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A Perfect Storm in Paediatric Anaesthesia: Uncorrected Transposition of Great Arteries with Goldenhar Syndrome Undergoing Non-Cardiac Surgery

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ABSTRACT

Background: *Coexisting uncorrected dextro-transposition of the great arteries (d-TGA) and Goldenhar syndrome is exceptionally rare and presents significant perioperative risk, particularly in non-cardiac surgery.*

Case Presentation: *Case presentation: Prior to cardiac surgery, a 5-year-old boy with d-TGA, a large atrial septal defect, patent ductus arteriosus, and Goldenhar syndrome craniofacial features was scheduled for full-mouth rehabilitation. On room air, the baseline oxygen saturation was between 62 and 72%. A thorough airway plan was created in anticipation of a challenging airway. Systemic vascular resistance was maintained during induction with ketamine and fentanyl, and tracheal intubation was accomplished successfully on the first try with a C-MAC D-blade video laryngoscope. Sevoflurane in 50% oxygen-air was used to maintain anesthesia, avoiding substances that could raise pulmonary vascular resistance. Intraoperative oxygen saturation improved to 92%, and hemodynamics stayed steady. After a smooth extubation, the patient was observed in the pediatric intensive care unit.*

Conclusion: *Careful preoperative assessment, multidisciplinary coordination, and advanced airway planning are essential in managing rare combinations of cyanotic congenital heart disease and craniofacial anomalies. Video laryngoscopy and ketamine-based induction can optimise both airway safety and cardiovascular stability in such high-risk paediatric patients.*

INTRODUCTION:

Managing anesthesia in children with congenital anomalies is rarely straightforward, and when cardiac and craniofacial issues overlap, the difficulty can significantly rise. One such challenge is dextro-transposition of the great arteries (d-TGA), a cyanotic congenital heart disease in which the pulmonary artery and aorta are attached to the wrong ventricles. Circulation hence takes place in parallel rather than sequential fashion. Oxygenated blood cannot effectively enter the systemic circulation if there is no intracardiac or extracardiac shunt, such as a patent ductus arteriosus (PDA) or an atrial septal defect (ASD). These patients' survival depends on these mixing lesions, and any change in vascular resistance or myocardial function while under anesthesia may be harmful [1].

The situation is complicated by Goldenhar syndrome, a craniofacial disorder that affects the development of the first and second branchial arches and often presents as abnormalities of the vertebrae, mandibular hypoplasia, and facial asymmetry. These anatomical features often lead to difficult airway conditions. Furthermore, a significant portion of patients with Goldenhar syndrome also have congenital cardiac defects, which are sometimes not discovered until symptoms start to show up following surgery. [2].

Goldenhar syndrome and uncorrected d-TGA rarely occur together, necessitating careful coordination between the surgical, cardiology, and anesthesia teams. In this case study, we detail the anesthetic treatment of a child with both disorders who received general anesthesia for full-mouth rehabilitation. A customized, step-by-step approach was necessary to guarantee airway control and hemodynamic stability during the procedure due to the patient's particular anatomical and physiological factors.

CASE REPORT:

A 5-year-old male child came to our hospital with history of dyspnoea on exertion, cyanosis with frequent cyanotic spells as informed by the mother. Clinical examination revealed central cyanosis with room air saturation of 62% preductal and 72% postductal. The child had facial dysmorphism (midfacial hypoplasia), microtia and retrognathia and was diagnosed to be Goldenhar syndrome. Cardiovascular examination revealed single loud second heart sound. 2D echo revealed dextrocardia with transposition of great arteries with atrial septal defect (ASD) and patent ductus arteriosus (PDA) with bidirectional shunt, good biventricular function with mild pulmonary hypertension. The child was initially scheduled for TGA repair, however due to the presence of dental caries and associated infection, the child was posted for full mouth rehabilitation prior to the cardiac procedure. Routine blood investigations like complete blood count, coagulation profile was within normal limit.

The child was posted for full mouth rehabilitation under general anaesthesia. Infective endocarditis prophylaxis was given 30 minutes before shifting the patient to operation theatre. Informed and high-risk consent was obtained. Baseline vitals were heart rate 109/min, BP 90/60 mm Hg, saturation of 72% room air. Since it is an anticipated difficult airway, difficult airway cart was arranged inside the operating room. Video laryngoscopes and different sizes of masks,bougie, oral and nasal airways were kept ready. Fentanyl 1mcg/kg was given to blunt the intubation response. Induction was done with ketamine 1mg/kg and the patient was paralysed with iv vecuronium 0.1mg/kg and bag mask ventilation was done for 3 minutes. C-MAC D blade video laryngoscope was used for intubation and we could able to intubate using a 4.5 uncuffed tube in a single attempt with minimal manipulations. Anaesthesia was maintained on 50% oxygen and 50% air with sevoflurane to maintain a minimum alveolar concentration (MAC) of 1. The saturation picked up to 92% intraoperatively. Hemodynamic parameters and SpO2 remained stable throughout the surgery. At the end of the procedure the patient was reversed with glycopyrrolate 0.01mg/kg and neostigmine 0.05mg/kg. After ensuring adequate neuromuscular blockade reversal, patient was extubated. The patient was observed in Post Anaesthesia Care Unit (PACU) for 1 hour and was shifted to Paediatric intensive care unit (PICU) for further management.

DISCUSSION:

Anaesthesia for children with complex congenital heart disease (CHD) and craniofacial syndromes presents significant perioperative challenges. In our case, the child had unrepaired d-transposition of the great arteries (d-TGA) and Goldenhar syndrome, a rare and high-risk combination that required meticulous planning.

In d-TGA, the systemic and pulmonary circulations run in parallel rather than in series. Systemic survival depends entirely on the presence of intracardiac or extracardiac shunts—such as an atrial septal defect (ASD), ventricular septal defect (VSD), or patent ductus arteriosus (PDA)—which allow mixing of

oxygenated and deoxygenated blood and help maintain systemic oxygen delivery [1]. In our patient, the presence of a large ASD and PDA with bidirectional shunting supported preoperative saturations of 62–72%. Anaesthetic goals in such settings revolve around maintaining systemic vascular resistance (SVR), avoiding increases in pulmonary vascular resistance (PVR), and preserving adequate preload and myocardial contractility [3].

Craniofacial abnormalities in goldenhar syndrome include vertebral anomalies, facial hypoplasia, retrognathia. These abnormalities can lead to airway difficulties.[2] In 58% of the patients with goldenhar syndrome there is association of congenital heart disease which will further complicate the anaesthesia care.[4] In our case, we successfully ventilated the child with a mask, even though the child's unusual facial features suggested potential airway issues. Anticipating difficulties with direct laryngoscopy, we decided to use a C-MAC D blade video laryngoscope. This choice allowed us to achieve successful intubation on the first attempt. Milne et al. have reported similar success using video laryngoscopy in Goldenhar patients, emphasizing the advantages of this approach.[5]

The choice of anaesthetic agents was individualized based on both cardiac and airway considerations. Ketamine was selected for induction because of its ability to preserve SVR and avoid myocardial depression—properties particularly beneficial in cyanotic CHD [6]. Fentanyl was used for its haemodynamic stability and to blunt sympathetic responses without increasing PVR. We deliberately avoided atracurium, which can cause histamine-mediated vasodilation and reduce SVR [7]. Anaesthesia was maintained with sevoflurane, and nitrous oxide was avoided due to its tendency to increase PVR and expand gas bubbles—posing a real risk of paradoxical embolism in patients with intracardiac shunts [6].

In order to reduce blood viscosity and avoid aggravating right-to-left shunting, we concentrated on preserving normothermia, normocarbia, and proper hydration throughout the procedure. With stable hemodynamics, the patient's intraoperative saturation increased to 92%. All intravenous lines and syringes were meticulously de-aired prior to use because paradoxical embolism is still a serious risk in children with intracardiac defects like ASD and PDA [8].

In this instance, preoperative planning was essential. To create a thorough perioperative plan, a multidisciplinary team comprising dental surgeons, anesthesiologists, cardiologists, and pediatric intensivists worked together. The American Heart Association advises patients with unrepaired cyanotic CHD undergoing invasive procedures to receive infective endocarditis prophylaxis 30 minutes prior to the procedure. [8].

This example adds to the limited literature on managing anesthesia in infants with cyanotic congenital heart disease who are syndromic and undergoing non-cardiac surgery. While the anaesthetic approaches to d-TGA and Goldenhar syndrome have been discussed individually, there are few published reports addressing their coexistence. The fact that this patient responded well to video laryngoscopy and a specially designed anaesthetic regimen highlights the significance of meticulous preoperative assessment, airway readiness, and vigilant hemodynamic management.

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