Anesthetic management in a Tessier cleft child with CHARGE syndrome: A new association?

Dear Editor,

A 1.5-year-old male 7 kg child diagnosed with CHARGE (coloboma, heart defects, choanal atresia, growth retardation, genital abnormalities, and ear abnormalities) syndrome and Tessier facial cleft with deficient lateral wall of orbit was posted for augmentation of the lateral orbital wall using rib cartilage graft with a cheek rotation flap. The child had an absent right eye, low set abnormal ears, developmental delay, and growth retardation [Figure 1]. Echocardiography showed a 6-mm ventricular septal defect with a left-right shunt.

An intravenous line was secured after sevoflurane-based inhalation induction and fentanyl 15 μg, ketamine 10 mg, and vecuronium 1 mg were administered intravenously. Mask ventilation was possible only with an oropharyngeal airway and two-hand mask ventilation. The Cormack–Lehane grading was IIb using a Macintosh laryngoscope and the child was intubated with a styletted uncuffed 4.5 mmID endotracheal tube in two attempts after external laryngeal manipulation. Controlled ventilation was initiated with a tidal volume (TV) of 60 mL and respiratory rate (RR) of 24/min. After 40 min, the oropharyngeal temperature of the patient started rising steadily from a baseline temperature of 35.5°C to a maximum of 37.5°C without any active heating measures. So, we removed the gamzee covering the child and initiated surface cooling. Simultaneously, end-tidal carbon dioxide ($EtCO_2$) started to rise and reached 72 mmHg. Despite hyperventilation (TV-12 mL/kg and RR-35/min), $EtCO_2$ was reduced to 52–56 mmHg. Tachycardia was seen and heart rate (HR) was persistently around 150–180/min. Anesthesia was deepened to 1.3 minimum alveolar concentration (MAC) and fentanyl 5 μg bolus was repeated twice. Arterial blood gas (ABG) showed mild acidosis (pH = 7.33, PCO_2 = 47 mmHg, bicarbonate = 19 meq/L, and lactate = 3.1 meq/L). Suspecting hypermetabolic response to inhalational agents, anesthesia machine was flushed out of inhalational agent and anesthesia circuit was replaced. Total intravenous anesthesia (TIVA) was initiated using propofol 125 μg/kg/h. Temperature and HR settled to 37.1°C and 130–140/min with these measures. Postextubation, HR was 90–100/min and the child remained stable in the high dependency unit. The parents did not give any history of

Figure 1: The child with CHARGE syndrome and Tessier's cleft

change in urine color suggestive of myoglobinuria. The creatine kinase (CK) enzyme level done the next morning was found to be elevated (470 IU/L).

Facial clefts are extremely rare congenital anomalies with an incidence of 1.43–4.85/100,000 births.[1] CHARGE syndrome (1/8,500–10,000 newborns) consists of coloboma of eye, heart disease, choanal atresia, growth retardation, and genital and ear abnormalities.[2] Upper airway abnormalities are detected in 56% of patients, which complicates airway management.[2,3] Mask ventilation could be difficult in these patients due to choanal atresia/stenosis and associated facial dysmorphic features. Choanal atresia was not present in the index case, but stenosis was not ruled out. There is a higher risk of adverse airway events perioperatively warranting close monitoring.[4] Presence of left-right shunting complicated the case due to the confounding effect of the direction of shunt on various parameters. The simultaneous rise of temperature, HR, and $EtCO_2$ suggested hypermetabolic etiology of the manifestations, which have never been reported in CHARGE syndrome. Chronically elevated baseline CK levels have been associated with MH susceptibility and may dictate the need for MH susceptibility evaluation.[5]

To conclude, the use of stringent monitoring and vigilance about the development of skeletal hypermetabolism and rhabdomyolysis is warranted in syndromic anesthetized children. TIVA may be preferred over inhalational anesthesia in these patients.

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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Conflicts of interest

There are no conflicts of interest.

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