P220 ABSTRACT WITHDRAWN

P221 SAFETY AND EFFICACY OF PROCEDURAL SEDATION AND ANALGESIA IN PEDIATRIC ONCOLOGICAL PATIENTS

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Background and Aim Procedural sedation and analgesia is the standard of care for painful procedures in children that require immobility. Children with cancer are subjected to many procedures for their treatment which are painful and cause anxiety in them. Our aim was to assess the safety and efficacy of procedural sedation and analgesia in pediatric oncological patients in a large tertiary care hospital in Karachi.

Methods A retrospective study was done and records were reviewed of children receiving PSA (procedural sedation and analgesia) for pediatric oncological procedures. This included patients for oncology procedures (lumbar puncture, intrathecal chemotherapy and/or bone marrow aspiration $\pm$ trephine). PSA was provided by non-anesthesiologists. These patients were assessed according to PSA protocol guidelines by American Society of Anesthesiology (ASA). Low dose Ketamine (0.5 mg/kg) and Propofol (2 mg/kg) were used.

Results A total of 1216 oncological procedures were performed out of which lumbar puncture was the commonest procedure performed (n=956; 78.6%) followed by bone marrow aspirate only (n=137, 11.3%) and both (n=123, 10.1%). A total of 565 children were enrolled in the study out of which majority (65.1%) were males and 34.9% were females (Table 1). Only eight (0.7%) of the patients were found to have hypoxia as an adverse effect of propofol-ketamine drug with 50% procedures utilizing propofol 1 mg/kg for sedation.

Conclusion This study concludes that the combination of Ketamine and Propofol is safe for procedures. There were no major complications. None of the patients required CPR or endotracheal intubation.

P222 ANALYSIS OF MEDICAL HELP ORGANISATION FOR INTERSEX-PEOPLE IN RUSSIAN FEDERATION. THE STUDY WAS FORMED BY THE LEADING CLINICIANS AND DOCTORS OF THE ST. PETERSBURG STATE PEDIATRIC MEDICAL UNIVERSITY

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Backgrounds and aims Intersex-people is one of the most stigmatized social group around the world. On the example of androgen insensitivity syndrome/variation we have analyzed medical help organization in Russian Federation. Androgen insensitivity syndrome/variation is a genetically determined anomaly/variation of sex development manifested by prenatal formation typical female phenotype with

XY genotype, because of changes in the structures of androgen receptors. There are no clear clinical standards in majority countries. We used clinical cases by different Federal Districts: Saint-Petersburg, Moscow, Ural Federal District, Central Federal District, The Republic of Bashkortostan.

Results we've taken anamnesis and studied medical documentation of seven intersex-people with androgen insensitivity syndrome/variation. In 100% cases first specialist, who diagnosed AIS/V was gynecologist. The most popular complaints were amenorrhea, delayed sexual development (all cases) and inguinal hernia (2 of 10 cases). Then intersex-patients were diagnosed by ultrasonography, karyotyping, without sequencing and determination the level of the sex hormones FSH and LH (all cases); diagnostic laparoscopy was performed in 2 cases. Every intersex-patient was performed with gonadectomy, and after that was appointed by estrogen replacement therapy. Only 2 of 7 intersex-patients sure that their gonads were diagnosed with histologic studying. According to the words, none of the intersex-patients have been correctly and fully informed about negative reactions and changes in their organism after gonadectomy or about the risks of rejection of the operation. None of them was informed about alternatives of treatments. Nobody was consulting with psychotherapist about critical states, gender choices and hormone replacement therapy or appearance choices. But 4 intersex-people of 7 decided to consult by themselves. So, only 2 intersex-patients of 7 use estrogen replacement therapy, but other rejected because of psychological, material or other reasons. None of intersex-patient has recommendations of consulting with other specialists in future, and because of negative past experience 50% avoid contacts with medical community. Only 3 of 7 intersex-patients control their coagulogram and 3 of 7 had fractures after rejection of hormone replacement therapy.

Conclusion we haven't found clear recommendations and clinical criteria about diagnostics, ways of treatments and social support for patients with androgen insensitivity syndrome/variation, and therefore the quality of care for intersex patients does not match for modern requirements and social challenges.

P223 TO BE OR NOT TO BE? – VARICELLA ZOSTER!

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Background A rash can create a difficult diagnostic dilemma for the General Paediatrician.

Aim/Method Our aim is to describe the clinical presentation with clinical photos, diagnostic pathway and result of microbiological investigations in a school age child who presented to our Paediatric Emergency department (PED) with an unusual rash ultimately proven to be Varicella Zoster despite vaccination.

Results A previously well 11 years old boy presented to the PED with a rash for four days and a one day history of pyrexia, coryza and cough. The rash started behind the ears and

then spread to the entire body including limbs. It was itchy. The family are originally from Quebec and had been residing in Ireland for the past 2 years in relation to the father's occupation. Contact with infectious disease and recent travel were denied. He had never been hospitalised and had no medical diagnoses. His mother was adamant that all vaccinations were up to date and included the Varicella vaccine at one year of age in Canada. He had no known allergies and was not taking any medications.

On examination he had multiple vesicular and pustular lesions with an erythematous base. Some lesion had necrotic centres. There were some vesicles on his lower lip, buccal mucosa and also on the throat.

His WCC was 2.08, Neutrophils 0.73, Lymphocytes 0.73, CRP 20. Influenza, RSV and Monospot were negative.

He was initially treated with IV Augmentin and Flucloxacillin for a presumed diagnosis of Impetigo. More lesions appeared over his trunk and abdomen over the subsequent 24 hours although he was not systemically unwell. The Dermatologist made a clinical diagnosis of 'Chicken Pox'. He was discharged home on an immunocompetent dose of oral acyclovir pending results of skin swab, throat swabs and Varicella titre.

At follow up one week later he was clinically well with multiple healing lesions. His Varicella titres were high confirming a diagnosis of Varicella Zoster infection. His mother brought with her his vaccination records from Canada which showed that he had received only one dose of the Varicella vaccine and had missed the booster. This is in contrast to his siblings who were fully vaccinated and did not develop Varicella despite close contact.

Conclusion Common conditions are common. Vaccines can fail. Parents should be encouraged to keep detailed records of all vaccinations including boosters and to follow through with booster vaccines when indicated.

P224 THE HAZARDS OF AN IRISH HEATWAVE; ECTHYMA

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Background and aims Ecthyma is a pyogenic, deep skin infection caused by Group A Streptococcus usually combined with Staphylococcus aureus and sometimes pseudomonas in the context of warm, moist weather and overcrowding, most commonly seen in extremes of age and immunocompromised patients resulting in healing in a few weeks with scarring.

Our aim is to report a case of this very rare dermatological condition in a three year old boy.

Methods The clinical presentation, examination findings with clinical photographs, laboratory investigations, natural history, treatment and outcome are described.

A review of the current available literature was undertaken.

Results A previously well and neurodevelopmentally normal three year old boy presented to the Paediatric Emergency Department (PED) during the summer months with a rash for 6 days and high grade fever for 2 days. The rash started as vesicular lesions on the abdomen before spreading to the

limbs. On examination, lesions were 1.5x1 cm in size, pustular with hard crust of dried exudate and erythematous base mostly on abdomen, genitals and lower limbs. Systemic examination was normal. His full blood count was normal. CRP was raised. After review by the Dermatologist a clinical diagnosis of ecthyma was made. Skin swab grew Staphylococcus Aureus sensitive to Flucloxacillin with which he was treated intravenously for 5 days. He was discharged home on oral erythromycin for 10 days, fucidin cream for one week, hydromel baths and paraffin gel for 3 weeks. At Follow up 2 weeks later he was found to be well with healing of residual lesions.

Conclusion Our case raises awareness of this extremely rare dermatological condition in a Paediatric and an Irish context. To our knowledge it is the first case of its kind ever seen by either the Paediatric or Dermatology services in our institution but may not be the last if our good summers continue

P225 CONGENITAL HYALINE FIBROMATOSIS SYNDROME: A CASE REPORT OF HETEROZYGOUS ANTXR2 AND LITERATURE REVIEW

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Introduction Juvenile hyaline fibromatosis is a rare hereditary disease characterized by deposits of a clear substance (hyaline) in the skin and other body tissues, it becomes apparent at birth or in infancy presenting with severe pain with movement, skin lesions & bumps, gingival hyperplasia, progressive joint contractures, and bone lesions.¹ Complications can be life threatening. The inheritance follows autosomal recessive pattern, Causing mutations in the ANTXR2 gene. Treatment is supportive and aims to alleviate pain and other symptoms of the condition. The prevalence is unknown; about 84 people with this disorder have been reported in 2018.²

Case We report a 23 month old boy, born at 38 weeks gestation, no neonatal complications. He was referred to paediatric clinic at 3 month old with excessive crying, feeding difficulty and failure to thrive crossing down on centiles from 9th to <0.4th centile. At 8 months noted to have limited movement of both shoulders. He then started to have gingivitis and gum swelling which progressed to gingival hyperplasia. Developmentally he sat unsupported at 8 months, not crawling or standing no concerns about hearing or vision, plays and interacts well with siblings.

Parents are non consanguineous of Syrian origin, He has two sisters and one brother all well. They had no skin lesions or joint problems. There was no history of similarly affected relatives.

Physical examination He appears symmetrically small, generally bright visually alert to his surroundings. Visible nodules over left ear and left nostril, gum hypertrophy. Decreased muscle bulk in his deltoids with limited passive movement of both shoulders and extreme pain on any shoulder movement. Cannot stand or walk independently.

Laboratory investigations Biochemical and metabolic work up was all normal DNA PCR: Heterozygous ANTXR2 gene detected.