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.

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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. 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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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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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.¹ Typically, 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.

Case A thriving, term five-month old infant was admitted with symptoms suggestive of bronchiolitis and lower respiratory tract infection. In her medical history she had mild eczema at eight weeks old, was exclusively breastfed from birth and was unvaccinated. Admission investigations showed an elevated c-reactive protein of 41 mg/L, and white cell count (WCC) of $7.8 \times 10^9/L$ with lymphocytes of $0.7 \times 10^9/L$. Chest radiography showed bilateral lung field opacifications. She was treated with intravenous (IV) amoxicillin and supplemental oxygen. Viral throat swab and stool samples respectively isolated rhino/enterovirus and norovirus. Cough swab and nasopharyngeal aspirate (NPA) isolated methicillin-sensitive staphylococcus aureus and scanty haemophilus influenzae respectively. Good clinical response to therapy was observed and she was discharged after 10 days.

Her respiratory symptoms returned following discharge; she developed a purulent ear discharge treated with oral co-amoxiclav by her GP. She was readmitted with persistent bilateral pneumonia shortly after this. Repeat investigations showed a WCC of $3.2 \times 10^9/L$ with lymphocyte count of $0.1 \times 10^9/L$. IV cefuroxime, PO azithromycin and oxygen therapy were commenced. Repeat NPA was positive for rhino/enterovirus and parechovirus. Immunoglobulin subsets showed severe deficiency of IgG, IA and IgM. Lymphocyte subsets demonstrated undetectable B cells, minimal T cells and deficient NK cells consistent with SCID. IV ceftriaxone and co-trimoxazole were commenced with aciclovir and fluconazole prophylaxis. Weekly intravenous immunoglobulin therapy (IVIG) was commenced and definitive treatment with haematopoietic stem cell transplant (HSCT) is planned. Genetic testing revealed RAG2 mutation. Serial viral load testing showed a gradual increase in the CT (crossing target) with weekly IVIG treatment indicating significant reduction in parechovirus viral load.

Discussion Without timely diagnosis and management, SCID is typically fatal within the first year of life. Although HSCT is the only definitive treatment option, IVIG and broad antimicrobial prophylaxis is life-saving. In this case, repeated IVIG therapy correlated with reduction in parechovirus viral load and clinical improvement.

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P377 COMPARISON OF THE IMMUNE RESPONSE TO BCG VACCINATION IN NEWBORN BABIES BORN FOLLOWING ASSISTED REPRODUCTION, WITH THOSE BORN BY NATURAL CONCEPTION: A PROSPECTIVE COHORT STUDY

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Background Various studies have reported conflicting findings regarding the growth, development, congenital anomalies and co morbidities in babies born following ART, in comparison to their naturally conceived (NC) peers. However hardly any studies so far have reflected on the immune function and response to routine newborn vaccination in these babies.

Study design/Methodology 58 term, AGA, Singleton babies born following assisted reproduction and weighing >2.5 kg were administered 0.1 ml of BCG vaccine within 48 hours of birth. Likewise 62 babies (matched cohorts) born following natural conception were also administered BCG in their left deltoid insertion. At 3 months of age during routine follow up visits their BCG scars were analysed by the same observer. At 6 months of age those babies in both the groups with visible BCG scars, were administered 5 TU PPD (0.1 ml) in their left forearm. After 72 hours the maximum transverse diameter of the indurations were measured (by a single observer). An induration diameter of >5 mm and <10 mm were considered a positive tuberculin conversion rate.

Results Out of the 58 babies [30 (51.72% were males, with a mean birth weight 2.56 kg (SD 0.05)] born following ART, 52 had developed clearly visible scar (Mean scar size 3.91 mm with a SD 0.36) marks. So the scar failure rate was 10.3% (6/58). Out of the 62 naturally conceived babies [32 (51.61%) were males with birth weight (mean 2.61 kg SD 0.06)] 55 had BCG scars (mean scar size 4.13 mm SD 0.40). The scar failure rate was 11.2% (7/62). The positive tuberculin skin test conversion rate in the naturally conceived group of babies were 49 out of 55 with BCG scars. Hence the scar conversion rate in this group was 88.71% (49/55). While in the ART group 36 out of 52 had a positive skin test conversion. So the scar conversion rate in this group was 69.23% (36/52).

Conclusion The size of BCG scars is significantly more in the NC group in comparison to the ART group. There was a significantly lower tuberculin skin test conversion rate in the ART group in comparison to the NC group, as per this study. The scar failure rates were almost identical in the two groups. Further bigger studies are needed to evaluate the implication (if any) of this finding. We need to explore whether this findings truly reflect any deviation in the cell mediated immune response in the ART group.

P379 CHILDHOOD BRUCELLOSIS IN AL BAH, KSA (EPIDEMIOLOGICAL AND CLINICAL OBSERVATIONS IN 209 CASES)

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Introduction and aim of the work

Saudi Arabia is hyper endemic for Brucellosis, with more than 8000 cases reported each year to public authorities.

The aim of the study is to document the epidemiological, clinical and laboratory pattern of brucellosis in children of Al-Baha, also to ascertain the efficacy of certain antibiotic regimen to treat the disease.

Patients and methods All patients less than 13 years old in Al-Baha whose diagnosis was brucellosis during the study period (October 2016-September 2017) were included in the study. Diagnosis was based on clinical grounds confirmed by serology and culture. The initial treatment for children <8 years old consisted of Rifampicin 15 mg/Kg/day and Co-trimoxazole (TMP/SMX) of 10/50 mg/Kg/day, given for 6 weeks. For older children, Doxycycline (4 mg/Kg/day) was substituted for co-trimoxazole. Those who were very ill and toxic were in addition administered gentamycin (5 mg/Kg/day) for 7 days.