Anesthesia for Pediatric Craniofacial Surgery

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Children with craniofacial abnormalities provide some of the most difficult challenges for anesthesiologists today. Whether the patient is scheduled for reconstructive surgery or a procedure unrelated to their anomaly, the care of these patients can be consuming and demanding. Since 1967 when the plastic surgeon Paul Tessier successfully demonstrated the correction of many deformities once thought to be uncorrectable, the field has continued to expand with new developments in both surgery and anesthesia. He found that the bones of the face and skull could survive as grafts when temporarily removed, remodeled and then replaced. It is important to understand the development and characteristics of the more common anomalies and their peculiar anesthetic challenges in order to construct a safe anesthetic plan.

Craniofacial abnormalities begin early in development. Fertilization and cell division are the initial embryological stages of development that eventually lead to the three divisional layers, the ectoderm, mesoderm, and endoderm. The mesoderm is the most influential to further development and specialization. It is the forerunner of the skull and upper face.

Facial prominences are formed from surface elevations of regional growth centers in the upper face. During the 30th to 37th gestational days, the bumps and valleys eventually form the nasal and oral cavity. The 50th through 60th gestational days consists of formation of the palatal shelves and eventually the hard and soft palate. Disturbances during the first phase give rise to facial clefts, while interruption in phase two results in cleft lips and palates.

Mesodermal bulges below the oral cavity are called branchial or visceral arches. The ectodermal grooves lead to branchial clefts and the endoderm will form branchial pouches. There are six branchial arches that will develop into the pharynx and oral cavity. The first arch is “involved” with development of the face, cranial nerve V, and the muscles of mastication. Malformations here often involve hearing loss, abnormalities in mouth opening and the mandible. The second arch involves development of the ear, the 7th cranial nerve and muscles of facial expression. The third arch involves the 9th cranial nerve and upper pharyngeal muscles. Fusion of the 4th and 6th arches forms all the laryngeal cartilages except the epiglottis, the pharyngeal and laryngeal muscles and the thyroid.

The first pouch forms the middle ear and eustachian tube, the 2nd forms the palantine tonsil; the 3rd forms the inferior parathyroid and thymus; the 4th forms the superior parathyroid.

The eye is formed by completion of neural tube closure, which was initially formed from the ectoderm. The tongue is complex development of mesodermal origin of the 1st and 3rd branchial arches. Tongue malformations are uncommon. The middle ear is formed during the 4th week of development. The external ear develops during the 3rd week.

Clefts occur due to a lack of fusion of facial prominences. They have an incidence of 1/1000 births. Tessier originated a numerical classification system for the various clefts. It uses the orbit as point 0, the reference point and
continues up to point 14. Point 7 is the most lateral. Number 0 cleft is known as telorbitism. Cleft number 3 is called oculonasal. Clefts number 6, 7, and 8 often occur together as a triad to form Treacher Collins Syndrome. Surgical correction of cleft lips and palates is usually done between 3 and 10 months of age. This is usually during the period of physiologic anemia of the infant. Excessive blood loss is uncommon. Close attention to placement of the throat pack and mouth gag by the surgeon is needed to avoid accidental extubation, advancement or compression of the oral endotracheal tube. Hand ventilation of the patient at this time is recommended. Post-operative swelling can cause difficulties, particularly in patients with Treacher Collins syndrome or Pierre Robin sequence as they already have compromised airways. Congenital heart disease is seen in up to 30% of patients with cleft deformities.

First arch malformations lead to hemifacial microsomia (Pierre Robin Syndrome) and mandibulofacial dysostosis (Treacher Collins Syndrome). Both are associated with cleft lips and palates as well as receding, narrow chins. Treacher Collins syndrome occurs in 1:10 000 births. In Europe it is known as Franceschetti-Klein syndrome. Its complex development involving facial clefts 6, 7, and 8 creates the fish or bird-like characteristics of mandibulofacial dysostosis. There is hypoplasia of the zygoma, maxilla and mandible as well as malformed ears. Cleft 6 causes a high palate, cleft 7 deforms the mandible. It is not uncommon for these patients to also have congenital heart disease. Treatment involves bone grafting with staged procedures. The technique of distraction osteogenesis of the mandible is often useful in these patients at around 4-5 years of age.

The triad of cleft palate, hypoplasia of the mandible and glossoptosis is called Pierre Robin syndrome or sequence. It is thought to be possible due to hemorrhage at the fusion of the carotid and stapedial arteries. The mandible does not reach its proper size during the 2nd month, thus the tongue is not brought into the oral cavity and closure of the palate is impeded. These patients often suffer from respiratory and feeding difficulties that may not become apparent till the 2nd month of life when rapid tongue growth appears. Mortality is 6% unless the infant is premature and then the mortality increases to 23%. Anterior glossopexy can help alleviate the airway obstruction caused by the tongue falling back when the mouth is closed. Unfortunately this will retard growth of the mandible. Placing the child in the supine position with the neck extended allows the tongue to fall back occluding the oropharyngeal airway. The upper airway obstruction will lead to hypoxia, pulmonary hypertension, cor pulmonale and failure to thrive secondary to difficulty swallowing and aspiration. With relaxation of the genioglossus muscles during sleep, the respiratory difficulties may worsen. Sleep studies using a polysomnogram are needed to identify the apneic episodes. Tracheotomy maybe needed to alleviate the airway obstruction, although this is not without risk. Positioning the infant prone and feeding in an upright position helps relieve the obstruction. By age 4, the retrognathia is usually self corrected due to a growth spurt of the mandible. Feeding these children in a 90° position helps to avoid aspiration. Airway management may be difficult due to the micrognathia and more posterior placed tongue.

Defective ossification that creates a malformation of the cranial base is known as craniofacial dysostosis. There are three clinical features seen: craniosynostosis, midface retrusion, and exorbitism. Skull sutures allow the bones to overlap during passage through the birth canal. They function as a “shock absorbing” site in childhood and are the principal sites of expansion of the skull. Craniosynostosis or premature fusion of one or more cranial sutures occurs in 1 per 2000 live births. When this occurs, the bones are no longer able to grow out from the suture as they should. Several theories have been proposed as to the origin, which leads to a probable multifactorial cause. Closure of the sagittal suture results in scaphocephaly or a boat shaped skull. Brachycephaly is the result of bilateral coronal suture closure forming a broadened skull. Turricephaly is due to closure of sagittal and coronal sutures forming a pointed skull. Metopic suture closure results in a triangular appearing skull. They all result in a decreased cranial cavity and increased intracranial pressure. Apert’s and Crouzon’s are two of the more common
syndromes involving craniosynostosis. Since they are syndromes with Mendelian inheritance, the parents should pursue genetic counseling. Surgical remodeling of the skull to correct these deformities is often begun at 6-12 months of age.

Apert’s syndrome or acrocephalosyndactyly is the result of craniosynostosis (usually coronal), mid-face hypoplasia due to hypoplastic maxillary development and syndactyly. It is of autosomal dominant inheritance. The nose is underdeveloped with a high arched palate that often has a cleft in the soft palate. They often suffer from chronic otitis media leading to deafness in 30% of the patients. Mental retardation is seen in 50% of patients. Treatment involves multiple, staged procedures involving cranial vault remodeling and a LeFort III procedure at age 4-5.

Crouzon’s syndrome, the most frequent syndrome involving craniosynostosis has significant premature suture closure (usually coronal sutures), maxillary hypoplasia and proptosis. It occurs in 1 per 25 000 and is due to autosomal dominant inheritance. These patients often have nasal airway obstruction due to mid-face hypoplasia and a high arched palate. Hearing loss is common (55%) and there is a 30% incidence of C2,3 spinal fusion. Mental capacity is normal. Treatment involves multiple staged procedures with a mid-face advancement or LeForte III at 4-5 years of age. Orbit manipulation during these procedures can cause marked bradycardia.

Anesthetic concerns in caring for these complicated patients are airway abnormalities, adequate vascular access for hemorrhage and monitoring and also heat loss. Preoperative sedation to a patient with a compromised airway should be individualized and used cautiously. Airway management difficulty should be predicted and planned for in advance to avoid urgent problems. Options should include availability of oral Airways, oversized masks, fiberoptic bronchoscope, a variety of laryngoscope blades and lighted stylets. Control of the airway may involve awake, fiberoptic, retrograde or digital intubation. Inhalation induction of anesthesia is preferred over intravenous. Paralysis with neuromuscular agents should be avoided until controlled mask ventilation is assured. Blood loss during these procedures can be excessive with 1 to 2 blood volumes often required. Adequate vascular access with heated intravenous solutions are manditory. The use of erythropoietin injections and additional iron supplements started 3 weeks prior to elective craniofacial surgery has been shown to modify transfusion requirements. An arterial line is required to monitor the hemodynamics and laboratory values in these patients. Use of central venous catheters to monitor volume status and precordial doppler to detect venous air emboli should be individualized to the procedure and patient. Postoperative care must continue in a closely monitored environment (intensive care unit) to continually access hemodynamic and respiratory parameters.

Pediatric patients with craniofacial abnormalities continue to provide the anesthesiologist with some of the most challenging cases that we see today. These difficult cases need careful planning and cooperation from the many professionals involved in the care of these patients.

References


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