symptoms ranged from birth to 15 years. A diagnosis of an AIS was made in 57% with PFAPA (periodic fever, aphthous stomatitis, pharyngitis, adenitis) accounting for the majority. An undetermined autoinflammatory syndrome was suspected in 40%. 3% who attended had no symptoms or features supportive of an AIS. Overall, 36% were found to have a confirmed genetic mutation, either benign or pathogenic and 28% had a positive family history of auto-immune or auto-inflammatory syndrome in a 1st degree relative. Colchicine monoclonal antibody (52%) was the first choice of therapy followed by anakinra monotherapy (19%) or a combination of both. All treated patients reported symptom improvement on therapy with the exception of 2.

**Conclusion** This study gives an overview on the spectrum of autoinflammatory disease presenting in Ireland and their current management. The majority of patients attending were appropriately referred to this clinic and had improvement in symptoms following initiation of treatment.

**REFERENCE**

**P374 LARYNGEAL SARCOIDOSIS RESPONSIVE TO TREATMENT WITH SIROLIMUS: A FIRST CASE REPORT**

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We report the case of a 15 year old girl with a four month history of gradual onset dysphonia and dysphagia. Endoscopy revealed a significantly enlarged epiglottis and arytenoids causing supraglottic airway obstruction (pinpoint airway). She progressed to emergency tracheostomy and a biopsy of the epiglottis was taken. Histology revealed lymphoid hyperplasia with focal non-necrotizing granulomata. A screen for infection was negative, as was a serum ACE. The patient was commenced on corticosteroids and methotrexate, but after three months, there was no improvement. Therafter, she was switched to sirolimus, which led to resolution of her epiglottic swelling, allowing the tracheostomy to be reversed and corticosteroids switched to sirolimus, which led to resolution of her epiglottis and arytenoids causing supraglottic airway obstruction (pinpoint airway). The majority of patients attending were appropriately referred to this clinic and had improvement in symptoms following initiation of treatment.

**Conclusion** This study gives an overview on the spectrum of autoinflammatory disease presenting in Ireland and their current management. The majority of patients attending were appropriately referred to this clinic and had improvement in symptoms following initiation of treatment.

**REFERENCE**