Management of Anesthesia in Goldenhar Syndrome: Case-Series Study

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ABSTRACT
Goldenhar syndrome or oculo-auriculo-vertebral dysplasia (OAVD) is characterized by a wide range of congenital anomalies including ocular, auricular, facial, cranial, vertebral and cardiac abnormalities. It is associated with the anomalous development of the first and second branchial arches. Patients with this syndrome usually suffer from unilateral maxillary and mandibular hypoplasia and vertebral anomalies which often result in limitation of neck movement. For this reason, intubation is very difficult in these patients and anesthesiologists usually face difficulty in airway management. Newborns with this syndrome often have premature internal organs, low birth weight and airway disorders. As a result, a safe anesthesia in such patients requires a complete knowledge regarding the metabolism and side effects of anesthetic drugs.

Our first case was a preterm 28-day old female with a fetal age of 28 weeks, birth weight of 1,200 g and gestational age of 32 weeks. At the time of admission, she weighed 1,500 g. She developed jaundice shortly after birth for which she underwent exchange transfusion. She was hospitalized in NICU for 14 days. Our second case was a 2 kg, 20-day old newborn with a fetal age of 37 weeks. Our third case was a full term 10-month old infant weighing 8 kg and our forth case was a 14 kg, 29-month old child who was a candidate for emergency surgery of dermoid cyst and bilateral upper lid coloboma.

Anesthesia induction was performed by inhalation anesthesia with N2O/O2= 50% and sevoflurane (0.5-3.5%); and in BIS (Bispectral Index) = 43, the appropriate size LMA (laryngeal mask airway) was inserted.

Anesthesia was maintained by using N2O/O2=50% and sevoflurane (1-2.5%). The BIS value was maintained in the range of 42-47. The surgical operations lasted for about 60-150 minutes. Patients regained consciousness 5 minutes after the completion of surgery and were transferred to the recovery room with stable vital signs. They were transferred to the ward 90 minutes later. Patients were discharged from the hospital the next morning with no post-up complications. (Tanaffos 2009; 8(4): 43-50)

Key words: Goldenhar syndrome, Prematurity, Anesthesia, Difficult intubation, Dermoid cyst, Lid coloboma, Sevoflurane
INTRODUCTION

Goldenhar syndrome or oculo-auriculo-vertebral dysplasia (OAVD) is characterized by a wide range of congenital anomalies including ocular, auricular, facial, cranial, vertebral and cardiac abnormalities. It is associated with the anomalous development of the first and second branchial arches (1-3). Although the etiology of this disease is not fully understood, autosomal recessive or dominant inheritance is possible. The disease occurs as sporadic cases and has a male predominance. Male/female ratio is 2:1(4); however, this ratio has reported to be 3:2 in one study. The average incidence rate of this syndrome is estimated to be 1/5,600 live births. This rate has reported to be between 1/3,000 and 1/5,000 live births in different studies (2).

Anomalies in patients with this syndrome include pre-auricular skin tags (90%), hemifacial microsomia (77%), vertebral anomalies of different size and shape (70%), microtia (52%), CNS malformations (47%), cardiac malformations (39%), epibulbar dermoid cyst (39%), unilateral maxillary hypoplasia, ocular anomalies such as lid colobomas, microphthalmia and corneal hypoesthesia, and genitourinary anomalies (5). This disease is associated with facial asymmetry and hypoplasia, micrognathia, external ear malformation, deafness and ocular dermoid cysts. Oral cavity malformations include high arch or cleft palate and tongue abnormalities (2,3,6). These deformities (especially ocular and palpebral abnormalities) often require surgical corrections under general anesthesia. Difficult intubation is expected in patients with this syndrome because of facial and oral abnormalities especially mandibular hypoplasia and limitation of neck movement resulting from vertebral anomalies. Anesthesiologists usually face difficulty in airway management of these patients. Therefore, maintaining an open airway is especially important in such patients (1,3,7-10). Since most patients are premature, they have significant health problems like post-up apnea, hypothermia, decreased metabolism of drugs, low weight and immature respiratory control (1-4). Therefore, it is recommended to avoid administration of parenteral drugs. In these patients, the method of anesthesia can affect the incidence of respiratory depression. Incidence of post-operative apnea is 12% in preterm infants who undergo a minor surgery. Therefore, post-op monitoring of respiration is necessary in these cases (1-4). Incidence of post-op apnea is associated with the gestational age (GA) and birth weight (11-14). Infants with the post conceptual age (PCA) of less than 44-46 weeks should be hospitalized for a minimum of 12 hours for respiratory monitoring after the operation (11,12). Six to 12 hours of respiratory monitoring is sufficient for infants with the PCA of 46-60 weeks and there is no need for further hospitalization. Elective surgery should be postponed until the PCA of 60 weeks (11-13). A surgical technique with minimum complications and a method of anesthesia with quick recovery time should be selected for such patients.

Case 1

Our first case was a 1,500 g, 28-day old newborn resulted from a twin pregnancy. She was conceived through in-vitro fertilization (IVF method) and delivered by C-section, with a birth weight of 1,200 g and fetal age of 32 weeks with negative familial history. She developed jaundice shortly after birth and underwent an exchange transfusion. She was hospitalized in NICU for 14 days because of respiratory distress as a consequence of preterm birth. She was fed with a formula designed for premature babies. Two weeks after discharge from NICU, she underwent an emergency surgery for correction of deformities of the eyes and eye lids.

Physical examination revealed several anomalies
including preauricular skin tags, nasal polyp, malformation of eyelids, cataract and microstomia. Hemoglobin was 11.5 g/dl, Hct was 32% and kidney and brain were normal in ultrasonography. The infant was pale and cachectic with the body temperature of 36.8°C and heart rate of 135 beats/min. The patient was NPO 4 hours prior to the operation. To prevent hypothermia in the patient, operating room temperature was set at 24°C and patient’s extremities, chest area and head were covered by surgical drapes. An intravenous line was started by inserting a 24 gauge catheter and 10cc/kg (15 cc) D5/S (dextrose 5% in normal saline) IV fluid was administered freely before the induction of anesthesia. The patient was under constant precordial, ECG, pulse oximeter and BIS monitoring. Anesthesia was induced by using inhalation anesthesia method with Mapleson F system, N2O/O2=50% and sevoflurane considering the patient’s condition (anemic, cachectic, and premature with difficult airway).

Sevoflurane was started with 0.5% concentration which increased by 0.25% after every 3-4 breaths. After 1-2 minute(s) when sevoflurane concentration reached 3% and HR was 152/min, we hyperventilated the patient for 15 seconds and then LMA (laryngeal mask airway) #1 was inserted for the patient while the pupils were midline, BIS was 43 and HR was 143/min. To make sure the LMA is inserted correctly with no air leakage, 2 cc of air was injected into the LMA cuff. The bag had a good compliance in ventilation. At this time, the operation was started and no parenteral drug was used during the procedure. Since the patient was stable, the other eye was also operated on. During the operation, dosage of sevoflurane (1-2.5%) was repeatedly adjusted based on heart rate, brachial pulse and precordial and BIS monitoring. Another 20 cc of D5/S IV fluid was administered during the operation. Patient’s respiration was assisted the whole time (30-35/min). 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 (BIS=90-97). LMA was removed when the patient started breathing spontaneously 1-2 minutes later. The patient regained consciousness and was transferred to the recovery room 4-5 minutes later while administering oxygen by mask. EKG and SPO2 monitoring and observation of breathing were started for her. After about 30 minutes, 15 cc of 1/3 2/3 IV fluid was administered and 30 minutes later she was fed her specific formula briefly. Since no complication was noted, she was transferred to the ward half an hour later. She was visited by a physician 2 and 4 hours later and she was discharged from the hospital the next day in good and stable condition.

Figure 1. First case, a 28-day old new born with Goldenhar syndrome.

Case 2

Our second case was a 2 kg, 20-day old female infant born by vaginal delivery with a fetal age of 36 weeks. Physical examination revealed several facial, ocular and palpebral anomalies, preauricular and nasal skin tags, an asymmetric face, mandibular hypoplasia and micrognathia. Cardiac and pulmonary auscultations were normal. Method of anesthesia selected for this patient was similar to the one chosen for our first case. The operation took about 90 minutes.
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Case 3

Our third case was an 8 kg, 10-month old baby girl with Hb=10 g/dl and Hct=29% born full term by C-section. She was anemic and had several facial, nasal and mandibular anomalies. Duration of operation was 135 minutes and method of anesthesia was similar to that of previous cases.

Case 4

The forth case was a 14 kg, 29-month old child with a history of surgery of bilateral choanal atresia at birth. Physical examination revealed several facial, nasal, ocular and mandibular anomalies, mandibular hypoplasia and micrognathia. The operation took about 90 minutes to complete. The same method of anesthesia was used for this patient as well.

DISCUSSION

Goldenhar syndrome or oculo-auriculo-vertebral dysplasia (OAVD) is characterized by a wide range of congenital anomalies including ocular, auricular, facial, cranial, vertebral and cardiac abnormalities. Mandibular hypoplasia and vertebral anomalies often result in limitation of neck movement (5-7, 9-11). During preoperative evaluation, a good method for airway management should be selected considering the patient’s special condition (12,13). Difficult mask
ventilation (13) has also been reported in these patients due to the lateral or mid-cheek extension of soft tissue cleft in the affected side (7,13). It was difficult to manage the anesthesia with a face mask for the whole duration of operation. Therefore, we used LMA for airway management in our cases. Okuyama in 1994 reported the anesthetic management of a 9 and a 14-year old with Goldenhar syndrome. He performed fiberoptic bronchoscopy-aided tracheal intubation under general anesthesia and controlled ventilation. Anesthesia and surgery went uneventfully (1). We did not use any parenteral drug considering the difficult airway and prematurity of our cases and also to prevent any trauma to the airway. We used pure inhalation with sevoflurane and N₂O/O₂ for the induction of anesthesia. In an optimal depth of anesthesia regulated by the BIS monitor, LMA was inserted for the patient. The operation took about 2.5 hours without any problems.

Cardwell and Walker in 2003 evaluated the difficult airway management in infants and children. In their study, they emphasized on the important role of LMA in the management of difficult pediatric airway. They considered sevoflurane as the drug of choice for the induction of anesthesia. They also avoided using any muscle relaxant in these patients (7).

As mentioned earlier, we used sevoflurane for induction of inhalation anesthesia and avoided using any parenteral drug. LMA was inserted under BIS monitoring.

Shukry et al. in 2005 reported the airway management of an 8-year old child with Goldenhar syndrome. He was a candidate for mandibular distraction surgery. He had a history of difficult tracheal intubation and laryngoscopy. Orotracheal intubation was not successful. By using #2.5 LMA an ideal condition for positive pressure ventilation was met and then by the application of Shikani Optical Stylet (SOS), tracheal intubation was performed successfully on the first attempt (Figure 5.) (8). Due to the successful insertion and placement of LMA, there was no need for endotracheal intubation in our cases.

Figure 5. The pediatric Shikani Optical Stylet.

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 (10).

Sukhupragarn in his case report in 2008 used a flexible laryngeal mask airway (FLMA) for the airway management of a 10-year old child with Goldenhar syndrome who was a candidate for eye surgery. He preferred FLMA over regular LMA and even tracheal intubation. We used LMA in our cases and inserted it without any trauma to the airway and we recommend this method (9). One of the advantages of LMA is its easy and smooth removal with no coughing or gagging; whereas, tracheal extubation in difficult airway patients causes serious problems for the patients and is usually associated with coughing and gagging which should be avoided following eye surgery (13,15). The most important point in these patients is to achieve the proper depth of anesthesia and using adequate dosage of inhalation drugs for insertion and removal of LMA. If these conditions are not met, LMA cannot be placed
properly and there would be a great risk for laryngospasm (13,15,17).

In these patients, it is crucial to stabilize the heart rate in order to maintain the cardiac output. Sevoflurane is able to do so and that is why this inhalation anesthetic was the drug of choice in our cases (17-23). Because of the difficult airway, it is necessary to maintain spontaneous breathing in these patients (11). Therefore, we avoided intravenous infusion of drugs (i.e. muscle relaxants, narcotics, local anesthetics) (24,25) to reduce the risk of post-operative apnea to a minimum (1-4). Considering the prematurity and low birth weight of our cases, sevoflurane was used for the maintenance of anesthesia because it has a quick recovery time (26-30).

Several studies have reported using fiberoptic bronchoscope, FLMA and video laryngoscope for maintaining an open airway in cases with Goldenhar syndrome. However, we had minimal manipulation or trauma to the airways and required the lowest dosage of anesthetic drug by using LMA. On the other hand, by using the BIS monitor for controlling the depth of anesthesia and regulating the dosage of sevoflurane accordingly (16,31), we prevented overdose or light anesthesia in our cases. As a result, perfect conditions were met for the surgery resulting in quick recovery of the patients (32).

Since eye is a very sensitive organ innervated by several nerve branches (33), depth of anesthesia in an eye surgery should be proportionate to the surgical stimulus and proper depth of anesthesia should be maintained throughout the operation. This goal was achieved by adding the BIS monitor to our standard monitors in the operating room (16,31,32).

CONCLUSION
- Premature and low birth weight patients have a higher risk of post-op apnea. In these patients, intravenous infusion or injection of any drug should be avoided. Also, post-op respiratory monitoring is necessary due to the immature respiratory control.
- Muscle relaxants should not be used in difficult airway cases. In these patients, it is necessary to maintain spontaneous breathing and for an elective surgery, proper insertion of LMA in an appropriate depth of anesthesia by an expert is recommended. In case of proper insertion, there would be no need for intubation and further trauma to the patient’s airway.

To achieve the adequate depth of anesthesia for insertion and removal of LMA and preventing drug overdose, it is necessary to use BIS monitor; because the risk of overdose is high in these ill, premature and low weight patients. Sevoflurane is the anesthetic drug of choice in these high risk patients because it has a better hemodynamic stability compared to other inhalation drugs. It maintains the cardiac output by stabilizing the heart rate and preventing bradycardia. It also has a quick recovery time which is one of the important goals of anesthesia management in high risk patients. Regular feeding starts in no time following quick recovery of the patient.

REFERENCES


