Case Report

A Child with Crouzon's Syndrome and Ventricular Septal Defect Posted for Ventriculoperitoneal Shunt Surgery

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Abstract

Quite often in our anaesthesia practice, we encounter syndromic children presenting to us for surgery either for correction of the primary deformity or for a secondary problem. A child with a craniofacial syndrome presents with a number of issues for the anaesthetist, with difficult airway being one of the most important one. The challenge increases with increasing complexity of the surgery. When pre-anaesthetic evaluation of such a child is done, other congenital anomalies also might come into picture. These may include various types of congenital malformations or congenital heart diseases which may be diagnosed incidentally while evaluation. Anaesthetic management of such a child presenting for surgery includes a thorough pre-anaesthetic evaluation, formulating a plan for intraoperative anaesthetic management and proper postoperative care. This case report describes a child with Crouzon's syndrome posted for ventriculoperitoneal shunt surgery.

Keywords: Congenital heart disease, crouzonal syndrome, hydrocephalus, ventriculoperitoneal shunt

Introduction

Crouzon's syndrome (CS) is a rare autosomal dominant disorder which presents with marked craniofacial abnormalities from birth.[1] Such patients may present to the hospital for reconstructive surgery of the craniofacial bones or for surgical procedures for unrelated issues.[2] A child with CS presenting with a co-existing congenital heart disease poses a double challenge for the anaesthetist. These may include management of difficult airway, hypothermia, difficult extubation, blood loss if the surgery is extensive and anaesthetic concerns related to the management of an acyanotic heart disease.[3,4] Here, we report a case of a child with CS who had a ventricular septal defect and was posted for a ventriculoperitoneal shunt surgery.

Case Report

A 1 year and 7-month-old female child (7 kg) presented to the paediatrics department in our hospital with the complaint of progressively increasing size of the head since birth. She was evaluated by the attending paediatrician and a diagnosis of CS was made based on the clinical features. These included enlarged and abnormally shaped head (craniosynostosis), hyperteleorism, proptosis and prognathism [Figure 1]. The child was posted for a ventriculoperitoneal shunt surgery.

The child was born of a full-term caesarean section and was the third in order. Her birth weight was 1 kg with delayed crying. She was intubated and put on ventilator support. There was no history of cyanotic or apnea spells. History of poor weight gain was present. Patient had frequent episodes of cough and cold but was never hospitalized. The child had delayed milestones and incomplete immunization. There was a history of consanguineous marriage between the parents, and the previous two children had expired in the neonatal period.

The preoperative vitals were heart rate (HR) 150 per minute, blood pressure (BP) was 90/60 mmHg and room air saturation (SpO_2) was 92%. Systemic examination detected no obvious abnormalities. Blood investigations done revealed a haemoglobin of 10.41 g/dl, total leucocyte count of 4,400 cells per cubic mm. All other investigations were within normal

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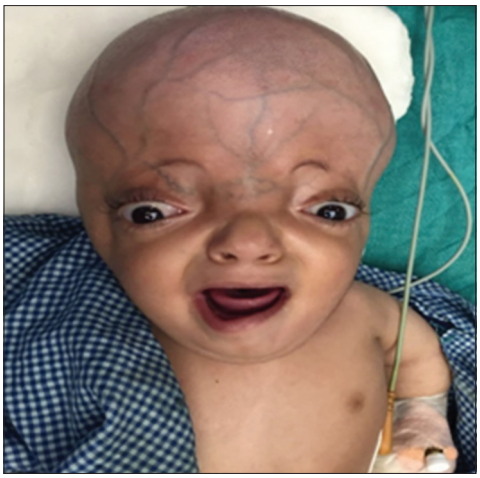

Figure 1: Child with craniosynostosis, hyperteleorism, proptosis and prognathism.

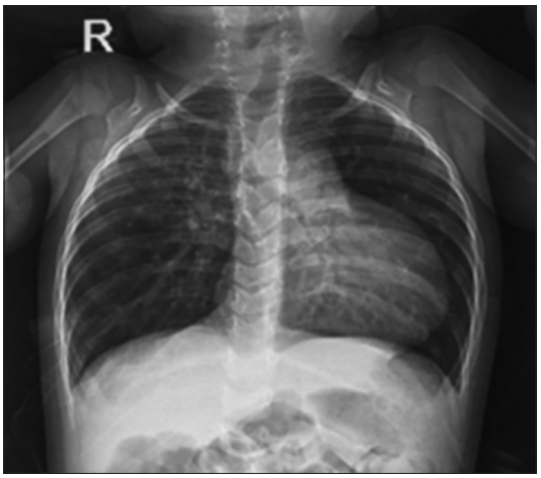

Figure 2: Chest X-ray of the patient showing cardiomegaly

limits. A chest X-ray revealed cardiomegaly [Figure 2]. The two-dimensional echocardiogram was reported as acyanotic congenital heart disease with a large non-restrictive 12 mm ventricular septal defect with left-to-right shunt, dilated main pulmonary artery with normal bi-ventricular function and an ejection fraction of 70%. A non-contrast computed tomography (CT) scan of the brain revealed a large obstructive hydrocephalus with dilated lateral and third ventricles [Figure 3].

The parents were explained the prognosis of the surgery and due risk consent was taken. Pre-operatively antibiotics regimen included intravenous (iv) injection cefotaxime 100 mg/kg in three divided doses and iv injection amikacin 15 mg/kg thrice a day. Difficult airway was kept ready. Monitors were attached and baseline vitals noted. SpO_2 was 92%. The child was induced with sevoflurane (starting from 8% and gradually decreased to 2%) and 100% oxygen. Intravenous access was established with a 24-G cannula. Fentanyl was given at a dose of 2 μg/kg. To facilitate smooth induction, lidocaine 1 mg/kg was given. Atracurium was used as a muscle relaxant and was given at a dose of 0.5 mg/kg body weight. After ensuring adequate ventilation and depth of anaesthesia, the child was intubated with a size 4 uncuffed endotracheal tube, and as there was an audible leak it was subsequently changed to 4.5 size uncuffed endotracheal tube. After ensuring adequate bilateral air entry, the tube was fixed and the child was connected to the ventilator. She was maintained on oxygen-air mixture (50:50), sevoflurane (1–3%), atracurium and intermittent positive pressure ventilation. Intra-operative period was uneventful. Iv injection paracetamol 20 mg/kg was given and fluid administration was done calculatedly. The child was kept warm throughout surgery.The surgery lasted an hour and the child was reversed and extubated after ensuring that she was fully awake. She was shifted to the intensive care unit (ICU) for observation and further management. She was shifted to the ward the next day and was discharged from the hospital after 3 days.

Discussion

Described by a French neurosurgeon Octave Crouzon in 1912, is characterised by craniofacial dysostosis, which includes a triad of skull deformities, facial anomalies and exophthalmos.[1,5] Other clinical features include hypertelorism, exophthalmos, strabismus, beaked nose, short upper lip, hypoplastic maxilla, and relative mandibular prognathism. Unlike some other forms of autosomal dominant craniosynostosis, no digital abnormalities are present. If such a child has a co-existing cardiac anomaly, the anaesthetic management becomes even more challenging. Among the congenital heart disease lesions, ventricular septal defects are the most common.[4] Moderate-to-large ventricular septal defects can cause delayed growth and development, decreased exercise tolerance, pulmonary infections and congestive heart failure during infancy.[6] Anaesthetic management in shunt lesions includes preventing hypoxia, hypercapnia, acidosis, hyperventilation, maintaining systemic vascular resistance, preventing increase in pulmonary vascular resistance, preventing air bubbles in the iv tubing, avoiding dehydration and ensuring an endocarditis prophylaxis before taking up the patient for surgery. This patient had received her regular dose of antibiotics before surgery. Infective endocarditis prophylaxis was not given to this child in accordance with the latest guidelines for infective endocarditis prophylaxis.[7] The most challenging aspect in a case of CS is airway management during any surgery performed under general anesthesia.[8] However, fortunately we did not encounter any problem during intubation and extubation. Due care was taken to maintain the haemodynamic stability intraoperatively and prevent all risk factors which could lead to worsening of the shunt. We chose an inhalational method of induction as it has been documented in cases of large ventricular septal defects.[7] For analgesia, we chose the intravenous route and not the rectal mode as the child had an imperforate anus. The child was extubated successfully and had an uneventful postoperative course.

Conclusion

A careful preoperative assessment, optimising the child for surgery, counselling the parents, taking a due risk consent

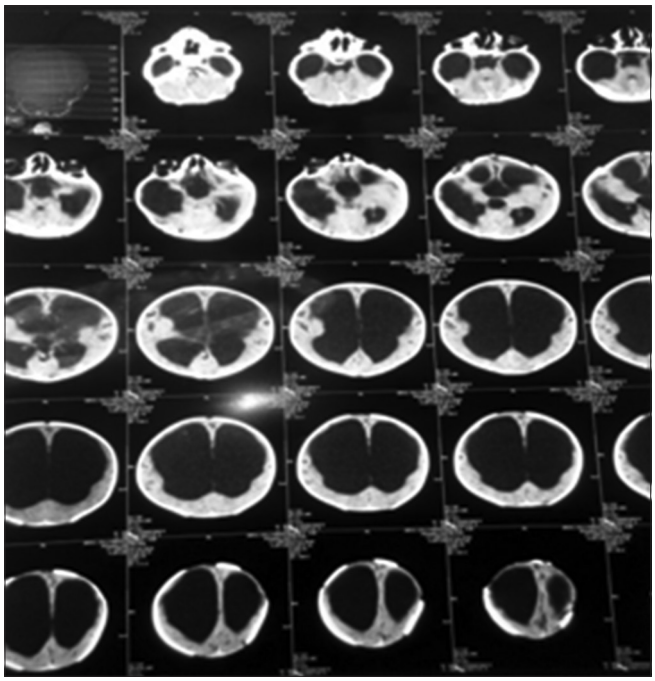

Figure 3: Preoperative CT scan of the patient showing ventriculomegaly

and a proper perioperative management is the basis for a good anaesthetic management of CS, thereby ensuring early recovery of the child and decreasing postoperative morbidity and mortality.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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