discharged. Immunology evaluation occurred at 4 months of age. During the intervening time the child had developed poor growth, intractable diarrhea, eczema, and resistant thrush. Exam was notable for eczematous rashes on chest, back and extremities. Flow cytometry revealed an absence of CD25+ T-cells and mild CD4 lymphopenia. Blood cell counts, immunoglobulin levels and vaccine titers were unremarkable. Next gen gene sequencing panel identified a homozygous pathological mutation (c.301C>T) resulting in a premature stop codon at Gln101 in the IL2RA gene. She was started on a treatment regimen that consisted of abatacept, rituximab, immunoglobulin replacement and sirolimus. She was transitioned to elemental formula for enteropathy, which improved. With treatment she clinically stabilized and was referred for transplant.

Discussion: IPEX and IPEX-like syndromes present early in life with evidence of marked immune dysregulation. IPEX-like syndromes can vary in the degree of immunodeficiency seen. Diversity of presentation can make early diagnosis challenging, requiring a high index of suspicion. Prompt immunologic evaluation and early use of genetic testing can accelerate diagnosis. Use of immunomodulators can be helpful in stabilizing patients pending definitive therapy by hematopoietic stem cell transplantation.

M142
RECURRENT ORAL CANDIDIASIS IN A PATIENT WITH VITAMIN B12 DEFICIENCY
D. Bravo Solarte1,2, L. Cuervo-Pardo2, S. Chiarella1, 1. Rochester, MN; 2. Gainesville, FL

Introduction: Vitamin B12 can regulate both cellular and humoral immunity. Here we report a patient with vitamin B12 deficiency and recurrent oral candidiasis who had a significant decrease in the frequency of oral candidiasis after vitamin B12 supplementation.

Case Description: A 56-year-old man with a history of chronic obstructive pulmonary disease and tobacco use was referred to our Immunology Clinic for recurrent oral candidiasis (Candida albicans and Candida glabrata in fungal cultures from throat swab) and concern for immunodeficiency. He reported multiple episodes of oral candidiasis in the past 2 years that required different antifungal regimens. The patient denied using inhaled corticosteroids or dentures. Laboratory results were remarkable for macrocytosis, mildly increased T and B cell numbers, low vitamin B12 level (170 ng/L), and negative HIV testing. IgG and IgM were mildly decreased, while IgA was normal. Lymphocyte proliferation to mitogens and antigens was also normal. After receiving vitamin B12 supplementation, his vitamin B12 level normalized and the patient reported a marked decrease in the frequency of oral candidiasis.

Discussion: Multiple risk factors have been associated with recurrent oral candidiasis, including smoking, use of inhaled corticosteroids, xerostomia, dentures, and defects in cellular immunity. The present case report suggests that vitamin B12 deficiency might be an additional risk factor for recurrent oral candidiasis. Further studies to evaluate the role of vitamin B12 in eliciting a normal immune response to Candida species are warranted.

M143
B CELL LYMPHOPENIA AND HYPOGAMMAGLOBULINEMIA: THE IMPORTANCE OF SCREENING FOR PRIMARY IMMUNODEFICIENCY BEFORE INITIATING IMMUNOSUPPRESSION
A. Navalpakam*, J. Kepes, Detroit, MI

Introduction: A 16-year-old male with hypogammaglobulinemia, absent B cells, and recurrent otitis media presents 7 years after rituximab treatment for Granulomatosis with Polyangiitis (GPA). However, a review of his chart showed these abnormalities were present prior to initiation of immunosuppressive therapies.

Case Description: A 16-year-old male with history of ANCA-positive GPA with severe pulmonary disease diagnosed in 2013 treated with rituximab who entered clinical remission presented to immunology clinic in January 2021 with persistent hypogammaglobulinemia (IgG 543) and undetectable CD19. He had a history of recurrent ear infections requiring 7 tympanostomy tube placements with subsequent hearing loss prior to any immunosuppression.

In 2014, he received cyclophosphamide for 6 months, then one course of rituximab, followed by low-dose azathioprine.

Review of studies obtained prior to rituximab in 2014 were significant for low IgG (467), low IgM (47), and an absolute B cell count of only 9. On two repeat evaluations in 2021, 7 years after rituximab was discontinued, he had undetectable CD19+ cells, persistently low IgG, and low/undetectable IgM. Pneumococcal titers were protective to only 18/23 serotypes. Tetanus and diphtheria titers were protective. B cell subsets showed only 16 CD19+ cells and no definitive populations of B cells were identifiable. Patient was started on IVIG for management of CVID.

Discussion: B cell recovery from rituximab typically occurs within 18 months of therapy cessation. Persistent hypogammaglobulinemia with B cell lymphopenia is concerning for primary immunodeficiency. Screening immunology studies prior to beginning immunosuppressive agents can prevent the masking of a primary immunodeficiency disorder.

Figure 1. Table describes persistently low IgG with low/undetectable IgM and low CD19+ B cells over the course of 7 years prior to and after rituximab treatment.

M144
TNFAIP3 DEFICIENCY AS AN UNCOMMON CAUSE OF HYPOGAMMAGLOBULINEMIA
C. Cotter, N. Hartog*, Grand Rapids, MI

Introduction: Heterozygous variants in TNFAIP3 cause an autosomal dominant haploinsufficiency of A20 (HA20). HA20 presents as an autoinflammatory disease characterized by oral/genital ulcers, recurrent fevers, skin lesions, gastrointestinal ulcers, and autoimmunity. While there is increasing recognition of monogenic etiologies of common variable immunodeficiency (CVID), this has not been classically described in TNFAIP3 deficiency. We present a case of HA20 presenting atypically as CVID and inflammatory bowel disease (IBD). This case demonstrates the importance of broad evaluation for monogenetic etiology in primary immunodeficiency as every patient does not present with a “classic presentation”.

Case Description: A 12-year-old male presented with IBD, multiple non-infectious pulmonary nodules, and splenomegaly. His work up revealed giardia infection decreased IgG (283), IgM (15), and undetectable IgA. He had non-protective post-Pneumovax titers, and an absolute lymphocyte count of 640. He was diagnosed with CVID and started on intravenous IgG replacement. At age 25, he developed inflammatory arthritis of the right ankle and left wrist treated with sulfasalazine. Genetic testing revealed a pathogenic heterozygous variant in TNFAIP3 (c.133C>T; p.R45*).

Discussion: Associated humoral immunodeficiency has rarely been documented with HA20, especially in absence of classic auto-inflammatory disease. This patient did not have oral/genital ulcers,