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CASE REPORT OPEN ACCESS

Anaesthetic management using nasal intubation in a child with CHARGE syndrome undergoing full-mouth rehabilitation: A case report

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Abstract

CHARGE syndrome is a rare, complex congenital disorder caused predominantly by mutations in the CHD7 gene, characterized by a multisystem involvement including Coloboma, Heart defects, Atresia of the choanae, Retardation of growth and development, genital anomalies, and ear abnormalities. Each patient presents a unique spectrum of anomalies, posing significant challenges for perioperative management. This case report presents the perioperative anaesthetic management of a 5 year old male child with genetically confirmed CHARGE syndrome (CHD7 mutation: c.6213delA) scheduled for full mouth rehabilitation under general anaesthesia. The child presented with multiple classical features including bilateral coloboma, atrial septal defect (ASD), bilateral sensorineural hearing loss, retrognathia, and global developmental delay. Given the known association with choanal atresia, nasal patency was assessed preoperatively. Airway examination revealed retrognathia and high arched palate, raising concerns for difficult intubation. A multidisciplinary strategy was planned. Inhalational induction with sevoflurane was used to preserve spontaneous ventilation, followed by intravenous propofol and atracurium to ensure smooth intubation while maintaining hemodynamic stability. Anticipating airway difficulty, nasal intubation was performed using a C-MAC video laryngoscope. Intraoperative management emphasized normothermia and normocapnia to ensure cerebral and hemodynamic stability. The surgery proceeded uneventfully, and the child recovered without complications. This case highlights the importance of a tailored, pathophysiology-driven anaesthetic plan and interdepartmental coordination in managing syndromic children. Key takeaways include early identification of airway and systemic risks, detailed preoperative planning, and the use of advanced airway tools to ensure safe outcomes in pediatric patients with complex congenital anomalies.

Keywords: CHARGE syndrome; congenital heart defects; CHD7 mutation; general anesthesia; difficult airway

Introduction

Coloboma, heart defects, atresia of choanae, retardation of growth and development, genital hypoplaisa,ear anomalies and deafness syndrome is a rare, multisystem congenital disorder characterised by a spectrum of anomalies, including coloboma of the eye, congenital heart defects, choanal atresia, growth retardation, genital hypoplasia, and ear abnormalities. First described in 1981 as an association of congenital malformations, it is now recognised as a genetically defined syndrome, most commonly caused by mutations in the CHD7 gene on chromosome 8q12.1. This gene plays a critical role in chromatin remodelling during embryogenesis, and its mutation results in defective development of neural crest-derived structures [1, 2]. The estimated incidence of CHARGE syndrome is approximately 1 in 10,000 to 15,000 live births [1]. Owing to its phenotypic variability and lack of uniform presentation, the clinical and anaesthetic management of affected individuals remains challenging [3, 4].

This case is significant as it describes the anaesthetic management of a 5-year-old child with genetically

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confirmed CHARGE syndrome undergoing full mouth rehabilitation under general anaesthesia. The perioperative period in such patients poses unique difficulties due to craniofacial anomalies such as retrognathia, micrognathia, cleft palate, and possible choanal atresia, all of which predispose to difficult airway management [5, 7]. Cardiovascular anomalies, particularly atrial and ventricular septal defects, are frequently observed and may influence anaesthetic drug selection and intraoperative haemodynamic stability [6, 8]. Additionally, sensorineural hearing loss and global developmental delay further complicate preoperative communication and cooperation [2, 3, 6].

Although individual features of CHARGE syndrome have been well documented in the paediatric literature, there is limited comprehensive guidance addressing its perioperative anaesthetic implications (Table 1), especially in the context of airway management. The anaesthesiologist must remain vigilant for airway obstruction, respiratory complications, and autonomic instability [6, 9].

Table 1: Flowchart summarising perioperative anaesthetic strategy and airway management plan in CHARGE syndrome.

Phase	*Considerations*	*Actions Taken*
Preoperative	Retrognathia, high-arched palate, ASD	Nasal patency checked (saline / suction); ENT consulted
Induction	Anticipated difficult airway, cardiac lesion	Inhalational induction with sevoflurane, propofol + atracurium IV
Intubation	Need for nasal access, limited mouth opening	Pediatric C-MAC, used 5.0 mm uncuffed nasal ETT
Intraoperative	Avoid increased PVR (glossoptosis), oedema	Dexamethasone IV, reintubation, nasopharyngeal airway
Post-op	Delayed airway compromise, neuro status	PACU monitoring, SpO_2 > 93%, discharge on POD 2

This case report contributes valuable insight into the multidisciplinary, precision-based anaesthetic planning required in children with CHARGE syndrome. It highlights the importance of thorough preoperative evaluation including choanal patency assessment anticipation of airway difficulty, judicious selection of induction agents, and the utility of video laryngoscopy to enhance safety [10, 11, 12]. Overall, the report reinforces the need for tailored perioperative care strategies aligned with the patient's underlying pathophysiology, with the goal of ensuring optimal outcomes and avoiding preventable complications in this high-risk paediatric population [9, 13].

This case report presents the perioperative anaesthetic management of a 5-year-old male child with genetically confirmed CHARGE syndrome (CHD7 mutation: c.6213delA) scheduled for full mouth rehabilitation under general anaesthesia.

Case presentation

A 5-year-old male child, weighing 16 kg with a BMI of 15.6 kg/m^2, presented with multiple dental caries, extensive enamel hypoplasia, and impaired oral intake secondary to pain at Sri Ramachandra Institute of Higher Education and Research. He was scheduled for comprehensive full mouth rehabilitation under general anaesthesia. The child had a genetically confirmed diagnosis of CHARGE syndrome, identified through chromosomal analysis revealing a heterozygous pathogenic mutation in the CHD7 gene (c.6213delA), consistent with contemporary diagnostic criteria.

CHARGE syndrome (Coloboma, heart defects, atresia of choanae, retardation of growth and development, genital hypoplasia, ear anomalies) is a rare multisystem disorder with an incidence of approximately 1:10,000 to 1:15,000 live births. In this patient, the phenotypic features, included bilateral iris and retinal coloboma, a haemodynamically significant ostium secundum atrial septal defect (ASD), bilateral sensorineural hearing loss requiring auditory amplification, retrognathia, high-arched palate, and global developmental delay. There was no history of seizures, vocal cord palsy, or confirmed choanal atresia, although neonatal feeding difficulties necessitated NICU admission in infancy.

The child had delayed motor and language milestones but showed stable social interaction and attended special education therapy. There was no consanguinity or family history of congenital anomalies. This was his first exposure to surgery and general anaesthesia.

Preoperative echocardiography revealed a 5 mm ostium secundum ASD with a left-to-right shunt, preserved biventricular function, and no pulmonary hypertension. Nasal patency was confirmed bilaterally using a saline drop test and suction catheter probing. Craniofacial evaluation showed retrognathia and a high-arched palate without cleft. Mallampati grading was difficult to assess due to limited cooperation and oral aperture was limited to 2.5cm, and was therefore partially attempted but not fully determined (Figure 1).

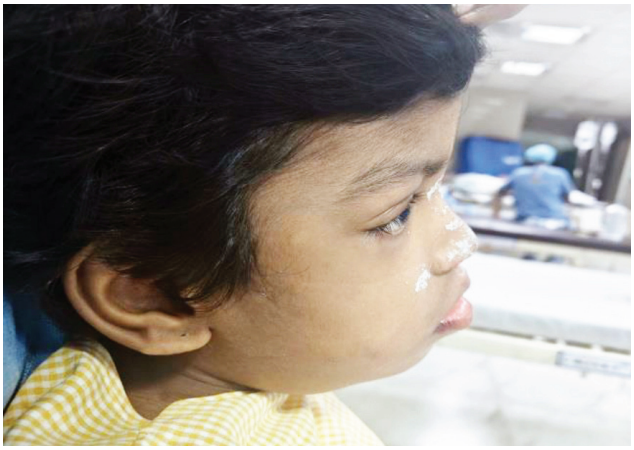

Figure 1: Preoperative facial profile showing retrognathia.

The child was fasted appropriately and premedicated with oral midazolam (0.5 mg/kg) for anxiolysis. In the operating room (OR), standard ASA monitoring was applied. Glycopyrrolate 0.06 mg IV was administered to reduce oral secretions. Given the anticipated difficult airway and underlying cardiac lesion, a spontaneous ventilation strategy was adopted. Preoxygenation was achieved with 100% oxygen via a well-fitted facemask for 3 minutes.

Induction commenced with 8% sevoflurane in 100% oxygen via a Jackson-Rees circuit. Following IV access, propofol 2 mg/kg was administered slowly, and atracurium 0.5 mg/kg IV was given for muscle relaxation. Bag-mask ventilation was adequate, though the oral aperture was limited.

A 2.5 mm C-MAC video laryngoscope blade was used. Laryngoscopy revealed a Cormack-Lehane Grade IIb view due to anterior airway distortion (Figure 2). A 5.0 mm uncuffed nasal endotracheal tube (ETT) was gently passed through the right naris under video guidance and confirmed with capnography and bilateral auscultation. The ETT was fixed at 16 cm, and a throat pack was placed.

Anaesthesia was maintained with sevoflurane in a 50:50 air-oxygen mixture under pressure-controlled ventilation. Analgesia included IV fentanyl (1 µg/kg) and paracetamol (15 mg/kg). Ringer's lactate was administered at 4 mL/kg/hr. Due to the ASD, fluid overload was avoided to prevent pulmonary overcirculation and right atrial strain.

The procedure lasted 2.5 hours. The child experienced desaturation episodes (SpO_2 falling to 84–86%), which improved within 1–2 minutes after airway repositioning and oxygen supplementation. $EtCO_2$ trends demonstrated transient elevation (45–50 mmHg) during desaturation, returning to baseline (35–38 mmHg) following corrective measures. Laryngospasm and bronchospasm were excluded.

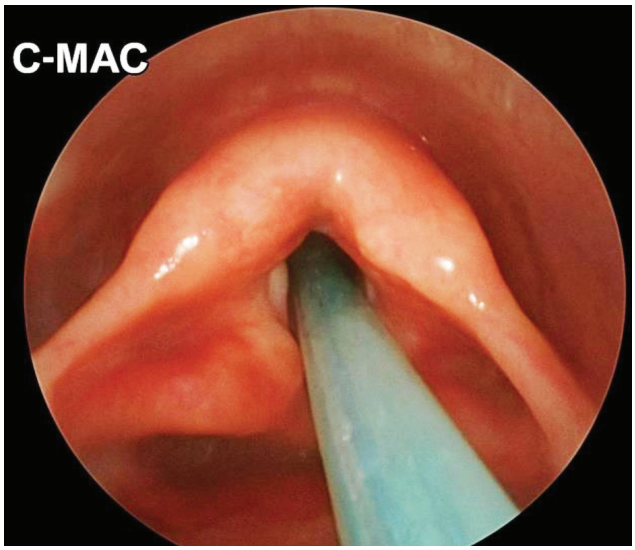

Figure 2: C-MAC video laryngoscopic view demonstrating Cormack-Lehane Grade IIb glottic visualisation with nasal ETT railroading.

Throughout surgery, core temperature was maintained between 36.8–37.0°C, $EtCO_2$ between 35–40 mmHg, and MAP between 55–65 mmHg. No arrhythmias were observed.

At the end of the procedure, spontaneous efforts returned. Neuromuscular blockade was reversed with neostigmine (0.05 mg/kg) and glycopyrrolate (0.01 mg/kg). Extubation failure occurred secondary to airway edema and desaturation. Reintubation was performed promptly (Figure 3). Postoperative monitoring in the pediatric ICU was continued to prevent recurrence and ensure airway stability. Dexamethasone (0.15 mg/kg IV) was administered.

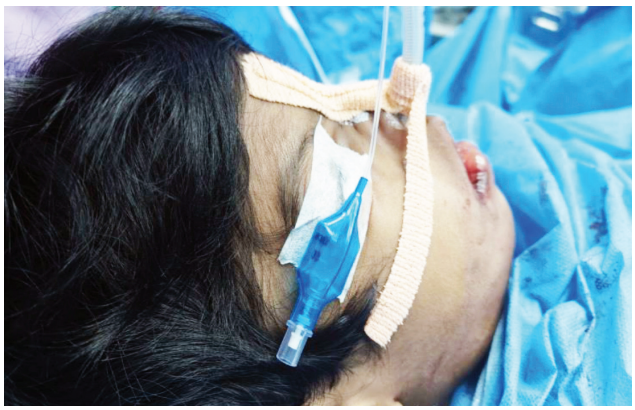

Figure 3: After successful nasal intubation.

After 20 minutes of elective ventilation and propofol sedation, the child resumed spontaneous respiration

with $EtCO_2$ 36 mmHg and stable haemodynamics. Re-extubation was performed with a lubricated nasopharyngeal airway (NPA) in place. Ventilation remained stable with SpO_2 >98% on 40% oxygen via facemask.

Post-extubation, the child was monitored in the post-anaesthesia care unit (PACU) for 4 hours. Respiratory rate remained between 22–26/min, and SpO_2 remained >98%. No signs of airway obstruction or retractions were observed. Oral intake was resumed 6 hours post-extubation, and the child remained clinically stable. Analgesia was maintained with IV paracetamol.

He was discharged on postoperative day two without respiratory or neurological complications. At 2-week follow-up, the child showed no delayed airway oedema, aspiration, or neurocognitive decline. Dental outcomes were satisfactory, with improved feeding and pain resolution.

Discussion

The decision to perform nasal intubation using a paediatric C-MAC video laryngoscope in this child with CHARGE syndrome was made after thorough assessment of the craniofacial anatomy, surgical requirements for full oral access, and the anticipated anaesthetic risks arising from multisystem involvement. CHARGE syndrome presents one of the most complex challenges in paediatric anaesthesia due to its variable phenotypic expression and multiple anatomical anomalies affecting airway, cardiac, and neurological systems [1, 2]. In this case, features such as retrognathia, high-arched palate, iris and retinal coloboma, atrial septal defect (ASD), sensorineural hearing loss, and developmental delay necessitated a tailored, multidisciplinary anaesthetic plan rooted in syndromic pathophysiology and paediatric airway principles [3, 4].

Children with CHARGE syndrome frequently present with craniofacial anomalies that complicate mask ventilation and direct laryngoscopy [2, 4]. Retrognathia reduces mandibular space, impeding anterior tongue displacement, while high-arched or cleft palate can interfere with mask seal and airway patency. Midfacial hypoplasia and narrow nasal passages further complicate nasal intubation [5]. Moreover, choanal atresia either unilateral or bilateral is a hallmark of the syndrome and mandates preoperative nasal patency assessment [6, 7]. In this child, bilateral choanal patency was confirmed using the saline drop test and gentle probing with a suction catheter, allowing us to safely proceed with the nasal route.

Nasal intubation was necessitated by the surgical requirement for unobstructed access to the oral cavity. The use of video laryngoscopy, specifically the C-MAC system, enhanced glottic visualisation while minimising cervical manipulation and trauma. Video laryngoscopy has demonstrated superior success rates and safety in difficult paediatric airways, especially in syndromic children with craniofacial malformations [11, 12]. Use of a lubricated bougie and under-vision advancement of a 5.0 mm uncuffed nasal endotracheal tube (ETT) facilitated atraumatic, first-pass success.

Inhalational induction with 8% sevoflurane in 100% oxygen was selected to preserve spontaneous ventilation a standard approach when airway difficulty is anticipated [12, 13]. Sevoflurane was preferred for its bronchodilatory effect, haemodynamic stability, and favourable induction profile. After securing intravenous access, atracurium was used due to its organ-independent Hofmann elimination, allowing predictable recovery in children with variable metabolic profiles [14]. Careful sizing and lubrication of the nasal ETT prevented resistance and mucosal trauma. Correct placement was confirmed by capnography, chest rise, and bilateral auscultation. A throat pack was inserted to minimise aspiration and facilitate a bloodless surgical field.

Intraoperative anaesthesia was maintained with sevoflurane in an air-oxygen mixture (FiO_2 0.5), using pressure-controlled ventilation. Analgesia was provided with IV fentanyl (1 µg/kg) and paracetamol (15 mg/kg). The child had a 5 mm ostium secundum ASD with a left-to-right shunt. Avoidance of factors that could increase pulmonary vascular resistance such as hypoxia, hypercarbia, and acidosis was critical to preventing shunt reversal and right-sided volume overload [15]. Excessive fluid administration and hyperoxia were also avoided to mitigate increased pulmonary blood flow. Fluid therapy was guided by heart rate, urine output, and mean arterial pressure (MAP).

Two intraoperative desaturation episodes (SpO_2 84–86%) occurred during intense oral suctioning and retraction. These events were promptly managed by suspending surgery, increasing FiO_2 to 100%, providing manual ventilation, and performing gentle alveolar recruitment. Such episodes underscore the fragility of the paediatric airway in syndromic children and the importance of protocolised responses in preventing further compromise [4, 16].

The most significant challenge occurred during extubation. Following neuromuscular reversal, despite spontaneous efforts and adequate tidal volumes, extubation led to immediate stridor, paradoxical chest movement, and desaturation. Bag-mask ventilation was successful, and reintubation was performed under video

laryngoscopic guidance. Airway inspection revealed glossoptosis and pharyngeal collapse, likely due to underlying hypotonia or cranial nerve dysfunction both associated with CHARGE syndrome [1, 4, 5]. IV dexamethasone was administered to reduce suspected oedema.

After 20 minutes of ventilatory support and spontaneous respiratory recovery, a second extubation was attempted with insertion of a lubricated nasopharyngeal airway (NPA). The NPA provided a stent for the posterior pharynx, preventing glossoptosis-related obstruction. This intervention was successful, and the child maintained SpO_2 >98% on 40% oxygen via facemask. Postoperative monitoring in a high-dependency setting ensured safe recovery.

This case adds to the growing literature on CHARGE syndrome anaesthesia, supporting the safety and efficacy of video-guided nasal intubation when anatomy and airway patency permit. While some reports advocate fibreoptic intubation or elective tracheostomy [6, 10, 18], our case highlights that anticipatory airway evaluation and use of paediatric video laryngoscopy can avoid invasive interventions. Moreover, proactive management of potential ASD-related haemodynamic shifts and a high index of suspicion for extubation-related obstruction contributed to a successful outcome.

The strength of this case lies in the anticipatory, multidisciplinary approach preoperative ENT and cardiology input, planned airway management with appropriate equipment, and peri-extubation strategies. The use of video guidance facilitated atraumatic nasal intubation, and post-extubation support with NPA mitigated pharyngeal collapse. These strategies are especially relevant for children with neurodevelopmental delay or cranial nerve involvement [1, 3, 14].

However, some limitations merit discussion. Desaturation episodes might have been mitigated by apnoeic oxygenation or high-flow nasal oxygen (HFNO) during suctioning [16].Furthermore, earlier prophylactic corticosteroid administration may have reduced extubation-related airway compromise [17]. These represent learning points for future similar scenarios.

Comparison with existing literature reveals that extubation failure and prolonged ventilation are not uncommon in CHARGE patients [6, 10, 18]. Our case demonstrates that with structured assessment and intraoperative vigilance, nasal intubation using C-MAC video laryngoscopy can be safely accomplished. Each CHARGE patient, however, presents a unique constellation of risks; hence, individualised anaesthetic planning remains essential [1, 2].

Conclusion

This case illustrates the necessity of a precision-based, interdisciplinary approach to anaesthetic care in CHARGE syndrome. It underscores the value of preoperative nasal patency assessment, real-time video laryngoscopic assistance for nasal intubation, ASD-focused haemodynamic management, and the proactive use of NPAs in the extubation process. As survival rates improve, the surgical needs of syndromic children are increasing, requiring tailored anaesthetic strategies to ensure safety and optimize outcomes.

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Conflicts of interest

Authors declare no conflicts of interest.

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