PRESSRELEASE

SHIONOGI

Shionogi Completes Acquisition of IntraBio Inc. and Adds AQNEURSA® (levacetylleucine) to Growing Rare Disease Global Portfolio

AQNEURSA is approved in the U.S. and Europe for neurological manifestations of Niemann-Pick disease type C and in the U.S. as the first and only treatment for Ataxia in patients diagnosed with Ataxia-Telangiectasia

Shionogi is committed to seamless continuity of care for patient communities in the U.S. and to achieving access in Europe

OSAKA, Japan, November 18, 2026 – Shionogi & Co., Ltd. (Head Office: Osaka, Japan; Chief Executive Officer: Isao Teshirogi, Ph.D.; hereafter "Shionogi") announced that it has completed the acquisition of IntraBio Inc. ("IntraBio"). The acquisition adds AQNEURSA® (levacetylleucine) and additional rare disease capabilities to Shionogi's growing global portfolio.

AQNEURSA was approved by the U.S. Food and Drug Administration in 2024 and by the European Medicines Agency (EMA) in January 2026 for neurological manifestations of Niemann-Pick disease type C (NPC) in adults and pediatric patients weighing at least 15 kg in the U.S. and 20 kg in Europe.[1,2] The FDA approved AQNEURSA as the first and only option for the treatment of Ataxia in adults and pediatric patients diagnosed with Ataxia-Telangiectasia (A-T) weighing at least 15 kg in September 2026.[3]

AQNEURSA is currently under review by the EMA for adult and pediatric patients diagnosed with A-T and it is currently being studied for the treatment of CACNA1A disorders in a Phase III, randomised, placebo-controlled, double-blind, crossover study. Shionogi will continue programs initiated by IntraBio and explore additional opportunities to research AQNEURSA in additional rare disease conditions.

"With the closing of the IntraBio acquisition, we welcome AQNEURSA to Shionogi's growing portfolio and strengthen our global capabilities with additional rare disease expertise," said Isao Teshirogi, Ph.D., CEO of Shionogi. "For nearly 150 years, Shionogi has grown our business by adapting to the changing needs of patients. This milestone is the latest representation of our steadfast commitment to deliver innovative medicines to protect the health and wellbeing of the patients we serve."

Under the terms of the agreement announced on October 5, 2026, Shionogi Inc., a New Jersey-based subsidiary of Shionogi, paid a lump sum of USD 2.0 billion to acquire all outstanding shares of IntraBio, including global intellectual property and commercialisation rights for AQNEURSA.

"Through acquisitions, integrations and strategic hires, Shionogi is building a strong, sustainable rare disease presence with a clear purpose: to make a meaningful difference for patients," said Nathan

McCutcheon, President and CEO, Shionogi Inc. “We have the infrastructure, portfolio, and commitment to deliver the medicines, patient support and services, and the scientific expertise and determination to advance new treatment options for tomorrow.”

"We look forward to partnering with patient groups representing people living with these devastating conditions, and learning from patients, families and carers about the challenges they face and how we can best help improve their lives," said Huw Tippett, CEO, Shionogi Europe.

“We look forward to working with the Shionogi team. The recent U.S. approval of AQNEURSA, the first FDA-approved treatment for ataxia in A-T, has brought new hope to children and families affected by this devastating disease,” said Brad Margus, founder of the non-profit A-T Children’s Project. “With Shionogi’s additional reach and resources, we hope that more families will be able to access this important new treatment.”

“For the Niemann-Pick community, this acquisition is an opportunity to build on progress to date for people living with NPC and their families. We are deeply grateful to IntraBio for securing approval for AQNEURSA and their support of our community. We look forward to working with Shionogi to expand access and ensure this important therapy reaches the families who need it most,” said Heather Teal, Executive Director, National Niemann-Pick Disease Foundation.

Following the acquisition of RADICAVA ORS® (edaravone) earlier this year, a treatment for amyotrophic lateral sclerosis (ALS), Shionogi has established a strong commercial and medical affairs foundation in rare disease, including expertise serving patients with neurodegenerative diseases. Bringing IntraBio to Shionogi expands that foundation with new capabilities to support the company’s growing rare disease pipeline, which includes clinical programs in Pompe disease, Fragile X syndrome and Jordan’s syndrome, as well as additional early-stage rare neurodegenerative disease programs.

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 manifestations 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.[6] 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 and CNS disorders. Shionogi's research and development currently targets 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. Today in North America, our FDA-approved infectious disease portfolio includes an antibiotic for serious infections caused by susceptible Gram-negative bacteria and an oral antiviral for post-exposure prophylaxis of COVID-19. Our FDA-approved rare disease portfolio includes therapies for amyotrophic lateral sclerosis (ALS), Niemann-Pick disease type C (NPC) and Ataxia in patients diagnosed with Ataxia-Telangiectasia (A-T). 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.

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.

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