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Case Report

Anaesthetic Management in a Child with Cutis Laxa for Bilateral Ureteric Reimplantation

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ABSTRACT

Cutis laxa is a rare congenital multisystem connective tissue disorder. Patients with cutis laxa can present with distinctive facial features, pulmonary emphysema and right-sided heart failure. Anaesthetic management is important because of difficult airway and respiratory/cardiac abnormalities. We have reported the preoperative evaluation and anaesthetic management of a child with Cutis laxa with right cleft lip and palate.

Cutis laxa is a rare condition characterized by excessive loose pendulous skin, associated to premature aging expression [1].
It may be associated with genitourinary and gastrointestinal diverticula, diaphragmatic hernia and emphysema leading to cor pulmonale, congenital dislocation of the hip, cranio stenosis, interventricular communication, cardiovascular anomaly like Peripheral pulmonary stenosis (most common -90%), coarctation of aorta and aortic aneurysm [2].
Main anaesthesia concerns are cardiac / respiratory system involvement, difficult airway, difficult IV access and neuroaxial block, hyperthermia, problems with coagulation and increased risk for peripheral nerve injury with intraoperative positioning. The literature is scarce about the response to various anaesthetic agents in such patients and to best of our knowledge this would be the first case report on cutis laxa patient undergoing long duration surgery.

Case Report

A four years old male boy weighing 12 kilograms diagnosed to be autosomal recessive cutis laxa with bilateral hydronephrosis and bilateral grade 5 vesicoureteric reflex had presented to our hospital for bilateral ureteric reimplantation. On the pre-operative anaesthesia visit, he was well oriented in time, space and person. He had senile looking face (as shown in Figure 1), loose and thin skin, right cleft lip and cleft palate, plagiocephaly, pectus excavatum with global development delay. He was afebrile and pink without respiratory distress and edema and ascites. This patient is the first child in his family and with healthy parents born from second degree consanguineous marriage. In the past he was operated for right inguinal hernia under GA (LMA) + caudal which was uneventful.

On general examination vitals were normal, Pectus excavatum (Figure 2) seen. Airway examination revealed a Mallampati Class III, and normal range of movements at atlanto-occipital joint with right cleft lip and palate with crowded teeth. Blood hemogram, coagulation profile, blood sugar and renal function tests were normal preoperatively. DMSA (Dimercaptosuccinic acid) scan showed left kidney to be only 5 % functioning and right being 95%. Chest x ray showed mild hyperinflation of both the lungs and normal shadow of heart. ECG and ECHO was normal.

OT was prepared in anticipation for malignant hyperthermia (since they are prone for hyperthermia). IV access (22G canula) was secured in wards. GA+ epidural anesthesia was planned. Anaesthesia concerns for our

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case were Airway (right cleft palate and lip), Renal (poor functioning of one kidney can lead to delayed recovery), Respiratory (Pectus excavatum) and others (like difficult IV access, malignant hyperthermia, peripheral nerve injury).

Child was premeditated with IV midazolam (0.5mg). Routine monitors were attached. General anaesthesia was initiated after adequate preoxygenation with IV thiopentone (60mg), IV fentanyl (25mcg). After establishing successful bag mask ventilation, IV atracurium(6mg) with IV propofol (20mg) was given. Cormack-Lehane score 1 (Figure 3) were assessed with direct laryngoscopy (macintosh blade, size 2), microcuffed endotracheal tube number 4.5mm internal diameter was safely placed into the trachea without trouble.

Another 22 G intravenous cannula was passed on the left dorsum of hand. A nasopharyngeal probe was inserted for Temperature monitoring.

Lumbar Epidural - was given at L1-L2 using 19 G cath, threaded upward, bolus of 5ml of 0.125% levobupivacaine given. Anaesthesia was maintained with air, O2, IV morphine (0.3mg) and TIVA (IV dexmedetomedine infusion+ propofol boluses). IV fluids was calculated and given appropriately. The procedure took about 5 hrs.

At the end of procedure, after adequate reversal and suctioning, patient was extubated and then he was sent to the recovery. During the procedure he remained haemodynamically very stable. The intra and postoperative course was uneventful. Consent for his case to be reported was received from the parents.

Figure 1

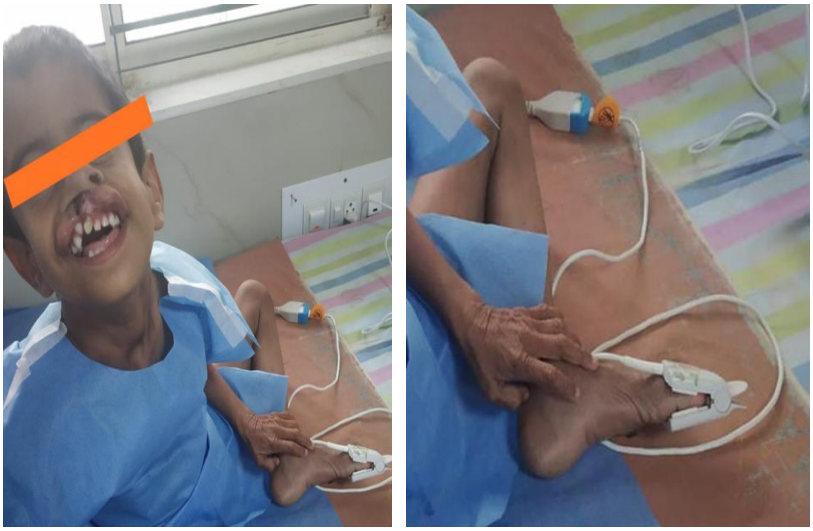


Figure 2

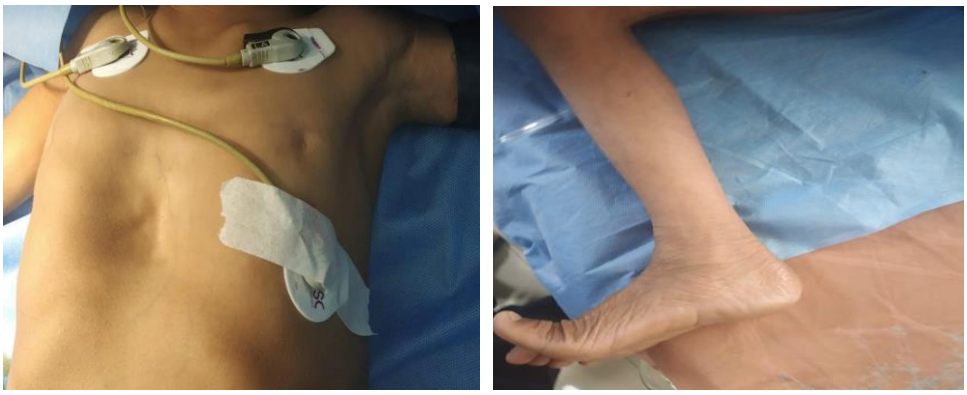
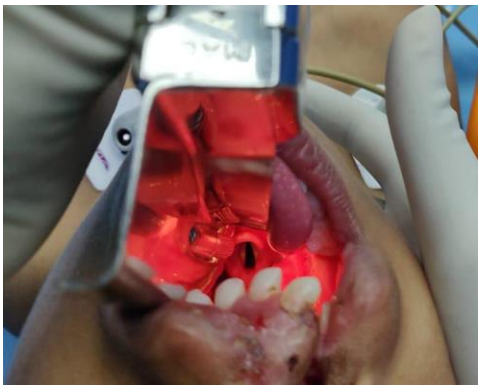


Figure 3



Discussion

Cutis laxa is a rare disorder of the connective tissue and are distinguished as congenital (autosomal dominant/ recessive) or acquired, localized or generalized [3].

In a case report, a 21-month-old baby with Cutis laxa who underwent B/L orchidopexy, had severe wheeze and respiratory distress after extubation. Immediately the trachea was reintubated and drugs were given for bronchospasm [4].

Also an another case report of a baby with Cutis laxa who underwent herniorrhaphy with pulmonary stenosis under general anesthesia with PLMA was reported [5].

W.M Hajjar et al reported a very rare and near fatal form of autosomal dominant cutis laxa in a 15 year old boy with acute bilateral pneumothorax treated with bilateral staged thoracoscopic bullectomy and pleurodesis [6].

De Barsy syndrome (autosomal recessive cutis laxa type 3) is a rare clinical syndrome characterized by cutis laxa, ophthalmic opacification, skeletal malformation, mental and growth retardation [7].

Warner et al conducted a case series on de Barsy syndrome and encountered episodes of intraoperative hyperthermia, difficult airway and difficult intravenous access (thick loose sagging skin) [8].

Differential diagnosis of cutis laxa includes Costello syndrome and Ehler Danlos syndrome type III C [9].

Although there is no evidence of malignant hyperthermia, intraoperative hyperthermia associated with tachycardia has been reported [10]. It is possible that these cases represent a form of non- malignant hyperthermia similar to that manifested in patients with other congenital diseases such as Costello syndrome and Osteogenesis imperfecta. [11-12]. Hence, close temperature monitoring is recommended intraoperatively.

The potential benefits of epidural anaesthesia included: decreased requirement of inhalational agents, improved respiratory mechanics, avoidance of parenteral narcotics, and adequate postoperative analgesia.

Conclusion

Cutis laxa is an extremely rare genetic disorder and one should be aware of associated cardio respiratory anomalies along with potential challenges with airway management, vascular access, and temperature monitoring.

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