

Shionogi Continues Rare Disease Expansion With Planned Acquisition of IntraBio Inc.

- Transaction will add AQNEURSA® (levacetylleucine) to Shionogi's expanding rare disease portfolio
- AQNEURSA was approved in the United States in 2024 and the European Union in 2026 for neurological manifestations of Niemann-Pick disease type C (NPC)
- Recent U.S. Food and Drug Administration (FDA) supplemental New Drug Approval for AQNEURSA makes it the first and only drug approved for the treatment of Ataxia in patients with Ataxia-Telangiectasia (A-T)
- Milestone builds on strong rare disease foundation established through the acquisition of RADICAVA® (edaravone) and strengthens Shionogi's capabilities in rare disease

OSAKA, Japan, October 5, 2026 - Shionogi & Co., Ltd. (Head Office: Osaka, Japan; Chief Executive Officer: Isao Teshirogi, Ph.D.; hereafter "Shionogi") announced that its Board of Directors, at a meeting held on October 5, 2026, approved Shionogi entering into an agreement to acquire IntraBio Inc. (Head Office: Austin, TX; hereinafter "IntraBio"), a biopharmaceutical company developing and commercializing therapies for neurodegenerative diseases. Pursuant to the agreement, which was signed by the parties today, IntraBio would become a wholly owned subsidiary of Shionogi Inc., a New Jersey-based subsidiary of Shionogi. The proposed transaction, which is subject to customary closing conditions, would add AQNEURSA® (levacetylleucine) and IntraBio's capabilities in the rare disease area to Shionogi's growing portfolio.

AQNEURSA was approved by the FDA in September 2024 for neurological manifestations of Niemann-Pick disease Type C (NPC) in adults and pediatric patients and by the European Medicines Agency (EMA) in January 2026.^{1,2} It was approved by the FDA for the treatment of Ataxia in adults and pediatric patients diagnosed with Ataxia-Telangiectasia (A-T) on September 18, 2026.³ AQNEURSA is currently under review by the EMA for adult and pediatric patients diagnosed with A-T.

"The planned acquisition of IntraBio actively demonstrates Shionogi's solid commitment to building a leading global rare disease business," said Isao Teshirogi, Ph.D., CEO of Shionogi. "Shionogi has a strong record of delivering innovative medicines for challenging infectious diseases, and we are applying that same focus and determination to rare diseases. Bringing AQNEURSA to Shionogi after our acquisition of RADICAVA will deepen our commitment to rare disease communities, expand our capabilities and strengthen our portfolio as we advance future innovation for patients with significant unmet needs."

"We look forward to bringing AQNEURSA to Shionogi as part of our expanding rare disease presence in the U.S., Europe and around the world. This milestone gives us the opportunity to build on the meaningful foundation IntraBio has established with the Niemann-Pick Type C community and to deliver the first and only therapy for Ataxia-Telangiectasia to a community that has lived for many years without options to treat the diverse and debilitating symptoms caused by this disorder," said Nathan McCutcheon, President and CEO, Shionogi Inc. "In addition to AQNEURSA, the capabilities and resources

gained through this acquisition would help to accelerate the development of new treatment options. With this transaction, we deepen our commitment to serving rare disease communities today and delivering innovation tomorrow.”

1. Background and Purpose of the Transaction

Shionogi is committed to providing the best possible medicines to protect people’s health. Building on its leadership in infectious disease, Shionogi is strengthening rare disease as a strategic area of growth and investing in solutions for diseases with significant unmet needs.

Since acquiring the global rights to edaravone, a treatment for amyotrophic lateral sclerosis (ALS), known as RADICAVA® in the U.S. and RADICUT® in Japan, in April 2026, Shionogi has established a strong commercial foundation in rare disease, including expertise in serving patients with serious neurological diseases.

The planned acquisition of IntraBio, a biopharmaceutical company specializing in the research, development, and commercialization of pharmaceuticals targeting rare neurodegenerative diseases and genetic neurological diseases, and its approved therapy, AQNEURSA, builds on that foundation. Following closing, Shionogi will be well positioned to support healthcare professionals and deliver AQNEURSA to patients who may benefit from treatment.

Through this transaction, Shionogi would acquire global rights, including intellectual property and commercialization rights, for AQNEURSA. AQNEURSA is approved in the U.S. and EU for the treatment of neurological manifestations of NPC, with commercialization well underway in the U.S. The FDA recently approved AQNEURSA for the treatment of Ataxia in patients with Ataxia-Telangiectasia (A-T) based on positive Phase 3 clinical trial results. AQNEURSA is currently under review by the EMA for adult and pediatric patients diagnosed with A-T.

In addition, IntraBio’s rare disease expertise would strengthen Shionogi’s capabilities across its rare disease pipeline, including clinical programs in Pompe disease, Fragile X syndrome, and Jordan’s syndrome, as well as additional early-stage rare neurodegenerative disease programs recently added to the pipeline. With this expanded expertise and portfolio, Shionogi would work to accelerate the development of innovative treatment options for patients living with rare diseases.

2. Overview of the Transaction

Under the terms of the agreement, Shionogi Inc. will acquire all outstanding shares of IntraBio for an upfront consideration of USD 2.0 billion payable to IntraBio shareholders.

3. Overview of the Subsidiary Subject to Change

(1) Name	IntraBio Inc.
(2) Address	201 W 5th Street, Suite 1100, Austin, TX 78701, United States of America
(3) Title and Name of Representative	Mallory Factor, President and CEO

(4)	Business Description	Research, development and sales of pharmaceuticals		
(5)	Established	2015		
(6)	Relationship with Listed Company	Capital Relationship	None	
		Personal Relationship	None	
		Business Relationship	None	
(7)	Financial Results and Financial Position for the Past Three Years (Note)			
	Fiscal Year	FY2023/12	FY2024/12	FY2025/12
	Net Assets	USD 35,411 thousand	USD 135,344 thousand	USD 144,549 thousand
	Total Assets	USD 42,787 thousand	USD 168,507 thousand	USD 173,701 thousand
	Net Sales	0	USD 3,535 thousand	USD 67,867 thousand
	Operating Income	USD (22,434) thousand	USD (56,215) thousand	USD (35,292) thousand
	Net Income	USD (26,995) thousand	USD (78,305) thousand	USD (35,800) thousand

(Note) The above financial information has been prepared in accordance with accounting principles generally accepted in the United States of America (U.S. GAAP).

4. Overview of the Counterparties to the Share Acquisition

The shares will be acquired from the existing shareholders of the target company (including individuals, trusts, and corporate entities). The identities of such shareholders are not disclosed at their request and in consideration of the protection of personal information. There is no capital, personal, or business relationship between Shionogi and such individual shareholders.

5. Number of Shares to be Acquired, Acquisition Price and Shareholding Before and After Acquisition

(1) Shares Held Before Acquisition	0 shares (Voting rights ownership ratio: 0.0%)
(2) Shares to be Acquired	All issued shares
(3) Acquisition Price	USD 2,000 million
(4) Shares Held After Acquisition	All issued shares (Voting rights ownership ratio: 100.0%)

6. Schedule

(1) Resolution Date of Board Meeting	October 5, 2026
(2) Agreement Execution Date	October 5, 2026
(3) Transaction Execution Date	Scheduled between November 2026 and December 2026 (planned)

The proposed transaction is subject to the expiration of all applicable waiting periods under competition laws in each relevant country, and to the satisfaction of conditions precedent to the execution of the proposed transaction, including the acquisition of approvals and clearances from relevant regulatory authorities.

7. Outlook

The impact of the proposed transaction on Shionogi's consolidated financial results for the fiscal year ending March 2027 is currently under review. We will promptly announce any matters requiring disclosure in the future.

Advisors

Cleary Gottlieb Steen & Hamilton LLP served as legal advisor to Shionogi. Goldman Sachs & Co. LLC provided financial advice to Shionogi. Centerview Partners LLC and Morgan Stanley & Co. LLC served as financial advisors and Kirkland & Ellis LLP and DLA Piper LLP served as legal advisors to IntraBio.

About AQNEURSA®

AQNEURSA® is approved in the U.S. for the treatment of neurological manifestations of Niemann-Pick disease type C (NPC) in adults and pediatric patients weighing at least 15 kg, and for the treatment of Ataxia in adult and pediatric patients diagnosed with Ataxia-Telangiectasia (A-T) weighing at least 15 kg. In Europe, it has also been approved for the treatment of neurological symptoms of NPC in adults and pediatric patients aged 6 years and older weighing at least 20 kg in combination with miglustat, or as monotherapy in patients where miglustat is not tolerated. AQNEURSA is currently under review by the EMA for adults and pediatric patients diagnosed with Ataxia-Telangiectasia (A-T).

About Niemann-Pick Disease Type C

Niemann-Pick disease type C (NPC) is an ultra-rare, progressive and life-limiting autosomal recessive lysosomal storage disorder caused by variants in the NPC1 or NPC2 genes.^{4,5} These variants impair the movement of cholesterol and other lipids within cells, leading to their accumulation in the brain and other organs.^{4,5} NPC can cause a broad range of neurological and systemic symptoms that significantly impair daily functioning.⁵ Symptoms may begin at any age, from infancy through adulthood, and the disease's variable presentation can contribute to delayed diagnosis. NPC has an estimated incidence of approximately one case per 100,000 live births.⁵

About Ataxia Telangiectasia

Ataxia-Telangiectasia (A-T) is a rare, inherited and progressive neurodegenerative disease caused by variants in the *ATM* gene that typically begins in early childhood.⁶ A-T is estimated to affect approximately one in 40,000 to 100,000 live births worldwide.⁶ The disease causes worsening problems with movement, coordination, speech and eye movements, and many people develop visible dilated blood vessels known as telangiectasias.⁶ A-T can also impair immune function and cause recurrent infections and serious pulmonary complications. A-T is associated with a markedly increased risk of cancer, particularly leukemia and lymphoma, and significantly reduced life expectancy.⁶

About Shionogi & Co., Ltd.

Shionogi & Co., Ltd. is a 148-year-old global, research-driven pharmaceutical company headquartered in Osaka, Japan, that is dedicated to bringing benefits to patients based on its corporate philosophy of "supplying the best possible medicine to protect the health and wellbeing of the patients we serve." The company currently markets products in several therapeutic areas including anti-infectives, pain, CNS disorders and cardiovascular diseases. Shionogi's research and development currently targets two therapeutic areas: infectious diseases and diseases with unmet medical needs, including rare diseases,

allergy/inflammation, oncology, hearing loss, and sleep apnea. For more information on Shionogi & Co., Ltd., please visit <https://www.shionogi.com/global/en>.

About Shionogi Inc.

Shionogi Inc. is a U.S. subsidiary of Shionogi & Co., Ltd., a global research-driven pharmaceutical company. Since 1878, Shionogi has been developing new treatments to improve patients' lives worldwide. Today in North America, our portfolio of FDA-approved products includes an antibiotic for serious infections caused by susceptible Gram-negative bacteria, an oral antiviral for post-exposure prophylaxis of COVID-19 and a therapy for amyotrophic lateral sclerosis (ALS). Our pipeline programs include investigational treatments across infectious disease, rare disease and additional high-need therapeutic areas. Together with our subsidiaries Qpex Biopharma Inc. and Shionogi Canada Inc., we are continuing to innovate for patients in the U.S. and worldwide. Learn more at [shionogi.com](https://www.shionogi.com).

About Shionogi B.V.

Shionogi B.V. is the European subsidiary of Shionogi & Co., Ltd., a global, research-driven pharmaceutical company headquartered in Osaka, Japan. In Europe, Shionogi is dedicated to bringing benefits to patients by advancing innovative medicines in areas of significant unmet medical need. Our work focuses on infectious diseases and rare diseases, building on Shionogi's long-standing expertise in infectious diseases and growing commitment to addressing rare conditions. Shionogi also works with stakeholders across the healthcare ecosystem to support public health and strengthen health security in Europe. For more information about Shionogi in Europe, please visit <https://www.shionogi.com/eu/en/>.

About Shionogi in Rare Disease

Shionogi is committed to changing the trajectory of rare disease. Our mission is to transform the lives of people living with rare diseases by advancing breakthrough science into transformative medicines. We focus on areas where patients face profound unmet needs, bringing together scientific innovation, a focused development approach, and a deep commitment to patients, families and communities. Today Shionogi is delivering a meaningful therapy for patients living with amyotrophic lateral sclerosis (ALS) and, through ongoing research and development, is advancing the next generation of rare disease medicines.

Forward-Looking Statements

This announcement contains forward-looking statements. These statements are based on expectations in light of the information currently available, assumptions that are subject to risks and uncertainties which could cause actual results to differ materially from these statements. Risks and uncertainties include general domestic and international economic conditions such as general industry and market conditions, and changes of interest rate and currency exchange rate. These risks and uncertainties particularly apply with respect to product-related forward-looking statements. Product risks and uncertainties include, but are not limited to, completion and discontinuation of clinical trials; obtaining regulatory approvals; claims and concerns about product safety and efficacy; technological advances; adverse outcome of important litigation; domestic and foreign healthcare reforms and changes of laws and regulations. Also for existing products, there are manufacturing and marketing risks, which include, but are not limited to, inability to build production capacity to meet demand, lack of availability of raw materials and entry of competitive products. 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.

For Further Information, Contact:

SHIONOGI Website Inquiry Form: <https://www.shionogi.com/global/en/contact.html>

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References

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3. [FDA Approves Therapy to Treat Ataxia in Patients with Ataxia-Telangiectasia, a Rare Genetic Disorder](#)
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