Tanabe Pharma
Creating hope for all facing illness.

Press Release

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Commencement of a Prospective Observational Study on the Long-Term Efficacy and Safety of Uplizna for IgG4-Related Disease

Tanabe Pharma Corporation (Head Office: Chuo-ku, Osaka; Representative Director, CEO: Akihisa Harada; hereinafter "Tanabe Pharma") announced the initiation of a prospective observational study 「4SigHT」（jRCT1031250749）to clarify the long-term efficacy and safety of "Uplizna® for I.V. Infusion 100mg" (generic name: Inebilizumab [genetically recombination], hereinafter "Uplizna®"), which obtained approval on November 20, 2025 for the treatment of the suppression of relapse of immunoglobulin IgG4-related disease (IgG4-RD), under real-world clinical conditions as of March 24, 2026, and has recently published the protocol paper for this study.

This study is planned to be conducted at 40 sites nationwide, targeting 100 subjects, and will continue until December 31, 2032. Tanabe Pharma will prospectively collect and analyze data on clinical relapse, treatment practices, adverse events, and the status of relapse and remission by organ.

IgG4-RD can occur in multiple organs and lead to fibrosis and permanent organ damage[1)]. It is a progressive disease that is characterized by periods of remission and unpredictable disease flares[2)3)]. The exact pathogenesis of IgG4-TD remains unknown, but B-cells, particularly IgG4-positive plasmablasts and plasma cells, might contribute to disease pathogenesis[4)].
As IgG4-RD is a rare disorder, estimated at 5 in 100,000 worldwide, ongoing observational research into its long-term efficacy and safety after approval is considered valuable not only for patient treatment but also for understanding the disease.

Under our MISSION, “Creating hope for all facing illness,” Tanabe Pharma will continue to strive to provide pharmaceuticals that address unmet medical needs, including rare diseases, through in-house development and partnerships with other companies, delivering these options to patients.

Contact:
Tanabe Pharma Corporation
CEO Office Communication Group
+81-6-6205-5119

About Uplizna®

Uplizna® is a humanized monoclonal antibody (mAb) that causes targeted and sustained depletion of key cells that contribute to underlying disease process (autoantibody-producing CD19+ B cells, including plasmablasts and some plasma cells). The patient receives a

second dose 14 days after the initial administration, followed by one dose every six months after the first administration.
In Japan, Uplizna® was approved and launched in 2021 for the indication of "prevention of relapse in neuromyelitis optica spectrum disorder (including neuromyelitis optica)." In November 2025, approval was additionally obtained for the indication of relapse suppression in IgG4-related disease.

About IgG4-RD

Immunoglobulin G4-related disease (IgG4-RD) is a chronic, systemic, immune-mediated, fibroinflammatory disease which can affect numerous and generally multiple organs of the body[1)]. The exact pathogenesis of IgG4-related disease remains unknown, but B-cells, particularly IgG4-positive plasmablasts and plasma cells, might contribute to disease pathogenesis[4)]. It is a progressive disease that can affect a variety of organ system and often affect multiple organs over time. It is characterized by periods of remission and unpredictable disease flares[2)3)]. IgG4-RD can cause permanent organ damage with or without the presence of symptoms[5)]. B cells are central to the pathogenesis of IgG4-RD[1)] In IgG4-RD, CD19-expressing (CD19+) B cells are thought to drive inflammatory and fibrotic processes and interact with other immune cells that contribute to disease activity[1)5)].
The prevalence is estimated at 5 in 100,000 worldwide[1)5)]. The typical age of onset of IgG4-RD is between 50 and 70 years old and, it varies depending on the location of the lesion, IgG4-RD is more likely to occur in men than women[2)].

1) Inebilizumab for Treatment of IgG4-Related Disease.
DOI:10.1056/NEJMoa2409712

2) IgG4-related disease: Changing epidemiology and new thoughts on a multisystem disease.
DOI:10.1016/j.jtauto.2020.100074

3) Predictors of disease relapse in IgG4-related disease following rituximab. DOI:10.1093/rheumatology/kev438

4) B lymphocytes directly contribute to tissue fibrosis in patients with IgG4-related disease.
DOI:10.1016/j.jaci.2019.07.004

5) IgG4-related disease: an update on pathophysiology and implications for clinical care. DOI:10.1038/s41584-020-0500-7