blockade was not reversed until just prior to transfer to the recovery room. With conversion from controlled ventilation, during which intrathoracic pressure is never less than atmospheric, to spontaneous ventilation, in which intrathoracic pressure is less than atmospheric during inspiration, the problem of venous air entrainment developed. Because the patient was transported to the recovery room without ECG monitoring, cardiac dysrhythmias were unrecognized until ECG monitoring was resumed in the recovery room.

Common causes of cardiac dysrhythmias such as hypoxia, hypercarbia, anemia, hypo- or hyperkalemia, and myocardial ischemia were immediately ruled out after arrival in the recovery room. Michenfelder has reported that common dysrhythmias associated with VAE are PVCs and bigeminy,[9] both of which occurred in our patient. However, only after excluding the common causes of dysrhythmias did we consider the possibility of VAE. The diagnosis of VAE was confirmed using a precordial Doppler. The precordial Doppler has been shown to be one of the most sensitive monitors for VAE.[10] Doppler sounds were most intense during inspiration and gradually cleared during expiration and between breaths. Although air was most likely being entrained by the patient during inspiration, changes in the intensity of Doppler sounds during the respiratory cycle may also have been due, at least in part, to positional changes of the Doppler relative to cardiac structures. Because air was no longer heard on the Doppler and the cardiac dysrhythmias resolved after the left anterior screw site was occluded, suggests that air entrainment occurred from scalp veins or diploic veins in this area.

This case demonstrates several important points. Episodes of venous air embolism are not restricted to the intraoperative period and can occur during spontaneous respiration in patients in the supine position. Also, one should consider VAE in the differential diagnosis of cardiac dysrhythmias in patients with exposed venous channels when other more likely etiologies have been ruled out. Finally, the importance of patient monitoring during transport is reemphasized.

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Fiberoptic Intubation Complicated by Pulmonary Edema in a 12-Year-Old Child with Hurler Syndrome

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Hurler syndrome (mucopolysaccharidosis storage disease, Type I-H) is an inherited progressive metabolic disorder.[1] The tongue, tonsils, adenoids, and nasopharyngeal tissues are hypertrophic in these children and may cause upper airway obstruction. Micrognathia, a very short neck, and restricted motion at the temporomandibular

joint may combine to make direct laryngoscopy difficult or impossible.[1,2] Hypoplasia of the odontoid and atlantoaxial subluxation may also occur.[3] In a recent report[3] it was also shown that direct laryngoscopy and intubation were easy in patients averaging 23 months, but that visualization of the vocal cords became difficult as children become larger and older.

Several authors[2,4] have suggested fiberoptic intubation for safe management of the airway in the older child with Hurler syndrome (HS). However, because of the mental retardation associated with this disease, the patient is likely to be frightened and uncooperative with this procedure. We could find only one report[4] of attempted fiberoptic intubation in the literature. The authors reported that during planned elective preoperative fiberoptic intubation in a 35-y-old woman with Hurler-Scheie syndrome, the patient suffered respiratory arrest induced by diazepam and fentanyl administered in preparation for intubation. Her lungs could not be ventilated *via* mask and bag and tracheostomy was required.[4] We now report a case of successful fiberoptic intubation in a 12-y-old child with Hurler syndrome with severe upper-airway obstruction. Even though the intubation was successful, postintubation pulmonary edema occurred. We believe that this was caused by forced inspiration against a closed glottis (Mueller maneuver) during endoscopy in a patient with decreased left ventricular function and mitral regurgitation.

CASE REPORT

The patient is a 12-yr-old, 25.6-kg female with Hurler syndrome. Her disease was manifest by dwarfism, with the characteristic facies: saddle nose, prominent brow, a large tongue, and limited mouth opening. Her vision was decreased due to corneal clouding and she was hard of hearing. Cardiac function, assessed by echocardiographic criteria, was markedly decreased with a shortening fraction of only 19% and mild mitral regurgitation. She had a history of recurrent cardiogenic pulmonary edema for which she was treated with furosemide and also paroxysmal atrial tachycardia treated with digoxin. Her liver and spleen were both enlarged causing her abdomen to be quite protuberant. Of note is the fact that her intelligence was well preserved.

She presented to our emergency room with an umbilical hernia containing incarcerated omentum. Dehydration, hypokalemia (K^+ = 3.2 mEq/l), and acidosis (HCO_3 = 15 mEq/l) were present and preoperative hydration and potassium replacement were instituted. Radiographic evaluation of the cervical spine demonstrated a hypoplastic odontoid with 5 mm of atlanto-axial subluxation (*i.e.*, instability present). After 15 h of rehydration (K^+ = 3.8; HCO_3 = 20) she was brought to the operating room for reduction of the incarceration and umbilical herniorrhaphy. Examination of the upper airway suggested that direct laryngoscopy for intubation was likely to be impossible. Furthermore, when she received thiopental and succinylcholine for another procedure 3 yr previously, her laryngeal inlet could not be visualized and the trachea was blindly intubated. Fiberoptic intubation was considered an option, however, especially since she was acceptably cooperative. The electrocardiogram and oxygen saturation (Sp_{O_2}) were monitored and a right radial arterial catheter was inserted. The patient was then sedated with atropine (20 μg/kg) and ketamine (0.4 mg/kg). The oropharynx was anesthetized with 4% lidocaine spray. Fiberoptic (Olym-

pus® 4-mm scope) intubation was then attempted with the patient spontaneously breathing oxygen. Several attempts were necessary to successfully enter the larynx. As the endoscope was introduced into the laryngeal inlet, the cords were seen to tightly oppose each other while the patient continued to make inspiratory efforts. Sp_{O_2} decreased to 70%. Eventually, her cords relaxed and the endoscope and a 4.5-mm (small size used because of anticipated mucopolysacchride related narrowing of trachea) endotracheal tube (ETT) were able to be passed through the larynx. It was immediately noted that she was difficult to ventilate, and frothy serous fluid was seen coming from the endotracheal tube. Continued positive pressure ventilation with 100% oxygen restored her Sp_{O_2} to 95%; with PEEP (10 cmH_2O) it improved to 100%.

After an otherwise uneventful repair of her umbilical hernia, controlled ventilation with PEEP was continued in the intensive care unit. A postoperative chest x-ray confirmed the diagnosis of pulmonary edema which improved with PEEP, mechanical ventilation, and furosemide. Sedation was discontinued 24 h later in preparation for discontinuing assisted ventilation and eventual extubation. As she became more alert, she also became increasingly agitated, fighting the ventilator and vigorously inspiring through the small ETT. Copious pink frothy fluid was again noted coming from the ETT. She was resedated and controlled ventilation reinstituted. A thermodilution pulmonary artery catheter was inserted to objectively assess her hemodynamic status. Steady state data revealed the pulmonary edema to be noncardiogenic in nature (CO = 3.5 l/min, PAWP = 11, RAP = 8, PAP = 32/23 mmHg). A bedside echocardiogram revealed that mitral regurgitation and cardiac contractility were unchanged from the preoperative study. However, because of the recurrent pulmonary edema, dopamine and dobutamine were begun to increase inotropy and nitroglycerine was administered for afterload reduction. Diuresis was continued with furosemide.

Two days later, while she was still moderately sedated, mechanical ventilation was discontinued and the trachea was extubated. Postextubation upper airway obstruction from copious secretions and redundant oropharyngeal soft tissue was treated with a nasopharyngeal airway, intermittent racemic epinephrine nebulization, oropharyngeal suctioning, and supplemental oxygen in helium. Anticholinergics were not used since the physical measures were adequate in clearing the airway.

Over the next 3 days, the vasoactive infusions were gradually discontinued and digoxin was reinstituted. She was discharged from the intensive care unit on her sixth postoperative day and from the hospital 2 days later.

DISCUSSION

Airway management in patients with Hurler syndrome can be very difficult. Perioperative mortality rates averaging 20%[5] have been reported for patients with this disease, with failure to control the airway as the largest single cause of mortality.[2]

Fiberoptic intubation has been suggested as an attractive alternative in those patients in whom direct laryngoscopy is either known to be impossible by history or strongly suspected to be so by physical exam.[2] However, successful intubation with this technique requires a cooperative quiet patient who will tolerate sedation and still maintain a patent upper airway. In a previous report[4] sedation resulted in total upper airway obstruction.

We report successful fiberoptic endoscopy for endotracheal intubation in a 12-y-old female with Hurler syndrome. Sedation with iv ketamine allowed spontaneous

respirations to continue, topical anesthesia with lidocaine to be applied, and introduction of the endoscope. However, the larynx and trachea were insufficiently anesthetized resulting in glottic closure during endoscopy. Bilateral superior laryngeal nerve and transtracheal anesthesia might have prevented this problem although this may have increased the risk from aspiration. Alternatively, topical spraying of the larynx through the bronchoscope prior to its introduction into the airway may have prevented glottic closure. The resultant pulmonary edema, after relief of airway obstruction, may have been aggravated by the patient's underlying cardiomyopathy. This case report highlights the problems accompanying fiberoptic endoscopy for intubation of children with Hurler syndrome.

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