Prior Authorization Letter for Intravenous Immunoglobulin (IVIG) Therapy

To Whom It May Concern:

I am writing to request prior authorization for intravenous immunoglobulin (IVIG) therapy for my patient, a ***-year-old with biopsy-proven pemphigus vulgaris (ICD-10: L10.0), who has severe, refractory disease despite multiple courses of standard and advanced immunosuppressive therapies.

Clinical History and Disease Severity:

The patient was diagnosed with pemphigus vulgaris in *** via skin biopsy. The disease involves the oral mucosa and scalp, causing significant morbidity and impairment of quality of life. Despite aggressive treatment, the patient has chronic, flaring disease that is not at treatment goal. Most recently, anti-desmoglein 3 antibody levels were markedly elevated at *** in the most recent measurement, with desmoglein 1 ***, confirming ongoing active autoimmune disease.

Prior Treatment Failures:

This patient has exhausted multiple standard and advanced treatment options:

1. **Rituximab (Truxima)**: The patient has received THREE complete courses of rituximab:

   - First course: April 2023 (1g IV x 2 doses, 2 weeks apart)

   - Second course: September 2024 (9/17/2024 and 9/23/2024)

   - Third course: March-April 2025

Despite three courses of rituximab, the patient continues to have active disease with persistently elevated anti-desmoglein antibodies.

2. **Mycophenolate Mofetil (CellCept)**: Currently on 1500mg twice daily (3000mg total daily dose) without adequate disease control.

3. **Prednisone**: The patient remains dependent on 40mg daily of prednisone and experiences disease flares when attempts are made to taper. This prolonged high-dose corticosteroid therapy has resulted in multiple adverse effects including leg swelling, sleep disturbance, and agitation, with ongoing risks of adrenal suppression, global immunosuppression, and increased infection risk.

Medical Necessity for IVIG:

IVIG is medically necessary and evidence-based for this patient's refractory pemphigus vulgaris. High-dose IVIG is specifically indicated for severe and recalcitrant pemphigus

according to international treatment guidelines and expert consensus. A randomized controlled trial demonstrated that IVIG was the only adjuvant therapy that significantly increased the number of patients achieving disease control in pemphigus unresponsive to prednisolone. Additionally, IVIG has been shown to have significant corticosteroid-sparing effects, which is critically important for this patient who is experiencing multiple adverse effects from prolonged high-dose prednisone therapy.

The advantages of IVIG in pemphigus include rapid onset of action, reduction in autoantibody production, and importantly, no increased risk of infection—a crucial consideration given this patient's already compromised immune status from multiple immunosuppressive medications. Studies have demonstrated that IVIG can produce long-term sustained clinical, serological, and immunopathological remission in pemphigus vulgaris patients. A systematic review reported disease remission rates of 82.8% with IVIG therapy in pemphigus patients, with favorable safety profiles.

Treatment Plan:

I am requesting approval for IVIG at a dose of 2g/kg administered over 2-5 days with monthly repetitions, which is the standard evidence-based dosing regimen for pemphigus vulgaris. The patient has normal IgA levels, confirming no contraindication to IVIG therapy.

Clinical Goals:

1. Achieve disease control as evidenced by reduction in anti-desmoglein antibody levels and clinical improvement

2. Enable tapering of high-dose prednisone to reduce corticosteroid-related adverse effects

3. Improve quality of life and functional status

4. Prevent disease progression and complications

Summary:

This patient has severe, refractory pemphigus vulgaris with documented failure of three courses of rituximab, high-dose mycophenolate mofetil, and prolonged high-dose corticosteroids. The patient remains steroid-dependent with active disease confirmed by markedly elevated anti-desmoglein antibodies (Dsg 3 & >200). IVIG represents an evidence-based, guideline-supported therapy for this exact clinical scenario—severe and recalcitrant pemphigus that has failed standard immunosuppressive therapy and rituximab. The denial of IVIG therapy leaves this patient without viable treatment options and at continued risk of disease progression and corticosteroid toxicity.

I respectfully request approval for monthly IVIG therapy for this patient and am available to provide any additional clinical information or documentation needed to support this request.