Abstract. – Children with congenital anomalies such as Goldenhar syndrome affecting the airway can be a problem for the anaesthesiologist. We present the case of an 18 month-old child with Goldenhar syndrome, in whom the ProSeal™ Laryngeal Mask Airway was successfully used for inguinal hernia surgery.

Key Words: Goldenhar syndrome, Laryngeal mask airway, Anaesthetics management.

Introduction

Airway and respiratory complications are the most common causes of morbidity during general anaesthesia in children. Patients with congenital anomalies affecting the airway can be a problem for the anaesthesiologist1.

Goldenhar syndrome also known as oculoauriculovertebral dysplasia is a rare congenital defect characterized by facial asymmetry and hypoplasia, micrognathia, hypoplastic zygomatic arch, external ear malformations, hearing loss, vertebral anomalies and ocular dermoids or lipodermoids. Other oral cavity anomalies may include a high-arch or cleft palate and abnormalities of the tongue2. The cause of Goldenhar syndrome seems to be heterogeneous. Most cases are sporadic. However, familial cases and concordant monozygotic twins have been reported, suggesting a genetic origin. In addition, several chromosomal abnormalities have been documented in affected individuals3,4. Tracheal intubation can be difficult because of mandibular hypoplasia, craniovertebral anomalies, and hemifacial microsomia. In addition, the difficult airway in Goldenhar syndrome typically becomes progressively worse with increasing age5. The choice of technique must be age- as well as airway-appropriate. Muscle relaxants should be avoided until the airway has been secured6.

The Laryngeal Mask Airway (LMA) was developed by Dr Archie Brain in the United Kingdom in the early 1980s. It can be used as a rescue airway and can be used during failed intubation scenario to maintain oxygenation and anaesthesia. It can also be used most effectively as a conduit to facilitate fiberoptic intubation6. The ProSeal™ Laryngeal Mask Airway (PLMA – Laryngeal Mask Co, Ltd, Henley-on-Thames, UK) is a modified LMA which has both an increased depth of bowl and a wedge shaped dorsal cuff to improve airway seal. It has an oesophageal drain to reduce gastric distension during positive pressure ventilation7.

Case Report

An 18 month-old, 15-kg, boy previously diagnosed with Goldenhar syndrome was scheduled for right inguinal hernia surgery. A detailed medical history was recorded based on available medical records and interview of the patient’s family. The patient had several features of Goldenhar syndrome including cleft palate, left mandibular hypoplasia, micrognathia, an epibulbar and limbal dermoid of the left eye, preauricular tags. He had no vertebral anomalies or cardiac disease. The patient’s neck extension was limited. Blood tests and chest radiography were normal.

The patient was not premedicated. Standard non-invasive monitoring was established. After allowing the patient to breathe 100% oxygen for 5 minutes, induction of anaesthesia was performed...
with voluntary inhalation of an increasing concentration of sevoflurane via a paediatric circle system. An intravenous catheter was then placed. After loss of consciousness, sevoflurane was adjusted to 2-3% and the same concentration of sevoflurane was maintained for several minutes until adequate jaw relaxation was attained for a PLMA. A size 2 PLMA was inserted without difficulty using the technique described in the LMA instruction manual. Anaesthesia was maintained with 2% sevoflurane in 60% nitrous oxide and 40% oxygen, and intermittent boluses of fentanyl (10 µg total dose) patient was ventilated with simultaneous intermittent mandatory ventilation. No muscle relaxant was administered. Two to 3 minutes before the completion of surgery, the concentration of sevoflurane was decreased to 0.5% and 1 minute prior to the completion of surgery, administration of anesthetic gases was discontinued and the patient was ventilated with 100% oxygen. LMA was removed when the patient started breathing spontaneously and returned protective reflexes. The patient’s operative and anaesthetic courses were uneventful, and he left the operating room fully awake, calm, and without nausea. His postanaesthetic course in the recovery room and patient ward was unremarkable, and he was discharged from the hospital the following day.

Discussion

Maintenance of a patent airway is the most important aspect of safe administration of anaesthesia. Craniofacial abnormalities often make airway management difficult. Preoperative evaluation of children with craniofacial abnormalities begins with evaluation of the upper airway, and formulation of a plan for airway management. Adequate preoperative planning and having the appropriate apparatus available are keys to success.

The principle of managing the difficult airway in children is to maintain spontaneous ventilation until the airway is secure. A spontaneous ventilation technique retains some muscle tone in the upper airway and allows the anaesthesist time to use alternative equipment. The use of an inhalational technique is favoured in paediatric practise. Airway muscle tonus is always decreased during inhalational induction and some degree of obstruction frequently occurs. A chin lift or a jaw thrust may be helpful. Some children continue to obstruct despite a correct technique. In these children, it is useful to apply assisted ventilation with continuous positive airway pressure (CPAP), 10-15 cm of water, by slightly squeezing the ventilation bag during the expiration phase, thereby expanding the airway. We maintained spontaneous ventilation by mask using carefully titrated sevoflurane until the appropriate level of anaesthesia was achieved.

 Awake intubation may be impossible because of a lack of cooperation in infants and children and also difficult airway devices are not available in paediatric patients at all times. For paediatric patients, fiberoptic intubation is more difficult and requires a higher level of speciality. Because of this patients are uncooperative with medical interventions which can produce a very challenging situation for the clinician. The LMA has been successfully used to manage difficult airways as a ventilatory device by itself and as a conduit for tracheal intubation. However, there are well known limitations classic LMA in paediatric patients. One of the main concerns is that the low-pressure seal may be inadequate for positive pressure ventilation (PPV), resulting in a risk of gas leakage into the stomach with the subsequent risk of gastric distension and regurgitation. This could put the patient at risk of pulmonary aspiration. The PLMA, a new LMA with a modified cuff and an oesophageal drainage tube, were shown to form a more effective seal, than the classic LMA as well as enable gastric tube placement in children. It is believed that these two features can contribute to improved protection against aspiration. It can also determine the correct positioning of the mask. In the current case, the insertion of a PLMA was a good option.

There are previous reports of anaesthesia in patient with Goldenhar syndrome pointing out adversities with airway management. A retrograde intubation method was used successfully in a 5-month-old infant with Goldenhar syndrome by Cooper and Murray–Wilson. A laryngeal mask airway insertion and fiberoptic intubation via the laryngeal mask method used one with Goldenhar syndrome by Bahk et al.

Conclusion

The PLMA may constantly be a good alternative when the anaesthesiologist with confined resources is faced with a child with congenital airway dimorphisms.
Airway management using the ProSeal™ laryngeal mask airway in a child with Goldenhar syndrome

References


