

Abstracts

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 X DEFICIENCY IN IRELAND

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Severe Factor X (FX) deficiency is a rare autosomal recessive bleeding disorder. Patients with FX:C level of <0.01–0.03 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 X 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 X 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 ($p\leq 0.001$), 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