P65

GITELMAN SYNDROME: CASE REPORT OF A TODDLER FOLLOWING SALBUTAMOL INHALER¹Vasanthee Sundram*, ¹Edwina Daly, ²Mary Waldron. ¹Tallaght University Hospital, Dublin, Ireland; ²Our Lady's Children's Hospital, Crumlin, Dublin, Ireland

10.1136/archdischild-2019-epa.420

Background Gitelman syndrome (GS), is an inherited autosomal recessive renal disorder characterized by hypokalemic metabolic alkalosis with significant hypomagnesemia and low urinary calcium excretion. It is a rare disorder which usually manifests in early adulthood with muscle weakness, fatigue, muscle cramps, and less commonly by abdominal pain, nausea and vomiting.

Case report We report on a 2 year old boy who presented to the Emergency Department with a two weeks history of coughing. His parents had been administering regular Salbutamol inhaler to control his symptoms.

He had a previous admission three months earlier and was discharged on inhaled Salbutamol to be used as required.

His weight was 11.6 kgs (25–50th percentile), height was 87.5 cm (50th percentile) and BMI 15.2 Kg/m²

He was miserable on examination with mild recession, bilateral expiratory wheeze with crackles in both bases.

Initial laboratory investigations showed a normal full blood count, urea and electrolytes, a normal venous blood gas with a C - reactive protein of 45 mg/L.

Chest x-ray reported patchy infective changes in the left lower lobe with diffuse perihilar inflammatory changes.

The patient was commenced on intravenous Amoxicillin and 2.5 mgs, oral steroids and nebulised Salbutamol to be administered every four hours.

On day 3 of admission, he was noted to be extremely lethargic and was barely responding to his mother's voice which was not in keeping with the improvement of his respiratory symptoms.

Repeated blood gas analysis showed a significant metabolic alkalosis (pH 7.552) and hypokalemia (2.7 mmols/L) He was started on intravenous fluids with added KCl 20 mmols/500ml. Nebulised Salbutamol was changed to the inhaled form and Inhaled Fluticasone was added.

ECG showed flattened T waves, possible U waves and a QTc of 460. Urinary creatinine, phosphate, sodium, calcium and Magnesium were requested. A Renal ultrasound and sweat test were arranged which were subsequently normal.

The nephrology Team were consulted who confirmed the diagnosis of Gitelman syndrome and recommended regular Ibuprofen for three days. He made an uneventful recovery and his potassium and blood gas normalised after two days.

Discussion Gitelman Syndrome is often difficult to diagnose in Paediatrics because it usually presents in adolescence or early adulthood. Sudden cardiac arrest has been reported and it is therefore an extremely important condition to recognise particularly when prescribing potassium lowering agents like Salbutamol and Steroids.

P66

A CASE OF PSEUDOHYPONATREMIA DUE TO HYPERCHOLESTEROLAEMIA IN A PATIENT WITH CHOLESTASIS

Hafiz Shafiq Butt*, Lubna Mahmood, Alfonso Herrera. St Luke's Hospital, Kilkenny, Ireland

10.1136/archdischild-2019-epa.421

Introduction Serum consists of water (93% of serum volume) and non aqueous components, mainly lipids and proteins (7% of serum volume) . Sodium is restricted to serum water. In states of hyperlipidaemia or hyperprotenaemia, there is an increased mass of non aqueous components of serum and a concomitant decrease in the proportion of serum composed of water.

The term pseudohyponatraemia refers to the apparently low sodium concentration found in plasma when blood specimens with high concentration of either lipid or protein are analysed by traditional methods.

Pseudohyponatraemia secondary to hypercholesterolaemia is a rare condition. In the diagnostic approach for the patients with hyponatraemia, it should always be differentiated during the initial steps. High concentration of intravascular protein or lipid ' dilute ' the plasma sodium concentration, but do not alter the solute concentrations of intracellular or interstitial fluid compartments. Thus hyponatraemia associated with hyperlipidaemia or hyperprotenaemia can be considered artifactual.

Case report Two years old boy was evaluated for seven weeks history of cholestatic Jaundice. During the workup for cholestasis, a significant drop in serum sodium, 120 mmol/L was noted. Child was asymptomatic apart from Jaundice. Serum Cholesterol level was significantly raised to 52.2 mmol/L (normal 2 - 5). Triglyceride and LDL was unavailable due to icterus, HDL was 0.45 (normal 1–2) . A concomitant Sodium level checked on Spot gas analyser was 139 mmol/L (normal).

Other Labs included raised ALT, AST, total bilirubin, alkaline phosphatase and Gamma GT, slightly low total protein and albumin. Viral Hepatitis screen was negative. Autoantibodies negative. Serum Ceruloplasmins, alphafetoproteins, and alpha1 antitrypsin levels were normal. Abdominal Ultrasound showed Fatty liver without biliary dilatation. Needle liver biopsy showed paucity of bile ducts. Comprehensive genetic testing did not reveal a cause for his cholestasis. Later on, he also developed xanthomata on his fingers.

A final diagnosis of Cholestasis with bile duct paucity of unknown aetiology was made. LFT remained abnormal six months into the illness and is under followup by Gastroenterologist. Repeat Liver biopsy showed disease progression. He is on Ursodeoxycholic acid, AquaDEK and oral vitamin K.

Conclusion This case shows that hypercholesterolemia can cause pseudohyponatraemia. It highlights that If there is any suspicion regarding a low serum Sodium in the presence of cholestasis, a serum Sodium should be re tested using blood gas analyser as this method is not subject to interference from hypercholesterolaemia.

P67

CASE REPORT: MANAGEMENT OF A NEONATE WITH BRACHIAL ARTERY THROMBOSIS¹Samy Allawendy*, ¹Cormac Duff, ¹Emmanuel Osakwe, ¹Dara Gallagher, ²Patricia Eadie, ²Saima Aslam. ¹SUH, Sligo, Ireland; ²OLCHC, Dublin, Ireland

10.1136/archdischild-2019-epa.422

Introduction Neonatal spontaneous arterial thromboembolism is a rare phenomenon with a high risk of morbidity and mortality. Currently, there is little information regarding common risk factors, diagnostic strategies, therapeutic interventions, and outcomes of this condition.

Case A 37+5 weeks female infant born at Sligo University Hospital presented at the time of birth with a right upper extremity consistent with ischemia with evidence of gangrene. Following stabilisation of the infant in NICU, immediate transfer to a tertiary Paediatric Hospital was requested. There was uncertainty as to which speciality would accept the baby and multiple contacts with Neonatology, General Surgery and Plastic Surgery were necessary. There was significant difficulty procuring a bed and transport. The referring hospital transferred the baby at 5 ½ hours of life to a tertiary hospital where the baby was diverted to the Emergency department and subsequently admitted under shared neonatal and surgical care. US Doppler examination identified no pulses distal to the elbow joint with small non occlusive thrombus in IVC. The limb was conservatively managed for two weeks at which point below elbow amputation of the right upper limb was done. Cranial ultrasound and hypercoagulable workup were normal and no identifiable cause for the thrombus has been found. The baby remains well with full early intervention support.

Discussion Neonates with a clinically significant arterial thrombosis that develops in utero or during delivery are often born with or quickly develop gangrenous changes, thus illustrating the importance of prompt intervention. However, management guidelines for neonatal arterial thrombosis have not been well established. When an arterial thrombosis occurs perinatally, it is difficult to determine the duration of occlusion, and limb ischemia and loss may be imminent. The neonate in this case was born with gangrenous changes in the right arm due to an extensive thrombus and was subsequently managed with below elbow amputation with rehabilitation. The lack of a clear pathway of referral for neonates with thrombosis is evident in this case.

Conclusion A clear pathway of referral for neonates with arterial thrombosis should be established nationally and should include priority bed allocation and emergency transport.

P68

WHEN A VIRUS HAS DIFFERENT FACES

¹Sofia Siena, ¹Rosa Canestrone, ¹Rossella Giorgio, ²Carmela De Meco, ²Irene Rutigliano, ²Pasquale Pio Maccarone, ¹Francesca Lotti, ¹Enrica Manca*, ¹Agostino Petrarco, ¹Michaela Sciacovelli, ¹Jessica Cinalski, ¹Isabella Patisso, ¹Annarita Pedico, ¹Loredana Dipasquale, ²Michele Carmine Sacco, ²Massimo Pettoello Mantovani. ¹Residency program in Pediatrics, University of Foggia, Foggia, Italy; ²Department of Pediatrics, Pediatric Unit, 'Casa Sollievo della Sofferenza' Scientific Institute, University of Foggia, San Giovanni Rotondo, Italy

10.1136/archdischild-2019-epa.423

Measles is a highly contagious viral illness, characterized by fever, malaise, cough, coryza and conjunctivitis, followed by maculopapular exanthema, which spreads cephalocaudally and centrifugally. Measles, mumps, rubella (MMR) vaccination has led to the interruption of measles virus transmission and gives protection to unvaccinated individuals via herd immunity. The morbilliform exanthema can be found in various conditions, including infectious mononucleosis. It is characterized by fever, pharyngitis, lymphadenopathy and a generalized maculopapular, urticarial or petechial rash occasionally can be present, especially after administration of beta-lactams. **CASE REPORT:** 17 months old male was admitted in our pediatric department for the appearance, 4 days earlier, of rash and fever (T 38,8°C). The exanthema consisted of an

erythematous, maculopapular, blanching rash, which began on the face and progressed to the trunk and extremities involving the palms and soles. In some areas it showed confluent and hemorrhagic features. The physical examination showed the presence of laterocervical lymphadenopathy, nonpurulent conjunctivitis and pharyngitis. About 10 days earlier it was administered antibiotic therapy with Amoxicillin for a fever associated with malaise, cough and coryza. The child had no history of allergies and the MMR vaccine was repeatedly delayed and eventually not carried out for multiple episodes of respiratory infections. The laboratory tests showed leucocytosis with a normal differential count, mild elevation of transaminases, elevation of inflammatory markers and LDH; the morphological evaluation of the peripheral smear showed some activated lymphomonocytoid cells. Given the rash characteristics and the strong suspicion of measles, the patient was located in isolation and infectiological tests were performed (TORCH, Monotest, Respiratory Multiplex PCR panel and a serology for measles). They all came back negative except for the anti VCA IgM for EBV infection. The patient was treated with IV fluids and antipyretics. Antibiotic therapy was administered in order to prevent bacterial superinfections. After 72 hours the rash started to darken and then to gradually fade. The patient was dismissed with the diagnosis of maculopapular exanthema in mononucleosis infection. Clinical manifestations of infectious mononucleosis can be similar to those of measles and, especially in unvaccinated patients, can sometimes be confused with it. Maculopapular exanthema can be found in various conditions, such as common viral or bacterial infections, IgA vasculitis, Kawasaki disease or drug eruption. For this reason, it is important to consider mononucleosis in the differential diagnosis of measles, especially in case of hemorrhagic and infiltrated rash, not much described in the literature.

P69

TAKAYASU ARTERITIS PRESENTING IN AN 11-YEAR-OLD BOY

¹Ana Louise Hawke*, ²Danielle O'Connor, ²Emma Jane MacDermott, ²Orla G Killeen. ¹Our Lady's Children's Hospital, Crumlin, Dublin, Ireland; ²Our Lady's Children's Hospital, Dublin, Ireland

10.1136/archdischild-2019-epa.424

Introduction We present a case of Takayasu arteritis in a Caucasian Irish 11 year old male.

Background Takayasu Arteritis, which usually involves the Aorta and it's main branches is an idiopathic granulomatous vasculitis of unknown aetiology with significant associated morbidity and mortality.

It is rare in children, usually presents in Asian adult females and has a worldwide incidence of 2.6/1000,000.

Case Presentation A previously well Caucasian eleven year old boy, with no past medical history of note, presented with an insidious onset of persistent, intermittent abdominal, flank and back pain, waking him from sleep and exacerbated by exercise.

Histologically confirmed appendicitis with appendectomy was made 4 weeks prior to presentation to his local hospital.

Examination was normal, including pulses and 4 limb Blood Pressure measurements. Inflammatory markers on presentation- CRP(90) and ESR(120)- were markedly raised. Full blood count, renal and liver profiles were normal.