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153 EFFICACY AND SAFETY OF ACYCLOVIR AS PREVENTION OF VARICELLA DISSEMINATION IN IMMUNOCOMPROMISED CHILDREN: AN EVIDENCE-BASED CASE REPORT

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Background Varicella infection is very common in children and easily transmitted. In immunocompetent, varicella infection is usually mild and self-limiting. However, in immunocompromised, varicella infection has the potential to disseminate and cause complications, one of which is pneumonia, due to impaired cellular immunity that the morbidity and mortality is much higher. Acyclovir is an antivirus that is effective for varicella in immunocompetent children.
Objectives Evaluate evidence exists to date regarding the efficacy and safety of acyclovir in reducing morbidity (disease severity, duration of illness and dissemination) and mortality of immunocompromised children with varicella.
Methods Literature searching was conducted on PubMed, Cochrane and MEDLINE with the keywords of *'acyclovir'*, *'varicella'*, *'immunocompromised'* and *'children'*. After filtering articles based on inclusion and exclusion criteria without time limitation, we found 3 relevant articles. One was excluded since it didn't apply blinding and thus result to the final 2 randomized clinical trials that were critically appraised based on the validity, importance and applicability criteria of Oxford Center for Evidence-Based Medicine (2011).
Results In terms of efficacy, 12 (48%) out of 25 placebo recipients were withdrawn from the double-blind randomized treatment to be treated by open intravenous acyclovir due to their worsening condition, only 1 out of 25 (4%) who were treated by intravenous acyclovir and only 2 out of 25 (8%) who were treated by oral acyclovir were similarly withdrawn ($p<0.001$). Consequently, intravenous and oral acyclovir both significantly reduce varicella dissemination and mortality. The use of intravenous acyclovir also significantly accelerates crusts formation ($p<0.001$), reduces the duration of varicella infection. In terms of safety, there were increase in blood urea nitrogen levels without any clinical manifestation and acute diarrhea without dehydration which was mild and self-limiting.
Conclusions All immunocompromised children who develop varicella should be considered for early treatment with either intravenous or oral acyclovir since both are safe and significantly prevent worsening condition of the patient due to varicella dissemination. Intravenous acyclovir also reduces the duration of infection. All patients need to be monitored closely by the physician, especially the one who receives oral therapy, that any whose condition does not show improvement should be considered for intravenous route.

156 USE OF ABDOMINAL X-RAY IN CHILDREN WITH CONSTIPATION PRESENTING TO ED IN A DISTRICT GENERAL HOSPITAL: A REVIEW OF OUR PRACTICE

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Background Constipation is common in childhood with UK prevalence ranging between 5–30%. Consequently, inpatient data statistics showed that 79% of children with constipation are admitted through emergency admission. Functional constipation is a clinical diagnosis using history and clinical examination. The national institute for health and care excellence (NICE) and our local trust guidelines clearly state that abdominal x-rays (AXR) should not be used in the diagnosis of constipation unless by specialist services.
Objectives To assess the number of children with constipation presenting to the Paediatric Emergency department(PED) who had Abdominal X-ray(AXR) against recommended guidelines, time spent by these patients in paediatric emergency department, patient flow, the cost of having the AXR and the amount of radiation exposure.
Methods A retrospective review of electronic notes of patients under the age of 16 who had a diagnosis of constipation was conducted over a 3 month period from 1 September 2019 to 30 November 2019.
Results 67 cases were identified.

28% (19) of the 67 children had AXR. Only 1 of the 19 patients who had imaging was admitted. The rest were discharged with no change in management.

The average time spent in the Emergency Department for those who had AXR was 3.92 hours compared to 2.62 hours (*P Value 0.008*) in children who had no imaging.

The estimated avoidable cost for the AXR was £2000 over a 3 month period (£100/AXR) with a total avoidable radiation of 0.03–0.11 mSv/AXR.
Conclusions Abdominal X-rays are still performed in children presenting to our PED with constipation despite our guideline recommendations against doing so. Performing X-rays in these children led to longer time spent in the department, increased cost and unnecessary radiation exposure without influencing change in management plan or need for admission. We recommend adhering to the national and local guidelines and avoid AXR where necessary.

160 CASE REPORT: MENKES DISEASE, AN ILLUSTRATION OF DISEASE RELATED PROGRESSIVE SKULL FRACTURE IN A PRETERM NEONATE

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10.1136/bmjpo-2021-RCPCH.89

Background Menkes disease is an X-linked progressive neurodegenerative disorder caused by abnormalities of the ATP7a transporter that result in abnormal copper transportation in the mammalian nervous system. Menkes disease results in neurological symptoms and connective tissue abnormalities.

Objectives Through this case report we aim to present a series of cranial images illustrating the progression of a lytic bone lesion and subsequent infiltration of tortuous arterial vasculature over the course of one year in a preterm neonate with Menkes disease.

Methods Retrospective case review of the management a patient with Menkes disease and a skull fracture over one year.

Results Our patient was born in good condition at 29+5 weeks gestation via forceps assisted vaginal delivery. There was a family history of maternal carriage of Menkes disease with a second degree relative having had the condition which was the reason for testing in this patient. For treatment of his hypocupremia the patient received intramuscular copper histidine injections, however in the first few weeks of his life serum levels were below the normal range.

Cranial ultrasound scans within the first 7 days of life demonstrated a left sided grade IV intraventricular haemorrhage. He was noted to have widely splayed sutures with head circumference on the 25th centile. Neurological examination throughout his neonatal admission was normal.

Over time, he developed a persistent boggy swelling over the left parietal region, an ultrasound probe placed over this region revealed an area of absent parietal bone with visualisation of underlying brain parenchyma. A skull x-ray demonstrated a left parietal skull fracture which was further demonstrated on CT scan. This confirmed a mid-left parietal fracture which extended posteriorly from the left coronal suture with a protruding soft tissue component.

Interestingly, the patient had previously had a cranial MRI performed as part of his diagnosis for Menkes disease. Retrospectively a protrusion of subarachnoid space in the left parietal region was seen which was not appreciated initially. He was transferred to a tertiary hospital for neurosurgical review and subsequently managed conservatively.

Over the course of a year there were neurological and respiratory deterioration secondary to his Menkes disease despite copper histidine injections, necessitating admission to paediatric intensive care. MRI scans performed during this time found an evolution of abnormalities in the left parietal area with diffuse arterial tortuosity and a left parietal pseudomeningocele. The patient passed away during his intensive care admission aged under 1 year.

Conclusions This case illustrated the evolution of a lytic parietal bone lesion with the progression of arterial tortuosity. Recent literature has described cases studies of neonatal skull fracture as a known presentation of Menkes disease. However, with this case study we demonstrate the changes of both a skull lytic lesion in combination with arterial tortuosity as a series of images over time from a single patient. We hope through this case to increase knowledge and awareness in recognising these changes in the context of a patient with Menkes disease.

165 PROLIDASE DEFICIENCY IN AN INFANT WITH AN INCIDENTAL FINDING OF METHAEMOGLOBINAEMIA

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10.1136/bmjpo-2021-RCPCH.90

Background A 4-week-old infant presented to hospital with diarrhea and vomiting. An initial diagnosis of cow's milk allergy was made and he was discharged with extensively hydrolysed formula. However the infant was readmitted within a week with ongoing symptoms and associated metabolic acidosis. He was suspected to have sepsis and treated with intravenous antibiotics. However, he deteriorated further with worsening metabolic acidosis despite treatment. Methaemoglobinaemia was then identified and he was treated with methylene blue and transferred to HDU for further management.

Objectives

1. Consider Methaemoglobinaemia as a differential diagnosis in infants with unexplained metabolic acidosis.
2. Raise awareness on Prolidase deficiency – a rare genetic condition affecting 1 in 1 million of worldwide population.

Methods Literature search on Ovid using Medline/Embase was undertaken.

Results

Conclusions Further investigations results of Imidopeptiduria and rapid exome sequencing confirmed diagnosis of Prolidase deficiency (PD) in this infant. PD is a rare autosomal recessive genetic condition caused by mutations in PEPD gene, which codes for Prolidase, an enzyme involved in the final stage of the degradation of collagen and other proline containing proteins including dietary proteins. PD affects approximately 1 in 1 million worldwide. The symptoms of PD include dysmorphic features, skin lesions, recurrent infections, hepatosplenomegaly and intellectual disability.

In this case, the child had symptoms of diarrhoea, likely secondary to PD. As the diarrhoea was persistent, he then

Abstract 165 Table 1

Blood gas pre-methylene blue	Blood gas post-methylene blue
pH 7.18	pH 7.30
pCO2 3.9	pCO2 4.0
HCO3 10.8	HCO3 19
Lactate 5.7	Lactate 2.0
MetHb >30%	MetHb 7%

Abstract 165 Table 2

Full Blood Count(FBC) (on admission)	Hb 120 g/l, WCC 18.6 × 10^9/l, Neutrophils 11.69 × 10^9/l, Platelets 544 × 10^9/l
FBC (on day of deterioration)	Hb 125 g/l, WCC 40.7 × 10^9/l, Neutrophils 39 × 10^9/l, Platelets 653 × 10^9/l
CRP	31 mg/l (on admission), increased to 75
Urea & Electrolytes	Na 133 mmol/l, K 4.1 mmol/l, Urea 0.9 mmol/l, Creatinine 17 umol/l
Blood culture	No growth after 5 days
Ammonia	36 umol/l
Plasma amino acid	Normal level of alanine, tyrosine, phenylalanine, lysine, ornithine
Cerebrospinal fluid	Culture -no growth, WCC 23, RBC <3, negative meningococcal/pneumococcal PCR, negative virology
Urine culture	No growth
Urine amino acids	Imidopeptiduria
Rapid trio exome sequencing	Homozygous pathogenic PEPD variant {c.978G>A, p.(Trp326*)}.