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Paediatric Anaesthetic Management of a Child with Goldenhar Syndrome Associated With Persistent Sub Aortic Vsd Posted For Cleft Palate Repair

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Abstract: Goldenhar Syndrome (GS), a rare congenital disorder characterized by craniofacial anomalies, presents unique challenges in surgical and anesthetic management, particularly when accompanied by complex cardiac defects. Here, we present the case of a 5-year-old child with GS undergoing cleft palate repair surgery, complicated by Tetralogy of Fallot (TOF) and a large subaortic ventricular septal defect (VSD). Anesthetic management focused on maintaining hemodynamic stability and optimizing oxygenation, considering both the craniofacial abnormalities and the underlying cardiac pathology. The case underscores the importance of tailored perioperative strategies and interdisciplinary collaboration in achieving favorable outcomes for patients with GS and associated cardiac anomalies.

1. Introduction

Goldenhar Syndrome (GS), also known as oculo-auricular-vertebral dysplasia (OAVD), is a rare condition that affects approximately 1 in 3000-5000 newborns. The syndrome is characterized by malformations of the eyes and ears, which are derived from the first and second branchial arches (1). In some cases, it can also be associated with bone defects, heart malformations, and central nervous system anomalies (2) The cause of this syndrome is not yet known, though it may be related to an anomaly in the blood supply of the branchial arches during the seventh week of pregnancy or an anomaly in the migration of neural crest cells (3-5). We would like to present the case of a 5-year-old child who underwent cleft palate repair surgery for facial deformities. The child also had tetralogy of Fallot with a large subaortic VSD, which had previously been operated on.

2. Case Report

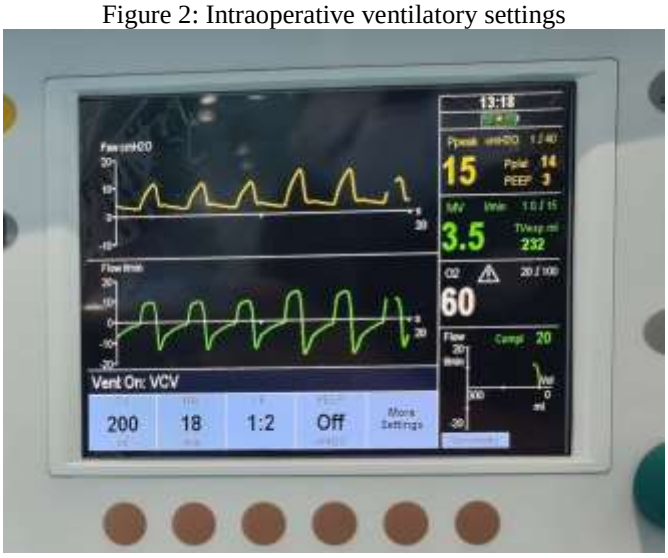
A 5-year-old boy of weight 20Kgs was referred to the hospital with Goldenhar Syndrome for cleft palate repair. On presentation, the patient had hemifacial microsomia. The patient had Tetralogy of Fallot with large sub-aortic VSD. TOF was corrected with persistent VSD. Echocardiography was done which showed an ejection fraction of 64% with corrected TOF with a large VSD with a big directional shunt. The patient had severe PAH with a pressure of 63mmHg. Room air saturation was maintained at 85 – 88%. The patient's family history was negative for any known birth defects. Both parents had normal craniofacial development. An airway assessment of the patient revealed a COPUR score of 6. All routine investigations were within normal limits. The patient was planned for cleft palate repair and circumcision.

The patient was shifted inside the OR, and routine monitors were connected, Basal heart rate was 110 bpm, and SpO2 was 86% at room air. Premedicated with Midazolam 1 mg and induced with propofol 40mg intravenously. Intubated with size 5.5 south pole Ring–Adair–Elwyn endotracheal tube and fixed at 18 cms after paralyzing with atracurium 10mg. Maintained with medical air and oxygen with FiO2 of 0.6 and 2% sevoflurane. The left radial artery was cannulated and invasive BP monitoring was done. Hemodynamic was maintained with a MAP of >70mmHg throughout the procedure. Saturation was ranging around 92-95% intraoperatively with 0.6 FiO2. The procedure lasted for 1 hour which went uneventful and an ABG was taken at the end of the procedure which showed metabolic acidosis, correction was given with sodium bicarbonate

intravenously. The patient was shifted to the PICU for elective ventilation. FiO2 was reduced to 21% and saturation was maintained at 92-94% and the boy was extubated 2 hours postoperatively in the PICU.

Figure 1: Intubated with south pole Ring–Adair–Elwyn endotracheal tube

Figure 2: Intraoperative ventilatory settings



3. Conclusion

The anesthetic management of the 5-year-old boy with Goldenhar Syndrome undergoing cleft palate repair and circumcision proved successful, despite the underlying complex cardiac condition of Tetralogy of Fallot with a large subaortic ventricular septal defect (VSD). The use of invasive blood pressure monitoring, along with meticulous attention to airway assessment, ensured a controlled intraoperative course.

The presence of severe pulmonary arterial hypertension (PAH) and a large VSD added complexity to the case, necessitating a delicate balance in maintaining oxygen saturation and hemodynamic stability. The decision to

utilize a combination of medical air and oxygen with sevoflurane was tailored to the patient's needs, contributing to a well-maintained MAP throughout the procedure.

The postoperative period in the Paediatric Intensive Care Unit (PICU) involved effective ventilation, gradual reduction of FiO2, and successful extubation, highlighting the importance of continued vigilance in the early recovery phase. The prompt correction of metabolic acidosis with intravenous sodium bicarbonate underscores the vigilance and adaptability of the anaesthesia team (11).

GS is characterized by malformations that remain unexplained by any single theory (6). Potential causes of GS include chromosomal abnormalities, disruptions in placental or embryonic blood supply due to vascular pathogenesis, disturbances in neural crest cells, environmental factors, exposure to teratogens, and maternal diabetes. Nevertheless, the exact cause of GS remains unidentified (6-10).

This case exemplifies integration of preoperative assessment, advanced monitoring, and skillful intervention to navigate the complexities of Goldenhar Syndrome and congenital cardiac anomalies.

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