

Anaesthesia Management in a Paediatric Patient with Goldenhar Syndrome and Hemifacial Microsomia Undergoing Distraction Osteogenesis

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Abstract: *This case report discusses the perioperative management of an 8-year-old girl diagnosed with Goldenhar syndrome and hemifacial microsomia, undergoing distraction osteogenesis of the right mandible. The patient presented with significant craniofacial and systemic anomalies, including congenital fusion of cervical vertebrae, cross-fused ectopic kidney, and a dysplastic temporomandibular joint. The surgical and anesthetic management emphasized careful preoperative assessment, airway management, and intraoperative monitoring to address the complexities associated with this rare condition. Postoperative care included ICU observation, ensuring a stable recovery. This report highlights the multidisciplinary approach required for managing Goldenhar syndrome, focusing on anesthesia and surgical considerations.*

Keywords: Goldenhar syndrome, Hemifacial microsomia, Paediatric anaesthesia, Distraction osteogenesis

1. Case Report

8 years old, weighing 25 kg, presented with Goldenhar syndrome and hemifacial microsomia. The planned surgical procedure was distraction osteogenesis of the right side of the mandible.

Birth and Development History: Full-term delivery via LSCS due to non-progress of labor (NPOL). NICU admission for 28 days due to intestinal obstruction. History of neonatal jaundice and pneumonia at 6 months, both managed conservatively. Febrile seizures occurred twice at 11 months and 2 years of age, with the last episode at 2 years.

Past Surgical History: Jejunojejunostomy with Ladd Procedure, Inversion Appendectomy, and Excision of Polydactyly (8/6/15): Postoperative ICU admission for 28 days. Excision of Skin Tag of Left Preauricular Region (22/6/15). Modified Woodward Procedure and Morcellation of Clavicle (18/4/18). Transcatheter Closure of PDA (3/7/16). Repair of Right Angular Cleft, Excision of Preauricular Tag (7/12/16).

Preoperative Assessment: Airway: Adequate mouth opening, Mallampati class II. Dentition: Loose lower right premolar, rest intact. Physical Examination: Deviation of the tongue to the right and right angle of the mouth. Investigations (31/3/24): Hemoglobin 13.1 g/dL, ALP 287 U/L, routine workup within normal limits. 2D ECHO (3/4/24): Normal biventricular systolic function, trivial TR, PDA device in LPA with no residual flow. CT Face (31/3/24): Facial cranial disproportion, dysplastic and hyperplastic right temporo-mandibular joint, hypertrophied bilateral inferior turbinates, congenital fusion of C2-C3 and C5-D1 vertebrae. USG Abdomen (1/4/24): Cross fused ectopic kidney on the right side.

Perioperative Management: Premedication: Midazolam 0.05 mg/kg and glycopyrrolate 0.004 mg/kg. Induction: Fentanyl 2 mcg/kg, propofol 2 mg/kg, atracurium 0.5 mg/kg. Intubation with uncuffed ET tube size 5 using videolaryngoscope. Maintenance: Atracurium and

sevoflurane. Intraoperatively, dexamethasone 2 mg, hydrocortisone 20 mg, and 100 ml packed cell volume were administered. Extubation: Neostigmine 0.05 mg/kg and glycopyrrolate 0.008 mg/kg. Postoperative Care: Patient was admitted to ICU for further observation and was shifted to the ward after 2 days.

2. Discussion

Goldenhar syndrome, also known as oculo-auriculo-vertebral (OAV) spectrum, is a rare congenital condition characterized by craniofacial anomalies, including hemifacial microsomia, microtia, and vertebral defects. It is associated with anomalies of the eyes, ears, and spine, and can also involve cardiac, renal, and central nervous system abnormalities. The etiology is believed to involve both genetic and environmental factors, although the exact cause remains unknown. This case report describes the anaesthetic considerations and perioperative management of an 8-year-old female patient undergoing distraction osteogenesis of the right mandible.

3. Summary

An 8-year-old with Goldenhar syndrome and hemifacial microsomia underwent distraction osteogenesis of the right mandible. She had multiple previous surgeries, cardiac and renal anomalies, and a challenging airway. Careful perioperative management focused on securing the airway, addressing comorbidities, and vigilant postoperative monitoring.



References

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