PULMONARY MANIFESTATIONS AND OUTCOMES IN ANCA ASSOCIATED VASCULITIDES: A SINGLE CENTER EXPERIENCE.

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Background: ANCA-associated vasculitides (AAV) are a group of systemic diseases most commonly reported to involve kidneys and lungs. The literature on pulmonary manifestations in pediatric AAV is variable. Information on long-term pulmonary outcomes is limited. We aim to describe pulmonary manifestations among different types of AAV, presentation and outcomes after therapy.

Materials/Methods: We performed a retrospective chart review of patients with ICD-10 associated with AAV. We included all patients <18 years of age at presentation, between 1/2008-2/2018. We reviewed data over the first three years after presentation. Data was described using mean and standard deviations for continuous variables and counts and percentages for categorical variables.

Results: Forty-nine patients met inclusion criteria. Most common ethnic group was Hispanic (46.9%) followed by Caucasian (32.7%). Mean age at presentation was 11.88±4.87 years. Twenty-three patients had microscopic polyangitis (MPA), 13 patients had granulomatosis with polyangiitis (GPA) and 2 patients had eosinophilic granulomatosis with polyangitis. 11 patients were unclassified. 36 patients (73.5%) had pulmonary manifestations at any point, with 29 patients symptomatic at presentation. Most common manifestations were pulmonary hemorrhage (62.2%) and cough (67.6%). Younger Hispanic patients with MPA were identified as a high-risk group for pulmonary “rebleed”. Respiratory failure was seen in 21.6% patients and 16.2% required intubation. 68 computed tomography scans of the chest of 30 patients were available for review with the majority showing some abnormality. On spirometry, obstructive and restrictive patterns were rare; diffusion defect was common and persistent throughout the three years. Follow up data at 6, 12 and 36 months after diagnosis and treatment showed pulmonary manifestations in 61.8%, 39.4% and 29% patients respectively. Treatment modality and response were comparable across different AAV

Conclusions: Pulmonary manifestations in AAV can present at diagnosis or later during the course of the disease. MPA was more common than GPA in our population. Diffuse alveolar hemorrhage is common and severe but responds well to therapy as demonstrated clinically and by CT finding improvement. Interestingly, a high risk group of MPA patients had more pulmonary involvement including relapse despite similar treatment modalities. There is a need for further studies on pulmonary manifestations of AAV specifically MPA which is not well described in the literature