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REVIEW ARTICLE

CASE OF CHILD WITH CATEL MENZKE SYNDROME POSTED FOR ORTHOPAEDIC SURGERY - ANAESTHETIC CHALLENGES AND PERIOPERATIVE MANAGEMENT

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ABSTRACT

Catel Menzke syndrome is a rare congenital bone disorder characterized by clinodactyly and bilateral hyperphalangy of the Index fingerwith Pierre Robin sequence (glossoptosis, micrognathia, and cleft palate). Frequently reported with valvular heart defects like ventricular septal defect (VSD), and Patent ductus arteriosus (PDA).[1] This case report reviews major anaesthetic challenges and successful perioperative management of a rare syndromic pediatric patient undergoing orthopaedic surgery.

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INTRODUCTION

Case of a 10-year-old child diagnosed case of Catel Menzke syndrome presented with Pierre robin sequence along with previously repaired VSD/PDA, cleft palate, and failure to thrive (15th percentile) on growth hormone therapy posted for Right knee arthroscopy and ACL reconstruction.

CASE REPORT

A 10-year-old child weighing 28kgs (15th percentile) and 149 cms in height (85th percentile)born out of a non-consanguineous marriage, delivered vaginally with uneventful birth history. Post-natally diagnosed with Catel Menzke syndrome was posted for arthroscopic right anterior cruciate ligament tear repair. He had history of growth failure and was on Somatotropin and Gn RH A for the same. He was operated for VSD patch closure and PDA closure at the age of 2 weeks and cleft palate repair at the age of 1 year. The patient had an alleged history of falla week ago which led to his knee injury and a massive ligament tear requiring arthroscopy and reconstruction. Airway examination revealed micrognathia along with retrognathia, a high arched palate and bifid uvula. Neck movements were normal. Mallampatti grade 2. All blood investigations were within normal limits, ECG was suggestive of RBBB (right bundle branch block), 2D Echo showed prolapse of atrial tricuspid valve leaflet with moderate tricuspid regurgitation (TR) Grade (II/III), Trivial aortic regurgitation, No residual shunt across the VSD patch with good LV function. Pediatric Endocrinologist and Cardiologist were involved for peri-operative management, who recommended Infective Endocarditis (IE) prophylaxis-administration of antibiotics 1 hour before and 6 hours after the surgery and withholding of rGH (recombinant growth hormone) injection 48 hours before the planned surgery. Parents were counselled. Written informed consent was obtained. 22G IV cannula was insitu. Adequately starved, child was taken into prewarmed OT. Inj Midazolam 0.5 mg was given in a controlled environment. Once inside the OT, all standard ASA monitors were attached. Preoxygenation was done through nasal prongs and simultaneously Sevoflurane-Oxygen was started (Fig 1).

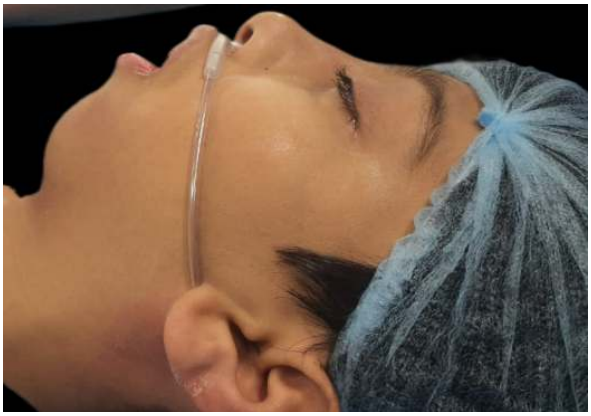

Fig. 1. Micrognathia and Retrognathia Pre-oxygention through nasal prongs.

Fig. 2. Post LMA insertion

Once sedated, anaesthesia further deepened with Sevoflurane-Oxygen through the pediatric ventilator circuit and small aliquots of IV propofol and fentanyl. Supraglottic airway device- LMA Supreme 2.5 size was inserted (Fig 2), carefully avoiding damage to the previously repaired Cleft palate. (LMA chosen over the endotracheal tube to surpass laryngoscopy response hence avoiding major hemodynamic fluctuations). Deeper plane of anaesthesia was maintained to achieve a MAC of 1.Intra-operative fluid was administered using the Holiday Segar formula and hours of NBM (nil by mouth).Once adequately anesthetised, antacid and IE prophylaxis weregiven. Intra-operative MAP of >70mmHg was maintained for adequate cerebral and tissue perfusion. Adequate measures were taken to maintain normothermia. The duration of the surgery was approximately 2 hours, our main goal was to maintain stable hemodynamics and depth of anaesthesia at all times. For postoperative analgesia, ultrasound-guided Femoral nerve block was given with 10ml of 0.2%Ropivacaine. At the end of the surgery, after the return of spontaneous respiration LMA was removed where the patient was hemodynamically stable and pain-free, subsequently shifted to the ward.

DISCUSSION

For the uneventful management and better outcome of the case, a multidisciplinary team should have good coordination during the perioperative period. A multidisciplinary team approach and in-depth knowledge about the syndrome and pharmacology of anaesthetic drugs are very important for the successful management of rare syndromic pediatric patients posted for orthopaedic surgery.

Catel Manzke syndrome has an autosomal recessive inheritance. The main feature being- bilateral hyperphalangy of the index fingerin which there is an accessory ossification centre at the MCP joint, resulting in radial deviation of the index finger. The digital abnormality is associated with the Pierre Robin sequence which combines micrognathia, glossoptosis, and cleft palate. Additional frequently reported congenital malformations include cardiac defects such as septal defects in ventricles (VSD) and interatrial communication like patent ductus arteriosus. Other features can be present but our patient didn't exhibit can be-Iris coloboma, mild facial dysmorphism like- hypertelorism, short palpebral fissures, full cheeks, low-set or posteriorly rotated ears, pectus excavatum, carinatum, scoliosis, bilateral brachydactyly, bilateral fifth finger clinodactyly, knee dislocation, talipes, short halluces, failure to thrive and an intellectual disability.[(2)]

Ehmke *et al*. recently implicated that **TGDS** (dTDP-D-glucose 4,6-dehydratase) homozygous or complex heterozygous as pathogenic variants - the causative factor in a series of seven unrelated patients with features of Catel–Manzke syndrome.[(3)]
A detailed review of all organs is vital during the pre-operative period along with a completely general and systemic examination before planning the mode of anaesthesia keeping in mind the pediatric age group of the patient. After reviewing all the above-mentioned systems, keeping in mind the associated congenital malformation- the decision to administer General anaesthesia along with ultrasound-guided femoral nerve block was taken for more patient comfort, better hemodynamic control and postoperative analgesia.

Conclusion

Specific studies related to this rare syndrome perioperative management are not conducted. Also, there are lacunae in management guidelines as well. Additional specific research should be encouraged with an aim to come up with proper guidelines for the management of CatelManzke syndrome posted for surgery.

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