Anesthetic management for surgery of esphagus atresia in a newborn with Goldenhar’s syndrome

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Abstract

Background and objectives: Goldenhar’s syndrome is a polymalformative condition consisting of a craniofacial dysostosis that determines difficult airway in up to 40% of cases. We described a case of a newborn with Goldenhar’s syndrome with esophageal atresia and tracheoesophageal fistula who underwent repair surgery.

Case report: We report the case of a 24-h-old newborn with Goldenhar’s syndrome. He had esophageal atresia with distal tracheoesophageal fistula. It was decided that an emergency surgery would be performed for repairing it. It was carried out under sedation, intubation with fibrobronchoscope distal to the fistula, to limit the air flow into the esophagus, and possible abdominal distension. Following complete repair of the esophageal atresia and fistula ligation, the patient was transferred to the intensive care unit and intubated under sedation and analgesia.

Conclusions: The finding of a patient with Goldenhar’s syndrome and esophageal atresia assumes an exceptional situation and a challenge for anesthesiologists, since the anesthetic management depends on the patient comorbidity, the type of tracheoesophageal fistula, the usual hospital practice and the skills of the anesthesiologist in charge, with the main peculiarity being maintenance of adequate pulmonary ventilation in the presence of a communication between the airway and the esophagus. Intubation with fibrobronchoscope distal to the fistula deals with the management of a probably difficult airway and limits the passage of air to the esophagus through the fistula.

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Introduction

Goldenhar’s syndrome (GS) or oculo-auriculo-vertebral dysplasia was first described by von Arlt in 1845.1 This is a polymalformative condition resulting from defects of the first and second branchial arches consisting of craniofacial dysostosis, where we observe abnormalities of the eyes and cervical vertebrae, malformations of the ears and unilateral microtia, facial asymmetry, micrognathia,1 epibulbar dermoid cysts1 and other changes of the oral cavity, such as cleft palate and lingual change.1,4 Although the incidence of GS is estimated at 1 in 5000 live newborns,2,4 the association of esophageal atresia (EA) and this condition only occurs in 5% of these patients,1 and is an exceptional combination.

Clinical case

We present a case of a 24-h-old newborn of 3.2 kg of weight with left unilateral microtia (Fig. 1), bilateral ureterocele and esophageal atresia with distal tracheoesophageal fistula (TEF). An emergency surgery was chosen to repair the EA and associated fistula. On examination of the airways, mild micrognathia and difficult opening of the mouth were found. As supplementary preoperative tests, a chest X-ray with nasogastric tube was performed, which confirmed the diagnosis (Fig. 2), along with blood count, biochemistry and coagulation study, transthoracic echocardiography and electrocardiography, which showed no changes.

In the operating room monitoring was performed with non-invasive blood pressure, pulse oximetry (SpO2), and electrocardiogram. Under sedation with 2% sevoflurane, 0.1 mg IV atropine through a venous access with epicutaneous catheter was given to reduce oropharyngeal secretions, as well as 10 µg fentanyl. With the patient under spontaneous ventilation the fibrobronchoscope was introduced by mouth up to the opening of the glottis, and a rigid tube of 3.5 mm in diameter was inserted, being positioned distal to the TEF and near the main carina. The proper placement of the endotracheal tube (ETT) was confirmed by capnography and pulmonary auscultation, starting volume-controlled ventilation with a tidal volume of 8 ml kg−1 and respiratory rate between 17 and 20. To maintain the anesthetic, the concentration of sevoflurane was increased to 3% and 1 mg cisatracurium in doses of 6–9 µg of fentanyl were given, according to the need for painkillers.

Figure 1 Patient with Goldenhar syndrome that has left unilateral microtia.
that Cameron Haight performed the first surgical correction of this problem. Surgery to repair EA and TEF has important anesthetic implications and is a challenge for anesthesiologists. Often, newborns may have respiratory diseases and associated cardiac malformations that, along with a weight of less than 2 kg, are risk factors for postoperative mortality and should be assessed prior to surgery.10,11

The main feature of the anesthetic management in this surgery is the maintenance of adequate ventilation in the presence of a communication between the airway and the esophagus, and desaturation episodes may occur during induction and during anesthetic maintenance. On the other hand, the performance of a thoracotomy may determine the presentation of hemodynamic and respiratory changes, and difficult maintenance of an appropriate level of analgesia.10 The anesthetic management depends on patient comorbidities, the type of EA with or without TEF, the usual hospital practice, and the skills of the anesthesiologist in charge.

Newborns with TEF are at increased risk of gastric distension with potential risk of pneumoperitoneum12 which can increase with the introduction of mechanical ventilation with positive pressure. During repair of TEF there are several alternatives for the maintenance of mechanical ventilation, such as left bronchial selective intubation,11 assuming the risk of atelectasis, and intubation with fibrobronchoscope-guided placement of ETT distal to the fistula, which was the technique used in this case.11 Both forms of ETT placement require close monitoring by the anesthesiologist in charge to detect a bad positioning of the tube during the procedure. Another technique that could be used is the occlusion of the fistula using a Fogarty catheter until its complete ligation.12

It is recommended that spontaneous ventilation is maintained through inhalation induction without neuromuscular blockade nor positive pressure ventilation until the correct placement of the ETT is achieved distal to the fistula to avoid gastric hyperinflation. Other authors recommend maintaining spontaneous ventilation to the complete closure of the fistula to minimize gastric distension associated with positive pressure ventilation, with some anesthesiologists even recommending the performance of a gastrostomy before anesthetic induction to decompress the stomach and improve mechanical ventilation.11,12

In the case of GS, the finding of a DAW associated with EA with TEF could seriously impair oxygenation in these patients during anesthetic induction, significantly increasing the risk of respiratory complications that are already high due to the surgical procedure itself.

**Conflicts of interest**

The authors declare no conflicts of interest.

**References**