

Whistling Face Syndrome: General Anesthesia and Early Postoperative Caudal Analgesia

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Cranio-carpotarsal dysplasia or Freeman-Sheldon syndrome is an uncommon congenital myopathy featuring abnormal facies with severe microstomia, camptodactyly with ulnar deviation, and talipes equinovarus.¹ Originally described by Freeman and Sheldon² in 1938, the term "Whistling Face" was applied by Burian³ in 1962 because of the peculiar facial appearance. These and other musculoskeletal and soft tissue deformities frequently require the attention of the surgeon. The anesthetic experience with this syndrome has been recently reported.^{4,5} The purpose of this case report is to further illustrate this interesting anomaly, discuss its implications for the anesthesiologist, and suggest a simple technique for early postoperative analgesia for operations on the lower extremities.

REPORT OF A CASE

A 7-month-old male infant with diagnoses of Freeman-Sheldon (Whistling Face) syndrome and sickle cell trait was scheduled for surgical correction of bilateral clubbed feet. At the time of admission for operation, the infant weighed 8.5 kg and had experienced normal growth and development. Characteristic dysmorphic features present included a large head with prominent cheeks and brow, a flat face with deeply recessed eyes and blepharophimosis, and a small mouth with the lower lip fixed in a tight, pursed position. The neck was short and its mobility was significantly limited. Mild pectus excavatum was present, and a faint systolic murmur was noted. Bilateral clinched hand deformities with digitalization of the thumbs were present. The spine was straight. Severe clubbed foot deformities were present bilaterally.

Screening laboratory studies, including electrocardiogram and echocardiogram, had unremarkable results. A cervical spine radiograph was normal in appearance, and the chest film showed an apparent increase in the transverse diameter of the ribs and a pectus deformity. The cardiodynamic silhouette and lung fields were normal.

In the operating room, the child was monitored with precordial

stethoscope, automated blood pressure device, electrocardiograph, temperature thermistor, and pulse oximeter. After intramuscular administration of atropine 0.1 mg, lidocaine was applied topically to the nostrils and upper airway. With the infant carefully restrained, attempts at awake blind nasotracheal intubation were made and were unsuccessful. During subsequent attempts at awake orotracheal intubation, the severe microstomia and immobile neck made visualization of the larynx impossible. Glimpses of the posterior portion of the arytenoids were briefly obtained, and tracheal intubation was accomplished by positioning a styletted endotracheal tube, curved at the tip, just anterior to these structures and advancing it when breath sounds were transmitted and coughing occurred. General anesthesia was then induced with halothane and nitrous oxide in oxygen 40%, and ventilation was controlled. A 22-G intravenous catheter was placed in the right antecubital fossa, and a maintenance infusion of dextrose 5% and 0.45% normal saline was begun. Additional intravascular volume requirements were provided by separate infusions of lactated Ringer's solution. The subsequent anesthetic course was uneventful. At the conclusion of the operation, the patient was carefully placed in a lateral position and 4 ml of bupivacaine 0.25% were introduced in sterile fashion into the caudal epidural space via a 23-G needle. Bilateral club foot splints were then applied, and the patient was taken to the recovery room while he was spontaneously ventilating with the endotracheal tube in place. His trachea was extubated when he was fully awake. Although specific dermatomal levels and weight bearing were difficult to assess in this infant, he appeared to be free of pain and maintained hemodynamic stability throughout his 2-h recovery room admission. No analgesics of any kind were required during this period. Convalescence was uncomplicated.

DISCUSSION

The three major clinical signs of cranio-carpotarsal dysplasia are microstomia with pursed lips, camptodactyly with ulnar deviation, and talipes equinovarus.¹ Other frequently associated findings include coloboma alae, convergent strabismus, blepharophimosis, high arched palate, flattened midface, kyphoscoliosis, and short stature.⁶ These are by no means always present, and in one instance⁷ the absence of microstomia was noted.

Studies of nerve conduction, electromyography, and muscle biopsy indicate that this syndrome is caused by a nonprogressive or slowly progressive myopathy.¹ Qualitative comparison of skeletal muscle fiber types shows structural changes that predominantly involve Type I fibers and suggests that the muscle lesion is a form of congenital fiber type disproportion. Facial, limb, and respiratory muscles are primarily affected. An autosomal dominant inheritance pattern is generally accepted,⁸ al-

Received from the Departments of Anesthesiology and Clinical Investigation, Naval Hospital, San Diego, California. Accepted for publication September 26, 1988.

The work reported herein was performed under the Navy Clinical Investigation Program, case report no. 84-16-1968-35.

The views expressed in this article are those of the author and do not reflect the official policy or position of the Department of the Navy, Department of Defense, nor the United States Government.

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Key words: Airway; anatomy; Freeman-Sheldon syndrome; Whistling Face syndrome. Anesthesia; pediatric. Intubation, tracheal; difficult.

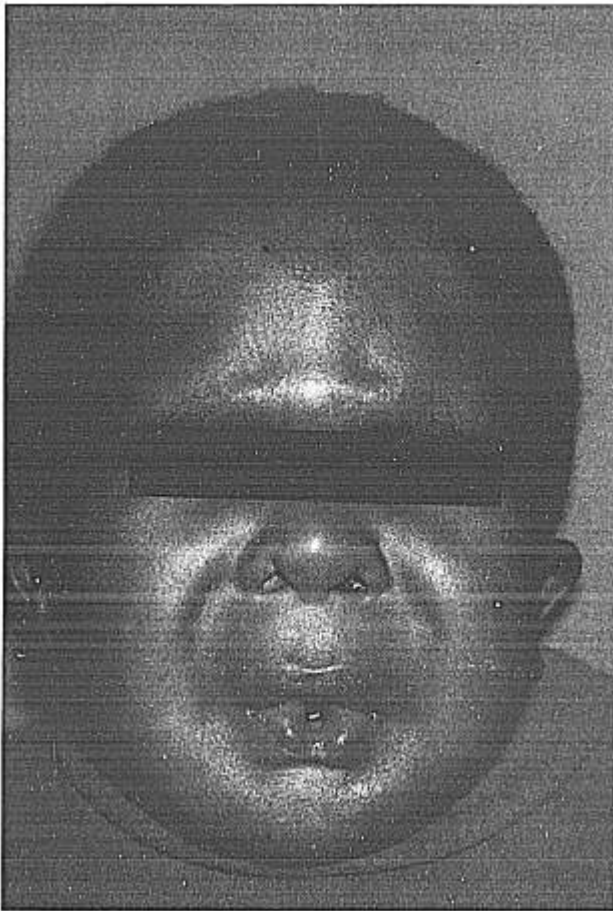


FIG. 1. Awake, full face view. Note the broad face, blepharophimosis, and pursed lower-lip deformity.

though there have been reports of a recessive gene,⁹ and isolated cases are considered to be new mutations.⁸ Intelligence is normal in most cases.⁸

The most striking feature and a source of major concern for the anesthesiologist is the severe microstomia. The oral intercommissural distance is significantly reduced, and the lips are fixed in a pursed or "whistling" position (fig. 1).⁶ This tight circumoral architecture results from diffuse fibrosis within the orbicularis oris muscle and from a dermal fibrous band along the vermilion border of the lower lip that prevents complete relaxation even with partial resection of the orbicularis oris.¹⁰ The presence of teeth in older children may further crowd an already inadequate oral aperture. An important point is that the circumoral architecture is not significantly changed by neuromuscular blockade,¹⁰ or general anesthesia (fig. 2), and the use of muscle relaxants should not be expected to improve intubating conditions. We chose an awake technique for tracheal intubation because we were unwilling to risk inducing general anesthesia in an infant whose airway might subsequently prove difficult to manage.

Although neither Laishley and Roy⁵ nor we encountered perioperative respiratory complications, a number of authors have reported significant pulmonary complications and mortality associated with this syndrome. Freeman and Sheldon² reported pneumonia and empyema after anesthesia and operation in their original case. MacLeod and Patriquin⁸ described a child in whom bronchopneumonia developed after anesthesia for an orthopedic procedure. Other authors^{1,11,12} refer to children who died of bronchopneumonia, respiratory insufficiency,

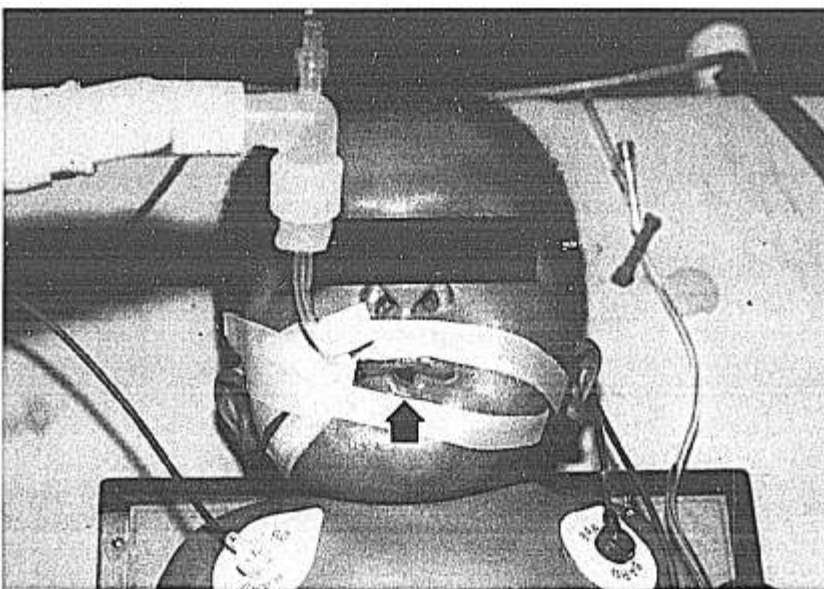


FIG. 2. After tracheal intubation and induction of anesthesia. Note that the circumoral architecture (arrow) is unchanged from that in the awake state.

or recurrent respiratory tract infections unrelated to anesthesia. The causes of respiratory difficulty in these patients are thought to involve intercostal myopathy¹³ and abnormal development of the bony thorax⁸ with loss of pulmonary volume. MacLeod and Patriquin⁸ studied their patient's respiratory cycle during a cinefluoroscopic procedure and noted an abnormal thoracic lordosis and a rigid, immobile thoracic cage, the diaphragm alone effecting tidal ventilation. On external physical examination, this patient had no detectable movement of his thorax. The combination of these myopathic and skeletal abnormalities, then, may predispose affected patients to significant postoperative respiratory difficulty.

Our decision to use caudal epidural local anesthetic for postoperative analgesia was based on our desire to avoid parenteral opioid in the early postoperative period, and it attained two goals: First, it eliminated the potential for interaction between opioid and residual volatile anesthetic and thus reduced the risk of somnolence after tracheal extubation. Second, it eliminated the depressant effect of opioid on pulmonary function and thus facilitated pulmonary toilet in the early postoperative period. The volume of local anesthetic administered, approximately 0.05 ml/kg dermatome, was calculated to produce a level of analgesia to the L1 dermatome after the method of Takasaki *et al.*¹⁴ Although we used bupivacaine 0.25%, a recent report¹⁵ indicates that bupivacaine 0.125% with epinephrine 1:200,000 yields equivalent analgesia with potentially fewer side effects. A continuous technique might be more efficacious, provided adequate supervision is available. Where appropriate (*e.g.*, for hand surgery), other forms of conduction anesthesia (*e.g.*, axillary block) might serve a similar function.

In conclusion, surgical procedures to correct the mouth deformity, as well as ophthalmologic, orthopedic, and other functional and cosmetic procedures, will continue to place the anesthesiologist in contact with affected children. Because the ability to manage the airway without an endotracheal tube in these children is uncertain, we felt more comfortable with awake endotracheal intubation before induction of general anesthesia. As an alternative to our method of awake intubation, fiberoptic nasotracheal intubation in infants has been described,¹⁶ and appears ideally suited for this situation. Many anesthesiologists will prefer an inhaled induction with halothane and oxygen followed by laryngoscopic examination and in-

tubation with the patient under general anesthesia. Each alternative has advantages, and management must be guided by the specific clinical circumstance. The risk of significant pulmonary complications makes conduction analgesia attractive as an alternative to opioids in the early postoperative period.

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