P648 ANONYCHIA CONGENITA: A RARE CONGENITAL ANOMALY

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Background To describe the case of a new born baby with absent nails on fingers.

Case report A term female baby was born to 27 years old G3P3 mother of Asian(Chinese)origin via spontaneous vaginal delivery with normal antenatal scans.During routine newborn examination it was found that the nails are absent on the middle and ring finger of both hands with shortening of distal phalanx of right ring finger.Rest of the newborn examination was within normal limits.Findings of fingers were consistent with Anonychia Congenita.There is a strong family history of similar findings in father,uncle and grandfather affecting the fingers of hands and feet.

Discussion Anonychia congenita is a rare congenital condition with unknown prevalence which manifest as absence of some or all nails.Inheritance is autosomal dominant.Mutations in R-spodin 4(RSPO4) gene which is located on short arm of chromosome 20 cause anonychia congenita.It can occur as isolated anomaly or as a part of syndromes that affect multiple parts of body like Coffin -Siris syndrome and nail-patella syndrome.

P649 UNCOMMON COMPLICATION AFTER CONGENITAL DIAPHRAGMATIC HERNIA REPAIR IN A NEWBORN

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Introduction Bowel intussusception is an extremely rare postoperative complication of congenital diaphragmatic hernia. Any delayed diagnosis can be life-threatening. Clinician has to evoke the diagnosis every time symptoms of bowel obstruction appear after surgical repair.

Methods We report the case of a newborn who presented an acute intussusception at the fourth post operative day of a congenital diaphragmatic hernia repair.

Results A female full term newborn was operated for a congenital diaphragmatic hernia at her second day of life. Four days after surgery she started vomiting bile-stained fluid. Abdominal ultrasound showed an ileoileal intussusception with the classic target sign. Laparotomy evidenced a jejunojejunal intussusception associated to multiple intestinal adhesences. The intussusception was manually reduced and the intestinal adhesions were released. The postoperative course was uneventful. She is now 2 years-old and she is healthy.

Conclusions In our case, intestinal intussusception would be secondary to postoperative intestinal adhesions and peristalsis disorders during the phase of its reactivation in the first post-operative days.

P651 TRANSIENT ACUTE CEREBELLITIS WITH HEPATITIS A VIRUS INFECTION IN A 3 YEARS OLD CHILD ON NITROFURANTOIN PROPHYLAXIS

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Background Hepatitis A Virus (HAV) Infection is prevalent worldwide. HAV is well known to cause extra hepatic involvement by autoimmune mechanisms. This case highlights the importance of how two autoimmune inciting factors working together could lead to transient cerebellitis, which has not been reported in association with either of them singly.

Case report A three year old female child presented with gait imbalance and jaundice for last three days. She had visited her GP two weeks previously with complaints of mild fever, vomiting and diarrhoea which had resolved with oral rehydration in a week time. She had also complained of mild upper abdominal pain in last 2 days. She had suffered 2 episodes of urinary tract infection in recent past and her GP had commenced her on nitrofurantoin prophylaxis since last 3 months. DMSA scan of the kidney was due in a month time. Past history was not relevant and had no sick contacts in recent past.

Examination revealed jaundice and tender right upper quadrant of the abdomen. Neurological examination revealed wide based mildly ataxic gait with past pointing of left hand on cerebellar examination. Rest of the neurological and other systemic examination were within normal limits.

Blood tests revealed normal full blood counts, electrolytes and inflammatory markers. Liver function tests (LFT) was deranged with total bilirubin 52 micromole/L, conjugated bilirubin 20micromol/L, GGT 36 IU/L, ALP 140 IU/L, ALT 74, AST 60 and Albumin 38. Prothrombin time and INR were within normal limits. Ultrasound of the abdomen revealed mildly echogenic liver suggestive of acute inflammatory changes with slightly oedematous gall bladder wall. MRI of the brain revealed small hyper intense signal in the right cerebellar hemisphere. Blood tests for viral serology were positive for anti HAV IgM antibody. Diagnosis of hepatitis A virus infection with cerebellitis was made based on all these results.

Subsequently nitrofurantoin was stopped and she was treated conservatively. LFT was repeated weekly which normalized completely within next three weeks and her ataxic gait improved gradually.

Conclusion Nitrofurantoin has been linked in past with autoimmune organ injury. Transient cerebellitis has well been associated with several infections and autoimmune processes. Neither Nitrofurantoin nor HAV in isolation has been reported to cause cerebellitis. Barring the possibility of chance association, this case adds to the medical literature, the association of cerebellitis with HAV infection in isolation or in combination with concomitant autoimmune triggering factors such as medication.