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

technique to detect nucleoprotein (NP) antigens and RT-PCR were positive for pandemic H1N1 influenza. The virus antigen was also detected by flow cytometry using monoclonal antibody specific for NP labeled with fluorescein isothiocyanate (FITC). Interestingly, most of positive cells were CD14+ cells and the amounts of positive cells were 10% of total marrow cells. His immunological profiles were normal.

Conclusion This infant demonstrated the evidence of disseminated pandemic H1N1 influenza prior having pneumonic symptoms. Infant is more susceptible to have disseminated manifestation and can present initially as clinical sepsis.

522 ASPERGILLUS NIGER: AN UNUSUAL CAUSE OF INVASIVE PULMONARY ASPERGILLOSIS

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Objective Report two cases of chronic granulomatous disease presenting with uncommon cause of invasive pulmonary aspergillosis (IPA).

Methods We report two cases of IPA in two 6-year-old twin girls with a family history of primary immunodeficiency and a personal medical history of recurrent bronchopneumopathy referred to our Pediatric Department in July 2011 for investigations regarding persistent pneumonia and biological inflammatory syndrome. IPA was confirmed by isolating Aspergillus niger from bronchoalveolar lavage and radiology results. Amphotericin B therapy was initiated intravenously for 1 month relayed then by oral Voriconazole for two months. Clinico-biological evolution was favourable. Follow-up computed tomography showed full regression of the pulmonary infiltrates and the thoracic wall mass after 3 months of antifungal therapy. Investigation for immune deficiency revealed chronic granulomatous disease. Our two patients started then on prophylactic antibiotics with co-trimoxazole. We are currently considering HLA identical bone marrow transplantation for them in a near future.

Conclusion Although is a well recognized clinical entity, invasive disease caused by aspergillus niger is less common when compared to aspergillus fumigatus and other Aspergillus species .These two case reports demonstrate the potentially aggressive nature of aspergillus niger and highlight the importance of looking for an immune deficiency particularly in the case of uncommon infection such as aspergillosis in early childhood.

523 A CASE REPORT; BRUCELLOSIS TRIGGERING HEMOLYTIC ANEMIA IN GLUCOSE-6-PHOSPHATE DEHYDROGENASE DEFICIENCY

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Background and Aims Glucose-6-phosphate dehydrogenase (G6PD) deficiency is the most common red blood cell enzyme deficiency in worldwide. It may lead to acute hemolytic anemia triggered by infection, drugs or broad beans (favism). To the best of our knowledge, this is the second reported case of a Turkish patient with brucellosis triggering hemolytic anemia in glucose-6-phosphate dehydrogenase deficiency.

Case A 5-year-old male was admitted our hospital with malaise, fever, abdominal pain, vomiting, jaundice, and darkening of urinary color of one week duration. There were no intake drug, infectious diseases or favism in patient's history. On physical examination conjunctivae were pale, sclerae were subicteric and the other system examinations were normal. Laboratory examinations revealed leukocyte count of 22800/mm^3, hemoglobin of 5.7g/dl, platelets of 596000/mm^3, total bilirubin:4.3, indirect bilirubin:3.9 mg/dl, lactate dehydrogenase:1440U/l, AST:190IU/L, ALT:181IU/L. The corrected reticulocyte count was 9.3% and serum haptoglobin level was low (< 10). There was hemoglobinuria in the urinary examination. Direct and indirect Coombs tests were negative. The value of G6PD was low (3.8 IU/g) where osmotic fragility was normal. Serum standard tube agglutination (Wright) test for *Brucella* sp was positive at a titer of 1/640. The patient was treated by rifampicin and gentamicin.

Conclusion We presented this case report to show that acute brucellosis might trigger an acute hemolytic attack in a patient with underlying G6PD deficiency. This is important especially in countries such as Turkey, where brucellosis is endemic and there is a high frequency of G6PD deficiency in the population.

524 TWO PEDIATRIC CASES OF CEREBRAL VENOUS SINUS THROMBOSIS WITH DIFFERENT PRESENTATIONS

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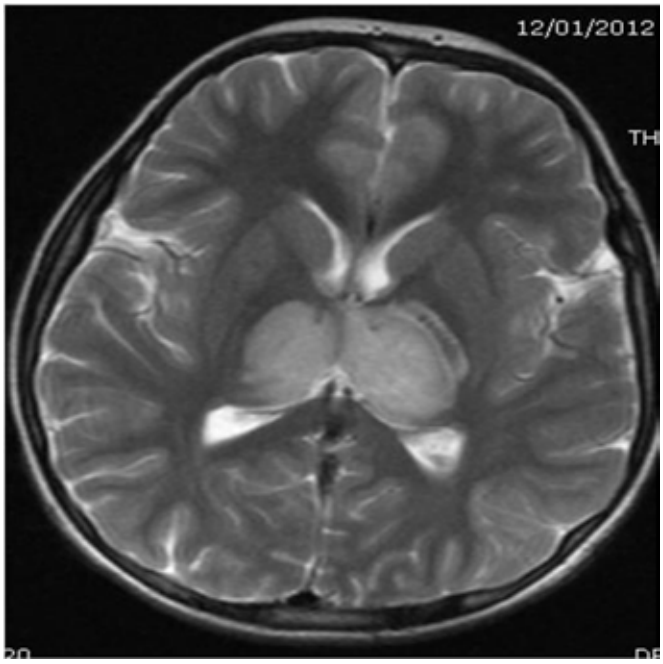
Background and Aims Cerebral Venous Sinus Thrombosis (CVST) is a serious and rare disorder in pediatric patients.

Case reports: Case 1: An 8-year-old girl admitted with left middle ear infection, bilateral sixth cranial nerve palsy and papilledema. The Magnetic Resonance Imaging (MRI) venography showed occlusion of superior and inferior sagittal sinus totally and left sigmoid sinüs parsial.Pansinüsitis, left otitis media and mastoiditis were also determined. Evaluation for thrombophilia and tests for specific predisposing conditions were normal. The patient was treated by third generation cephalosporin, clindamycin, acetazolamide and low-molecular weight heparin after the initial diagnosis.

Case 2: A 12-year-old boy presented with aphasia and lethargy after a week of dental infection. Brain MRI and MRI venography demonstrated thalamic enfarct, thrombosis of deep cortical veins, left transverse and sigmoid sinuses.



Abstract 524 Figure 1