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Anesthesia for patients with cleft lip and palate

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Cleft lip and palate are common defects that may be a part of a syndrome or associated with other anomalies that are of real significance during anesthesia care. Accompanying facial or head and neck anatomical variations may result in difficulties with airway maintenance, endotracheal intubation, and postoperative ventilation, which can vary from mild to extreme. Concurrent congenital heart disease or other systemic illness may require special considerations during the perioperative period.

Cleft lip and palate are the most common of the craniofacial anomalies and are present in various combinations in as many as 1 of every 800 live born infants. Associated anomalies are common, especially in children with a cleft palate, and many of these have significant implications for the anesthesiologist. The incidence of associated anomalies has been reported to be 8% in patients with isolated cleft lip, 22% in patients with cleft palate, and 28% in patients with cleft lip and palate.[1] The more severe the cleft, the higher the incidence of associated anomalies. The most common associated anomalies affect the limbs or vertebral column (33%) and the cardiovascular system (24%). A very large number of named syndromes have also been described which include a cleft palate with/or without cleft lip. Some of these syndromes are very rare and are associated with limited survival (eg, Walker-Warburg syndrome); however, others are relatively more common (eg, Klippel-Feil syndrome). Many of these latter conditions are associated with an almost normal lifespan, and such patients very frequently present for anesthesia care. Craniofacial and head and neck anatomic anomalies associated with syndromic cleft lip and palate may create difficulties with airway maintenance and endotracheal intubation that vary from mild to extreme.[2,3] Associated anomalies in other major organ systems may require very careful peri-operative management if optimal outcomes are to be achieved. The anesthesiologist caring for cleft lip/palate patients should be very familiar with the anatomical and physiological disturbances that may be present in these patients.

The surgical approach to patients with facial clefts has evolved over the years, and some aspects have been controversial, especially those relating to the timing of repair operations. An enthusiasm for neonatal repair of cleft lip has waned, but the eventual possibility of fetal surgery to correct clefts is a continuing topic in experimental surgery, offering the attraction of minimal or no scarring. At the present time, the multidisciplinary cleft clinic has emerged as the optimal care provider, and the anesthesiologist has an important place in this team. Depending on the extent of the lesion, comprehensive surgical care for these patients might include early active orthopedic manipulation of the dental arch followed by lip repair, palate repair, a procedure for velopharyngeal incompetence, and later a maxillary advancement procedure (Lefort 1 operation). Dental interventions may be necessary as the incidence of dental caries is high and many children require prolonged orthodontic care.

The patient with cleft lip/palate

The neonate with cleft lip/palate has difficulty feeding from the time of birth; suckling and deglutition are impaired,

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and the infant may tire before a feed is completed. This commonly results in a poor nutritional state and anemia. In countries with plentiful medical and nursing care, nutrition can be improved by the use of modified equipment and feeding routines. Help for the parents and supervision of the patient's progress by nursing and dietetic professionals has been demonstrated to improve the weight gain and health of the infant.[4] Anemia may also be corrected relatively rapidly by appropriate oral iron therapy. However, many patients are underweight and some may still be anemic when they present for surgery, especially in regions with less than optimal medical care. Experience with general anesthesia for repair of clefts has suggested that peri-operative complication rates are not increased by mild degrees of anemia (Hb 8-10 G/dl).[5]

Chronic nasal and sinus infections are common in the child with cleft lip/palate, and these may predispose to lower respiratory tract infections and ear disease. Acute upper respiratory infection (URI) also seems to occur with increased frequency in these patients. The presence of URI at a carefully conducted preoperative evaluation is an indication to postpone surgery, especially in the patient with a more severe cleft. The incidence of perioperative respiratory complications has been reported to be significantly higher in patients with URI.[6]

The cleft lip/palate patient may have other congenital malformations; in one reported series, 78 of 284 patients (27.5%) had associated malformations,[7] most frequently involving the face (25.9%) or the cardiovascular system (16.5%). In another series of 220 cleft lip/palate patients, congenital heart disease (CHD) was present in 9.5%,[8] as compared with less than 1% in the general infant population. The presence of CHD did not correlate with the type of cleft; however, patients with a syndrome had a higher incidence of CHD (21.4%) compared with the others (8.5%).[8] The type of CHD present was most commonly a simple lesion (VSD, ASD, or mitral valve prolapse), but some had cyanotic CHD.

Syndromes are present in approximately 10% of patients with an isolated cleft palate or a cleft lip and palate.[9] A wide range of different syndromes are involved and some of these that have been described in patients coming for general anesthesia are listed in Table 1, together with the implications for their anesthesia care.

Preoperative evaluation of the patient

The general health of the patient must be assessed and the presence or absence of an associated condition or syndrome determined. The features of a syndrome may affect the course of anesthesia; in particular, if the face, head, and neck are involved, the ease of airway maintenance and endotracheal intubation.[2,3] Mandibular hypoplasia in the Pierre Robin sequence, Goldenhar, or Treacher Collins syndrome may result in extremely difficult airway maintenance after induction of anesthesia. Laryngoscopy and intubation may also be extremely difficult, and the deformity may predispose to postoperative obstructed ventilation. In patients with the Pierre Robin sequence, perioperative ventilatory difficulties are reported to occur more frequently in those who had neonatal difficulties with ventilation.[10] In those patients with an abnormality of the cervical spine (eg, Klippel Feil syndrome), the limitation of head movement may make positioning for intubation difficult. Patients with possible unstable cervical spine (eg, Down syndrome) should be positioned carefully to avoid neurologic damage. Every patient should be carefully examined to assess such conditions. The size and shape of the mandible, the extent of mouth opening, size, and mobility of the tongue, and the range of neck movement should be examined carefully. The Mallempati scoring system, predictive of difficult intubation of the adult, may be less reliable in children. The records of previous anesthesia administrations should be reviewed, but be careful not to place too much reliance on previous experience as far as the ease of intubation is concerned. This changes as the patient grows: for example, patients with isolated cleft palate tend to become easier to intubate as they grow older,[11] and patients with Goldenhar syndrome may become more difficult.

Congenital heart disease (CHD) may be present, especially in those children with an associated syndrome or with isolated cleft palate.[5] Some children will have been seen by a cardiologist and have their history well documented. Otherwise, evidence of CHD should be sought (small stature, cyanosis, murmurs, etc.), and if found, preoperative referral to a cardiologist for assessment (ie, echocardiography) is required. All children with CHD require prophylactic antibiotics even if the lesion has been repaired (exceptions: Isolated ASD, mitral valve prolapse without regurgiation, ligated PDA, closed ASD or VSD more than 6 months postoperative without residua). Children with cyanotic heart disease, surgical shunts, prosthetic valves, or conduits are at especially high risk for sub-acute bacterial endocarditis and accordingly should be carefully treated.

Infants and small children with cleft lip and palate frequently have colds and upper respiratory infections which may predispose to peri-operative respiratory complications.[6] Such complications are increased in those with more severe cleft deformities. In such a case, surgery should be deferred for 2 or 3 weeks until the child has fully recovered and the airway reactivity has returned to normal.

Management for dental procedures

The multidisciplinary approach to patients with a cleft may involve preliminary dental splinting and orthopedic maneuvers in the neonatal period. Impressions of the palate may be taken with the infant awake, but general endotracheal anesthesia will be required for the placement of dental orthopedic devices. This should be administered with due con-