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

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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

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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.

CT Angiogram showed upper periaortic soft tissue surrounding the origin of the coeliac axis and superior mesenteric artery (SMA) causing marked stenosis of the SMA.

A working diagnosis of an inflammatory or infective Aortitis involving the proximal intra-abdominal Aorta, extending from the diaphragm to the renal vessels was made.

An extensive infectious work up was performed which proved negative including exclusion of tuberculosis.

C3 and IgA were marginally raised. Serum Anti Nuclear Antibodies, Anti-Double Stranded DNA Antibodies, Rheumatoid factor and urinary catecholamines were all within normal limits.

PET scan confirmed uptake in the proximal abdominal aorta with associated periaortic soft tissue suggestive of Aortitis.

A diagnosis of Takayasu arteritis- large vessel granulomatous vasculitis, was made.

Treatment was instigated with high dose intravenous methylprednisolone for three days followed by high dose oral prednisolone and subcutaneous methotrexate at a dose of 15 mg/m² weekly.

Inflammatory markers slowly began to normalise with immunosuppressive treatment. Follow up ultrasound at one month showed interval improvement in the aortic mass with increase in the aortic lumen size. On corticosteroid wean a further ultrasound 6 weeks later showed no improvement in the mass and was associated with a rise in inflammatory markers. Biologic therapy with adalimumab subcutaneously has since been added with plan for serial imaging to assess response to therapy.

Conclusion This is a rare presentation of a large vessel vasculitis- Takayasu arteritis in a male child of Caucasian origin.

P70

TWIST AND SHOUT: DELAYED PRESENTATION OF TESTICULAR TORSION IN A 14 YEAR OLD

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10.1136/archdischild-2019-epa.425

Introduction Testicular torsion is an acute surgical emergency, with an approximate 6 hour window from onset of symptoms, to irreversible damage to the testicle.¹ Trauma associated testicular torsion is a well recognised complication of blunt scrotal trauma, accounting for 4–8% of all torsions.^{2,3} Delays in recognition of the condition, and subsequent delayed surgical exploration, may necessitate orchidectomy, and risks impairment of future fertility.⁴ Patient factors associated with delay in presentation include embarrassment around pain in the genitals, and lack of awareness of the condition.⁵

Case description A 14 year old Irish boy, with a previous history of bilateral retractile testes, presented to the Emergency department with a three day history of severe left sided scrotal pain and swelling following a traumatic injury to the perineum. The patient was cycling off-road three days prior to presentation, and suffered a cross-bar related injury to the scrotum and perineum. He felt immediate left testicular pain, however believed this to be related to bruising of his testicle. The pain progressively worsened over the following three days, and presented to hospital once the pain had become unbearable.

Examination demonstrated a tender, erythematous, and swollen left hemiscrotum with an absent cremasteric reflex on the left side. The right testicle was retractile, but present within the right hemiscrotum, with a positive cremasteric reflex. US of the scrotum demonstrated an enlarged, oedematous left testicle, with absent internal vasculature, and a normal appearing right testicle. At operation, the left testicle was found to be necrotic and non-viable, and so we proceeded with a left orchidectomy. After discussion with his parents, we decided to proceed with a right sided orchidopexy with three-point fixation of his remaining testicle. The patient was discharged home later that day on a course of oral antibiotics.

Discussion Time is of utmost importance in cases of testicular torsion, with early presentation, and early surgical exploration the most important factors in testicular salvage.³ It is easy to attribute post-traumatic testicular pain and swelling to the initial insult, however, a high index of suspicion is required to out-rule trauma associated testicular torsion. Early education around testicular health, an understanding that scrotal pain requires swift medical evaluation, and that delays in evaluation may impair future fertility, are important messages to convey to paediatric populations, and should be included in sexual health education for males.

P71

SKELETAL DYSPLASIA- CASE REPORT OF AN INFANT WITH THANATOPHORIC DYSPLASIA

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10.1136/archdischild-2019-epa.426

Background An infant with respiratory failure and features of skeletal dysplasia

Case report A male infant dichorionic twin weighing 2.3 Kg with antenatally suspected skeletal dysplasia had initial examination findings of frontal bossing, a flat nasal bridge, a wide anterior fontanelle and shortened upper and lower limbs along with a markedly narrow chest and short ribs. His systemic exam revealed no other abdominal or cardiovascular abnormalities. His twin had normal growth parameters and phenotypic appearance.

He was intubated within a few minutes of life and commenced on high frequency oscillatory ventilation. A skeletal survey confirmed his thorax to be narrow with shortened ribs and handlebar clavicles. He also had vertebral abnormalities with platyspondyly and U-shaped vertebra. His limbs showed generalised micromelia with short bowed femurs and humeri. He had no pneumothoraces.

The infant displayed progressive respiratory failure despite maximum oxygen concentration at 50 hours of life and interventions were discontinued due to futility. His clinical and radiological findings were most in keeping with Thanatophoric dysplasia I and this was confirmed genetically on microarray showing heterozygous FGFR3 mutation.

Discussion Thanatophoric dysplasia is the most frequent form of lethal osteochondrodysplasia with an estimated incidence of 1 in 60,000 births.¹ There are two subtypes: Type ¹ is characterised by a short, curved femur, and a straighter femur with cloverleaf skull characterises type II.² Affected infants show marked underdevelopment of the skeleton and short limbs due to sporadic mutations of fibroblast growth factor receptor 3 gene (FGFR3).³ Infants are usually stillborn or die shortly