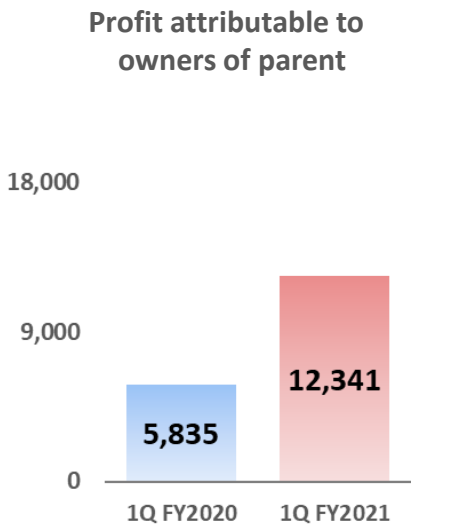
- **Net sales** : 41,157 million yen (+37.6%)
- **Operating profit** : 16,448 million yen (+118.6%)
- **Ordinary profit** : 16,598 million yen (+114.9%)
- **Profit attributable to owners of parent** : 12,341 million yen (+111.5%)

(Million yen)

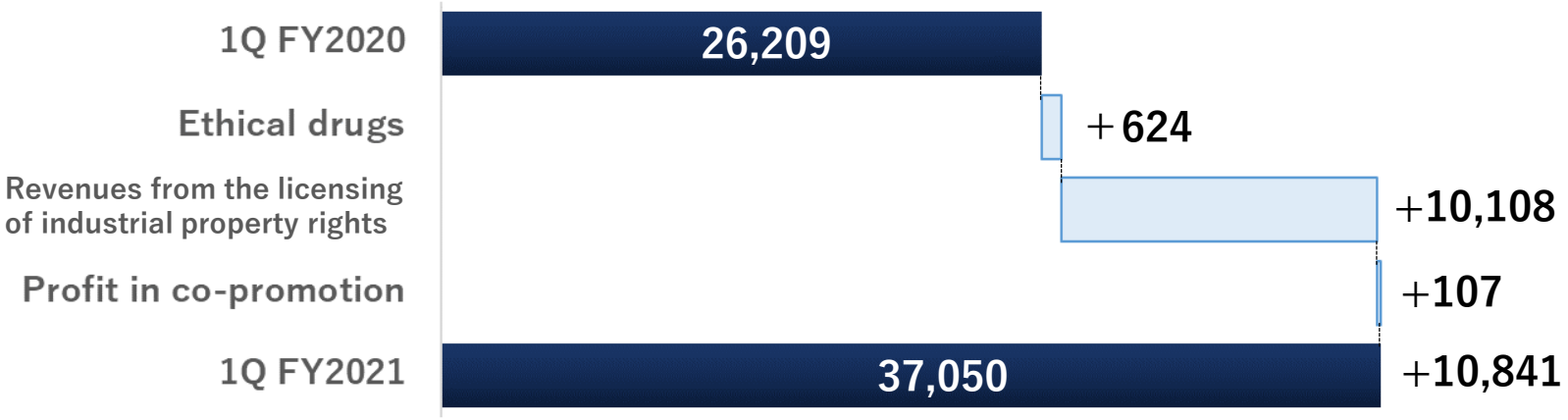








Segmental Review - Pharmaceuticals -



(Million yen)	1Q FY2020		1Q FY2021		YoY Change	
	Results	Ratio	Results	Ratio	Amt	%
Ethical drugs	18,919	72.2%	19,543	52.7%	+624	+3.3%
Revenues from the licensing of industrial property rights	5,105	19.5%	15,214	41.1%	+10,108	+198.0%
Profit in co-promotion	2,184	8.3%	2,292	6.2%	+107	+4.9%
Net sales	26,209	100.0%	37,050	100.0%	+10,841	+41.4%

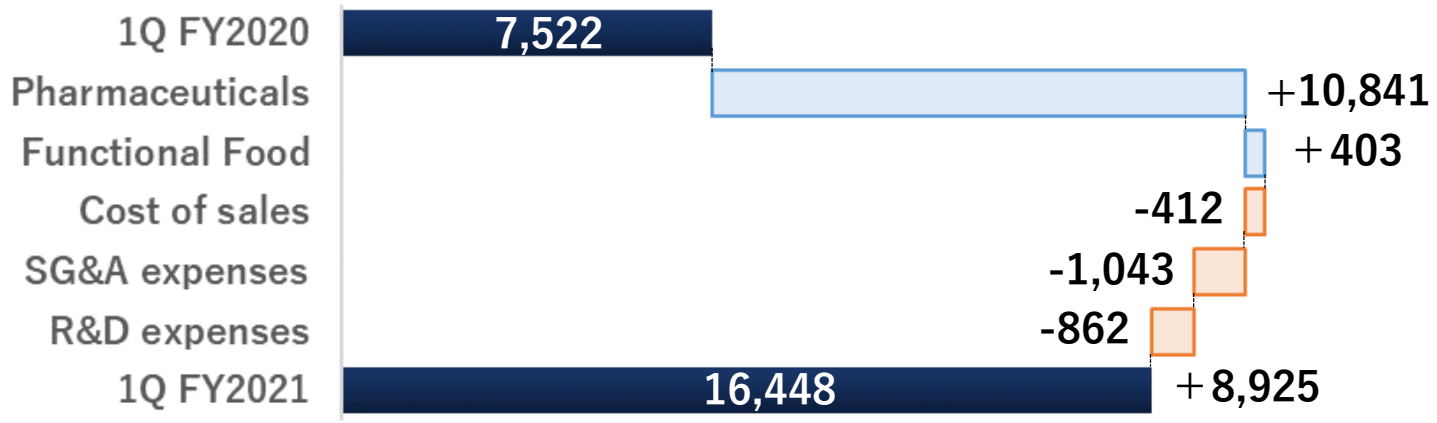
Net sales increased by 41.4% through sales of Ethical drugs and Revenues from the licensing of industrial property rights which contains of royalty revenue from Uptravi's overseas sales and gain on sales from the priority review voucher.

Segmental Review - Functional Food -

(Million yen)	1Q FY2020		1Q FY2021		YoY Change	
	Results	Ratio	Results	Ratio	Amt	%
Protein preparations	2,477	66.9%	2,759	67.2%	+282	+11.4%
Preservatives	610	16.5%	653	15.9%	+42	+6.9%
Health food ingredients	250	6.8%	275	6.7%	+24	+9.7%
Others	364	9.8%	418	10.2%	+54	+14.8%
Net sales	3,703	100.0%	4,106	100.0%	+403	+10.9%

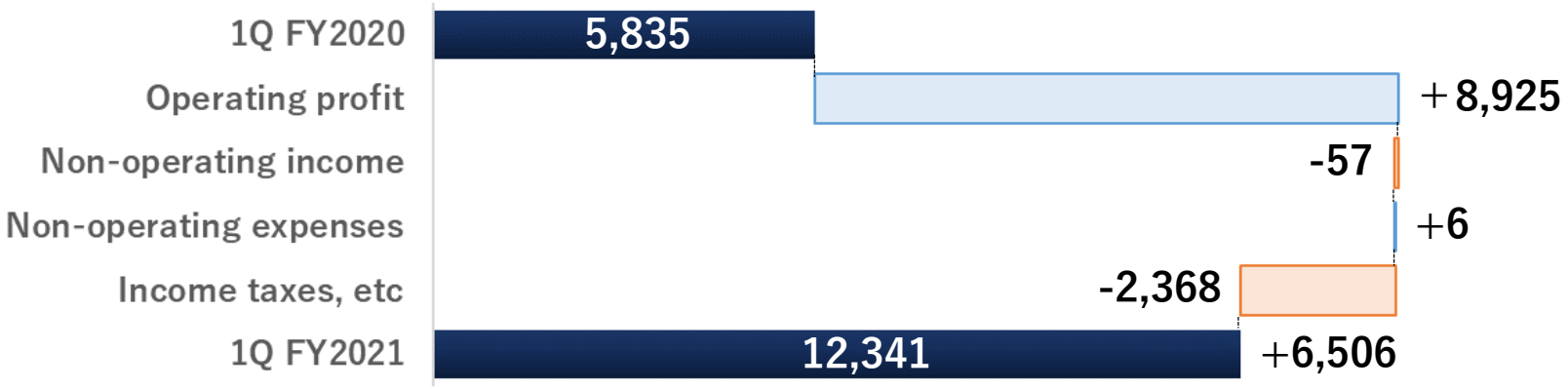
Net sales increased by 10.9% through sales from Protein preparations and Preservatives.

Operating profit



(Million yen)	1Q FY2020		1Q FY2021		YoY Change	
	Results	Ratio	Results	Ratio	Amt	%
Net sales	**29,913**	**100.0%**	**41,157**	**100.0%**	**+11,244**	**+37.6%**
(Pharmaceuticals)	(26,209)	(87.6%)	(37,050)	(90.0%)	(+10,841)	(+41.4%)
(Functional Food)	(3,703)	(12.4%)	(4,106)	(10.0%)	(+403)	(+10.9%)
Operating expenses	**22,390**	**74.9%**	**24,709**	**60.0%**	**+2,319**	**+10.4%**
Cost of sales	**12,818**	**42.9%**	**13,231**	**32.1%**	**+412**	**+3.2%**
SG&A expenses	**6,734**	**22.5%**	**7,778**	**18.9%**	**+1,043**	**+15.5%**
R&D expenses	**2,836**	**9.5%**	**3,699**	**9.0%**	**+862**	**+30.4%**
Operating profit	**7,522**	**25.1%**	**16,448**	**40.0%**	**+8,925**	**+118.6%**

Profit attributable to owners of parent



(Million yen)	1Q FY2020 Results	1Q FY2021 Results	YoY Change Amt	YoY Change %
Operating profit	7,522	16,448	+8,925	+118.6%
Non-operating income	427	369	-57	-13.4%
Non-operating expenses	226	220	-6	-2.9%
Ordinary profit	7,723	16,598	+8,874	+114.9%
Income taxes, etc	1,887	4,256	+2,368	+125.4%
Profit attributable to owners of parent	5,835	12,341	+6,506	+111.5%

Business Forecast for FY2021

(Million yen)	FY2020		FY2021			
	1Q Results	FY Results	1Q Results	Progress for 1H	1H Forecasts	FY Forecasts
Net sales	**29,913**	**121,885**	**41,157**	**59.2%**	**69,500**	**135,000**
(Pharmaceuticals)	(26,209)	(106,478)	(37,050)	(60.0%)	(61,800)	(119,300)
(Functional Food)	(3,703)	(15,406)	(4,106)	(53.3%)	(7,700)	(15,700)
Operating profit	**7,522**	**26,134**	**16,448**	**91.4%**	**18,000**	**28,000**
Ordinary profit	**7,723**	**26,760**	**16,598**	**91.2%**	**18,200**	**28,500**
Profit attributable to owners of parent	**5,835**	**20,702**	**12,341**	**94.9%**	**13,000**	**21,000**

Net sales and each profit have progressed toward achievement of 1H, FY forecasts.

Status of Product Pipeline

NIPPON SHINYAKU CO., LTD.

R&D Compounds (Domestic)

Code No. (Generic name) <Origin>	Application type	Indications	Preparation for development	Preparation for PI	PI	PII	PIII	NDA	Launch
NS-065/NCNP-01 (viltolarsen) <in-house>	NME	Duchenne muscular dystrophy					PIII in progress		
NS-304 (selexipag) <in-house>	New indication	Chronic thromboembolic pulmonary hypertension							
		Arteriosclerosis obliterans							
		Lumbar spinal stenosis							
	New dose	Pediatric pulmonary arterial hypertension							
NS-32 (ferric derisomaltose) <in-license>	NME	Iron deficiency anemia							
ZX008 <in-license>	NME	Dravet syndrome Lennox-Gastaut syndrome							
NS-580 <in-house>	NME	Endometriosis							
NS-87 <in-license>	New combination	Secondary acute myeloid leukemia							
NS-229 <in-house>	NME	Inflammatory diseases							
NS-917 <in-license>	NME	Relapsed/refractory acute myeloid leukemia							
NS-401 (tagraxofusp) <in-license>	NME	Blastic plasmacytoid dendritic cell neoplasm							

R&D Compounds (Overseas)

Code No. (Generic name) <Origin>	Application type	Indications	Preparation for development	PI	Preparation for P II	PII	PIII	Launch
NS-065/NCNP-01 (viltolarsen) <in-house>	NME	Duchenne muscular dystrophy					PIII in progress	
NS-304 (selexipag) <in-house>	New indication	Chronic thromboembolic pulmonary hypertension						
NS-018 (ilginatinib) <in-house>	NME	Myelofibrosis						

In June, 2021, a New Drug Application for NS-065/NCNP-01 (Viltolarsen) was submitted in China.

Reference Materials

NIPPON SHINYAKU CO., LTD.

Consolidated Balance Sheet

(Million yen)	End of FY2020	End of 1Q FY2021	Change Amt		End of FY2020	End of 1Q FY2021	Change Amt
Assets	197,028	200,114	+3,085	Liabilities	34,485	29,648	-4,836
Current assets	139,090	143,004	+3,914	Current liabilities	31,514	26,590	-4,923
Fixed assets	57,937	57,109	-828	Long-term liabilities	2,970	3,058	+87
				Net assets	162,543	170,465	+7,921
Total Asset	197,028	200,114	+3,085	Total liabilities and net assets	197,028	200,114	+3,085

＝Assets＝

Cash and deposits	＋1,644
Notes and accounts receivable	＋2,029
Investment and other assets	-1,637

＝Liabilities and Net assets＝

Accounts payable	-2,518
Accrued tax payable	-2,359
Provision for bonuses	+1,534
Retained earnings	+8,974

NS-065/NCNP-01 (viltolarsen)
- Treatment for Duchenne muscular dystrophy -

Development Phase	・Japan: Launch ・USA : Launch ・China : NDA filing ・Global PIII
Origin	Co-development: National Center of Neurology and Psychiatry
Development	Nippon Shinyaku
Mechanism of action	Exon 53 Skipping
Indication	Duchenne muscular dystrophy
Dosage form	Injection
Feature	・Improvement in symptoms and prevention of the disease progression by recovery of dystrophin protein expression ・Morpholino based oligonucleotide with possible high safety profile and maximized activity

NS-304 (selexipag)

- Treatment for pulmonary hypertension, arteriosclerosis obliterans, lumbar spinal stenosis -

Development Phase	<CTEPH> Japan: NDA filing Overseas: PIII <ASO> Japan: PIIb <LSS> Japan: PIIa <Pediatric PAH> Japan: PII
Origin	Nippon Shinyaku
Development	・Co-development in Japan: Janssen Pharmaceutical K.K. (CTEPH / Pediatric PAH) ・Overseas: Johnson & Johnson (CTEPH / Pediatric PAH) ・Nippon Shinyaku (ASO) ・Nippon Shinyaku (LSS)
Mechanism of action	Selective IP receptor agonist
Indication	・Chronic thromboembolic pulmonary hypertension (CTEPH) ・Arteriosclerosis obliterans (ASO) ・Lumbar spinal stenosis (LSS) ・Pediatric pulmonary arterial hypertension (Pediatric PAH)
Dosage form	Tablet
Feature	Long-acting oral drug

NS-32 (ferric derisomaltose)
- Treatment for iron deficiency anemia -

Development Phase	Japan: NDA filing
Origin	[Dec. 2016] Licensed-in from: Pharmacosmos A/S
Development	Nippon Shinyaku
Mechanism of action	Iron
Indication	Iron deficiency anemia
Dosage form	IV bolus injection or IV drip infusion
Feature	・Can be administered in high doses allowing full iron correction in the majority of patients ・Good safety profile with no dose dependent ADRs ・Minimal potential toxicity from release of labile iron due to tight iron binding in a matrix structure of interchanging isomaltoside and iron ・No profound hypophosphatemia

ZX008
- Treatment for rare intractable epilepsy -

Development Phase	Japan: PIII
Origin	[March. 2019] Commercial rights from: Zogenix, Inc.
Development	Zogenix, inc.
Mechanism of action	Serotonin agonist
Indication	Dravet syndrome and Lennox-Gastaut syndrome
Dosage form	Oral liquid agent
Feature	・Effective for Dravet syndrome and Lennox-Gastaut syndrome patients refractory to existing treatment options ・ZX008 can be used in combination with other drugs, as standard of care for intractable epilepsy based on combination therapy

NS-580
- Treatment for endometriosis -

Development Phase	Japan: PIIa
Origin	Nippon Shinyaku
Development	Nippon Shinyaku
Mechanism of action	Inhibition of membrane-associated prostaglandin E synthase-1
Indication	Endometriosis
Dosage form	Oral agent
Feature	Treatment for endometriosis without hormonal effect and with possible analgesic potency

NS-87
- Treatment for secondary acute myeloid leukemia -

Development Phase	Japan: PI/II
Origin	[Mar. 2017] Licensed-in from: Jazz Pharmaceuticals plc
Development	Nippon Shinyaku
Mechanism of action	Liposomal combination of cytarabine and daunorubicin
Indication	Secondary acute myeloid leukemia (secondary AML)
Dosage form	Injection
Feature	・NS-87 is the first therapy for the treatment of secondary AML in Japan ・The enhancement of antitumor activity and reducing adverse events are expected by NS-87 accumulated in bone marrow

NS-229
- Treatment for inflammatory diseases -

Development Phase	Japan: PI
Origin	Nippon Shinyaku
Development	Nippon Shinyaku
Mechanism of action	JAK1 inhibitor
Indication	Inflammatory diseases (to be determined)
Dosage form	Oral agent
Feature	・Potent and highly selective JAK1 inhibitor ・High efficacy and good safety profiles are expected in the treatment for inflammatory diseases

NS-917

- Treatment for relapsed or refractory acute myeloid leukemia -

Development Phase	Japan: Preparation for PI
Origin	[Mar. 2017] Licensed-in from: Delta-Fly Pharma, Inc.
Development	Nippon Shinyaku
Mechanism of action	DNA strand-break by incorporating itself into DNA
Indication	Relapsed or refractory (r/r) acute myeloid leukemia (AML)
Dosage form	Injection
Feature	・Significant anti-leukemic activity with unique mechanism of action from other nucleoside analogs at low dose continuous infusion ・Tolerable safety profile available to elderly patients with r/r AML

NS-401 (tagraxofusp)

- Treatment for blastic plasmacytoid dendritic cell neoplasm -

Development Phase	Japan: Preparation for Clinical Development
Origin	[Mar. 2021] Licensed-in from: The Menarini Group
Development	Nippon Shinyaku
Mechanism of action	Induction apoptosis of cells by inhibiting protein synthesis by specifically targeting cancer cells expressing CD123
Indication	Blastic plasmacytoid dendritic cell neoplasm (BPDCN)
Dosage form	Injection
Feature	・Composed of diphtheria toxin (DT) fusion protein and recombinant human IL-3 ・Novel targeted therapy directed to CD123 on tumor cells ・IL-3 binds to CD123-expressing tumor cells and delivers the cytotoxic diphtheria toxin to the cells, resulting in the blockage of protein synthesis in the cell and causing cell death in CD123-expressing cells

NS-018 (ilginatinib)
- Treatment for myelofibrosis -

Development Phase	Overseas (USA): Preparation for PII
Origin	Nippon Shinyaku
Development	Nippon Shinyaku
Mechanism of action	JAK2 inhibitor
Indication	Myelofibrosis
Dosage form	Tablet
Feature	・Potent and highly selective JAK2 inhibitor ・High efficacy and safety are expected for myelofibrosis (MF) patients with low platelet count, for whom QOL improvement can’t be obtained because no treatment is available.

Safe Harbor Statement

- Materials and information provided during this presentation may contain so-called “forward-looking statements.” These statements are based on current expectations, forecasts and assumptions that are subject to risks and uncertainties which could cause actual outcomes and results to differ materially from these statements.
- Risks and uncertainties include general industry and market conditions, and general domestic and international economic conditions such as interest rate and currency exchange fluctuations. Risks and uncertainties particularly apply with respect to product-related forward-looking statements. Product risks and uncertainties include, but are not limited to, technological advances and patents attained by competitors; challenges inherent in new product development, including completion or failure of clinical trials; claims and concerns about product safety and efficacy; regulatory agency’s examination, obtaining regulatory approvals; domestic and foreign social security reforms; trends toward healthcare cost containment; and governmental laws and regulations affecting domestic and foreign operations.
- Also, for products that are approved, there are manufacturing and marketing risks and uncertainties, which include, but are not limited to, inability to build production capacity to meet demand, unavailability of raw materials, and competition with others.
- The Company disclaims any intention or obligation to update or revise any forward-looking statements whether as a result of new information, future events or otherwise.
- This English presentation was translated from the original Japanese version.
  In the event of any inconsistency between the statements in the two versions, the statements in the Japanese version shall prevail.