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To whom it may concern:

I am writing to request reconsideration of LCD A59101 regarding the use of rituximab to include the diagnosis code D89.84. IgG4-related pancreatitis (D89.84) is a rare form of autoimmune pancreatitis characterized by inflammation and fibrosis of the pancreas, often associated with elevated IgG4 levels. If not managed properly, it can result in irreversible organ damage, exocrine and endocrine insufficiency, and obstructive jaundice due to biliary involvement.¹

Corticosteroids are the first-line treatment for autoimmune pancreatitis; however, many patients experience relapses or require long-term steroid sparing therapy due to medical status (e.g., diabetes or osteoporosis). For patients where corticosteroids are ineffective or inappropriate, rituximab provides an essential therapeutic option.

Rationale for Rituximab in IgG4-Related Pancreatitis:

For patients with steroid-refractory or who are corticosteroid-intolerant, rituximab provides an evidence-based, effective and safe alternative.

- Patients with IgG4-related diseases generally show an incomplete response to glucocorticoids. Rituximab depletes CD-20 positive B cells, which are responsible for producing IgG4 antibodies, and the use of this agent has led to prompt clinical and serologic improvement in patients with refractory IgG4-related diseases.²
- A published meta-analysis and systematic review demonstrate that rituximab is efficacious in the treatment of IgG4-related pancreatitis, with durable responses.³⁻⁴
- Cohort studies have demonstrated complete remission in 80 – 97% of patients who are steroid intolerant/resistant treated with rituximab, with sustained benefit over long-term follow up.³⁻⁶
- The United European Gastroenterology guideline recognizes rituximab as a viable option for steroid-refractory IgG4-related pancreatitis.¹
- The International Consensus Guidance Statement on IgG4-related disease (2015) recommends rituximab as an alternative in instances where corticosteroids are contraindicated.⁷

Given the supporting clinical evidence and guidelines supporting the use of rituximab in steroid-refractory IgG4-related pancreatitis (D89.84), I request reconsideration of LCD A59101 to include D89.84 as a covered diagnosis code for rituximab. Expanding coverage to include this patient population aligns with current best practices and ensures access to medically necessary, evidence-based treatment.

Thank you in advance for your consideration.

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References:

1. L  hr JM, Beuers U, Vujasinovic M, et al. European Guideline on IgG4-related digestive disease - UEG and SGF evidence-based recommendations. *United European Gastroenterol J*. 2020;8(6):637-666
2. Khosroshahi A, Bloch DB, Deshpande V, Stone JH. Rituximab therapy leads to rapid decline of serum IgG4 levels and prompt clinical improvement in IgG4-related systemic disease. *Arthritis Rheum*. 2010;62(6):1755-1762.
3. Lanzillotta M, Della-Torre E, Wallace ZS, et al. Efficacy and safety of rituximab for IgG4-related pancreato-biliary disease: A systematic review and meta-analysis. *Pancreatology*. 2021;21(7):1395-1401.
4. Nikolic S, Panic N, Hintikka ES, et al. Efficacy and safety of rituximab in autoimmune pancreatitis type 1: our experiences and systematic review of the literature. *Scand J Gastroenterol*. 2021;56(11):1355-1362.
5. Hart PA, Topazian MD, Witzig TE, et al. Treatment of relapsing autoimmune pancreatitis with immunomodulators and rituximab: the Mayo Clinic experience. *Gut*. 2013;62(11):1607-1615.
6. Carruthers MN, Topazian MD, Khosroshahi A, et al. Rituximab for IgG4-related disease: a prospective, open-label trial. *Ann Rheum Dis*. 2015;74(6):1171-1177.
7. Khosroshahi A, Wallace ZS, Crowe JL, et al. International consensus guidance statement on the management and treatment of igg4-related disease. *Arthritis & Rheumatology*. 2015;67(7):1688-1699.

Your partner for good

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