CASE REPORTS

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Anesthetic Considerations for Patients with Larsen's Syndrome

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The association of multiple congenital dislocations of major joints and characteristic facial abnormalities were first recognized as a syndrome by Larsen and co-workers in 1950.[1] Children with Larsen's syndrome may be born with dislocations of the knees, hips, and elbows, as well as equinovarus deformities of the feet. They have widely spaced eyes, long cylindrical fingers, prominent forehead, and a depressed nasal bridge. Other associated defects include mental retardation, hydrocephalus, hearing loss, palate defects, abnormal segmentation of the cervical spine, kyphoscoliosis, spina bifida, tracheomalacia, and cardiovascular abnormalities.[2–5] Larsen's syndrome children undergo a wide variety of surgical treatments for their orthopedic abnormalities. Since their life span may be normal, it is important that both pediatric and adult anesthesiologists be aware of the anesthetic implications of Larsen's syndrome.

CASE REPORTS

Case 1. This 4-month-old full-term girl presented for operative treatment of congenital knee dislocation. The patient's abnormal facies, in association with multiple joint dislocations, allowed the diagnosis of Larsen's syndrome.

Preoperative examination revealed a vigorous 4.3-kg child. In addition to the characteristic facies of Larsen's syndrome, the child had a high arched palate and an extremely short neck. The hemoglobin and white blood cell counts were normal. Atropine 0.15 mg was given intramuscularly 45 min prior to the induction of anesthesia. Induction of anesthesia was accomplished using nitrous oxide, oxygen, and incremental doses of halothane. The airway was easily managed by mask. An intravenous catheter was inserted, and 0.5 mg pancuronium administered.

After muscle relaxation was complete, oral intubation was attempted but was found to be extremely difficult due to the short neck and relatively cephalad position of the larynx. After several attempts, visualization of the extreme posterior aspect of the glottis was accomplished using a #1 Miller blade and a right lateral approach to the larynx, along with external posterior laryngeal pressure. At the end of the surgical procedure, neuromuscular relaxation was antagonized and the trachea extubated without difficulty. The child had no difficulties during the recovery room stay and was discharged home 3 days later.

Case 2. This 23-month-old, 11-kg girl presented for halo application for immobilization of the cervical spine. At 18 months of age, cervical spine x-rays documented dramatic cervical kyphosis with hypoplastic vertebral bodies. The spinal canal was markedly narrowed at the C5 level. The child was treated conservatively with a soft cervical collar until 23 months of age. At that time, minor trauma sustained after falling from a chair resulted in transient quadriplegia lasting 3 h. Magnetic resonance imaging revealed hypoplastic cervical vertebral bodies causing kyphosis at the cervical level with C5–C6 dislocation (fig. 1). Preoperative examination of the child revealed an awake, alert patient with Larsen's facies and a soft cervical collar in place. The external appearance of the airway appeared normal, and the child was able to open her mouth widely. With the exception of orthopedic abnormalities of the extremities, the rest of the physical examination was normal. The white blood cell count, hematocrit, and urinalysis all were normal.

Thirty minutes prior to anesthetic induction, 0.3 mg atropine and 10 mg meperidine were administered intramuscularly. Anesthesia was induced with 45 mg ketamine intramuscularly. Following intravenous catheter insertion, anesthesia was maintained with nitrous oxide and oxygen administered by mask. There was constant observation of the patient's head position by both the anesthesiologist and the neurosurgeon to ensure that movements of the cervical spine were minimized. During the subsequent 3 months, the child was again anesthetized on three separate occasions for readjustment of the halo. Each time, the anesthetic was accomplished with ketamine as the primary anesthetic agent and without intubation.

Ten months after the original halo placement, the child underwent anterior spinal fusion. Anesthesia was induced with halothane, nitrous oxide, and oxygen. After the airway was ascertained to be easily managed, intravenous pancuronium was given. Intubation was accomplished without difficulty using a Miller #1 blade. At the end of the 6-h procedure, neuromuscular relaxation was antagonized and the trachea was extubated without difficulty. Two months later, with the child still in halo traction, posterior cervical fusion was accomplished without difficulty using a similar anesthetic technique.

Case 3. This 4-month-old, 5.6-kg girl presented for bilateral club foot reconstruction under general anesthesia. Cervical radiographs and computerized axial tomography showed hypoplastic vertebral bodies and narrowing of the cervical canal at the C5–C6 level. Although the child demonstrated no neurologic deficit, the neck was judged unstable, and the child was treated with a soft cervical collar. Anesthetic evaluation revealed an active child wearing a cervical collar. The external airway appeared normal. Examination of the mouth revealed a large cleft palate. Heart and lung examinations were normal. The hemoglobin and urinalysis were normal.

The patient received 0.2 mg atropine 60 min preoperatively. In the operating room, the cervical collar was removed, and the neck was stabilized manually in the neutral position. Induction was accomplished with nitrous oxide, oxygen, and halothane by mask with ease. Succinylcholine 10 mg was given intramuscularly. After adequate neuromuscular relaxation, an attempt was made to pass a 3.5-mm ID oral endotracheal tube. Although visualization of the larynx was not difficult, the 3.5-mm endotracheal tube would not pass the cricoid level easily. A 3.0-mm endotracheal tube subsequently passed with mild resistance. The anesthetic was maintained with halothane, oxygen, nitrous oxide,

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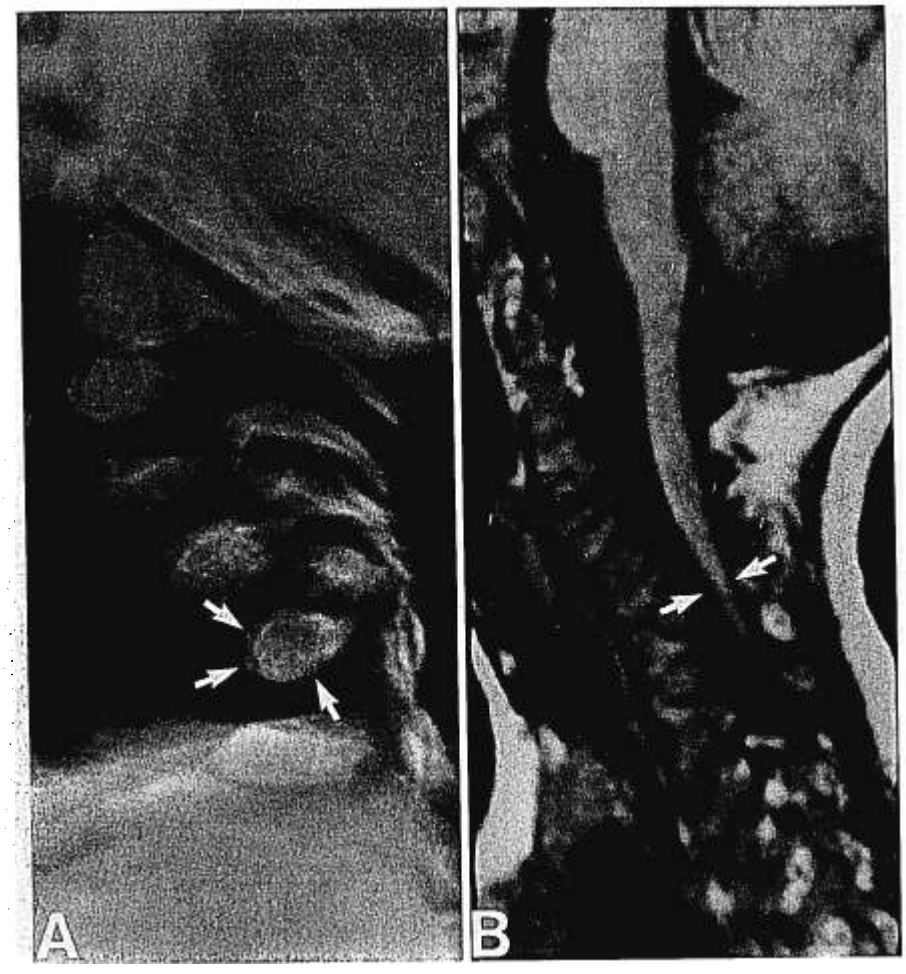

FIG. 1. Cervical spine radiograph (*A*) of patient 2, showing severe kyphotic deformity associated with hypoplastic vertebral bodies, most obvious at the C5–C6 level (*arrows*). Nuclear magnetic resonance imaging (*B*) of the same patient, showing posterior prolapse of C5 vertebra with resultant compression of the cervical canal and cord (*arrows*).

and pancuronium. At the end of the 4-h procedure, neuromuscular blockade was antagonized; the child was awakened; and the trachea was extubated.

The child returned 1 month later for open reduction of bilateral knee dislocations. In view of the airway difficulty noted during the prior procedure, endoscopic evaluation by an otolaryngologist also was planned. The child received atropine intramuscularly, followed by induction with halothane, nitrous oxide, and oxygen. The vocal cords and larynx were sprayed with 2% lidocaine, and the larynx was exposed with a slotted Holinger laryngoscope. Using a 2.8-mm telescope, the subglottis appeared narrow and abnormally elliptical in shape. The diagnosis of subglottic stenosis with an elliptical cricoid was made. A 3.0-mm oral endotracheal tube was placed. The anesthetic was maintained with halothane, nitrous oxide, oxygen, and pancuronium. Intravenous dexamethasone was given to decrease postmanipulation airway swelling. At the end of the 3-h procedure, the trachea was extubated. Only minimal croup was noted, requiring no treatment except cool mist. The child was discharged on the fourth postoperative day.

DISCUSSION

In 1950, Larsen *et al.*[1] described six patients suffering from a rare connective tissue disorder characterized by multiple large joint dislocations in association with a characteristic abnormal facies (depressed nasal bridge, bulging forehead, and widely spaced eyes). All six of these patients also had abnormal cylindrical shaping of the hands and phalanges, foot deformities, and palatal defects. Since its original description, several other abnormalities have been found to be associated with Larsen's syndrome (table 1).

Anesthetic management of patients with Larsen's syndrome has not been well documented; case reports have appeared only in nonanesthesia literature. It is of note that one of the children originally described by Larsen died on the evening of surgery from a "respiratory anesthetic complication."[1]

The three cases described in this report underscore some potential difficulties of airway management of children with Larsen's syndrome. Laryngeal visualization of one of the three children (case 1) described was extremely difficult. Only after multiple attempts could the trachea be intubated, using a lateral laryngeal approach. The entire larynx was never visualized. This appears to be the first description of difficult intubation associated with Larsen's syndrome. The other two cases here presented did not involve difficulty in laryngeal visualization; however, these patients also required caution during intubation, due to the potential for cervical subluxation. Cervical kyphosis secondary to hypoplasia of the vertebral bodies and associated mechanical instability frequently has been

TABLE 1. Larsen's Syndrome: Associated Features[1,3-15]

Airway
Characteristic facies* (depressed nasal bridge, prominent forehead, hypertelorism)
Cleft palate
Difficult intubation
Subglottic stenosis
Tracheomalacia
Bronchomalacia
Cardiothoracic
Congential heart disease (atrial and ventricular septal defects, patent ductus arteriosus, mitral valve prolapse)
Pectus excavatum
Musculoskeletal
Characteristic hands* (long, cylindrical fingers)
Cervical spine instability
Kyphoscoliosis
Spina bifida
Multiple joint dislocations (hips, knees)
Club foot
Neurologic
Hydrocephalus
Developmental delay
Hearing loss
Miscellaneous
Poor wound healing

* Universal features.

reported in Larsen's patients.[2,8,11] Quadriplegia and death have been reported after minor trauma.[2] In view of this, flexion–extension radiographic cervical spine evaluation should be performed on all Larsen's patients, with computerized axial tomography or magnetic resonance imaging as necessary. If cervical instability is documented, the neck should be maintained firmly in the neutral position throughout intubation efforts.

During induction of anesthesia in patients with Larsen's syndrome, muscle relaxants should be avoided until the ability to manually ventilate the lungs has been confirmed. Had manual ventilation been difficult after induction of our patients, muscle relaxation would have been avoided and laryngoscopy performed with the patient maintaining spontaneous ventilation. Muscle relaxants were used in these cases, only after ensuring that the lungs could be ventilated by mask, in order to provide optimal conditions for laryngeal visualization and eliminate potential for laryngospasm. Alternatives for intubation, including various blades, an Oxyscope, a Howland lock, a Bullard laryngoscope, a fiberoptic laryngoscope, and means for cricothyrotomy and transtracheal jet ventilation, all were readily available in the operating room suite but were not needed. Regardless of intubation technique used, the head and neck must be maintained in neutral position in cases with known cervical instability.

One child described had an abnormally shaped cricoid cartilage associated with subglottic stenosis. Even when an unusually small endotracheal tube was used, mild postoperative croup developed but responded to mist treatment. The degree of posttraumatic croup observed in this child possibly was lessened by the use of intraoperative steroids.[16]

There is significant variability in the precise type and degree of severity of defects seen in patients with Larsen's syndrome.[2,12] In view of the potential for multiple system involvement, caution should be exercised in the preoperative evaluation and anesthetic management of patients with Larsen's syndrome, and special attention should be paid to potential airway and neurologic problems, as well as orthopedic abnormalities.

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