ANESTHESIA FOR CRANIOFACIAL SURGERY

Of all the plastic surgical procedures, correction of craniofacial deformities in children offers perhaps the greatest challenge to anesthesiologists. Every since John Snow published the first report of giving ether to a seven year old boy for cleft lip repair in the *Lancet* in 1847 (1), anesthesiologists have been striving to perfect the safety of craniofacial operations.

In 1937, Phillip Ayre described the first use of the T-piece during cleft lip repair in infants to prevent hypercarbia, a technique that became a standard method for many years. Still, craniofacial operations carry a high risk of complications, among them venous air embolism (2-4), difficult intubation, hypercarbia, massive exsanguination, hypothermia, cerebral edema, loss of airway, and airway obstruction (5).

In spite of the potential problems associated with craniofacial surgery, 87% of patients report a subjective improvement in appearance (6). 91% of parents of small children and 77% of adolescents would repeat the decision to undergo surgery (7,8).

SURGICAL CONSIDERATIONS

Craniofacial surgery is a broad term referring to both cranial and facial surgery for the correction of cranial dysostosis or craniofacial dysmorphism. Craniosynostosis refers to premature fusion of cranial sutures, the most common form of which is scaphencephaly (saggital suture). Other recognized types include trigonocephaly (metopic), plagiocephaly (lambdoid) and brachycephaly (coronal). Early detection and surgical correction correlate with improved cosmetic outcome.

Surgical positioning varies with suture abnormalities, but can be either prone or supine. The table is rotated 90^0. Surgical duration is in the 4-8 hour range. Mortality should be < 1-2%(13), with operative injury to the superior saggital sinus among the most catastrophic surgical complications. Major complications are reported in 14.3% of patients (13). Morbidity includes meningitis, CSF rhinorrhea, bone infection, intracranial hypertension, and venous thrombosis. Blindness has been reported in < 1% of patients (14).

PRE-OPERATIVE PREPARATION

Craniofacial deformities are associated with 58 recognized syndromes; Apert's and Crouzon's syndrome are the most common. Associated CNS, pulmonary and cardiac anomalies may occur (9). Others include Pfeiffer's syndrome (mid-face hypoplasia, seizure, Arnold-Chiari malformation may be associated) and Carpenter's syndrome (often associated with obesity and CHD). Isolated nonsyndromic craniosynostosis occurs in 6 per 10,000 births, 57% affecting the sagittal and 20% the coronal suture. Apert's syndrome occurs in approximately 1

per 100,000 births and is so severe that many of the patients require preoperative tracheostomy due to airway obstruction.

Up to 23% of craniofacial patients in one study had intracranial hypertension during sleep, even without hydrocephalus. Hydrocephalus is common, especially in Apert's and Crouzon's Syndrome, due to low cranial volumes. However, clinical signs and symptoms of elevated intracranial pressure are uncommon. Intracranial pressure has been shown to normalize two to eight weeks after surgery.

The pre-operative exam should focus on the airway, heart and lungs. 20-37% of craniofacial patients exhibit airway anomalies and, as many as 53% have intra-operative airway problems (5, 11). The old chart should be reviewed, with special reference to prior intubations, and parents advised that peri-operative airway compromise might necessitate tracheostomy. Choanal atresia, macro-glossia, micrognathia and facial asymmetry may contribute to the sudden loss of airway during induction; awake fiberoptic intubation may be required. These patients are frequently left intubated post-operatively, due to massive fluid resuscitation intra-operatively, as well as airway anomalies; arrangements for post-operative sedation and ventilation in the PICU should be anticipated.

A pre-operative history of snoring, daytime somnolence or morning headaches may be signs of nighttime sleep apnea in older children. Pre-operative sedation should be withheld unless close monitoring is feasible.

Chronic severe airway obstruction with hypoxia can lead to pulmonary artery hypertension and subsequent cor pulmonale; tracheostomy is usually performed before this condition develops.

Significant heart murmurs should be evaluated by echocardiograms pre-operatively. If an intracardiac shunt exists, air emboli may enter the coronary or cerebral circulation with catastrophic consequences.

The presence of an upper or lower respiratory infection may render an already difficult situation disasterous; thus the procedure for these children should be postponed.

The placement of central venous and arterial catheters is routine in many of these procedures, and the risks and complications of these invasive monitors should be appreciated and discussed with the parents pre-operatively.

Laboratory testing should include a hemoglobin and coagulation studies (PT, PTT, platelets) as well as a type and cross.

ANESTHETIC MANAGEMENT

Associated respiratory, cardiac, and neurological disorders will influence the anesthetic technique. Usually, an inhalation induction is satisfactory, unless

airway obstruction is deemed imminent. Awake or sedated, fiberoptic intubation in children can be difficult. A wide variety of airway equipment should be available, including laryngeal mask airway (LMA), lightwand, fiberoptic laryngoscope, guidewires and, of course, the ability to perform a rapid tracheostomy.

After intubation, two large IV's and an A line are mandatory, as these are procedures which may involve large blood loss (as defined: greater than 20% of the blood volume). Blood loss of up to 65% of blood volume can accompany even a simple craniectomy. Longer cases may involve transfusion of greater than one blood volume ("massive transfusion"), necessitating platelets and fresh frozen plasma to prevent coagulopathy. The Hemocue® is a useful tool to monitor serial hemoglobin in these situations. A recent study compared 13 patient (average age 10 months) randomized to get aprotinin or placebo, and found that aprotinin markedly decreased rbc transfusion (Ahmed, SPA 2007).

Since the procedure may last eight to ten hours, the patient's position and temperature must be carefully monitored. The wide-open cranium presents a large area for heat loss. Warm IV fluids and other perioperative warming devices are indicated.

Routine methods of reducing cranial hypertension, such as hyperventilation, may be necessary. Although control of $PaCO_2$ and mean arterial pressure is often adequate, mannitol, furosemide, and dexamethasone should be available; Dr. Duncan prefers that patients receive mannitol and furosemide at induction.

Air embolism with circulatory collapse is a risk during craniectomy, even in the supine position. Precordial doppler monitoring is optimal (2,3,4), but generally not utilized except in the sitting position.

Finally, severe post-operative swelling of the face often occurs after longer procedures, and post-operative ventilation and sedation may be deemed necessary to provide a safe airway until the swelling resolves.

In summary, severe complications including intraoperative death from loss of airway, massive blood loss and air embolism may occur during craniofacial surgery. Post-operative cerebral edema, brain damage and loss of airway have also occurred. At our institution, the anesthetic management of these specialized cases is handled by an experienced team of pediatric anesthesiologists and operating room personnel. These skilled professionals call upon the full scope of their skills to provide not only excellent perioperative care, but to ensure smooth transfer to the pediatric intensive care unit for postoperative management.

BIBLIOGRAPHY

1. Jones RG: A short history of anesthesia for hare-lip and cleft palate repair. *Br J Anesth* 43: 796, 1971.

2. Harris M, Strafford M, Rowe R et al: Venous air embolism and cardiac arrest during craniectomy in a supine infant. *Anesthesiology* 65:547-550, 1986.

3. Harris M, Yeman T, Strafford M: Venous air embolism during craniectomy in supine infants. *Anesthesiology* 67: 816, 1987.

4. Cucchiara R, and Bowers B: Air embolism in children. *Anesthesiology* 57:338-339, 1982.

5. Delegue L, Guilbert M: Management of airway problems during the repair of craniofacial anomalies in children. *Craniofacial Surgery*, Little Brown, Boston, P. 141 en Carnni EP/Ed.

6. Lefebvre A, Barclay S: Psychosocial impact of craniofacial deformities before and after reconstructive surgery. *Can J Psychiatr* 27:579, 1982.

7. Phillips J, Whitaker LA: The social effects of craniofacial deformity and its correction. *Cleft Palate J* 16:7, 1979.

8. Pertschuk MJ, Whitaker LA: Psychosocial outcome of craniofacial surgery in children. *Plast Reconstruct Surg* 82:741, 1988.

9. Cohen MM Jr (Ed): Craniosynostosis: diagnosis, evaluation and management. Raven Press New York, 1986.

10. Gault DT, Renier D, Marthae D, Jones BM: Intracranial pressure and intracranial volume in children with craniosynostosis. *Plast Reconst Surg* 90:377, 1992.

11. Handler SD, Beaugard MD Whitaker, LA, Potsic WP: Airway management in the repair of craniofacial defects. *Cleft Palate J* 16:16, 1979.

12. Whitaker LA, Munro IR Salyer, KE, et al: Combined report of problems and complications in 793 craniofacial operations. *Plast Reconstr Surg* 64:198, 1979.

13. Jaffe RA and Samuels SI (editors):*Anesthesiologist's Manual of Surgical Procedures*. Raven Press, New York, 1994

14. McCarthy JG, Thorne CHM, Wood-Smith D: Principles of craniofacial surgery: Orbital hypertelorism. *Plastic Surgery*, Vol 4. McCarthy JG, ed. WB Saunders Co, Philadelphia: 1990, 2974-3012

CME Question

All of the following are feared complications during craniofacial surgery except:

(1) venous air embolism
(2) saggital sinus tear
(3) loss of airway
(4) bradycardia due to oculocardiac reflex

the answer is (4) – although bradycardia can certainly occur during craniofacial surgery due to a variety of factors – including brainstem traction - the oculocardiac reflex is *usually* associated with traction on extraocular muscles during strabismus surgery