for serving on a Scientific Advisory or Data Safety Monitoring board for ArgenX, Belgium. Dr. Nobile-Orazio has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for LFB, France. Dr. Nobile-Orazio has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Roche, Switzerland. Dr. Nobile-Orazio has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for CSL Behring. Dr. Nobile-Orazio has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Janssen, USA. The institution of Dr. Nobile-Orazio has received research support from Takeda/Shire. Dr. Peric has received personal compensation in the range of $5,000-$9,999 for serving as a Consultant for Argenx. Dr. Peric has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Mylan. An immediate family member of Dr. Katzberg has received personal compensation for serving as an employee of Sanofi Genzyme. Dr. Katzberg has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Argenx. Dr. Katzberg has received personal compensation in the range of $500-$4,999 for serving as a Consultant for UCB. Dr. Katzberg has received personal compensation in the range of $5,000-$9,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Octapharma. Dr. Katzberg has received personal compensation in the range of $500-$4,999 for serving on a Speakers Bureau for UCB. Mrs. Cadour has nothing to disclose. Miss Van de Walle has received personal compensation for serving as an employee of argenx bv. Mrs. Van Hoomissen has received personal compensation for serving as an employee of argenx. Mr. Jacquemyn has received personal compensation for serving as an employee of argenx. Dr. Van de Steen has received personal compensation for serving as an employee of argenx. Dr. Van de Steen has stock in argenx. Dr. van der Pol has nothing to disclose.

Vibrance-mg: Clinical Trial of Nipocalimab in Pediatric Myasthenia Gravis

Sindhu Ramchandren, Shawn Black, Hong Sun

Objective

We describe an open-label study of nipocalimab to determine the effect of nipocalimab in pediatric participants with gMG.

Background

Nipocalimab is a high affinity, fully human, aglycosylated, effectorless IgG1 anti FcRn monoclonal antibody that targets the neonatal Fc receptor (FcRn) with high affinity, thereby lowering IgG pathogenic antibodies in autoimmune disease. Data from Vivacity-MG, a Phase 2, multicenter, randomized, double-blind, placebo-controlled study of nipocalimab demonstrated safety, tolerability, and efficacy of nipocalimab in adult generalized myasthenia gravis (gMG) (clinicaltrials.gov NCT03772587).

Design/Methods

This global study will enroll at least 12 participants, aged 2 to <18, with a diagnosis of gMG and an insufficient clinical response to ongoing, stable standard-of-care therapy, as reflected by a Myasthenia Gravis Foundation of America (MGFA) Class of IIa/b through IVa at screening. Participants must have a positive serologic test for either acetylcholine receptor or Muscle Specific Tyrosine Kinase pathogenic autoantibody. The study will consist of a screening period of up to 4 weeks, a 24-week open-label Active Treatment Phase where participants will receive nipocalimab intravenously every two weeks, and a Long-term Extension phase; after last dose, a safety follow-up assessment will be conducted at 8 weeks. The primary outcome is the effect of nipocalimab on total serum IgG, safety and tolerability, and PK in pediatric participants with gMG.

Results

Study enrollment will begin in 2022.

Conclusions

The vibrance-mg study will assess the PK/PD, safety and activity of Nipocalimab in pediatric gMG.

Disclosure: Dr. Ramchandren has received personal compensation for serving as an employee of Janssen Pharmaceutical Companies of Johnson & Johnson. Dr. Ramchandren has a non-compensated relationship as a Scientific Advisory Board Member with CMT Research Foundation (CMTRF) that is relevant to AAN interests or activities. Mr. Black has received personal compensation for serving as an employee of Janssen. Mr. Black has stock in Janssen. Dr. Sun has received personal compensation for serving as an employee of Janssen.

Vivacity MG Phase 3 Study: Clinical Trial of Nipocalimab Administered to Adults With Generalized Myasthenia Gravis

Sindhu Ramchandren, Panna Sanga, Michel Burcklen, Hong Sun

Objective

We describe Vivacity-MG3. our pivotal Phase 3, Multicenter, Randomized, Double-Blind, Placebo-Controlled Study to Evaluate the Efficacy, Safety, Pharmacokinetics (PK), and Pharmacodynamics (PD) of Nipocalimab Administered to Adults With gMG (NCT04951622).

Background

Nipocalimab is a high affinity, fully human, aglycosylated, effectorless IgG1 anti FcRn monoclonal antibody that targets the neonatal Fc receptor (FcRn) with high affinity, thereby lowering IgG pathogenic antibodies in autoimmune disease. Data from Vivacity-MG, a Phase 2 randomized placebo-controlled study of nipocalimab in adult generalized myasthenia gravis (gMG), demonstrated safety, tolerability, and efficacy of nipocalimab (clinicaltrials.gov NCT03772587).

Design/Methods

This global study will enroll approximately 180 participants with gMG, aged 18 and older, with an insufficient clinical response to ongoing, stable standard-of-care therapy, as reflected by a Myasthenia Gravis-Activities of Daily Living (MG-ADL) score of >/= to 6 at screening and baseline, and a Myasthenia Gravis Foundation of America (MGFA) Class of IIa/b–IVa/b at screening. The study will consist of a screening period of up to 4 weeks, a 24-week double-blind placebo-controlled phase where participants will be randomly assigned in a 1:1 ratio to receive either placebo or nipocalimab intravenously every two weeks, and an open label extension phase of variable duration. The primary outcome is the average change in MG-ADL score from baseline to weeks 22, 23 and 24 of the double-blind placebo-controlled phase.

Results

Study enrollment began in July 2021 and is ongoing.

Conclusions

The ongoing Vivacity MG Phase 3 study will assess the efficacy, safety, and PK/PD of Nipocalimab in adult gMG.

Disclosure: Dr. Ramchandren has received personal compensation for serving as an employee of Janssen Pharmaceutical Companies of Johnson & Johnson. Dr. Ramchandren has a non-compensated relationship as a Scientific Advisory Board Member with CMT Research Foundation (CMTRF) that is relevant to AAN interests or activities. Panna Sanga has received personal compensation for serving as an employee of Janssen Research & Development. Panna Sanga has stock in Johnson & Johnson. Michel Burcklen has received personal compensation for serving as an employee of Actelion Pharmaceuticals, a Janssen pharmaceutical company of Johnson & Jonhson. Michel Burcklen has received stock or an ownership interest from Johnson & Johnson. Dr. Sun has received personal compensation for serving as an employee of Janssen.

Response of Sjogren-related Small Fiber Neuropathy to rituximab

Anita Venkatesh, Suraj Muley

Objective

Small fiber neuropathy typically presents as loss of temperature and pain sensation and may be associated with other autonomic symptoms. Sjogren's-related ganglionopathy can selectively affect small fibers. Current treatment options for small fiber neuropathy secondary to

Sjogren's syndrome include glucocorticoids, IVIG, or immunomodulatory agents such as rituximab. However, these treatments are of uncertain effectiveness and can have variable responses. We present a case of Sjogren's-related small fiber neuropathy that had complete symptom resolution with rituximab.

Background

A 54 year old female presented to the clinic with neuropathic pain starting in her legs, which ascended to her trunk and arms after one week. She also reported five years of dry eyes. On initial presentation, physical exam showed normal sensation but a diagnosis of small fiber neuropathy was made by skin biopsy. The neuropathy was attributed to seronegative Sjogren's syndrome diagnosed by salivary gland biopsy. She had previously failed treatment with prednisone 40 mg daily and IVIG. After presentation to the clinic, she was started on rituximab every 4 months. A log of symptoms kept by the patient clearly demonstrated a reduction in symptoms following each Rituxan dose. She achieved complete symptom resolution after 19 months with Rituxan 10 mg/mL every 4 months.

Design/Methods

NA.

Results

NA.

Conclusions

In summary, our patient responded dramatically to rituximab after failing treatment with glucocorticoids and IVIG. Treatment of Sjogren's-related small fiber neuropathy has consisted of empiric trials of glucocorticoids and IVIG, although response to these treatments has been variable. Rituximab has been useful in Sjogren's-related neuropathy, although it remains unclear whether it is effective in small fiber neuropathy. Our case illustrates that patients with Sjogren's-related small fiber neuropathy, even if seronegative, may respond dramatically to rituximab. We concluded that rituximab can be considered in treatment of this condition, especially in patients who don't respond to first line immunotherapies.

Disclosure: Dr. Venkatesh has nothing to disclose. Dr. Muley has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Argenx. Dr. Muley has received personal compensation in the range of $10,000-$49,999 for serving on a Speakers Bureau for Alexion. Dr. Muley has received personal compensation in the range of $500-$4,999 for serving on a Speakers Bureau for CSL Behring. Dr. Muley has received personal compensation in the range of $500-$4,999 for serving on a Speakers Bureau for Takeda. Dr. Muley has received personal compensation in the range of $500-$4,999 for serving on a Speakers Bureau for Catalyst. Dr. Muley has received publishing royalties from a publication relating to health care.

The First Randomized, Double-Blind, Placebo-Controlled Phase 2 Study to Evaluate the Efficacy and Safety of an FcRn Inhibitor, Rozanolixizumab, in Patients With Leucine-Rich Gliomainactivated 1 Autoimmune Encephalitis

Divyanshu Dubey, Maarten J Titulaer, Anjana Dhar Koul, Stephen Yates, Sarosh R Irani

Objective

To evaluate efficacy and safety of rozanolixizumab for treatment of leucine-rich glioma inactivated 1 (LGI1) autoimmune encephalitis (AIE).

Background

LGI1 AIE is a clinically homogeneous syndrome mediated by autoantibodies, predominantly of the IgG4 subclass, characterized by seizures, cognitive impairment and neuropsychiatric symptoms. No approved treatment options are available, and current treatment paradigms are variable. Rozanolixizumab is a fully humanized monoclonal antibody that can be delivered subcutaneously and inhibits the IgG binding region of the neonatal Fc receptor (FcRn), reducing the concentration of circulating IgG-antibodies, including pathogenic IgG-autoantibodies.

Design/Methods

This multicenter, randomized, double-blind, placebo-controlled LEGIONE study (NCT04875975) is the first Phase 2 study to evaluate efficacy and safety of FcRn inhibition as treatment for LGI1 AIE. The study is recruiting adults with serum LGI1-autoantibodies, considered for intravenous methylprednisolone treatment, with new-onset disease (0-12 months prior to study entry). At screening, patients with prior diagnosis of epilepsy unrelated to LGI1 AIE, IgG level =5.5 g/L, clinically relevant infection or history of neoplastic disease will be excluded. Alongside a typical steroid taper, ~68 patients will be randomized 1:1 to subcutaneous infusion of rozanolixizumab or placebo for 24 weeks, stratified by time from disease onset and cognitive function (measured by the Repeatable Battery for the Assessment of Neuropsychological Status) at randomization. Primary endpoint is measured by seizure freedom (28 consecutive days of no seizures maintained until the end of the treatment period). Secondary endpoints are change in cognitive function, use of rescue medication, time to onset of seizure freedom and safety and tolerability of rozanolixizumab. Exploratory pharmacokinetic/pharmacodynamic and biomarker-based endpoints are anticipated.

Results

Study background, rationale and design will be presented.

Conclusions

The LEGIONE study is the first randomized, double-blind, placebo-controlled Phase 2 study to evaluate the efficacy and safety of an FcRn inhibitor, rozanolixizumab, in patients with leucine-rich glioma-inactivated 1 autoimmune encephalitis and is enrolling patients.

Disclosure: The institution of Dr. Dubey has received personal compensation in the range of $500-$4,999 for serving as a Consultant for UCB. The institution of Dr. Dubey has received personal compensation in the range of $5,000-$9,999 for serving as a Consultant for Astellas. Dr. Dubey has received personal compensation in the range of $0-$499 for serving on a Speakers Bureau for AGRIMS. Dr. Dubey has received personal compensation in the range of $0-$499 for serving on a Speakers Bureau for Advances in Neurology. Dr. Dubey has received personal compensation in the range of $0-$499 for serving on a Speakers Bureau for Moffit Cancer Center. Dr. Dubey has received research support from Department of Defense. Dr. Dubey has received intellectual property interests from a discovery or technology relating to health care. Dr. Dubey has received intellectual property interests from a discovery or technology relating to health care. Dr. Dubey has received intellectual property interests from a discovery or technology relating to health care. The institution of Dr. Titulaer has received research support from Dutch Epilepsy Foundations (NEF 14-18 and 19-08). The institution of Dr. Titulaer has received research support from CSL Behring. The institution of Dr. Titulaer has received research support from UCB. The institution of Dr. Titulaer has received research support from Netherlands Organisation for Scientific Research (ZonMW, Memorabel initiative and E-RARE UltraAIE). The institution of Dr. Titulaer has received research support from Horizon Therapeutics. The institution of Dr. Titulaer has received research support from Dioraphte (charity). The institution of Dr. Titulaer has received research support from Guidepoint Global LLC. Dr. Koul has stock in UCB. An immediate family member of Dr. Koul has received intellectual property interests from a discovery or technology relating to health care. An immediate family member of Dr. Koul has received personal compensation in the range of $0-$499 for serving as a Scientific advisory board member with CSIR. Dr. Yates has received personal compensation for serving as an employee of UCB Biosciences, Inc. Dr. Yates has stock in UCB Biosciences, Inc. Dr. Irani has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Immunovant. Dr. Irani has received personal compensation in the range of $5,000-$9,999 for serving as a