To whom it may concern:

I am contacting you as a dermatologist caring for @NAME@ with regard to the patient's diagnosis of Pemphigus Vulgaris (L10.0). As you know, this is an extremely morbid condition that causes severe blistering and painful erosions in the skin and mucous membranes.

I recently prescribed this patient Rituximab at the weight-based dosing, 375 mg/mg2 weekly x4 weeks and am writing a letter of medical necessity for this dosing.

When treating a patient with Pemphigus Vulgaris (L10.0) it is necessary to have access to the full spectrum of accepted treatments as patients may not be able to use one particular treatment due to lack of response, the potential for side effects or even an allergic reaction. It can become a serious safety issue for the patient if I am not able to prescribe a wide variety of treatments for this condition.

I strongly believe @NAME@ needs access to Rituximab at the weight-based dosing. Prior to oral steroids, pemphigus vulgaris (L10.0) was uniformly fatal. However, oral steroid has increased survival but is only a short term solution that can cause hypertension and hyperglycemia, as well as chronic toxicities such as disturbance of the Hypothalamic–pituitary–adrenal axis, osteoporosis with devastating spinal compression and hip fractures, and cataracts. Off label use of various immunosuppressive therapies has facilitated longterm treatment of pemphigus vulgaris (L10.0), but these treatments have to be used outside their FDA indication which usually is focused on other rheumatic diseases or organ transplantation. They also do not lead to remission, but only suppress the disease. Rituximab can suppress the CD-20 B-cells that keep the disease active, is the only targeted therapy and can lead to remission, sometimes after a number of treatment courses.

A recent randomized controlled trial published in Lancet found that those assigned to Rituximab (Rituxan) plus oral prednisone were more likely to be in complete remission at month 24 than those who were initially treated with prednisone alone (89 vs 34%). In addition, the Rituximab (Rituxan) plus prednisone group had a shorter median delay to achieve complete remission off therapy compared to the prednisone alone group (277 vs 677 days). Importantly, the Rituximab (Rituxan) plus prednisone group had less frequent severe adverse events.

Subsequently, a study found that the weight-based dosing was 2.7 times more likely to lead to clinical remission than the FDA approved dosing. For this reason, I would like to proceed with 375 mg/m2 weight-based dosing. Additionally, my patient previously received with dose with excellent results and clinical remission for several years.

Additionally, I request that you review the following evidence showing how this medication can be effectively utilized to treat Pemphigus Vulgaris (L10.0):

1. Joly P, Maho-Vaillant M, Prost-Squarcioni C, Hebert V, Houivet E, Calbo S, Caillot F, Golinski ML, Labeille B, Picard-Dahan C, Paul C, Richard MA, Bouaziz JD, Duvert-Lehembre S,

Bernard P, Caux F, Alexandre M, Ingen-Housz-Oro S, Vabres P, Delaporte E, Quereux G, Dupuy A, Debarbieux S, Avenel-Audran M, D'Incan M, Bedane C, Bénéton N, Jullien D, Dupin N, Misery L, Machet L, Beylot-Barry M, Dereure O, Sassolas B, Vermeulin T, Benichou J, Musette P, French study group on autoimmune bullous skin diseases. First-line rituximab combined with short-term prednisone versus prednisone alone for the treatment of pemphigus (Ritux 3): a prospective, multicentre, parallel-group, open-label randomised trial. Lancet. 2017;389(10083):2031. Epub 2017 Mar 22.

2. Lunardon L, Tsai KJ, Propert KJ, Fett N, Stanley JR, Werth VP, Tsai DE, Payne AS. Adjuvant rituximab therapy of pemphigus: a single-center experience with 31 patients. Arch Dermatol. 2012;148(9):1031.

3. FDA package insert

4. Kushner CJ, Wang S, Tovanabutra N, Tsai DE, Werth VP, Payne AS. Factors Associated With Complete Remission After Rituximab Therapy for Pemphigus. JAMA Dermatol. 2019 Dec 1;155(12):1404-1409. doi: 10.1001/jamadermatol.2019.3236. PMID: 31642878; PMCID: PMC6813574.

On behalf of @NAME@, I would appreciate your prompt reconsideration of this denial. Please feel free to contact me below for any additional information you may require. I look forward to receiving your response and approval of coverage for this medication.

Sincerely,