blood results revealed platelet count of 1. The rest of his blood results were essentially normal.

On examination, extraoral facial, limb and trunk bruising were noted. Lips were crusted with blood and surrounded petechiae. Intraoral examination revealed soft palate petechiae, areas of ulceration/petechiae in the left and right buccal mucosa and a minor bleed from the lower left lateral primary incisor. The findings were consistent with a diagnosis of Immune Thrombocytopenia Purpura (ITP) precipitated by a viral infection. Initial treatment was with tranexamic acid for five days. On review, there was a marked improvement noted with only minimal blood crusting of the lower lip and fewer intraoral petechiae evident. No further treatment has been required to date. The child is being monitored by haematology and dental specialists with regard to signs, symptoms and progress.

ITP is a haematological disorder defined by low platelet count. Children often develop this following a viral infection. Spontaneous bleeding or bleeding induced by trauma, may be the first clinical signs of ITP. This interesting case highlights a primary presentation of ITP with oral signs. This situation may present to a General Dental Practitioner or General Medical Practitioner in community or another Healthcare Professional and it is important to recognise the signs and symptoms and intervene appropriately

P341 REVIEW OF PAEDIATRIC PATIENTS WITH SEVERE FACTOR $\times$ DEFICIENCY IN IRELAND

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Severe Factor $\times$ (FX) deficiency is a rare autosomal recessive bleeding disorder. Patients with FX:C level of $<0.01\text{--}0.03$ iu/ml have a severe bleeding phenotype with haemarthrosis occurring in 69% of patients and intracranial haemorrhage accounting for 15% of all bleeding events. Prophylactic FX replacement is challenging and usually requires a central venous access device (CVAD). This review included 4 paediatric patients with severe FX deficiency, aged between 1 and 16 years. Three were born to consanguineous parents. Factor $\times$ mutations have been identified in all patients. 2/4 of this patient cohort were diagnosed at birth due to a previously identified family history, 1/4 was diagnosed at day three of life when he presented with an intracranial haemorrhage and 1/4 presented at day three of life with epistaxis. All 4 patients commenced Prothrombin Complex Concentrate (PCC) prophylaxis in the first week of life. Dosing regimens range from 25 – 60 units per kilogram once to twice weekly to maintain trough factor $\times$ level >0.05 iu/ml. There have been no spontaneous life or function threatening bleeding episodes while on prophylaxis. All patients had CVAD inserted with PCC support. Three patients had CVAD removed because of Staph aureus septicaemia. One of these three patients developed CVAD related iliofemoral thrombosis, necessitating anticoagulation. In conclusion, with early recognition and diagnosis of severe FX deficiency, bleeding symptoms can be effectively treated and managed.

P342 CLINICAL COURSE IN CHILDREN DIAGNOSED WITH HEREDITARY SPHEROCYTOSIS IN ALBANIA

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Objective To describe the clinical spectrum of children diagnosed during 15 years with hereditary spherocytosis, classify them on 4 groups based on their clinical data and evaluating their outcome and giving our recommendations for their management.

Methods This is a analytic, observational, retrospective and case-control study analyses of 30 children diagnosed and followed up at our center during 15 years (2003–2018). The patient were diagnosed with hereditary spherocytosis based on their clinical history, clinical examinations, and on a positive osmotic fragility curve. They were divided based on the clinical form on 4 groups : mild, moderate, moderate to severe and severe group. The groups were compared between them regarding the laboratory data, the need for transfusion, cholelithiasis, splenic sequestration, aplastic crises and splenectomy.

Results In our study 3 patients had mild H.S (10%), 10 patients had moderate H.S (33.3%), 10 patients had moderate to severe H.S (33.3%) and 7 patients had severe H. S (23.4%). The mean age at diagnosis was 4.7 years Patient with severe form were younger than patients with moderate form ($p=0.016$) and had lower hemoglobin level ($p\leq0.001$), higher reticulocyte count ($p=0.049$) and also needed more transfusions ($p=0.002$). There was not found a correlation between the level of bilirubin and clinical gravity ($p=0.873$). Splenic sequestration was commonly found, 63.3% of patients experienced a splenic sequestration and its frequency was significant higher in severe patients ($p=0.011$). Cholelithiasis was present in half of the patients but its development was not related with disease's severity ($p=0.391$). Aplastic crisis was relatively rare, only one patient developed aplastic crisis and it was due to parvovirus infection. Splenectomy was performed in 16 patients (53.3%) with the main indication transfusion dependence and splenic sequestration.

Conclusion We have a high number of children with severe clinical forms because as they do severe forms they came at our center and are diagnosed compare with mild forms which are asymptomatic and remain undiagnosed. The clinical course of our patients was relatively benign, but we still have a high frequency of splenic sequestration 63.3% (most of them due to recurrent infections) a high rate of blood transfusion (54%) and a high number of splenectomised patients.

P344 A RARE ASSOCIATION: ACUTE DISSEMINATED ENCEPHALOMYELITIS IN CHILD AFFECT BY AUTOIMMUNE HAEMOLYTIC ANAEMIA AND AUTOIMMUNE THROMBOCYTOPENIA

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Acute disseminated encephalomyelitis (ADEM) is an inflammatory demyelinating illness, characterized by a

monophasic course frequently associated with infections or previous immunization. Regarding pathogenesis, encephalomyelitis is associated with some immunological mechanisms. Post-infection or post-vaccination inflammatory reactions in the perivenular region in the central nervous system (CNS) may be explained by a molecular mimicry mechanism. An antecedent infection was identified in 72–77% of paediatric patients.

We present a case of a 5 year-old girl in follow up for autoimmune thrombocytopenia (AT) and autoimmune haemolytic anaemia (AHA) in steroid treatment, with history of left hemiparesis, language delay, from as a consequence of perinatal suffering, who has developed ADEM.

This patient presented with progressive weakness of limbs, neck pain associated with fever from one day. It was acute in onset and gradually progressive. She became quickly hypotonic and hyporesponsive and had reduced level of consciousness. Within 12 hours of admission, the patient developed sphincter incontinence and dysphagia.

After exclusion of papilloedema, lumbar puncture has been performed. An urgent MRI was performed, which showed multiple subcortical lesions of varying size showing hyperintensities in TR at the bridge and in T2 at the medulla.

Cerebrospinal fluid (CSF) study showed cell count of 40 cells/mm³, protein 58 mg/dL, glucose 54 mg/dL. No oligoclonal bands were present in CSF. Blood and CSF cultures were negative like other infectiological analyzes. Also CSF-PCR for the presence of bacteria and virus was negative. Her autoimmune profile with antinuclear antibody was also negative.

The clinical features and the MRI findings were suggestive of ADEM. Partial quadriplegia, and reduction of reflexes, as seen in the myelitic form of ADEM, were present. She had developed ADEM while on the maintenance dose of prednisolone.

After diagnosis, intravenous methylprednisolone was given at 30 mg/kg daily for 5 days. After 48 hours there was a significant improvement in the patient's clinical condition. A regimen of oral steroid was advised after intravenous therapy. The patient responded well to steroid therapy. No residual lesion was found on follow-up.

AT and AHA are relatively uncommon. There are studies postulating the possibility of a combination of several autoimmune diseases. Very few cases have been found with this rare association in the literature. Follow-up of these patients is essential for detecting the development of other autoimmune disease in such cases.

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AN UNUSUAL PRESENTATION OF HAEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS

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Introduction We present a case of an unusual clinical presentation of Haemophagocytic Lymphohistiocytosis (HLH). HLH is a rare, life-threatening hyperinflammatory condition resulting from dysregulation of normal innate and adaptive immune responses. The common presenting features of HLH are fever, rash, neurological symptoms, hepatosplenomegaly,

lymphadenopathy, cytopenias, high serum ferritin and liver function abnormalities(2).

Case We present a case of HLH in a 5 year old girl presenting to a large regional secondary paediatric unit with unusual clinical features. Although pyrexia is a well-recognised presenting feature of this condition, the predominant features in our case were persistent left-sided pleuritic chest pain with a loud flow murmur and radiological evidence of lower lobe atelectasis with an associated pleural effusion and cardiomegaly. An initial diagnosis of a lower respiratory tract infection on a background history of a yet-to-be-determined cardiac structural defect was made and she was commenced on broad spectrum IV antibiotics and supportive care.

She remained pyrexial and unwell despite 48 hours of IV antibiotics and a repeat chest x-ray revealed radiological deterioration, precipitating a decision to refer her for tertiary paediatric care. Further investigations confirmed a diagnosis of HLH.

Conclusion Although rare this serious medical condition can be life threatening and increased awareness is needed, particularly when clinical features at presentation are not characteristic of the condition, to improve early clinical detection, intervention and prognosis.

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BLOOD TRANSFUSION IN NICU

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Introduction Blood products transfusion is of critical importance to the care of sick and premature infants. Considering the significant drop in hemoglobin level due to the physiological factors after birth till the tenth to twelfth week of life, this study aimed to evaluate the volume of transfused blood in NICU at privet hospital, Mashhad, Iran during 6 years.

Materials and methods In this cross sectional study, all the infants admitted to NICU of the hospital (2011–2017) were evaluated in terms of the volume of transfused blood. 34% of 24183 patients in this ward were female. 5% of them were preterm. More than 6% weighted less than 2500 gram.

Results Of all the admitted patients in this ward, 569 subjects received blood and the related products, most of which 51% (291 neonates) received FFP, and the least 14% (80 neonates) needed PLT. Meanwhile, more than 27% (159 neonates) received PC. It should be noted that most of the neonates (35%) were reported with blood type O⁺ and the least (1.5%) with AB⁻.

Conclusion Regarding the high volume of transfused blood in NICU, it is essential to focus our attention on the appropriate use of blood and blood products as well as the prevention of transfusion-associated infections.

Although almost more than 70% of NICU patients require less than 20^{cc} blood, the adult blood bags are used for this purpose and the fairly large quantities of the blood are discarded. Therefore, it is suggested a special blood bag is designed for neonatal transfusion.