infection-free despite IgG trough <500 mg/dL. At his 2-year visit, CD19 counts normalized while CD8 and CD16/56 counts remained unchanged. He was cleared for live vaccines and will receive varicella first with his pediatrician.

**Discussion:** Active immunosuppression, neutropenia, and T cell lymphopenia or dysfunction are contraindications to live vaccination. CDC recommends deferring live vaccines for minimum of 8 months after last IVIG. Allergist/immunologists are well-poised to co-manage acquired immunodeficiency by performing immune evaluation and interpreting guidelines, enabling patients and providers to use shared decision-making to maximize patient outcomes.

**M148**

**A CASE OF CUTANEOUS BOTRYOMYCOSIS IN A PATIENT WITH X-LINKED AGAMMAGLOBULINEMIA**

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**Introduction:** Botryomycosis, a rare granulomatous inflammatory response to bacteria, commonly involves the skin. Botryomycosis is reported with various immunodeficiency states, particularly acquired cell-mediated immunodeficiency disorders. We describe, to our knowledge, the first case of cutaneous botryomycosis in a patient with X-linked agammaglobulinemia (XLA).

**Case Description:** The patient is a 28-year-old man who was diagnosed with XLA at age 3 years due to recurrent pneumonia, very low serum immunoglobulin levels, and a maternal uncle with XLA. His history was notable for Campylobacter bacteremia and tibial osteomyelitis but was without overt infections on immunoglobulin replacement for the past five years. Two years ago, he started to develop scattered noduloulcerative skin lesions on forearms, lower back, and abdomen. Lesions were worse in the summertime. He worked in construction and attributed these skin lesions to mosquito bites incurred at his workplace. He had no systemic symptoms. Skin biopsy revealed numerous dermal basophilic clusters of cocci with surrounding inflammatory infiltrates of neutrophils, histiocytes, and eosinophils. Tissue culture grew methicillin-sensitive *Staphylococcus aureus*. He declined intravenous antibiotics and oral cephalaxin was attempted but discontinued because of an allergic reaction. A 3-month course of trimethoprim/sulfamethoxazole resulted in the resolution of skin lesions.

**Discussion:** Botryomycosis results from a combination of host immune factors, bacterial load, and virulence. Cutaneous botryomycosis requires repetitive skin trauma as seen in our patient’s construction work. Previously reported in patients with cell-mediated immunodeficiencies, cutaneous botryomycosis should also be considered in XLA patients especially those with repetitive skin trauma. Targeted antibiotic therapy can be curative.

**M149**

**PARTIAL DIGEORGE SYNDROME WITH PROLONGED T-CELL LYMPHPENIA**

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**Introduction:** The clinical presentation of DiGeorge Syndrome (22q deletion) is highly variable, particularly with respect to the immunologic manifestations. The length of time required for recovery of immune system abnormalities is not well-defined in current literature. This case demonstrates spontaneous immune reconstitution in a patient with partial DiGeorge syndrome following a prolonged period of T-cell lymphopenia.

**Case Description:** A one-month-old female had abnormal newborn TREC screening. 22q11.2 deletion was identified by FISH. Manifestations included severe conotruncal heart defects, bronchomalacia, facial dysmorphisms, and lymphopenia. Immunoglobulin levels were also low. Initial flow cytometry revealed markedly low T, B, and NK cell counts. Mitogen proliferation studies were normal. She received immunoglobulin replacement and antimicrobial prophylaxis. Only at 28 months of age were her immunoglobulin levels, CD4 lymphocyte and NK cell counts all normal for age. Total CD3 and CD8 lymphocyte counts remain low but continue to increase.

**Discussion:** This patient meets the definition of partial DiGeorge syndrome based on TREC copies indicating decreased thymic output with normal mitogen proliferation studies. She demonstrated gradual improvement but did not reach functional immune reconstitution until well after two years of age. Time to full immune reconstitution in partial DiGeorge Syndrome may be highly variable as indicated in this case. Frequent follow up is essential to monitor needs and progress in these patients.

**M150**

**FAMILIAL MEDITERRANEAN FEVER IN A 6-YEAR-OLD FILIPINO CHILD: A CASE REPORT**

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**Introduction:** Familial Mediterranean Fever (FMF) is an auto-inflammatory disease characterized by periodic episodes of fever and recurrent serositis with characteristic symptom-free intervals. It is classically known to be an ethnically restricted disease predominantly affecting those of Mediterranean descent, until recently, when high prevalence (31.9%) of FMF was reported in Europe and Asia on patients presenting with unexplained fever. Migration pattern from the Mediterranean basin was linked to the spread of the disease throughout the world.

**Case Description:** We present a case of a 6-year-old female, Filipino, seen at the outpatient department with a 2-year history of periodic fever accompanied by abdominal pain occurring for 2-4 days, every 19-25 days interval, noted to be asymptomatic in between episodes. Acute phase reactants (ESR and CRP) were elevated during febrile episodes and declines to normal once afebrile. Our patient was given Colchicine 500mcg BID which resulted in a significant decrease in duration and frequency of febrile episodes.

**Discussion:** FMF results from mutations in the MEFV (Mediterranean Fever) gene which encodes the pyrin protein. MEFV gene analysis showed a heterozygous c.442G>C, p.E148Q mutation without mutations found with the mother and a homozygous variation E148Q from the father. To date, this is the only confirmed case of FMF in the Philippines.

**M151**

**A RARE CASE OF AUTOIMMUNE AUTONOMIC GANGLIONOPATHY**

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**Introduction:** Autoimmune autonomic ganglionopathy (AAG) is a rare disease mediated by antibodies causing severe orthostatic-hypotension. Due to its unique presentation, patients can go undiagnosed causing them profound disability and poor-quality of life. Our case of AAG presenting in a 69-year-old-female illustrates the importance of including AAG in the