negative (LTCD3+ TCR αβ+ CD4- CD8-) about 27% (control < 2.5%). The study of FAS gene allowed the identification of a mutation in exon 9.

Conclusion ALPS is an underestimated entity that must be considered in non malign lymphoproliferation, autoimmunity and expansion of an unusual population of a/ß CD3+CD4-CD8- (double-negative T cells>1%).

515 A RARE CASE OF LANGDON DOWN SYNDROME WITH COMPLETE ENDOCARDIAL CUSHION DEFECT, TETRALOGY OF FALLOT, DEFICIENCY OF FACTOR VII

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Aims We sought to summarize a very rare association between multiple rare incidence diseases in a patient with Langdon-Down syndrome and also to correctly document each pathology and use the best course of treatment.

Background Factor VII deficiency has an incidence of 1 in 500.000 reported cases. Complete endocardial cushion defect [ECD] occurs in 2% percent of all congenital heart defects. Additional cardiac abnormalities (persistent ductus arteriosus and tetralogy of Fallot [ToF]) may occur in 10% of all ECD's. Associated defects are rare in children with Down syndrome.

Methods A 5 weeks old infant with a Down phenotype was admitted in the Intermediate Care Unit for severe tonic-clonic seizures and an unexplored heart murmur. A computed tomography scan revealed a massive hemorrhaging in the fronto-parieto-occipital left cerebral region. Trauma was excluded and the prothrombin time was prolonged with the activated partial trhomboplastin time normal so we sent a blood sample for the factor VII activity.

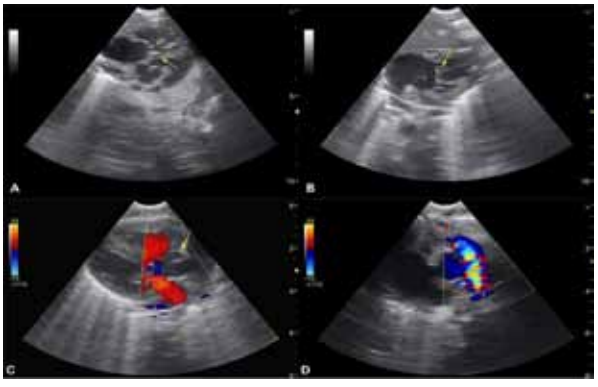
We performed an echocardiography.

A karyotype study was carried out.

Results

- Complete ECD with the common atrio-ventricular valve in dextroisomerism, left ventricle hypoplasia, associated with ToF.
- The factor VII activity showed a 2% activity level
- classical 21 trisomy

Conclusion We provided a good documentation of a very rare association between separate severe pathologies and we showed that when faced with a congenital malformative syndrome one should never stop looking for other abnormalities.



Abstract 515 Figure 1

A- subcostal view- the arrow points to the insertion of anterior left leaflet on a papillary muscle, the X's show a ventricula

B- subcostal view- the common AV valve and the atrial septal defect;

C- parasternal long axis- the over-riding aorta;

D- parasternal short axis- turbulent flow through the pulmonary valve (pulmonary artery stenosis)

516 A VERY RARE CASE REPORT OF HERMANSKY-PUDLAK SYNDROME TYPE II

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Introduction Hermansky-Pudlak syndrome type 2 (HPS-2) is a very rare multi-system disorder characterized by oculocutaneous albinism, reduced visual acuity, horizontal nystagmus, bleeding diathesis and recurrent infections due to neutropenia and impaired cytotoxic activity. HPS-2 is caused by mutations in the AP3B1 gene (5q14.1) and is transmitted in an autosomal recessive manner. The gene product is the Beta 3A subunit of adaptor protein 3 (AP3), involved in vesicle formation and protein sorting. Here we report a very rare case of HPS-2 who admitted because of fever.

Case Report A 5 months-old female patient admitted to our hospital because of fever. She is the first child of a consanguinous parents. She had mild facial dysplasia, whitish-yelllow hair and horizontal nystagmus. Opthalmological evaluation showed oculocutaneous albinism. Moderate hepatosplenomegaly was revealed. Anemia (Hb; 8.3 gr/dl) and neutropenia (0.4×109/μL) with normal platelet count were documented. Bone marrow aspiration yielded hemophagocytosis. Triglyceride, ferritin and fibrinogen levels were in normal limits. She was treated with proper antibiotic treatment and discharged to follow-up in outpatient clinic. Neutropenia was subsequently fluctuated. She had been hospitalized six more times due to febrile neutropenia and at each admission cytopenia including thrombocytopenia (15×109/μL) in addition to hepatosplenomegaly were revealed. Time to time increased triglyceride levels were documented. All episodes were resolved with proper antibiotic and r-HuG-CSF treatment,. without requiring HLH treatment. Genetic analysis revealed homozygous nonsense mutation in exon 18 of the AP3B1 gene.

Conclusion Patients with albinism and opthalmological complaints should be evaluated for Hermansky-Pudlak syndrome.

517 THALASSEMIA PREVENTATION AND ACTIVITY OF PEDIATRIC HEMATOLOGY AND ONCOLOGY DEPARTMENT AT BANGABANDU SHEIKH MUJIB MEDICAL UNIVERSITY IN BANGLADESH

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Background and Aims Thalassemia is a genetic and crucial disease. Approximately 240 million peoples are suffering from this disease. Every year 10 million children are suffering from this disease. Hb-E dieases is available in south east Asia, north east India and Bangladesh. Originalally Hb-E disease is 5 times more than Beta Thalassemia in Bangladesh.

Method The outbreak of this disease is not calculated at this moment but carrier is 15 million. We are collected experimental data from 3 hundred voluntiers in our center. They have not family history of Thalassemia. In that experimental data Beta Thalassemia carrier 2.33% and Hb-E Carrier 10%. If we are experiment among the people who have history of Thalassemia, this disease is increased no doubt. This disease have actually no curable treatment except BoneMarrow tranplantation. Treatment cost is excessive and unbearable. Only time to time Blood transfusion and costly drug is given for the increasing of life span. Treatment cost of every 30kg child need 4 lac taka every year. If 2 bag blood need every patient in every month, 1 lac 20 thousand bag blood will be need every month in Bangladesh.

Result Bonemarrow tranplantation is the only curable treatment of this disease. About 3 million to 10 million taka need for this treatment. Above this situation we learn that prevention is only way to reduce this diseases. Screening system NESTROFT is available in our center. Only 10 taka need. Some cases DNA analysis will be need.
Conclusion Antenatal diagnosis is important and available in our center. If the foetus suffering from this disease legal termination of pregnancy should be need. This way we can free from the disease.

518 HEMOLYTIC ANEMIA ASSOCIATED WITH INTRAVENOUS IMMUNOGLOBULIN

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Introduction Intravenous immunoglobulin (IVIG) associated hemolytic anemia is a potentially serious complication that is often overlooked. Here we describe a case of Kawasaki disease (KD) who recurrently developed coombs positive hemolytic anemia following IVIG administrations.
Case Report A three-years-old girl admitted with the complaint of fever, swelling of the hands and feet with palmar erythema. Investigations revealed the diagnosis of KD and she was treated with IVIG (2 gr/kg) and aspirin. The fever subsided within a day but restarted after 6 days. A second course of IVIG was administered. On day 4 after second course of IVIG, laboratory evaluation revealed hemoglobine level of 8.3 gr/dL. Her red blood cells became positive on polyclonal IgG Coomb's testing (DAT). Aspirin was stopped and steroid was started. Her original signs had resolved and hemoglobine level gradually increased up to 11.7 gr/dL. Eight months later, she readmitted with significantly enlarged servical lymph nodes in parallel with previuos symptoms consistent with the diagnosis of recurrent KD. Hemoglobin level was 11 gr/dL and DAT was negative. Following single dose of IVIG treatment, hemoglobine level gradually decreased and became 6.6 gr/dL on 30th day of treatment with DAT positivity. Within first week of steroid treatment, hemoglobine level incresaed to 8.9 gr/dL. The patient is now free of any symptom with an hemoglobine level around 11.5 gr/dL.
Conclusion It is important that physicians using high dose IVIG are aware of the risk of hemolysis. Careful monitoring of hemoglobin levels during IVIG treatment may provide proper diagnosis and early intervention.

519 CLASSIC KAPOSI SARCOMA WITH PULMONARY INVOLVEMENT MIMICKING ENDOBRONCHIAL TUBERCULOSIS IN A CHILD

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Kaposi' sarcoma (KS) is a low-grade vascular neoplasm and classic KS, a subtype of KS, is extremely rare in children. Childhood pulmonary involvement in classic KS has not been reported in the literature. We describe an HIV-seronegative pediatric case with a fulminant course of classic KS with pulmonary involvement mimicking endobronchial tuberculosis.

520 DIAGNOSIS AND TREATMENT PECULIARITIES IN AN INFANT WITH BLEEDING DISORDER

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Background and Aims One of bleeding causes due to vitamin K deficiency is gut flora destruction secondary to antibiotic treatment early in life. Authors emphasize diagnosis and treatment difficulties for an infant with massive uncontrolled bleeding.
Methods Authors present a 5 weeks-old breastfed infant transferred in pediatric clinic for severe anaemia. Family history: healthy parents, no consanguinity. Case history: recent respiratory infection treated with antibiotics; no recent trauma or surgery. Clinical exam: skin pallor, petechiae, ecchymoses, jaundice, huge haematoma (20/14 cm), wide-spread from neck to lumbar area.
Results Blood investigations: severe anaemia (Hb=3.5g/dl), severe hyponatremia, normal liver function, negative serology for celiac disease. Negative test for cystic fibrosis. Hemostasis evaluation: normal values for bleeding time, platelets and fibrinogen; significant prolongation for prothrombin time and activated partial thromboplastin time.

Evolution: Infant developed fulminant seizures secondary to hyponatremia and bleeding at venous puncture sites, justifying urgent initiation of anticonvulsant therapy and recombinant human coagulation factor VII, even before first hemostasis evaluation. Despite of therapy, bleedings symptoms persisted and became more severe. According to hemostasis investigations, we diagnosed vitamin K deficiency and we reconsidered the treatment using K vitamin. Prompt improvement of bleeding after vitamin K therapy confirmed vitamin K deficiency. After blood transfusion authors noticed haemoglobin(Hgb) improvement (at discharge Hgb=14.1 g/dl).

Conclusions

1. Authors emphasize diagnosis and treatment difficulties in an infant with severe bleeding because of vitamin K deficit;
2. In cases with severe bleeding, it's mandatory to consider vitamin K treatment;
3. Antibiotic treatment should be carefully considered in infants.

521 UNUSUAL PRESENTATION OF DISSEMINATED PANDEMIC INFLUENZA A (H1N1) 2009 IN AN INFANT

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Background Children are the risk group for severe disease of Pandemic influenza A (H1N1) 2009 infection. A case of disseminated manifestation of pandemic H1N1 influenza has been rarely reported.
Methods We present a case of a-3-month-old male infant who manifested with clinical sepsis and can be demonstrated the evidence of disseminated pandemic H1N1 influenza in bone marrow prior having respiratory symptoms.
Results The patient presented with high fever for 1 day. The initial diagnosis was sepsis but he had persisted fever with hepatosplenomegaly. Complete blood count persistently showed pancytopenia. Bone marrow aspiration and biopsy on day 8 showed predominant population of maturing myeloid precursors. In contrast, erythroid precursors were virtually absent. PCR tested in serum was negative for Epstein-Barr virus, cytomegalovirus, dengue virus and parvovirus. On day 11, he developed respiratory distress and required ventilator support. Bronchoalveolar lavage was positive for pandemic H1N1 influenza by both RT-PCR and viral culture. The staining marrow specimens performed on day 8 with immunofluorescence