

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 syndromes 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 monotherapy (32%) 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

1. Ter Haar NM, Oswald M, Jeyaratnam J, *et al.* Recommendations for the management of autoinflammatory diseases. *Ann Rheum Dis* 2015;**74**:1636–1644.

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

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10.1136/archdischild-2019-epa.720

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. Thereafter, she was switched to sirolimus, which led to resolution of her epiglottic swelling, allowing the tracheostomy to be reversed and corticosteroid therapy to be discontinued.

Conclusion This is the first case, to our knowledge, of sirolimus being successfully used in treatment of laryngeal sarcoidosis. Sirolimus could be considered a treatment option in these instances, particularly in the presence of lymphoid aggregates on histology.

P375 QUALITY OF LIFE ASSESSMENT IN PATIENTS WHO RECEIVES SCIG AND IVIG

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10.1136/archdischild-2019-epa.721

Introduction Primary immunodeficiency diseases (PIDs) describe a group of clinically and genetically heterogeneous disorders that afflict lymphoid and myeloid lineages.

Immunoglobulin replacement therapy has been the standard treatment for patients with antibody production deficiency since the 1950s. Human serum immunoglobulin manufactured from pools of donated plasma can be administered intramuscularly, intravenously or subcutaneously. Subcutaneous route is a safe and efficacious method to prevent serious bacterial infections in patients experiencing difficulties with IVIG.

Aim Treatment satisfaction and HRQOL in patients with PIDs was evaluated upon switching from intravenous (IVIG) or subcutaneous immunoglobulins (SCIGs) to 10% SCIG (Kiovig®).

Material and methods Twelve patients who received SCIG and 14 patients who received IVIG were included in the study. THE WHOQOL-BREF questionnaire was applied to all cases. The WHOQOL-BREF https://www.who.int/substance_abuse/research_tools/en/english_whoqol.pdf.

Results Male to female ratio was 8/4 in SCIG group and 8/6 in intravenous IgG (IVIG) group. The median age was 31.2 years (minimum=5 years, maximum=49 years) in SCIG group and 23.2 years (minimum=5 years, maximum=47 years) in IVIG group. All patients who received SCIG were taking IVIG before. When the median scores of the responses to each question were examined according to before and after SCIG treatment; improvement was seen in 13 questions, deterioration in 2 questions. When the total quality of life score was evaluated before and after SCIG and in the IVIG group, the highest median score belonged to the patients receiving SCIG (median=104, minimum=44, maximum=130). In addition, the total quality of life score was found to be 10 points higher in SCIG patients than before. The questions with deterioration in the answers were 'How much do you need any medical treatment to function in your daily life?' and 'Are you able to accept your bodily appearance?'. We think that the worsening of these questions can be due to recurrent injections and local side effects. No serious side effects were seen in both groups. SCIG was performed without any problem in a female pregnant patient who had anaphylactoid reactions to different IVIG brands.

Conclusion The number of studies which investigated the effects of the SCIG treatment on quality of life in Turkey were limited. In this study, SCIG was found to be a safe treatment modality that improves the quality of life.

P376 PARECHOVIRUS INFECTION AS THE PRESENTING ILLNESS IN AN INFANT WITH SCID

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10.1136/archdischild-2019-epa.722

Introduction Severe Combined Immunodeficiency (SCID) is a heterogeneous group of primary immunodeficiency disorders with impaired cellular and humoral immunity. This leads to increased susceptibility to life-threatening bacterial, viral and fungal infections. Incidence is approximately 1 per 58,000 live births.¹Typically, SCID presents in the first months of life with severe, recurrent infections, failure to thrive, dermatologic manifestations or chronic diarrhoea. We present a case of newly diagnosed SCID and examine parechovirus viral loads before and after treatment.