Airway management in Goldenhar syndrome: See it big, Keep it simple!!

Sir,

Goldenhar syndrome (GS) is a rare congenital disease caused by abnormal development of first and second branchial arches, causing unilateral maxillary and mandibular hypoplasia along with vertebral anomalies resulting in limitation of neck movement. It can pose significant challenges in airway management.[1]

We report the case of an 8-month-old, 6 kg, male infant, who was scheduled to undergo cataract surgery in the left eye. The mother gave a history of gestational diabetes when she was pregnant with this child. On examination, the patient had various features of GS

such as micrognathia, retrognathia, malformed left ear and facial asymmetry [Figure 1]. His developmental milestones were delayed. He had limited movements of the head and neck, which suggested the possibility of difficult laryngoscopy and intubation. His two-dimensional echocardiography showed a small atrial septal defect (4 mm).

A written informed parental consent was taken. All preparations were made for an anticipated difficult airway (DA) including a video laryngoscope and a fibreoptic bronchoscope. After 6 h, the child was transferred to the operation theatre and standard monitors were attached including pulse oximetry, non-invasive blood pressure and electrocardiogram. The child was not premedicated in view of the anticipated DA. Inhalational induction was initiated with 8% sevoflurane in an air–oxygen mixture. Venous access was then established and 1 μg/kg fentanyl was administered. An I-gel#1.5 was inserted after attaining adequate jaw relaxation. Anaesthesia was maintained with oxygen: air (1:1) and sevoflurane 2–3%, targeting a minimum alveolar concentration (MAC) of 1–1.3. At the end of an uneventful surgery, the child was awakened and extubated in the operating theatre.

GS, also known as oculo-auriculo-vertebral dysplasia, was first described by Maurice Goldenhar in 1952. The incidence of this syndrome ranges from 1:3500 to 1:45000 live births with a male: female preponderance of 3:2. The clinical manifestations of this syndrome include ear anomalies, hearing loss, epibulbar dermoids, upper eyelid colobomas, subconjunctival lipomas, unilateral facial hypoplasia, micrognathia, cleft palate, congenital heart disease, renal anomalies, mental retardation and vertebral abnormalities. This syndrome poses a challenge to the anaesthesiologist in terms of difficult mask ventilation and intubation due to facial asymmetry, micrognathia, cleft palate, unilateral facial hypoplasia and malocclusion. The limitation of neck movement due to vertebral abnormalities makes intubation difficult. In a paediatric patient, cardiac anomalies, mental retardation and secondary respiratory problems further compound the problem. Hypothyroidism is common and may lead to unusual delays in extubation. Further, anatomical abnormalities of the bronchial system, such as the tracheal bronchus, have been described. In one case, there was a tracheal bronchus that ended blindly; the endotracheal tube entered this blind pouch leading to the desaturation of the patient and cancellation of the surgery.[2] Fibreoptic intubation may be preferable in such cases. C-MAC video laryngoscope has also been used for these patients.[3] Although the use of video laryngoscope and fibreoptic intubation has been described, it can be difficult and is associated with high failure rates.[4] Computer-generated virtual imaging of the airway can generate video output similar to the views obtained from a fibrescope and may be useful in these cases.[5] In view of a DA, maintaining spontaneous breathing remains a vital technique in the anaesthetic management. Since both mask ventilation and intubation can be difficult, it may be advisable to manage the airway by insertion of a supraglottic device (SGD) as a primary airway device, similar to what we did in our case. There is still limited literature regarding the use of SGD in such patients.[6] This technique led to a favourable outcome in our patient despite the anticipated DA.

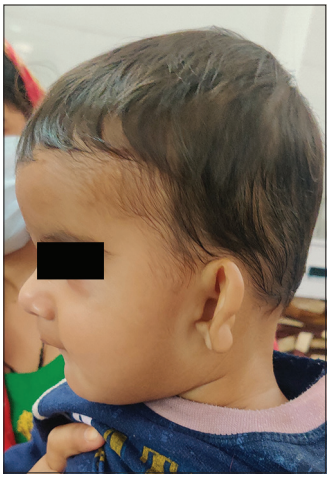

Figure 1: Clinical image showing features of Goldenhar Syndrome

To summarise, awareness regarding this rare syndrome and the nuances of its anaesthetic management may be vital in patients with GS.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

Ranju Singh, Pooja Singh[1], Anshu Gupta, Kumar Kaustubh[1]

Department of Anaesthesia, Lady Hardinge Medical College and Associated Shrimati Sucheta Kriplani Hospital, New Delhi, [1]Department of Anaesthesia, Lady Hardinge Medical College and Smt Sucheta Kriplani and Kalawati Saran Children's Hospital, New Delhi, India

Address for correspondence:
Dr. Pooja Singh,
Department of Anaesthesia, Lady Hardinge Medical College and Smt Sucheta Kriplani and Kalawati Saran Children's Hospital, New Delhi, India.
E-mail: dr.pooja260887@gmail.com

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