Since January 2020 Elsevier has created a COVID-19 resource centre with free information in English and Mandarin on the novel coronavirus COVID-19. The COVID-19 resource centre is hosted on Elsevier Connect, the company's public news and information website.

080 Effects Of The COVID-19 Pandemic On A Group Of Patients With Pathogenic Variant of Cytotoxic T-Lymphocyte Associated Protein 4 (CTLA-4) in a Tertiary Center in Florida

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RATIONALE: Immune dysregulation and tendency for inflammation are risk factors for severe COVID-19 as described in several autoimmune disorders. Similarly, the clinical course of patients with immune dysregulation secondary to inborn errors of immunity in COVID could also be severe as shown in NFKB1 and NFKB2 deficient patients. However, data is limited on clinical and laboratory features of patients with COVID-19 and/or SARS-CoV-2 immunization and cytotoxic T-lymphocyte associated protein 4 (CTLA-4) deficiency, which is the focus of our study.

METHODS: We performed retrospective chart review and telephone interviews of genetically confirmed CTLA4 deficiency patients for demographic information, clinical history including acute infection, and infectious exposure mitigation, immune phenotype and immune response after SARS-CoV-2 immunization. Antibody response to SARS-CoV-2 were measured by ELISA and photonics ring resonance assay.

RESULTS: Our patient cohort included 2 children and 6 adults (median age 23.5 years, range 15-51). All adults have clinical disease and required treatment for hypogammaglobulinemia and immune dysregulation. Patients demonstrated varying levels of exposure mitigation. So far, only one patient was infected—a young adult who had benign course of disease. All adults, and one out of 2 vaccine eligible children, are fully immunized. SARS-CoV2 antibody levels of 2 patients tested thus far (1 post-vaccination, 1 post natural infection) were < 6.25% that of vaccinated healthy controls.

CONCLUSIONS: During this pandemic, most of our CTLA-4 patients have not experienced COVID-19. Antibody response to the SARS-CoV-2 vaccine and/or infection were suboptimal which can increase their vulnerability. T cell studies are needed to further understand immune responses.

081 New onset of autoimmune diseases in patients with primary antibody deficiency and CD4 lymphopenia after COVID-19

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RATIONALE: Patients with primary antibody deficiency (PAD) have increased susceptibility to infectious and non-infectious complications including autoimmune diseases. It is speculated that SARS-CoV-2 can disturb self-tolerance and trigger autoimmune responses in COVID-19. Here, we present two cases of patients with PAD who developed new onset of autoimmune disease after COVID-19.

METHODS: We performed chart review of two cases using electronic medical records.

RESULTS: Case 1: A 58 year old female with common variable immune deficiency (CVID) was hospitalized for COVID-19 pneumonia which required high dose steroid. As the steroid was tapered, she developed significant dyspnea. EKG revealed new onset of left bundle branch block and echocardiogram showed ejection fraction of 48%. Cardiac MRI was consistent with myocarditis with abnormal T2 mapping. Upon immune work up, CD4 count was 239 cells/mm^3.

Case 2: A 66 year old female with a history of selective IgG subclass 2 deficiency and CD4 lymphopenia (111 cells/mm^3) developed increasing dyspnea over the two months after SARS-CoV-2 infection. This did not improve with multiple courses of antibiotics. CT chest showed multiple enlarged mediastinal lymph nodes, biopsy of which revealed non-necrotizing granulomatous inflammation, consistent with sarcoidosis.

CONCLUSIONS: We believe these are the first reported cases for new onset of autoimmune diseases after COVID-19 in PAD patients. New onset autoimmune disease should be considered in patients with PAD especially with CD4 lymphopenia who experience prolonged dyspnea after COVID-19.

082 Patient with AR-CGD Presenting with Disseminated Aspergillosis During Pregnancy Associated with Suppression of the Th17 Axis

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RATIONALE: Pregnancy is associated with suppression of the Th17 axis to induce tolerance to the fetus. Here, we report a patient with chronic granulomatous disease (CGD) presenting with disseminated Aspergillosis during pregnancy.

METHODS: Genetic sequencing was performed at GeneDx. p47-phox expression was assessed by immunoblot. T helper cell phenotyping was performed via flow cytometry and intracellular cytokine staining.

RESULTS: 35-year-old G2P1 female presented at 36 weeks of gestation with fever, lower back pain, and weight loss. Infectious workup revealed vertebral osteomyelitis and CNS, breast, mediastinal abscesses due to Aspergillus fumigatus. Histograms from multiple dihydrorhodamine (DHR) tests were consistent with autosomal recessive (AR) CGD. Targeted sequencing revealed a single heterozygous pathogenic variant (c.75_76delGT) in NCF1. Further analysis by immunoblot confirmed deficient p47-phox expression. The patient improved on long-term antifungal therapy with voriconazole and caspofungin and immunomodulation with interferon-gamma and G-CSF. T helper cell polarization shifted over time, demonstrating a 4.8-fold decrease in Th1/Th17 ratio and no significant change in Th1/Th2 ratio from 1 to 7 months postpartum.

CONCLUSIONS: This is the first reported case of AR-CGD presenting as an invasive fungal infection during pregnancy. The Th17 axis is critical for controlling fungal infections including Aspergillus. We hypothesize that our patient's susceptibility to disseminated Aspergillus in the setting of AR-CGD was augmented by a prolonged immunosuppressed state secondary to two pregnancies within 24 months. We confirmed her relative Th17 suppression in the immediate postpartum period and demonstrated recovery of the Th17 compartment 6 months later.