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Anaesthetic Management of Achilles Tendon Tenotomy in a 2-Year-Old Child with Goldenhar Syndrome Using Videolaryngoscopy-A Case Report

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KEYWORDS

Cataract surgery. Age-related macular degeneration. Visual outcomes.

ABSTRACT:

Background: Cataract and age-related macular degeneration (AMD) are major causes of visual impairment in the elderly and often coexist. The role of cataract surgery in improving visual and functional outcomes in patients with AMD remains an area of clinical interest and debate.

Aim: To evaluate the outcomes of cataract surgery in patients with AMD with respect to visual acuity, quality of life, and disease progression.

Methods: A systematic review of 16 studies published between 2000 and 2024 was conducted using PubMed, MEDLINE, EMBASE, Cochrane Library, and Google Scholar. Eligible studies included randomized controlled trials and observational studies assessing visual outcomes, quality of life, and AMD progression after cataract surgery. Data were extracted and synthesized using random-effects models, with heterogeneity quantified using I^2 statistics.

Results: Across the 16 included studies, cataract surgery was associated with significant improvements in visual acuity (pooled SMD ≈ −0.17 logMAR) and vision-related quality of life (pooled SMD ≈ 0.50). Functional outcomes, including reading speed and contrast sensitivity, also improved consistently. No convincing evidence was found that cataract surgery accelerates AMD progression, particularly in patients receiving modern anti-VEGF therapy. However, heterogeneity among studies was high (I^2 > 70%), reflecting variations in study design, AMD stage, and outcome measures.

Conclusion: Cataract surgery in AMD patients results in clinically relevant improvements in visual function and quality of life without significantly increasing the risk of disease progression. Careful patient selection and counseling remain crucial, particularly in advanced AMD where postoperative visual gains may be limited.

ABSTRACT

Background: Goldenhar syndrome, a rare congenital condition characterized by craniofacial anomalies and potential multisystem involvement, poses significant anaesthetic challenges, particularly related to airway management. We report the case of a 2-year-old boy with Goldenhar syndrome undergoing right Achilles tendon tenotomy for isolated equinus contracture. Preoperative evaluation ruled out systemic anomalies, allowing focused planning for an anticipated difficult airway. General anaesthesia was induced with intravenous agents, and successful intubation was achieved using videolaryngoscopy on the first attempt. The intraoperative and postoperative courses were uneventful. This case highlights the importance of comprehensive preoperative assessment, preparedness with advanced airway devices, and individualized anaesthetic strategies in syndromic children. It also expands the phenotypic spectrum of Goldenhar syndrome to include isolated orthopaedic manifestations and demonstrates that routine procedures can be safely performed with meticulous planning.

INTRODUCTION

Goldenhar syndrome, also known as oculo-auriculo-vertebral spectrum, is a rare congenital condition arising from aberrant development of the first and second branchial arches [1-3]. The male to female ratio is 2:1 and the defect occurs in 1:3500-1:5600 live births. The aetiology of Goldenhar syndrome is multifactorial and related to genetic and environmental factors [4,5]. Affected children often present with a spectrum of craniofacial anomalies ranging from hemifacial microsomia and

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mandibular hypoplasia to microtia and maxillary hypoplasia that can complicate perioperative management.

Beyond the obvious facial asymmetry, Goldenhar syndrome often carries multisystem associations, including cardiac defects (such as ventricular septal defects or outflow tract abnormalities), renal malformations, and vertebral anomalies, each of which may remain occult until a thorough preoperative evaluation is undertaken [6,7]. The main anaesthetic concerns are difficult intubation and difficult bag-mask ventilation. The facial asymmetry may make mask ventilation difficult. Difficult intubation is due to an asymmetrical mandibular hypoplasia, hemifacial microsomia and tracheal deviation to one side [8]. Vertebral anomalies may include fused vertebrae which may make neck extension difficult [9]. The video laryngoscopes, fibreoptic bronchoscopes, and laryngeal mask airways have been used successfully to secure airway [10]. Consequently, every anaesthetic plan for these patients must be supported by meticulous airway assessment, readiness with advanced airway devices, and a robust strategy to manage unexpected complications.

In addition to the craniofacial features that dominate the clinical picture, Goldenhar syndrome can sometimes be associated with subtle orthopaedic manifestations, such as club foot deformity, limb defects etc.[11]. While the literature largely focuses on procedures involving the craniomaxillofacial region e.g., cleft lip and palate repair, ocular surgeries, or neurosurgical interventions such as brain abscess drainage, and even caesarean section under anaesthesia, there is limited evidence available on anaesthetic management in orthopaedic procedures among these patients [12-15].

We report the case of a 2-year-old boy with Goldenhar syndrome undergoing right Achilles tendon tenotomy for correction of congenital equinus contracture. However, there are no published reports specifically detailing anaesthetic management for equinus contracture correction or Achilles tendon tenotomy in this population.

This case illustrates the importance of integrating a thorough preoperative assessment with a carefully devised anaesthetic plan made suitable to the child's unique anatomical characteristics. By highlighting our perioperative approach from induction through emergence and detailing how faced anticipated airway challenges using videolaryngoscopy, this case report is made to highlight practical considerations that can guide anaesthesiologists when managing similar paediatric patients with craniofacial syndromes. Thus, this case emphasizes that, with meticulous planning and utilization of advanced airway techniques, even children with complex syndromic presentations can safely undergo routine orthopaedic procedures.

CASE PRESENTATION

A 2-year-old male child,11 kg body weight was brought to our tertiary care hospital by his parents for the surgical correction of a congenital right foot deformity, diagnosed as equinus contracture, for which a right Achilles tendon tenotomy was planned. The child was a known case of Goldenhar syndrome, diagnosed in early infancy based on clinical features with no familial tendency, and was under regular paediatric follow-up. The parents reported that the child had difficulty placing the right foot flat on the ground and was unable to walk with proper balance, prompting orthopaedic referral and recommendation for surgical intervention.

The child was born at term by normal vaginal delivery, with a birth weight of 2.6 kg. There was no history of birth asphyxia, NICU admission, or neonatal complications. Antenatal scans had detected mild facial asymmetry, but no gross structural anomalies were reported at birth. Developmental milestones were appropriate for age. There was no family history of congenital syndromes or facial anomalies. The child had no history of previous surgeries, hospital admissions, seizures, cyanotic spells, or recurrent respiratory infections. Immunizations were up to date as per the national immunization schedule.

Physical Examination

On general examination, the child was active and playful, with normal anthropometric parameters. The patient had various features of Goldenhar Syndrome like right-sided facial asymmetry, microtia (Grade II, right side), mandibular hypoplasia and micrognathia and mild deviation of the mouth and nasal base toward the right (figure 1).

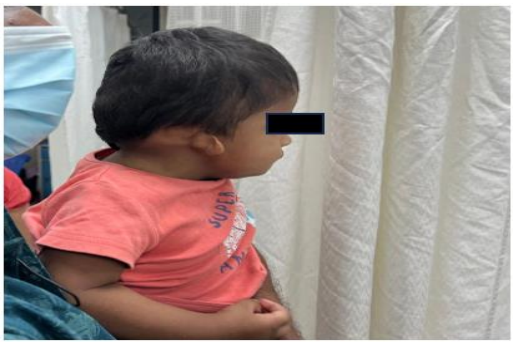

Figure 1: Craniofacial anomalies in a 2-year-old child with Goldenhar syndrome showing right-sided microtia, facial asymmetry, and mandibular hypoplasia.

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The airway assessment was challenging due to poor cooperation, but obvious micrognathia and limited mandibular space indicated an anticipated difficult airway. Neck mobility was within normal limits, and there was no obvious cervical spine abnormality or restriction. Cardiac and respiratory examination were clinically normal. Abdominal and neurological examinations were within normal limits.

The lower limb examination revealed tightness and shortening of the right Achilles tendon, limiting ankle dorsiflexion, consistent with equinus deformity. No neurovascular deficit was noted in the limb.

Laboratory Investigations

Routine blood investigations were conducted as part of the preoperative work-up. Complete blood count, serum electrolytes, blood urea, and serum creatinine were found to be within normal limits. The coagulation profile, including prothrombin time (PT), activated partial thromboplastin time (aPTT), and international normalized ratio (INR), was also within normal limits.

Diagnostic Procedures

To assess for systemic anomalies commonly associated with Goldenhar syndrome, a series of targeted diagnostic evaluations were performed. Echocardiography revealed normal cardiac structure and function, with no evidence of septal defects, valvular abnormalities, or outflow tract obstructions. Renal ultrasonography demonstrated bilaterally normal kidneys, with no signs of hydronephrosis, malrotation, or structural malformations. In addition to this a spinal X-ray and cervical spine screening were conducted, which showed no vertebral anomalies or cervical spine instability.

Pre-Anaesthetic Assessment

In the pre-anaesthesia clinic, the child was evaluated. On immediate preoperative assessment, the child was otherwise healthy without any history of fever or upper respiratory tract infections. Given the facial dysmorphism and airway-related anatomical challenges, a difficult airway was anticipated. The Mallampati classification could not be reliably assessed, but based on the external features (micrognathia, facial asymmetry), airway difficulty was expected.

The planned anaesthesia approach was general anaesthesia with endotracheal intubation using videolaryngoscopy. A difficult airway cart was prepared in advance, including alternate airway devices (laryngeal mask airway, bougie, and fibreoptic bronchoscope). The case was discussed with the surgical and paediatric teams, and written informed parental consent was taken after discussing risks and management of difficult airway with the parent.

Intraoperative Course

On the day of surgery, the child was brought to the operating room fasting and in good general condition. Standard ASA monitors were applied, including ECG, non-invasive blood pressure (NIBP), pulse oximetry, and capnography.

The child was preoxygenated with 100% oxygen for 3 minutes. Intravenous induction was achieved using propofol (2.5 mg/kg) and fentanyl (2 µg/kg). After confirming adequate mask ventilation, neuromuscular blockade was administered with atracurium (0.5 mg/kg). Tracheal intubation was performed using a videolaryngoscope, which significantly improved glottic visualization. A 4.5 mm uncuffed endotracheal tube was successfully inserted under direct vision, and correct placement was confirmed by bilateral chest auscultation and capnography.

Anaesthesia was maintained with sevoflurane (2%) in a 50:50 air-oxygen mixture. Intraoperatively, IV paracetamol (15 mg/kg) was administered for analgesia. Vital signs remained stable throughout the 30-minute procedure. There was no significant blood loss, and fluid management was minimal.

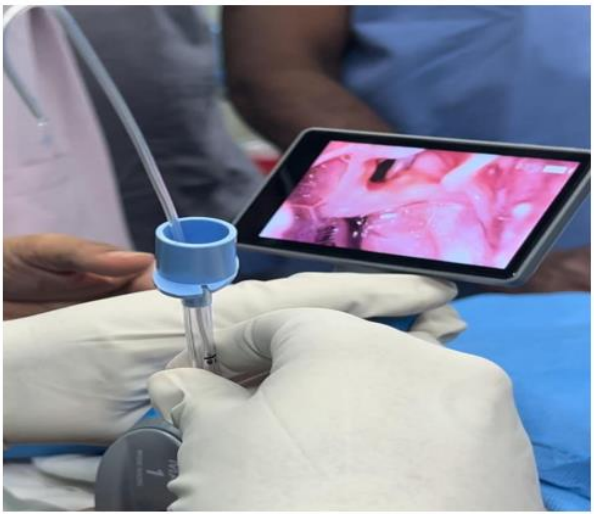

Figure 2: Intraoperative use of paediatric videolaryngoscope showing improved glottic visualization.

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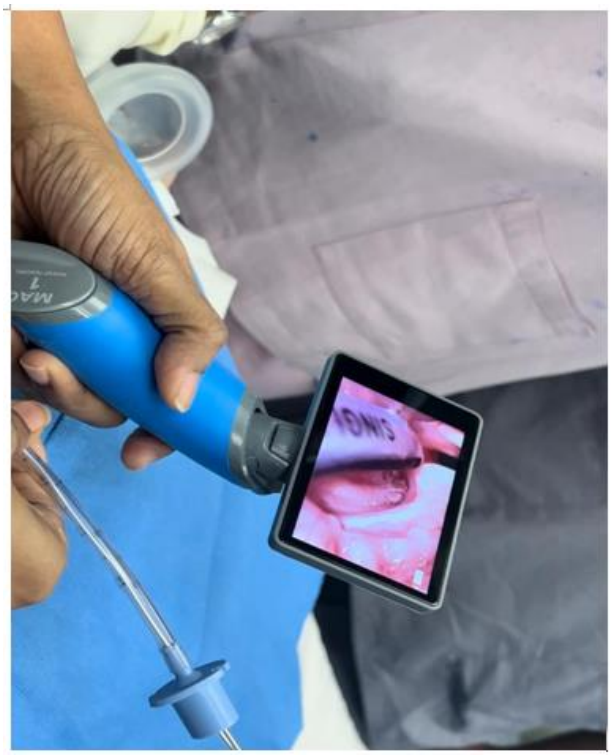

Figure 3

Postoperative Course

At the end of surgery, neuromuscular blockade was reversed using neostigmine (0.05 mg/kg) and glycopyrrolate (0.01 mg/kg). The child resumed spontaneous breathing, responded to verbal commands, and was successfully extubated once fully awake. Post-extubation, the child maintained good oxygen saturation and airway tone.

The patient was transferred to the post-anaesthesia care unit for observation. Recovery was smooth and uneventful. Pain was well controlled, and the child resumed oral intake after 4 hours. The patient was sent to the ward the same evening, and subsequently discharged home on postoperative day 2 in stable condition.

DISCUSSION

In Goldenhar syndrome, a multitude of systemic and anatomic anomalies converge to create a uniquely challenging perioperative environment[6,7]. While the literature suggests a male predominance, our patient's presentation right-sided facial asymmetry, mandibular hypoplasia, and microtia aligns with the classic craniofacial manifestations of Goldenhar syndrome[4,16]. However, many published cases demonstrate bilateral involvement or more extensive vertebral and ocular anomalies[17,18]. The absence of these systemic defects in our patient highlights the phenotypic variability of the syndrome and emphasizes the necessity for individualized assessment and management in each case.

Isolated equinus contracture in Goldenhar syndrome is conspicuously underrepresented in the literature. Equinus deformity defined by restricted ankle dorsiflexion due to a shortened gastrocnemius-soleus complex has been documented primarily in conjunction with neuromuscular disorders (e.g., spina bifida, cerebral palsy)[19]. Its occurrence as a standalone musculoskeletal anomaly in Goldenhar syndrome appears exceptionally rare. In this context, our patient's developmental milestones were age-appropriate and there was no evidence of neuromuscular disease, distinguishing his profile from more complex orthopaedic cases. This case therefore broadens the known phenotypic spectrum and alerts clinicians that Goldenhar syndrome may occasionally present with isolated limb deformities, not merely the classic craniofacial and vertebral anomalies.

Congenital heart disease affects about 35% of cases and vertebral deformities about 40% in Goldenhar syndrome [20]. Our thorough preoperative screening including echocardiography, renal ultrasonography, and cervical spine imaging revealed no abnormalities.

Airway management represents the cornerstone of anaesthetic planning in Goldenhar syndrome, where micrognathia, mandibular hypoplasia, and facial asymmetry greatly increase the risk of difficult mask ventilation and failed intubation [6].

Videolaryngoscopy was used, providing visualization to a Grade I–II view and enabling successful placement of a 4.5 mm uncuffed endotracheal tube on the first pass. Compared with fibreoptic intubation long considered the gold standard in difficult paediatric airways but hindered by the need for deep anaesthesia and technical complexity, videolaryngoscopy offers a wider angle of view, less requirement for neck manipulation, and shorter intubation times, thereby reducing the risk of desaturation [21,22]. The intraoperative stability of our patient's hemodynamic and absence of desaturation or bradycardia further support videolaryngoscopy's efficacy as a first-line tool in this setting. Sugino et al. reported a 5-year-old girl with Goldenhar syndrome who was a candidate for emergency surgery. Because of the difficult intubation, he intubated her by using video laryngoscope and reported it as a safe means [21].

Our induction regimen consisted of intravenous propofol (2.5 mg/kg) and fentanyl (2 µg/kg), followed by atracurium (0.5 mg/kg), consistent with protocols reported in similar patient cohorts [6]. Sevoflurane maintenance in a 50:50 oxygen–air mixture was likewise chosen for its rapid titratability, minimal airway irritation, and cardiovascular stability critical

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considerations in syndromic patients who might harbour occult cardiac disease [24]. In the paediatric population, the utilisation of an inhalational induction is favoured over intravenous induction, we elected intravenous induction because mask ventilation was confirmed to be easy despite facial asymmetry, minimizing the risk of airway loss during inhalation [25].

Recognizing the potential for airway obstruction, we prepared a comprehensive difficult airway cart containing a paediatric videolaryngoscope, fibreoptic bronchoscope, supraglottic airway devices (including SLMA), and emergency tracheostomy equipment. If primary videolaryngoscopy failed, fibreoptic-guided intubation was planned as the next step, with SLMA insertion as a rescue strategy.

During surgery, the child was placed on a flat table with a soft pad under the ankles to keep the neck straight and reduce the risk of spinal injury. A mid-thigh tourniquet provided a bloodless field; inflation time was limited to under 20 minutes to reduce the risk of ischemia reperfusion injury. Bleeding was minimal (<5 mL) and did not require transfusion, highlighting the importance of careful monitoring during even minor procedures in syndromic children.

Meticulous interdisciplinary planning and comprehensive preoperative investigations played a crucial role in anticipating and managing potential challenges, which contributed significantly to the patient's uneventful postoperative course. Continuous monitoring in the PACU ensured early detection of any airway compromise or respiratory distress, which did not occur.

This case shows some important points to keep in mind when giving anaesthesia to children with Goldenhar syndrome. Goldenhar syndrome exhibits significant phenotypic variability, occasionally manifesting as isolated orthopaedic anomalies; comprehensive preoperative screening including echocardiography, renal ultrasonography, and cervical spine imaging is indispensable but may reveal no systemic involvement, which, in turn, refines anaesthetic priorities; videolaryngoscopy can be considered a first-line advanced airway modality in syndromic paediatric patients, given its efficiency and high success rates; anaesthetic induction techniques must be made suitable to individual airway assessments, balancing the risks of inhalational versus intravenous approaches; and even seemingly routine orthopaedic procedures in syndromic children demand meticulous interdisciplinary planning to prevent potential complications.

CONCLUSION

Goldenhar syndrome presents significant perioperative challenges due to its phenotypic variability and associated multisystem anomalies. Comprehensive multisystem screening was performed to exclude occult cardiac, renal, or vertebral defects, thereby confirming that the primary perioperative concern would be airway management. Anticipation of a difficult airway led us to choose videolaryngoscopy as our first-line intubation technique, which provided a clear glottic view and enabled first-pass success. Anaesthetic induction with propofol and fentanyl, followed by atracurium, and maintenance with sevoflurane ensured stable hemodynamics and adequate analgesia throughout the procedure. Meticulous preparation and interdisciplinary coordination were important strategies to manage potential challenges effectively. The child's uneventful postoperative recovery further shows that with careful assessment and individualized anaesthetic planning, children with complex craniofacial syndromes can safely undergo routine surgical procedures.

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