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A case report of cardiac echinococcosis in a child with disseminated hydatid cystic disease

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Background: A 3-year 4-month-old child presented with a one-month history of non-productive cough, post-tussive vomiting and fever. Test for IgG Echinococcus was positive. CXR showed multiple cystic lesions and chest CT showed multiple hydatid cysts in the lungs, liver and pericardium. The child was transferred to Red Cross War Memorial Children's Hospital and managed medically for 5 weeks whilst awaiting surgery.

Echocardiography: The TTE showed an hydatid cyst measuring 2 x 3 cm in the anterior myocardium of the LV with no other abnormalities.

Perioperative management: Intravenous midazolam and fentanyl were followed by gas induction and pressure-controlled ventilation in the operating theatre. Invasive lines were placed before surgery. The major anaesthetic concerns were: high alert for an anaphylactic reaction due to rupture of cyst during intra-operative period; maintaining sinus rhythm, adequate cardiac output and perfusion pressure during the perioperative period. Surgery was via a median sternotomy. The patient was supported on CPB and a core temperature of 27°C was reached. The myocardium around the cyst at the base of the left ventricle was dissected and the ruptured/complicated cyst was removed under circulatory arrest. The patient came off bypass uneventfully on Adrenaline 0.04 mcg/kg/min which was stopped 3 hours later in PICU. The child was discharged from PICU the following day. TTE was done on day 3 and at 6 weeks post-surgery.

Conclusion: Cardiac echinococcosis is rare, comprising 0.01 – 2% of all registered cases. Hydatid cyst of the left ventricle is usually localised sub-epicardially and rarely ruptures into the pericardial space.

A retrospective study to evaluate the anaesthetic choices and complications for patients with osteogenesis imperfecta at a quaternary referral hospital

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Background and aim: Osteogenesis imperfecta (OI) is an inherited genetic syndrome affecting connective tissue. Patients

often undergo surgery due to an increased susceptibility to bone fractures. Anaesthesia is associated with many perioperative challenges. The study aimed to describe and evaluate the perioperative management of OI paediatric patients presenting for surgery at Inkosi Albert Luthuli Central Hospital.

Methods: A retrospective chart review of children under 18 years who had OI and underwent surgical procedures from 2000 to 2017 at a quaternary referral hospital was conducted. Patients were identified from the electronic hospital database. The following variables were extracted: demographic data, preoperative history, examination, investigations, chronic medications, intraoperative management, postoperative plan and perioperative complications. Simple descriptive statistics were performed using Excel.

Results: 38 patients who underwent 93 surgeries were included. The majority (72%) had severe type III OI and had elective orthopaedic surgery. Anaemia was identified in 64.5% of patients and 40% had a spinal deformity and respiratory abnormality. A supraglottic airway device (SGAD) was used in 91% of patients, with only three airway complications. 87% of cases had combined general (GA) and regional anaesthesia (RA). No children had documented signs suggestive of hypermetabolism or malignant hyperthermia.

Conclusions: Despite the majority of patients in our sample having severe OI, few of the complications and difficulties described in the literature were identified. Combined GA and RA technique with a SGAD was shown to be a safe anaesthesia technique. Improved perioperative investigation, especially full blood count, due to the high incidence of anaemia, should be encouraged to improve overall care.

Anaesthetists' knowledge of South African law pertaining to informed consent

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Background: Anaesthetists require knowledge of informed consent laws to mitigate the risk of litigation. The knowledge of South African anaesthetists regarding informed consent law is unknown.

Methods: Participants from an academic anaesthetic department anonymously completed a researcher-developed questionnaire with 23 questions. Factors affecting questionnaire performance were recorded: years after graduation from medical school (YGMS); professional designation (PD); years of anaesthetic

experience (YAE); and attendance at formal postgraduate training on informed consent (APGT).

Results: A subgroup analysis of data pertaining to the paediatric population (less than 18 years old) from 167 participants was conducted. The mean questionnaire score (SD) achieved was 60.08% (12.61%). Questions assessing The Children's Act No 38 of 2005 achieved the lowest mean score percentage overall, with a score of 51.82 ± 17.84 . Knowledge of the Children's Act correlated positively with PD ($p=0.0004$), YAE ($p=0.0180$) and APTG ($p=0.0080$). Mean questionnaire score improved with APTG ($p=0.0161$) and higher PD ($p=0.0163$). Of the 23 questions, question 7.1 was the best answered question, with 152 (91%) anaesthetists answering it correctly. It dealt with consent for medical and surgical treatment of a child. Question 6.1 was the most poorly answered question, with 32 (19%) anaesthetists answering it correctly. It dealt with gaining consent for blood transfusion for a child Jehovah's Witness.

Conclusions: Anaesthetists have suboptimal knowledge of informed consent laws. Anaesthetists should attend training and postgraduate education institutions should run these courses regularly. Comparative studies should be conducted in other anaesthetic academic departments countrywide and should include surgical staff.

Better safe than sorry; emergent intubation of a child with hereditary angioedema

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Background: Hereditary angioedema (HAE) is a rare autosomal dominant condition caused by a deficiency in the C1-esterase inhibitor. It has a reported incidence of 1:50 000 and it is estimated that there are approximately 800 affected individuals in South Africa. Acute attacks are potentially life-threatening if they involve the airway; in a case series of 60 patients in South Africa published in 2017, the incidence of airway involvement was 44.2% with a mortality rate of 4.7%.

Methods: We report on patient LN, who is known with HAE and is being managed by the Department of Allergology at the Red Cross War Memorial Children's Hospital. This was the patient's second acute attack: it involved the face and upper airways. On presentation to the paediatric emergency department, he was noted to have marked facial swelling but no evidence of airway obstruction.

Results: After administration of a bradykinin inhibitor called Icatibant, urticaria was noted at the injection site and the patient complained of difficulty breathing. It was decided to perform an emergent intubation in theatre and to ventilate him in ICU until the swelling subsided. The patient underwent a Sevoflurane gas induction and was kept breathing spontaneously. No significant

airway oedema was noted on videolaryngoscopy and the patient was intubated uneventfully with a nasal endotracheal tube.

Conclusion: While HAE is a rare condition, it is one of the few potential causes of acute upper airway obstruction and anaesthetists should be aware of the risks involved and have a management plan thereof.

Case report: Anaesthetic management of a hypotonic, syndromic infant with pulmonary hypertension

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Background: A 16-month-old female patient with dysmorphic features, hypotonia (cause unknown) and delayed milestones had been admitted for pneumonia. She was found to have upper airway obstruction secondary to adenoid hypertrophy with pulmonary hypertension, cor pulmonale and a patent PDA: she needed an adenotonsillectomy.

Methods: A TIVA technique was used, intubation assisted with videolaryngoscopy.

Results: The anaesthetic was complicated by a pulmonary hypertensive crisis following a difficult intubation, which responded to treatment. The patient was extubated to high-flow nasal cannula after the procedure and discharged 3 days later, clinically much improved.

Conclusion: The undiagnosed hypotonic, syndromic child with significant pulmonary hypertension presents multiple anaesthetic concerns and considerations with a high risk for complications if not well managed.

Case report: Bilateral erector spinae catheter placement for bilateral nephrectomy in a paediatric patient

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Background and aims: The erector spinae plane (ESP) block is a novel regional anaesthetic technique with reported use in thoracic, abdominal and hip surgery. Increasing use is also reported in the paediatric surgical population. Placement of indwelling catheters in the fascial plane can provide prolonged analgesia and may be useful in supplementing postoperative analgesia.

Methods: We report a case with the use of continuous, bilateral ESP blocks with catheters for analgesia in an infant undergoing resection of bilateral Wilm's tumours in a resource-constrained environment and provide a review of the literature on the use of ESP blocks in children.

Results: ESP blocks are proposed to have a safer approach and reduced side effect profile as compared to other regional and neuraxial anaesthetic techniques.

Conclusions: The ESP block allows for the provision of good analgesia and opioid-sparing effects.

Case report: Management of the difficult airway in an infant

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Background: This case study details a 5-month-old baby with temporomandibular joint ankylosis and a unilateral hypomandible, requiring release of the pterygomandibular ankylosis. Airway manipulation and management of paediatric temporomandibular joint ankylosis has previously been fully described in the literature. What makes this case so unique is the age and size of the child, as well as unusual other anatomical abnormalities such as a significant fat pad around the neck and sinus below the chin.

Methods: The case was expedited by the surgeons as there was a concern regarding obstruction of the airway by the growing tongue. Due to the baby's extremely difficult airway, the only possible radiographic investigation was a CT scan and sonar done with gentle restraint. It was not possible to sedate this child for an MRI. The CT showed a severely abnormal airway with no epiglottis and abnormal vocal cords situated on the right lateral wall of the pharynx. A fat pad was situated over the anterior part of the neck extending from the chin to the sternal notch. The sinus did not appear to communicate with the interior of the mouth.

Results: As previously described in the literature, the airway was managed with a nasopharyngeal airway and flexible bronchoscopic intubation. The ENT surgeons were on standby for a surgical airway. Postoperatively the child was ventilated for 72 hours in ICU and brought back to theatre, via MRI, to be extubated.

Conclusion: A multidisciplinary approach for such a case is imperative.

Case report: Managing an unusual case of a threatened spinal cord in a child

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Background: A 20-month-old boy with Conradi-Hünermann syndrome presented for fusion of his cervical spine. He presented with asymptomatic complete atlantoaxial dislocation and dislocation of the dens tip with accompanying spinal

cord stenosis and compromise. We performed an intravenous induction using propofol and rocuronium. Manual in-line stabilisation (MILS) of the c-spine was performed for intubation with the use of a videolaryngoscope (CMAC) with a D-blade. Surgeons placed Mayfield clamps on the patient whilst supine and maintained MILS during proning. The clamp was secured to the table in a neutral position. No spinal cord monitoring was used so isoflurane was used for maintenance of anaesthesia with multi-modal analgesia. Postoperatively the patient was extubated and admitted to the intensive care unit (ICU) where he had an uneventful stay.

Discussion: Conradi-Hünermann syndrome is a rare x-linked genetic disorder characterised by skeletal malformations, skin abnormalities, cataracts and short stature. It is a form of chondrodysplasia punctata which is characterised by the formation of small, hardened spots of calcium on the epiphyses of the long bones or inside other areas of cartilage in the body. Atlantoaxial instability presents some unique challenges to the anaesthetist. In children, cervical-spine instability is caused by ligamentous laxity, hypoplasia of the odontoid or inflammatory diseases.

Conclusion: Careful positioning of the patient and maintaining c-spine immobility during airway manipulation and positioning for surgery, particularly prevention of flexion, is essential to prevent further neurological fallout. These cases are especially dangerous in the asymptomatic child.

Effect of pre-warming on paediatric patients presenting for magnetic resonance imaging under general anaesthesia

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Background and aims: The combination of anaesthesia and environmental factors in the magnetic resonance imaging (MRI) suite increases patient susceptibility to hypothermia. This study aimed to determine whether pre-warming paediatric patients presenting for MRI under general anaesthetic prevented them from developing hypothermia.

Methods: A prospective, quasi-experimental research design was followed. A total of 102 patients were enrolled during the 4-month study period and were divided equally between pre-warmed and control groups. Inclusion criteria were patients aged 6 months to 6 years, ASA physical status I or II and with written informed consent from a caregiver. The pre-warmed group received 30 minutes of active warming with a forced-air warmer. The MRI examination was conducted using a volatile-based general anaesthetic. Tympanic temperatures were measured at baseline, pre-scan and post-scan.

Results: The pre-warmed group received an average (SD) of 34.8 (4.3) minutes of warming. The median (IQR) time spent inside the MRI unit was 60 (50 to 77) minutes. The incidence of hypothermia was 1.96%. There was a statistically significant change in CBT (core body temperature) between the pre-warmed and control groups (0.15°C; $p = 0.0001$). A weak negative correlation between patient weight and temperature change ($r = -0.220$, $p = 0.026$) and between length of time of pre-warming and temperature change ($r = -0.268$, $p = 0.006$) was found.

Conclusions: The effect of pre-warming to prevent hypothermia under general anaesthetic in the MRI unit could not be established due to the low incidence of hypothermia. Pre-warming patients for 30 minutes prevented a statistically significant decline in CBT, however, this finding was not clinically relevant.

Extracorporeal membrane oxygenation (ECMO) as a bridging therapy after ALCAPA repair: Red Cross War Memorial Children's Hospital's first success story

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Background: Anomalous left coronary artery (LCA) originating from the pulmonary artery (ALCAPA syndrome), also known as Bland-White-Garland syndrome, is a rare form of acyanotic congenital heart disease. It affects 1:300 000 live births and has a 90% mortality in the first year if left untreated. Patients usually present with left ventricular dilatation and failure, secondary to chronic ischaemia. Mitral incompetence is a common occurrence. Symptomatology and age of presentation depends on the degree of collateral flow between the right and left coronaries and stenosis of the proximal LCA.

Methods: This poster will discuss one such case seen at the Red Cross War Memorial Children's Hospital (RCWMCH) in September 2019. A 3-month-old female presented with ALCAPA syndrome, with mitral incompetence and left ventricular failure.

Results: Surgical repair was uneventful, with adequate coronary flow afterwards. However, after 3 unsuccessful attempts to wean from cardiopulmonary bypass (CPB), the patient was placed on venoarterial ECMO as a bridging therapy. After 3 days the patient was decannulated with good left ventricular function on adrenaline and milrinone infusions. Ventilatory parameters were acceptable.

Discussion: ALCAPA syndrome is a rare disease. At RCWMCH 4 ALCAPA cases were seen and operated on since January 2018. This was the second time ECMO was utilised at RCWMCH (a first for ALCAPA repair) and the first ECMO case with a favourable outcome. Future cases will benefit from the experience gained by all staff involved with this life-saving therapy.

Panendoscopy and anaesthesia for debulking of juvenile onset recurrent respiratory papillomata (RRP) using target control infusion (TCI) and the Manujet III for high-pressure source ventilation (HPSV)

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Background: Laryngeal papillomata causes upper airway obstruction. Airway papilloma results from exposure to the human papillomavirus (HPV) at birth (HPV Types 6; 11), a history of genital warts and other immunological risk factors. Presenting features include respiratory difficulties and voice changes and diagnosis is often delayed as illnesses like asthma, croup and foreign body aspiration are excluded. Diagnosis requires laryngoscopy or fibre optic bronchoscopy. Debulking of airway papillomata has unique challenges and timely intervention is lifesaving. Issues relate to a shared airway, unimpeded surgical access, adequate depth of anaesthesia, maintaining ventilation and overall cardiovascular stability. Available best practice guidelines for HPSV ensures safe practice but hypoxia, laryngospasm and other complications may occur. Papillomata regresses around puberty, but dysplasia occurs in 3%. Adjuvant therapies (acyclovir, monoclonal antibodies) to increase the time between repeat surgical procedures is being evaluated. HPV vaccination with good sexual health education will reduce disease prevalence, as will the delivery practice of mothers with active genital warts. Parental education remains important due to the risk of complete airway obstruction.

Case study: A child of 6 years with known RRP was scheduled for elective surgery. Monitoring included (NIBP), (ECG) and (SaO₂). End-tidal (ETCO₂) is not possible in the open system used and transcutaneous CO₂ monitoring is not done. Blood gas measurement for arterial PH and PCO₂ is mandatory as hypercarbia and a respiratory acidosis can occur. Induction began with sevoflurane and 100% oxygen. Manual assistance is given, and chest movements observed to ensure an uncompromised airway. TCI Propofol (Kataria model) (Ce) (2-4) µg/ml was started and titrated. Muscle relaxation (mivacron 0.1 mg/kg) and a short-acting opiate (Rapifen, 10-20 µg/kg) given. Ketamine (0.5 mg/kg) is added for its analgesic and sedation benefits. The Manujet III is connected to a side arm of the surgeon's laryngoscope. Jet-ventilation is supraglottic and intermittent and pressure varies from 0.5-2.5 Barr for children. Chest movements are observed to ensure oxygenation. Ventilation is halted when the laser used to minimise fire hazards, but saturation remains above 90%. Decasone (0.1mg/kg) to reduce surgical swelling and peralgan (15 mg/kg) given for pain relief. Post-surgery patients given 4-hourly nebulisation with adrenalin and pulmicort. Ward monitoring occurs over 24 hours.