

# Airway Management of Patients With Craniofacial Abnormalities: 10-year Experience at a Teaching Hospital in Taiwan

Ying-Lun Chen\*, Kuo-Hwa Wu

*Department of Anesthesiology, Mackay Memorial Hospital, Taipei, Taiwan, R.O.C.*

**Background:** Airway management for patients with craniofacial abnormalities poses many challenges. It potentially has a high rate of morbidity and even mortality.

**Methods:** We reviewed our experience in administering anesthesia to patients with a diagnosis of mucopolysaccharidosis or Pierre Robin sequence in the past 10 years (July 1998 to October 2008). The anesthetic procedures, methods of airway management, and events of morbidity and mortality were evaluated.

**Results:** Thirty patients with mucopolysaccharidosis and 53 patients with Pierre Robin sequence were placed under general anesthesia. Although the anesthesiologists always encountered difficulties in managing the airway, most of the anesthetic procedures were performed safely except for 1 case that resulted in mortality.

**Conclusion:** Managing the airway of patients with craniofacial abnormalities can potentially be difficult. It should be carried out by experienced anesthesiologists, with assistance from an otolaryngologist when necessary. A variety of different airway devices should be available if needed. [*J Chin Med Assoc* 2009;72(9):468–470]

**Key Words:** airway, endotracheal intubation, laryngeal mask airway, mucopolysaccharidosis, Pierre Robin sequence

## Introduction

Maintenance of a patent airway is the most important aspect of safe administration of anesthesia. Craniofacial abnormalities often make airway management difficult. The exact challenge depends on the specific abnormality, which includes certain congenital conditions.

The mucopolysaccharidoses (MPS) are a group of genetic disorders characterized by deficiencies in enzyme production that lead to accumulation of mucopolysaccharides throughout the body. Patients have macrocephaly, a prominent forehead, and short neck. Infiltration by mucopolysaccharide deposit in the airway may result in tongue enlargement, thickening and redundancy of the oropharyngeal mucosa, as well as blockage of nasal passages. These abnormalities, along with stiff temporomandibular joints and an anteriorly positioned larynx, make mask ventilation and endotracheal intubation extremely difficult.<sup>1–4</sup>

Pierre Robin sequence (RS) is a congenital malformation characterized by micrognathia and glossoptosis. Cleft palate is often present. These abnormalities may cause severe respiratory and feeding difficulties in the newborn. The condition is most severe at birth and tends to improve with age.<sup>5</sup> Tracheal intubation in these infants may be very difficult or even impossible.

We retrospectively analyzed our experience at Mackay Memorial Hospital in administering anesthesia to patients with MPS or RS.

## Methods

The anesthetic records of patients who received anesthesia between July 1, 1998 and October 31, 2008 at Mackay Memorial Hospital in Taipei, Taiwan, a tertiary teaching hospital, were reviewed. We selected the records of all patients with a clinical diagnosis of MPS



\*Correspondence to: Dr Ying-Lun Chen, Department of Anesthesiology, Mackay Memorial Hospital, 92, Section 2, Chung-Shan North Road, Taipei 104, Taiwan, R.O.C.

E-mail: alexingchaning@yahoo.com.hk • Received: December 23, 2008 • Accepted: July 13, 2009

**Table 1.** Patient characteristics and surgical procedures\*

	MPS (n = 30)	RS (n = 53)
Sex (M/F)	27/3	47/6
Age (yr)	12.3 (5–25)	6.85 (3 d–20 yr)
Weight (kg)	37.73 (28–56)	22.9 (3–60)
Hernia repair	5	4
Bronchoscopy	1	17
Myringotomy	8	10
Tonsillectomy	4	
Gastrostomy	10	8
Ventricle-peritoneal shunt	2	2
Tracheostomy		4
Palatoplasty		8

\*Data presented as n or mean (range). MPS = mucopolysaccharidoses; RS = Pierre Robin sequence.

or RS. Data extracted from the records included the operative procedure, anesthetic management, and perioperative course. Regional anesthesia for the operations was excluded from further analysis.

## Results

General anesthesia was administered to 30 patients with MPS and to 53 with RS. An overview of the demographic data and surgical procedures is shown in Table 1. A variety of airway management techniques was used. One patient with MPS died—who was in fact the first patient with this disorder whom we managed. Intravenous induction was given, but the patient could then be neither intubated nor ventilated by mask. He became cyanotic and had a cardiac arrest. Tracheostomy was difficult because of his short neck, and required about 20 minutes to complete. He was resuscitated but died 2 days later. None of the remaining patients with MPS had this disastrous outcome, although manipulation of the airway was frequently difficult (Table 2). After this, we used inhalation induction or regional anesthesia whenever possible. Methods of intubation included laryngeal mask airway (LMA) and awake fiberoptic intubation.

In the RS group, there was only 1 serious event—in an infant—again after intravenous induction. Mask ventilation was very difficult, and the infant became cyanotic. Fortunately, we were able to pass a rigid bronchoscope immediately to secure the airway. As with the MPS group, there was some difficulty manipulating the airway in a number of cases (Table 2). The most useful devices in our experience were LMA or a rigid bronchoscope. Awake fiberoptic intubation and a lightwand were also used.

**Table 2.** Airway management for patients with MPS and RS undergoing general anesthesia\*

Airway management	MPS	RS
Intubation after intravenous induction	1 <sup>a</sup>	10 <sup>f</sup>
Intubation after inhalation induction	10 <sup>b</sup>	8 <sup>g</sup>
LMA placement after inhalation induction	11 <sup>c</sup>	5 <sup>h</sup>
LMA placement after intravenous induction		7 <sup>i</sup>
Rigid bronchoscopy after inhalation induction	1 <sup>d</sup>	
Rigid bronchoscopy after intravenous induction		17 <sup>j</sup>
Awake fiberoptic intubation	7 <sup>e</sup>	4
Intubation with lightwand after intravenous induction		2

\*Data presented as n. <sup>a</sup>Cardiac arrest occurred because the patient could neither be mask ventilated nor intubated after induction; <sup>b</sup>more than 2 attempts at intubation in 4 cases; <sup>c</sup>more than 2 attempts at LMA placement in 3 cases; <sup>d</sup>failure of rigid bronchoscopy placement; <sup>e</sup>more than 10 minutes required for successful fiberoptic intubation in 2 cases; <sup>f</sup>more than 2 attempts at intubation in 3 cases; <sup>g</sup>more than 2 attempts at intubation in 4 cases; <sup>h</sup>2 attempts at LMA placement in 2 cases; <sup>i</sup>2 attempts at LMA placement in 2 cases; <sup>j</sup>1 infant became cyanotic after intravenous induction because he could not be ventilated by mask ventilation, but a rigid bronchoscopy was rapidly and successfully placed. MPS = mucopolysaccharidoses; RS = Pierre Robin sequence; LMA = laryngeal mask airway.

## Discussion

This review of our experience with anesthesia in patients with certain craniofacial abnormalities indicates that the airway can generally be adequately managed. One must be prepared, however, to make repeated attempts to secure the airway if necessary.

Following the death of the first MPS patient we encountered, we avoided intravenous induction with muscle relaxants prior to endotracheal intubation. Thereafter, we maintained spontaneous ventilation by mask using carefully titrated inhalation anesthetic agents (halothane or sevoflurane) until the appropriate level of anesthesia was achieved, at which point tracheal intubation with direct laryngoscopy was performed. Movement of the glottis and the appearance of air bubbles during spontaneous ventilation helped guide placement of the endotracheal tube.

LMA and a flexible fiberoptic bronchoscope were also useful. Although the LMA has earned a place in the management of difficult airways, its role in MPS has not yet been defined.<sup>6,7</sup> We had some difficulty using it, with repeated attempts at placement necessary in 3 of 11 cases. However, an LMA is still a helpful temporary ventilatory device.

Awake fiberoptic intubation is more difficult in patients with MPS because most are also mentally retarded. They may become very agitated during intubation because they do not understand and have difficulty cooperating. The judicious use of sedative agents in MPS is therefore worth considering. Excessive secretions can add to the difficulty in these cases.<sup>4</sup> Emergency placement of a surgical airway is particularly difficult in these patients because of their short neck. In general, regional anesthesia is preferred if possible. If general anesthesia is unavoidable in a patient with MPS, the airway should be managed by experienced personnel.

In our patients with RS, ventilation by mask was generally not a problem except in 1 case. Either intravenous or inhalation induction was acceptable in our series. When visualization of the glottis was difficult, we found that an LMA, flexible fiberoptic bronchoscope, or lightwand were helpful adjuncts. Jones and Derrick described a 2-anesthetist technique for intubating a patient with RS.<sup>5</sup> One laryngoscopist manipulates the larynx with the right hand by pressing firmly below the cricoid cartilage under direct vision until the arytenoids is visible. The second anesthetist is then able to insert the endotracheal tube. We found this to be a very useful technique in 3 cases.

Rigid bronchoscopy was performed by an experienced otolaryngologist in 16 patients with RS to examine their airway. The patients needed to be anesthetized for successful bronchoscopy. After the scope was placed in the trachea, we connected the breathing tube to the side-hole of the scope to ventilate the patients. As mentioned above, 1 infant in this group could not be ventilated by mask after induction, but the otolaryngologist was able to place the scope immediately.

The LMA was useful in securing the airway in these patients. It can even be used in awake infants.<sup>8</sup> An endotracheal tube can be passed through it with the aid of a flexible fiberoptic bronchoscope.<sup>9</sup> Direct laryngoscopy is very difficult in patients with RS. An alternative is blind intubation using a lightwand, a lighted stylet passed into the endotracheal tube and then advanced toward the airway. If the lightwand is in the glottis, the bright glow can be seen easily through the soft tissue of the anterior neck. If it is in the esophagus, no transillumination is observed. Regular use of and practice with the lightwand have been reported to increase the rate of successful intubation.<sup>10</sup>

Our experience in managing the airway in patients with MPS and RS demonstrates that it can be done

successfully. The challenges posed by these craniofacial abnormalities are best handled by experienced personnel and team work. Adequate preoperative planning and having the appropriate apparatus available are keys to success.

Finally, we suggest some points for this difficult airway management: (1) have an organized plan; (2) remain calm; (3) call 2 or more senior colleagues for help early; (4) prepare several types of airway devices such as gum elastic bougie, LMA, lightwand and flexible fiberoptic bronchoscope; (5) endeavor to become expert in each new airway device and technique, and practice with a normal airway; (6) pre-oxygenate the patient with 100% oxygen for at least 5 minutes; (7) remember that the airway in MPS cases is the most difficult; (8) avoid use of muscle relaxant before successful intubation in MPS patients; (9) use awake fiberoptic intubation or intubate after inhalation induction in MPS patients; (10) the airways in RS patients become less difficult as they grow up; (11) use a laryngoscope for a quick look at the glottis under light sedation in RS patients; (12) if the glottis or epiglottis can be seen, the RS patient can almost certainly be intubated after induction; (13) ask a rigid bronchoscopist to stand by if necessary.

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