Anaesthetic management for cleft palate in a child with digeorge syndrome: Look before you leap!

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ABSTRACT

DiGeorge syndrome is a multisystem disorder associated with various cardiac anomalies, aplasia or hypoplasia of thymus and parathyroid glands, palatal defects, immune deficiency, and characteristic facial features. Here, we are reporting the case of a known DiGeorge syndrome posted for the repair of cleft palate in a 3-year-old female child. Anaesthetic management for any surgery in a child with DiGeorge syndrome is challenging for anaesthesiologist in terms of the associated complications. Hence, sound knowledge about DiGeorge syndrome is required for safe anaesthetic management.

Keywords: *DiGeorge syndrome, Hypocalcemia seizure, Immune deficiency, Palatal defects.*

DiGeorge syndrome is the most common microdeletion syndrome with a prevalence of one in 4000 live births [1-3]. It is associated with craniofacial anomalies, cardiac anomalies, parathyroid aplasia or hypoplasia, and immune dysfunction. About 2.4% of the individuals of DiGeorge syndrome have developmental disabilities [4] and approximately 10% to 15% have Tetralogy of Fallot (TOF) [5]. Severe hypocalcemia, difficult airway, hemodynamic instability and increased susceptibility for infection are some issues which need special intra and postoperative care in these patients. As the DiGeorge syndrome is not so common, case reports are required to provide knowledge regarding perioperative challenges in its management.

CASE REPORT

A 3-years-old female child was posted for cleft palate repair. She was a known case of DiGeorge syndrome. Birth history revealed that the child was a term baby delivered vaginally and born to non-consanguineous parents. She had a history of birth asphyxia for which she was put on ventilator support for 24 hours. At six months of age, she was hospitalised for breathlessness and bluish discoloration on crying and treated. She also had complaints of poor weight gain, feeding difficulty in the form of choking. Echocardiography done at that time reported a small patent ductus arteriosus (PDA) with ventricular septal defect (VSD) right to the left shunt, overriding of the aorta and the left pulmonary artery atresia with right ventricular hypertrophy. PDA stenting was done at that time. At the age of two and a half years, she got operated for correction of VSD. At that time, serum calcium was also low for which she received treatment and was diagnosed as a case of DiGeorge syndrome.

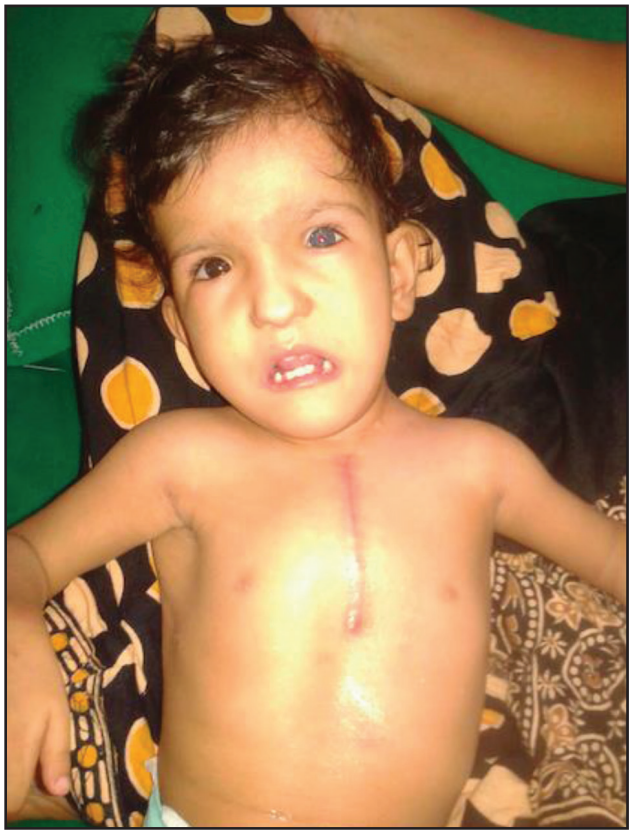

Figure1: Facial features of ocular hypertelorism, narrow palpebral fissures, low set ears, malar flatness, broad nasal tip and long face. Previous surgical thoracotomy scar mark over chest.

The child reported to our department for cleft palate repair. On examination, the child weighing 7 kg, had abnormal facies and a thoracotomy scar over chest [Fig. 1]. On systemic examination, the respiratory system was normal.

Preoperatively, the parathormone level was 10pg/ml (reference range 11-54pg/ml) and calcium level was 9.5 mg/dL (reference range: 9-11 mg/dL). The preoperative investigations like haemoglobin was 11.3gm/dL, prothrombine time 12 seconds, International Normalized Ratio (INR) 1.0, and total leucocyte count was $8.0x10^9$/L. 2D echocardiography revealed normal cardiacfunction.

In operation theatre, intravenous (IV) access was done with a 24G cannula and fentanyl 15 microgram and ondansetron 1 mg IV was given as premedication. Monitor for electrocardiogram, non-invasive blood pressures, and pulse-oximetry were attached. Anesthesia was induced with sevoflurane and nitrous oxide in oxygen. Endotracheal intubation was done with 3.5 mm reinforced tube which was facilitated by atracurium 3.5 mg IV. End-tidal carbon dioxide monitor was attached and right radial artery was cannulated for invasive blood pressure monitoring and arterial blood gas analysis.

Anesthesia was maintained with isoflurane (1%) in oxygen and air in 1:1 ratio and atracurium. Ventilation was done with pressure control mode to maintain tidal volume between 40 to 55 ml/kg and respiratory rate of 20/min and Fio2 of 0.5. Paracetamol 210mg suppository was inserted for postoperative analgesia. Surgery (palatal repair) lasted for about 60 minutes and there was about 30ml of blood loss. The surgery was uneventful, neurovascular blockade was reversed with injection neostigmine 0.5mg and glycopyrrolate 0.1mg IV. The patient was shifted to the recovery room after extubation and then to the ward after few hours. The patient was discharged after 5 days of hospital stay and was asymptomatic on subsequent follow-up after one month of hospital discharge.

DISCUSSION

DiGeorge syndrome is caused due to micro-deletion in chromosome 22q11. It is also known as CATCH22 syndrome. (C- Cardiac defect, A: Abnormal facies, T-Thymic hypoplasia, C-Cleft palate and H-Hypocalcaemia). This patient was a classic case of DiGeorge syndrome having all these features. DiGeorge described this syndrome in 1968 in a child with recurrent infections and hyperparathyroidism [6].

There are very few cases in the literature describing anesthetic management of DiGeorge syndrome [7,8,9]. These patients have an underdeveloped parathyroid gland which causes hypocalcemia. This can be aggravated by hyperventilation and the resultant alkalosis. The hypocalcemia may cause seizure which would be masked by the general anesthesia. Also, hypocalcemia causes hemodynamic instability. Though not required for our patient, rapid blood transfusion results in the reduction of serum calcium level [7]. So, perioperative monitoring of serum calcium should be made in every patient.

Craniofacial anomalies like micrognathia, retrognathia and cleft palate lead to difficult intubation. There is often difficulty in positioning the head for endotracheal intubation and obtaining internal jugular vein access due to tortuous carotid arteries. There are chances of tracheomalacia after extubation due to compression of the trachea by the vascular rings if present. Oxygen saturation and breathing should be monitored postoperatively as these patients may have retrognathia and velopharyngeal abnormalities [9].

Intravenous induction should be preferred in uncorrected TOF cases. As there is thymus aplasia or hypoplasia in 10% cases of DiGeorge syndrome [8], there is an increased risk of infection due to immunosuppression. So strict asepsis is technique is warranted. Also, irradiated blood product should be used to prevent transfusion-associated graft versus host disease.

CONCLUSION

Anaesthetic management for any surgery in a child with DiGeorge syndrome is challenging for anaesthesiologist in terms of cardiovascular complications, hypocalcemia with a risk of seizure, airway difficulty or postoperative complications like infection or hemodynamic instability. We should always be prepared to deal with for any kind of perioperative complications.

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