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Case Report

Anaesthesia Section

Tailored Sedation for Oesophageal Dilatation in Goldenhar Syndrome: Avoiding Airway Instrumentation with High-flow Nasal Cannula (HFNC) Support

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ABSTRACT

Goldenhar syndrome, or oculo-auriculo-vertebral spectrum, is a rare congenital anomaly involving craniofacial dysmorphia, vertebral deformity, and atypical outer ear, and is classically complicated by challenging airway management. In this case report, the anaesthetic management of a nine-year-old male with Goldenhar syndrome who also had a recurrent benign oesophageal stricture with odynophagia has been described. The patient also had many previous oesophageal dilatations and had generalised anatomical deformities such as asymmetrical facial deformity, mandibular hypoplasia, absent pinnae, scoliosis, and tracheal shift. Due to the presence of extremely high-risk airway complications, sedation was the procedure of choice without general anaesthesia. Intravenous sedation by ketamine, fentanyl, and midazolam was employed for the procedural sedation, and the maintenance of adequate ventilation as well as oxygenation during the procedure was with a High-Flow Nasal Cannula (HFNC). The anaesthetic team had a difficult airway cart and rescue airway devices on hand, even though invasive airway management was not employed. The procedure proceeded successfully with stable haemodynamics and satisfactory spontaneous ventilation. This case sets an example of the need for individualised anaesthetic planning, appropriate use of sedation, and the value of HFNC in the management of syndromic children with difficult airways.

Keywords: Anaesthesia, High-flow nasal cannula, Ketamine, Oxygen inhalation therapy, Paediatric anaesthesia

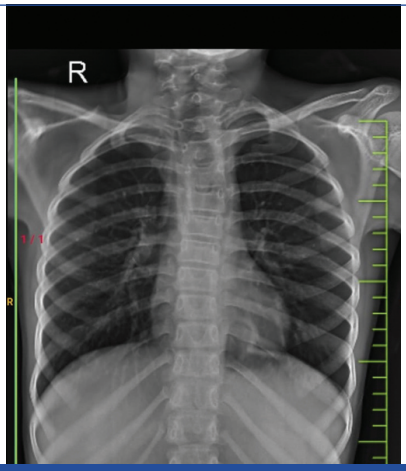
CASE REPORT

A nine-year-old boy with diagnosed Goldenhar syndrome came with the complaint of odynophagia for eight days. The pain was severe in nature and non-radiating. The child was described as being well by the mother until he developed one episode of generalised tonic-clonic seizure for about two minutes, a week back, after which he was worked up and found to have a benign stricture of the oesophagus on endoscopy. He was treated with oesophageal dilatation and was discharged on syrup levetiracetam, on which he was maintained regularly afterwards.

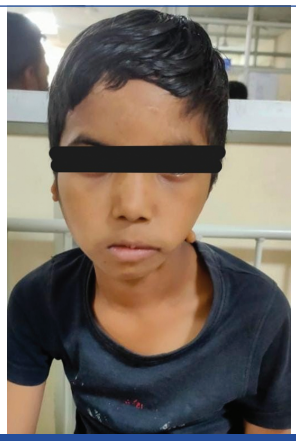
Over time, the child continued to develop intermittent bouts of dysphagia and odynophagia, requiring multiple admissions for dilatation of the oesophagus. Previous endoscopic studies always revealed a benign lower oesophageal stricture with herniated hiatus, Hill's grade IV [1]. Serial dilatations were advised at intervals to prevent recurrence.

The kid had several congenital anomalies typical of Goldenhar syndrome. These include bilateral anotia (complete absence of outer ears), facial asymmetry, hypoplastic mandible with a smaller left condyle, and scoliosis. Tracheal deviation to the right, most likely secondary to spinal curvature, was noted on the radiographs. CT head and neck scan showed a bilateral absence of external auditory canals, abnormal pinnae, and asymmetrical pneumatisation of the mastoid air cells. A 2D echocardiogram revealed no structural cardiac abnormalities.

For preoperative evaluation before sedation for oesophageal dilatation, the child underwent a complete assessment. His weight was 20 kg, height was 118 cm, and body mass index was 14.4. He was asymptomatic for active respiratory or cardiac disease. On clinical examination, he presented various findings indicative of a difficult airway- tracheal shift [Table/Fig-1], scoliosis-related restricted neck movement, microstomia, hypoplastic mandible [Table/Fig-2], and facial dysmorphism [Table/Fig-2]. With these observations, the



[Table/Fig-1]: Preoperative radiograph showing scoliosis and rightward tracheal deviation.



[Table/Fig-2]: Clinical photograph demonstrating facial asymmetry, bilateral anotia, and mandibular hypoplasia.

anaesthesiology team anticipated a difficult airway and took the necessary steps in anticipation.

A standardised and individualised anaesthetic regimen was planned. A difficult airway cart was prepared, including paediatric-sized laryngeal mask airways, various sizes of face masks to accommodate anomalous facial anatomy, a paediatric video laryngoscope, a fibreoptic bronchoscope, and an emergency tracheostomy kit. Sedation was considered ideal for reducing airway manipulation and allowing spontaneous respiration. An HFNC oxygenation was chosen to maximise oxygenation and apnoeic oxygenation support throughout the procedure.

On the day of the procedure, the child was kept nil per os for six hours. In the operating room, routine monitors were applied, including Electrocardiography (ECG), Non-invasive Blood Pressure (NIBP), SpO_2, and end-tidal CO_2. HFNC was initiated at 30-35 L/min with a FiO_2 of 0.5. This method was selected to maintain oxygenation, provide a minimal positive end-expiratory pressure, and extend the safe apnoea time without invasive ventilation.

Premedication was done with intravenous glycopyrrolate 0.08 mg to reduce secretions. Sedation was initiated with intravenous midazolam 1 mg and followed by fentanyl 40 mcg for analgesia. Ketamine was titrated in three increments (10 mg + 10 mg + 5 mg), taking advantage of its ability to maintain spontaneous respiration, airway reflexes, and cardiovascular stability. The combination of ketamine and fentanyl was sufficient to provide sedation and pain relief with a decreased risk of airway collapse or apnea.

Throughout the 30-minute procedure, the child maintained spontaneous ventilation and was haemodynamically stable. There was minimal airway obstruction with deep sedation, which was readily managed with jaw thrust and positioning of the head. No adjunct to the airway or supraglottic device was required. The HFNC provided efficient oxygenation throughout, with oxygen saturation greater than 98% and end-tidal CO_2 within the normal range.

Post-procedure, the child was shifted to the recovery room and observed. He regained consciousness within 30 minutes with normal vital signs and respiratory effort. Feeding was restarted orally once the child was awake and alert. There was minimal post-procedure pain, and no further analgesia was needed. The child was discharged home on the fourth day following the procedure and was in good condition. The family was counselled about the need for follow-up for ongoing oesophageal monitoring and possible repeat dilatation. At follow-up three weeks later, the child remained asymptomatic with no recurrence of odynophagia or dysphagia. Clinical evaluation revealed good recovery, and no post-procedural complications were observed, indicating effective and sustained symptom relief.

DISCUSSION

Goldenhar syndrome, or oculo-auriculo-vertebral spectrum, is a congenital anomaly that occurs rarely and is characterised by a triad of craniofacial dysmorphology, vertebral malformations, and ear malformations. It is marked by a broad phenotypic variability that frequently encompasses hemifacial microsomia, mandibular hypoplasia, anotia or microtia, and vertebral malformations like scoliosis. Airway malformations and challenging anatomical landmarks make it highly difficult to manage anaesthesia, especially in those cases involving sedation or general anaesthesia [2].

A male child with Goldenhar syndrome presented with chronic benign oesophageal stricture and worsening odynophagia. His airway presented with multiple anticipated problems with craniofacial asymmetry, mandibular hypoplasia, microstomia, scoliosis with associated tracheal deviation, and non-detectable external ears. These defects truncated standard airway instruments and traditional airway device use, necessitating accurate preoperative planning. Anaesthetic management of syndromic children with expected difficult airways is a sensitive balancing act between ensuring satisfactory sedation and not blunting spontaneous respiration. The anaesthetic team used procedural sedation rather than general anaesthesia with endotracheal intubation. The rationale was to prevent failed or traumatic airway instrumentation complications such as airway oedema or airway loss control within an already compromised anatomical configuration [3,4].

The sedation regimen utilised a mix of midazolam, fentanyl, and ketamine. Midazolam was utilised as an amnestic and anxiolytic agent, fentanyl was utilised for analgesia, and ketamine was utilised to provide sedation without interfering with airway reflexes or respiratory drive. The dissociative anaesthesia provided by ketamine enabled the procedure to be done smoothly while preserving protective airway reflexes- an advantageous characteristic in patients with difficult airways. Ketamine possesses cardiovascular stability, which is particularly beneficial in paediatric cases [5].

One of the unique aspects of this case was the application of HFNC during sedation. HFNC is increasingly being seen as a helpful option in managing patients with compromised airways or undergoing sedation for short procedures. By delivering high-rate humidified and warmed oxygen, HFNC allows a constant FiO_2, supports some degree of continuous positive airway pressure level, and permits apnoeic oxygenation in hypoventilation. With HFNC used here, efficient oxygenation for the procedure, especially concerning the unforeseeable dynamics of the depth of sedation and transient airway obstruction, was adequately facilitated. This technique is beneficial in paediatric syndromic patients where airway instrumentation is challenging or should be avoided. Anaesthetic challenges often involve maintaining oxygenation amid unpredictable sedation depth and airway obstruction. HFNC offers a non-invasive solution by ensuring consistent oxygen delivery and partial airway support [6].

The anaesthetic team had a proper setup with a difficult airway cart, including paediatric supraglottic devices, video laryngoscopes, fiberoptic bronchoscopes, and an emergency tracheostomy setup. However, none of the adjuncts were required due to successful sedation and spontaneous ventilation. The decision not to instrument the airway unless necessary adhered to the principle of "airway minimalism" in syndromic patients with known facial and skeletal deformities [7].

Facial asymmetry and the absence of external ears posed additional problems with mask ventilation. The team overcame these issues by selecting the appropriate face mask size and modifying the monitor application techniques. Scoliosis and tracheal deviation are also predisposed to positional hypoventilation or desaturation, emphasising the need for ongoing end-tidal CO_2 monitoring, even with sedation, which was easily achieved by the use of modified nasal prongs for sampling exhaled CO_2 [8,9]. [Table/Fig-3] lists the anaesthetic management of patients with Goldenhar syndrome [10-12].

Author(s)	Case description	Anaesthetic management	Takeaway points
Current case	A 9-year-old boy with Goldenhar syndrome undergoing oesophageal dilatation; features included bilateral anotia, micrognathia [Table/Fig-2], tracheal deviation [Table/Fig-1], scoliosis [Table/Fig-1], facial asymmetry [Table/Fig-2]; history of seizures and recurrent odynophagia due to benign oesophageal stricture.	Sedation with midazolam, fentanyl, and titrated ketamine; spontaneous ventilation maintained; HFNC oxygenation used throughout (30-35 L/min, FiO_2 0.5); difficult airway cart prepared; no airway adjuncts required; procedure completed uneventfully.	HFNC is a safe and effective method for sedation in paediatric patients with difficult airways; ketamine preserves airway reflexes and ventilation; sedation reduces airway manipulation risks; pre-operative planning and readiness for airway emergencies are key.

Sun YH et al., [10]	A 5-year-old girl with Goldenhar syndrome undergoing cochlear implant surgery; presented with facial asymmetry, micrognathia, retracted jaw, microtia; Mallampati i.v. airway; normal neck mobility.	Induction with sevoflurane, followed by i.v. propofol, fentanyl, and rocuronium; video laryngoscope (McGrath®) + fibreoptic bronchoscope used after failed intubation attempts; successful transoral fibreoptic intubation achieved.	Combined video laryngoscope and fiberoptic bronchoscope improves success in challenging pediatric airways; awake intubation is impractical in children; careful pre-operative evaluation and multi-modal airway planning are essential.
Singh R et al., [11]	An 8-month-old male infant (6 kg) with Goldenhar syndrome undergoing cataract surgery; micrognathia, retrognathia, malformed ear, limited neck movement, facial asymmetry; minor atrial septal defect noted on echo.	Inhalational induction with sevoflurane; i.v. fentanyl; supraglottic airway (I-gel #1.5) used instead of intubation; spontaneous breathing maintained; anaesthesia with sevoflurane and oxygen-air mix; uneventful surgery and recovery.	Supraglottic airway devices can be a safe primary airway strategy in Goldenhar syndrome patients with difficult airways; maintaining spontaneous breathing is crucial; detailed anatomical and cardiac evaluation aids in planning; awareness of possible airway anomalies like tracheal bronchus is essential.
Koo JU et al., [12]	An 8-month-old boy with Goldenhar syndrome, undergoing excision of conjunctival dermoid and ureteral reimplantation, had typical craniofacial features and vertebral anomalies associated with Goldenhar syndrome.	General anaesthesia with thiopental, rocuronium, and sevoflurane; successful tracheal intubation without complications; surgery and recovery were uneventful.	Despite complex airway features, standard induction and intubation may be feasible in some Goldenhar syndrome cases; careful airway assessment and preparation remain essential; congenital anomalies may not always preclude successful airway management.

[Table/Fig-3]: Comparison of anaesthetic management of patients with Goldenhar syndrome [10-12].

CONCLUSION(S)

Anaesthetic management of children with Goldenhar syndrome must be tailor-made and cautious, given the minimal craniofacial and airway anomalies in the condition. For this patient, HFNC oxygenation with ketamine sedation was a valuable and safe method to provide spontaneous ventilation without complications of airway manipulation. Preoperative rationalisation, readiness with the presence of a difficult airway trolley, and the use of non-invasive modes of sedation are guidelines for managing syndromic children's cases. The case emphasises innovation, flexibility, and collaboration among many specialities in delivering safe and effective anaesthesia treatment to children with complex anatomy.

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