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**Results:** A total of 17,234 patients with a PIDD were hospitalized from 2004-2018. There were 2.8 new PIDD diagnoses and 6.3 hospitalizations of PIDD patients per 1,000 discharges. These metrics were unchanged during the study period. The number of new diagnoses for B cell and antibody defects significantly increased over time. The number of new PIDD diagnoses significantly increased in adolescents/adult and decreased in infants. T cell disorders had the highest number of ICU admissions and highest healthcare expenditures. There were 747 PIDD patients who underwent HSCT; complications of HSCT significantly decreased over time. Age at transplant decreased for combined immunodeficiency patients. Mortality rates significantly decreased in all PIDD patients and in patients receiving HSCT.

**Conclusions:** The total hospitalizations and incidence of PIDD within the hospitalized pediatric population were unchanged. There were significant changes in the class of PIDD diagnosed, the age at diagnosis, and healthcare utilization metrics. Mortality significantly decreased over time within the PIDD cohort.

### A043

#### ORAL CORTICOSTEROID–SPARING EFFECT OF MEPOLIZUMAB IN PATIENTS WITH EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS

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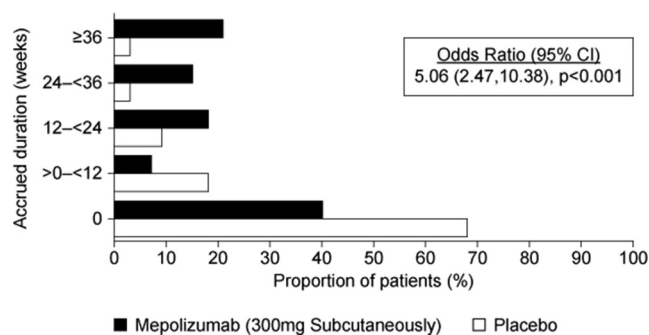


**Introduction:** Mepolizumab is an anti-interleukin-5 monoclonal antibody, previously demonstrated to increase time in remission in patients with eosinophilic granulomatosis with polyangiitis (EGPA). We investigated the oral corticosteroid (OCS)–sparing effect of mepolizumab using data from the Phase III MIRRA study.

**Methods:** In MIRRA, patients with relapsing/refractory EGPA, receiving stable prednisolone/prednisone  $\geq 7.5$ – $\leq 50$ mg/day for  $\geq 4$  weeks before baseline, were randomized (1:1) to mepolizumab 300mg or placebo subcutaneously every 4 weeks for 52 weeks, plus standard of care. OCS was tapered per physician judgment, following a recommended protocol. Mean OCS daily dose during Weeks 48–52 and cumulative OCS exposure over the treatment period were analyzed; accrued weeks of OCS  $\leq 4$ mg/day and proportion of patients receiving OCS  $\leq 4$ mg/day at Weeks 36 and 48 were assessed post hoc.

**Results:** Overall, 136 patients participated (mepolizumab[n=68]/placebo[n=68]). During Weeks 48–52, more patients received lower average daily prednisolone/prednisone doses (categories: 0, >0– $\leq 4$ , >4– $\leq 7.5$ , >7.5mg/day) with mepolizumab versus placebo (odds ratio [95% CI]: 0.20[0.09,0.41],  $p < 0.001$ ). Mean (SD) cumulative OCS exposure over the treatment period was also reduced with mepolizumab (3286.9[2095.83] vs 4710.0[2625.82] mg with placebo). Accrued weeks of OCS  $\leq 4$ mg/day over the treatment period were increased with mepolizumab versus placebo (odds ratio [95% CI]: 5.06[2.47,10.38],  $p < 0.001$ ; **Figure**). The proportion of patients receiving  $\leq 4$ mg/day OCS at both Weeks 36 and 48 was also greater with mepolizumab than placebo (41% vs 10%; odds ratio [95% CI]: 6.63[2.57,17.12],  $p < 0.001$ ).

**Conclusions:** In patients with EGPA, mepolizumab is associated with reduced OCS use, an important treatment objective owing to OCS-associated adverse effects.



**Figure.** Proportion of patients with accrued duration of OCS  $\leq 4$ mg/day over the 52-week treatment period.

### A044

#### COVID-19 IN CVID: A LARGE HOSPITAL EXPERIENCE

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**Introduction:** Despite common variable immunodeficiency (CVID) being a relatively common primary immunodeficiency that predisposes patients to respiratory disease, little is known about the outcomes of patients with CVID who are infected with SARS-CoV-2 and how CVID patients respond to SARS-CoV-2 vaccinations. The goal of this study was to report the outcomes of CVID patients infected with SARS-CoV-2 at a single institution

**Methods:** We used a chart review methodology to identify all patients with common variable immunodeficiency who were infected with SARS-CoV-2 from March 1, 2020–May 20, 2021 at Mayo Clinic.

**Results:** There were nineteen patients with CVID who were infected with SARS-CoV-2 at our institution. All patients were PCR tested and were positive. All patients survived. Five patients (26%,  $n=5/19$ ) patients were hospitalized. Four patients (21%,  $n=4/19$ ) received monoclonal antibodies in the outpatient setting. Post-infection (anti-nucleocapsid) seroconversion occurred in 5/6 patients assessed (83%). Post COVID-19 infection, 32% ( $n=6/19$ ) received vaccinated and tolerated it without significant adverse events. Post-immunization (anti-spike) serology was positive in the one patient in which it was assessed.

**Conclusion:** This is one of the largest case series from the United States that describes the experience of a tertiary care center of managing patients with CVID and COVID-19 infection. In our institutional experience, COVID-19 disease course in CVID patients is similar to most other patients and the presence of CVID *per se* does not seem to increase the risk of mortality with COVID-19. Additionally, vaccination seems to be safely tolerated in CVID patients post COVID-19 infection.

### A045

#### INADEQUATE SARS-COV-2 ANTIBODY RESPONSE TO MRNA SARS-COV-2 VACCINE IN ADOLESCENT KIDNEY TRANSPLANT RECIPIENTS

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**Introduction:** Community spread of SARS-CoV-2 is waning with vaccination. It is unclear if kidney transplant recipients (KTR) remain at significant risk of contracting COVID19 due to inadequate antibody response. We investigated immunological