with widespread port wine stain on his face, lower lip, arms, legs and trunk were noted. Brain magnetic resonance imaging revealed vascular malformations located periventricularly and adjacent to the corpus callosum. On brain magnetic resonance angiography ectatic, varicose deep veins (venous malformations) were detected. Lower extremity MRI showed hypertrophy of bones and soft tissues on the left side. As the child has capillary malformations (port wine stain), soft tissue and bone hypertrophy and vascular malformations a diagnosis of KTS was made. KTS consists of two major features; congenital vascular malformations and disturbed growth. For diagnosis presence of either capillary or venous malformations with disturbed growth of the bone or soft tissues is required. Children with the diagnosis of CP should be carefully examined for any finding suggesting a genetic disease.

567 NEAR FATAL CASE OF AMLODIPINE POISONING IN AN INFANT

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Background and Aims Infants contribute about 5.25% of total poisoning exposures in humans. We report first case of infant to have survived Amlodipine intoxication.

Methods 11 month old infant received 12.5 times the maximum therapeutic dose of amlodipine as a result of a medication error. He presented with vomiting, lethargy, breathlessness, muffled heart sounds and progressed to hypotensive shock within hours of admission. He received mechanical ventilation, fluid therapy with normal saline and inotropes. Peripheral pulses remained feeble and blood pressure was 70/40 mm of Hg. High dose insulin infusion at 0.5 u/kg/hr with intravenous 25% dextrose and intramuscular glucagon for maintaining euglycemia was given. Continuous infusion of calcium gluconate at 0.5 mEq/kg/hr was started simultaneously. To manage prerenal failure, oliguria and congestive cardiac failure induced pulmonary oedema peritoneal dialysis was initiated.

Result Infant improved rapidly after insulin and dextrose infusions (for 15 hours) along with glucagon and calcium gluconate infusion (for 72 hours) was initiated. Calcium channel blockers (CCB) are phenylalkylamines. CCB act by binding to the α unit of the L Type calcium channels. Insulin secretion being calcium dependent, blockage of the L type channel results in impaired insulin secretion causing hypoglycemia. CCB poisoning results in insulin resistance. A continuous increase in blood sugar levels may predict sudden decline in hemodynamic variables rather blood pressure and pulse measurements.

Conclusion Hyperinsulinaemia/euglycaemia therapy is beneficial in seriously intoxicated patients with CCB induced hypotension, hyperglycemia, and acidosis. There may be benefit in using them in combination with other standard therapies.

568 NEONATAL HEMOLYTIC UREMIC SYNDROME

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Haemolytic uremic syndrome (HUS) is characterised by the triad of non immune haemolytic anaemia, thrombocytopenia and renal failure. It is rare in newborns. Only 5 cases of neonatal HUS have been reported to-date. We describe a series of 2 cases of neonatal HUS where one had definite history of birth asphyxia and another was an abandoned baby without clinical details about perinatal period. The known modalities of treatment include transfusion of plasma and plasmapheresis. In first baby haematological and renal parameters improved after double volume exchange transfusion while second

baby responded to transfusion of fresh whole blood (packed red cells + plasma). Exchange transfusion essentially provides plasma exchange besides augmenting the haemoglobin levels. Arguably this is the first report of neonatal HUS treated with exchange transfusion.

569 THORACOSCOPIC SYMPATHECTOMY FOR LONG-QT SYNDROME IN A SICKLE CELL ANEMIA PATIENT

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Background and Aim Long QT syndrome (LQTS) is a relatively common disease, however, it is rarely described in Black African patients. We would like to present a west African patient with sickle cell disease and LQTS who was treated with thoracoscopic left sympathectomy and 1/3 lower stellectomy.

Methods An 11-year old boy with sickle cell anemia (Hb=7.2g/dl) was also found to have LQTS. He has had >15 defibrillations in 5 years. He was on high dose beta-blockers but required a number of shocks despite it. Implantable defibrillator was rule out due to his home country lack of cardiac surgical expertise in case of complications. He was considered to be a suitable candidate for sympathectomy. This was done through a 3 trocar approach to the left upper chest. Sympathetic chain including stellate ganglion was exposed. 20mls of 0.5% lignocaine was sprayed over the ganglions. Prior to excision of sympathetic chain, all visible nerves traveling medially (to the heart) was excised with hook diathermy. Lower 1/3 of stellate ganglion as well as T2-T4 ganglia was resected en-bloc and submitted for histology.

Results There were no per- or post-operative complications and no evidence of Horner's syndrome post surgery. The patient was discharged 72 hours later. QTc interval before surgery was 532ms and reduced to 448ms prior to discharge.

Conclusion Sympathectomy including lower portion of stellate ganglion is a viable alternative for patient with LQTS.

570 PULMONARY HYPERTENSION AS A COMPLICATION OF VON RECKLINGHAUSEN DISEASE

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Introduction Neurofibromatosis (NF) is occasionally complicated by systemic vasculopathy such as myocardial infarction, renovascular hypertension etc. The appearance of pulmonary embolism (PE) is not described until yet.

Case Report 15-year-old female patient with neurofibromatosis type I. Progressive dyspnoea, general fatigue and chest pain beginning 6 days before hospital admission. Anamnestic no clinical sign for deep vein thrombosis. No oral contraceptive, non smoker. Laboratory findings: Elevated troponin, plasma brain natriuretic peptide (BNP) and D-dimer. Normal ECG, echocardiogram showed right ventricular and atrial enlargement with moderate tricuspid regurgitation and estimated right ventricular systolic pressure at 45 mm Hg and dilated main pulmonary artery. Left ventricular function was normal.

Chest x ray showed right heart enlargement but normal lung structure. Lung CT revealed massive PE in left and right branches and a mediastinal tumor. Lysis with Alteplase was given for 4 days. Normalisation of laboratory and echocardiogram. Because of the tumor angiographic-MRI was done for follow-up and showed normal flow pattern in pulmonary vasculature. The tumor was classified as a ganglioneuroma. Genetic thrombophilia testing was negative,

the ganglioneuroma is surgically removed. The child was discharged with low weight heparine.

Conclusion Von Recklinghausen disease generally has a good prognosis. Major risks for morbidity and mortality are vascular complications. In case of pulmonary hypertension in these patients early diagnosis and sufficient therapy is essential to avoid major complications. Pulmonary embolism and NF as a cause for pulmonary hypertension has not been described before.

571 MALIGNANT PERTUSSIS IN THE YOUNG INFANT: A CASE REPORT

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Infections by *bordetella pertussis*, has resurged in many countries around the world including developed regions. Where as new vaccination strategies for adolescents, pregnant women and adults have been recommended, mortality affecting young infants is still significant. Patients with severe cases present with extreme leukocytosis and develop refractory hypoxemia and pulmonary hypertension that is unresponsive to maximal intensive care.

Objective To report a case of malignant pertussis in a 4 weeks old infant.

Design a descriptive case report.

Patient a 4 weeks old boy was admitted to the intensive Care Unit with respiratory distress and a diagnosis of bronchiolitis. Both parents and a 19 months old sibling had upper respiratory infection symptoms. Four days before admission he started with coryza and cough. The cough was not paroxysmal and cyanosis and whooping were never present. He presented a marked leucocytosis with lymphocytosis. Respiratory failure occurred rapidly from the fourth day with subsequent deterioration due to the development of severe pulmonary hypertension and multiorgan failure which caused the infant's death after 4 days of intensive care therapy.

Although *Bordetella Pertussis* was not confirmed by PCR, culture or serology the findings on the autopsy were suggestive of malignant pertussis.

Comments mortality due to malignant pertussis remains superior to 75% regardless life support measures. Adults are the main source of contamination of non-immunized infants, thus immunization of older adolescents and adults who will have close contact with infants aged < 12 months is the most effective preventive measure.

572 INTRACTABLE DIARRHEA FROM CYTOMEGALOVIRUS COLITIS IN AN IMMUNOCOMPETENT ADOLESCENT

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Introduction We reported CMV colitis in an immunocompetent adolescent who was critically ill with septicemia and significant intractable diarrhea that responded to specific CMV treatment.

Case A 15-year-old boy who previously known hereditary spherocytosis was referred to our hospital because of anemia, increased transaminase level, and massive cholestasis. The abdominal ultrasonography was detected cholelithiasis, choledoch stones, and dilated proximally bile ducts. External biliary drainage tube was placed into the choledoch. On the following days, splenic rupture and sepsis developed. Therefore, the patient was underwent splenectomy and cholecystectomy. After closed of external biliary drainage tube, patient was developed severe dehydration and malnutrition due to watery stool. The colonoscopy and colonic biopsies was performed. It was shown macroscopic colitis and CMV intranuclear inclusion

bodies in rectosigmoid colon. Moreover, PCR for CMV DNA in blood (6142 copy/mL) and colonic biopsy specimens was positive. The immunologic screen tests were normal. Parenteral gancyclovir for 21 days and oral gancyclovir therapy was continued two weeks. The patient resolved completely, serum PCR for CMV DNA was detected negative after two months.

Conclusion CMV colitis, although rare in immunocompetent adolescent, should be considered in the differential diagnosis of severe colitis when other causes fail to explain the course of disease.

573 UNCOMMON PRESENTATION OF UMBILICAL SEPSIS IN NEWBORN CAUSED BY HERPES INFECTION

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Herpes simplex virus (HSV) is a widespread pathogen. Comparatively neonatal HSV infections are rare but frequently severe with high mortality and morbidity. Early detection and treatment, has been shown to decrease the mortality significantly.

We present a rare presentation of neonatal herpes in an 8 day old neonate who presented with fever and pustules around the umbilicus. Baby was treated with intravenous benzylpenicillin and flucloxacillin. However, baby was still febrile after 48 hours of treatment with vesicular lesions now appearing around the umbilicus. There was no history of genital herpes or cold sores in family. Viral swab was taken and acyclovir started. Wound swab and blood PCR was positive for HSV 2. Lumbar puncture done at day 14 was clear. Baby was treated with two weeks of intravenous acyclovir. There were problems with intravenous access with difficult access and later central line causing thrombosis.

Baby represented at 5 weeks age with vesicular umbilical lesions and was treated with 2 weeks course of oral acyclovir.

Many issues were highlighted in this case.

Lumbar puncture was not done early on as umbilical sepsis was thought to be focus of infection. It was difficult to establish whether this neonate had CNS herpes.

Long term intravenous treatment is easily said than can be given. This neonate had difficult access; central line was not without complication. What is the best way forward?

The lesions recurred within 2 weeks of the baby going home. Suppressive Acyclovir therapy - Would it be beneficial for him?

574 VARICELLA ZOSTER INFECTION COMPLICATED BY MENINGOCOCCAL MENINGITIS, SEPSIS AND SUBDURAL EMPYEMA IN AN INFANT, A CASE REPORT

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Aims To highlight possible and may be fatal complications of Varicella Zoster virus (VZV) infection.

Method We report a case of an infant with VZV infection who developed meningococcal sepsis, meningococcal meningitis, VZV encephalitis and subdural empyema.

Results A 3 month, previously well and developmentally normal male child was admitted with skin rash. He had contact with varicella zoster infection. His initial inflammatory markers were high. He was treated as Chicken pox. His blood culture grew *Neisseria Meningitidis*, treated with intravenous antibiotics. He remained unwell with poor response to antibiotics. A lumbar puncture was then performed. This revealed a picture consistent with bacterial meningitis. CSF PCR was positive for *N. Meningitidis* as well as for VZV. A week after admission he started to have seizures. This required intubation and PICU admission. A brain CT showed to