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CHAPTER 12.8

Surgery for Craniofacial Malformations

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[1]Craniosynostosis

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Surgical Correction of Craniosynostosis

Surgical Considerations

Description: Premature fusion of cranial sutures, or **craniosynostosis,** causes various well-recognized patterns of cranial vault and facial deformities. Rarely, these are related to conditions such as Crouzon, Apert, Saethre-Chotzen, and Pfeiffer syndromes. Single or multiple sutures can be involved, the most common being the sagittal suture. This condition is called **scaphocephaly,** in which the cranial vault is bitemporally narrow, with AP elongation. **Anterior or posterior plagiocephaly** involves a single coronal suture or lambdoid suture and is characterized by flattening of the forehead on the affected side. **Oxycephaly** ('tower-head deformity') involves bilateral coronal sutures, with a flat, high forehead, whereas **brachycephaly** also involves the cranial base sutures, and results in bitemporal bulging, midfacial hypoplasia, an anterior open bite, and hypertelorism. These patients may have severe sleep apnea and can pose a challenge for airway management. **Trigonocephaly** (triangular head shape) (Fig. 12.8-1), with a keel-shaped forehead and hypoteloric tendency, involves the metopic suture.

Surgical correction of these craniofacial anomalies requires a combined plastic surgery and neurosurgery team approach involving the release or resection of the affected suture and simultaneous correction of the asymmetric skull by **bone-flap repositioning or advancement.** Frontal/orbital abnormalities are addressed with **bifrontal craniotomy and floating forehead advancement,** along with advancement of the supraorbital bar **(fronto-orbital advancement)** (Fig. 12.8-2). For example, in plagiocephaly, because of the unilateral coronal synostosis, the frontal bone is retruded and the superior orbital rim is elevated and retruded on this side. Craniectomy is performed, the forehead is removed, the involved coronal suture is resected, and the supraorbital bar is cut above the orbit and down to the lateral orbital wall across the midline. The bar is bent, advanced on the involved side—sometimes up to 1.5 cm—and fixed in this position. Additional bone strips are taken from the posterior cranium and split for use as graft material; the other bone pieces are replaced and fixed with wires, suture, or restorable plates.

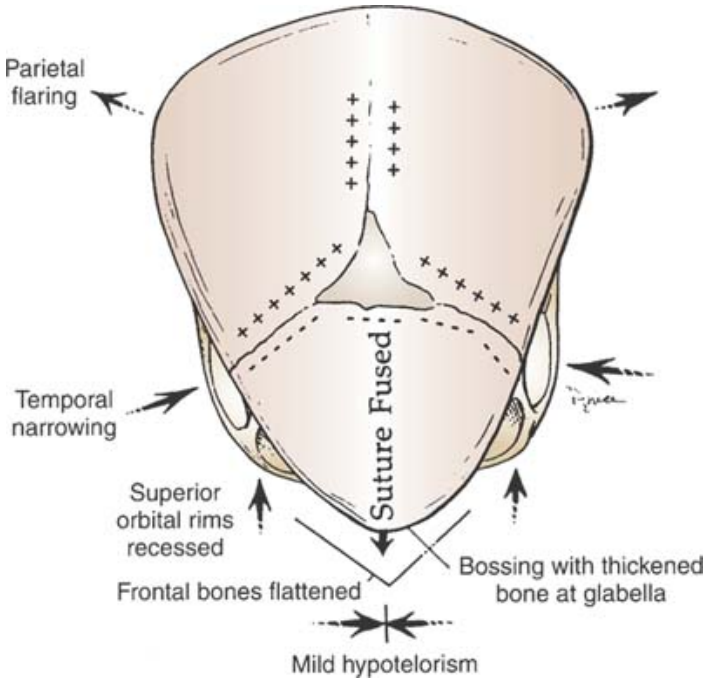

Figure 12.8-1. 1. Skull shape abnormalities in metopic synostosis: Regions of reduced bone deposition (—). Regions of compensatory increased bone deposition (+++). (From Belfrey ME, Pershing JA, et al: Surgical Treatment of Metopic Synostosis. In *Neurosurgical Clinics of North America*. Pershing JA, Jane JA, eds. WB Saunders, Philadelphia: 1991, used with permission.)

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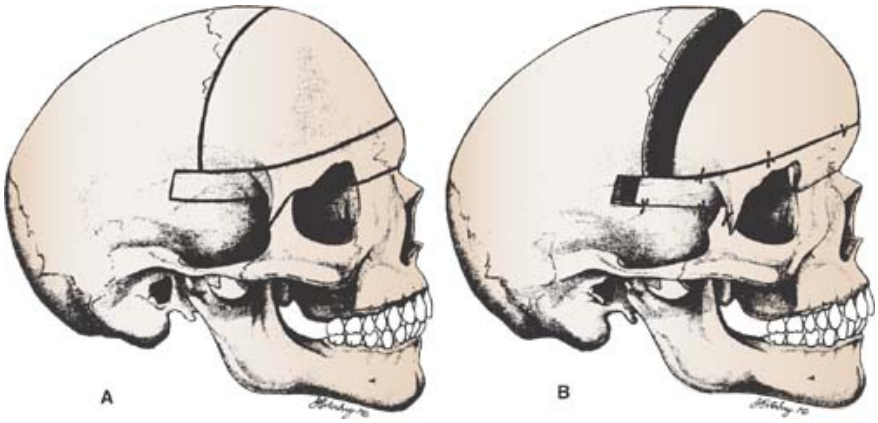

Figure 12.8-2. 2. Fronto-orbital advancement. **(A)** Lines of osteotomy for forehead and supraorbital bar advancement. **(B)** Fronto-orbital advancement in a tongue-in-groove manner and fixation with wires. (Reproduced with permission from Aston SJ, Beasley RW, Thorne CH, eds: *Grabb and Smith's Plastic Surgery,* 5th edition. Lippincott-Raven, Philadelphia: 1997.)

Patient positioning, type of headrest, and incision all vary, depending on location of the suture abnormality. If the deformity is mostly posterior (e.g., a sagittal or plagiocephaly case), a prone approach with biparietal or midsagittal incision can be used. Resection of the involved suture and **barrel staving** of the cranium, with grafting for reshaping, works well. Reconstruction as above usually is accomplished at 3–6 months of age. Brain mass doubles in size the first 6 months and triples by 3 years of age, when 80% of the brain growth is completed (the driving force for cranial vault growth).

All procedures are extradural. Dural tears, if they occur, are repaired to prevent CSF leak and CNS infection.

In the syndromic cases, the cranial synostosis deformity is treated similarly to the procedure described, usually before the age of 12 months, with a view to midfacial advancement of the primary dentition at maturity either by **LeFort III or monobloc advancement.** (See p. 1116 LeFort Osteotomies, and p. 1403 Major Secondary Craniofacial Surgical Procedures.)

Postsurgical orthopedic distraction devices like the **Delaire mask** also can be used to encourage midfacial growth. The syndromic cases may require repeat craniotomy and reshaping at a young age if signs of ↑ ICP appear. Other synostotic cases also should be

monitored for ↑ ICP and the need for urgent secondary craniotomy. Marchac and Renier detected ↑ ICP in 13% of single-suture synostosis and 42% of multisuture synostosis. ↑ ICP may increase the risk of dural breach during craniotomy 2° cranial bone resorption and thinning.

Blood loss can be significant at first incision through the vessel-rich scalp. Rainey clips are applied immediately to minimize blood loss. In anticipation of major blood loss, transfusion should be started with the first incision. Severe life-threatening blood loss can occur if the sagittal sinus is breached, and neurosurgical repair must be accomplished quickly. Elevation of the bone flaps usually causes diffuse bleeding, which is stopped easily with irrigated bipolar cautery and thrombin-soaked sponges. It is useful to have the patient in the reverse Trendelenburg position from the start. Diffuse bleeding at the cut bone edges and over the bone surfaces can be further controlled with bone wax. Focal bleeding from around the orbit and in the temporal fossa region subperiosteally also can be controlled with bone wax. A LeFort/monobloc component to the surgery increases blood loss, especially during the initial mobilization of the facial segment. Local anesthetic with epinephrine injected, and/or on sponge packs for pressure, will control the diffuse mucosal bleeding. BP control is also paramount. Electrocautery and Ligaclips to larger vessels (e.g., descending palatine pedicles) may be necessary.

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Variant procedure or approaches: In older children (> 2 yr), split cranial bone grafts may be required to correct defects caused by bone-flap advancement. Excision of skull segments is commonly accompanied by rigid fixation.

Usual preop diagnosis:
Craniosynostosis (sagittal, coronal, metopic, lambdoid); syndromes such as Apert and Crouzon syndromes; ↑ ICP

Summary of Procedures

Position	Usually supine; prone for correction of lambdoidal suture synostosis or posterior sagittal synostosis. If entire cranial vault reshaping (multiple sutures), special padded occipital-cheek headrest, because pin fixation is not safe until > 2 yr.
Incision	Usually bicoronal, biparietal, Meisterschnitt, midsagittal
Special instrumentation	Horseshoe headrest, usually pediatric; occasionally, Gardiner tongs; Midas Rex craniotome; resorbable plates and screws
Unique considerations	Control of ICP — spinal drain may be placed. Blood in room, if transfusion anticipated.
Antibiotics	Pediatric: cefazolin 25 mg/kg q 8 h, or vancomycin 10–15 mg/kg q 6 h, and cefotaxime 25–50 mg/kg q 6 h for oropharyngeal contamination and following dural tears
Surgical time	2–6 h
Closing considerations	Blood loss with Rainey clip removal. Full head-wrap dressing causes head/neck movement → bucking.
EBL	200–800 mL; may be formidable.
Postop care	ICU: 1–2 d (further transfusion often necessary in 1st 2 h to replace drain losses)
Mortality	0.6–1.6%
Morbidity	Major complications: 14.3% Bone infection: 3–7% Meningitis CSF leak: 4.5% ↑ ICP Air embolus: < 1% Blindness: < 1% Massive bleeding: < 1% Venous thrombosis Neurologic injury: Rare
Pain score	4

Patient Population Characteristics

Age range	2–24 mo (primary correction)
Male:Female	1:1
Incidence	Non syndromic: 1/10,000 births (most sporadic; few familial patterns) Crouzon syndrome: 1/25,000 Apert syndrome: 1/100,000
Etiology	Idiopathic; however, some are associated with specific genetic conditions (e.g., Crouzon and Apert syndromes). Other cases may be 2° ↓ brain growth.
Associated conditions	Hydrocephalus; ↑ ICP; mental retardation; airway problems; ocular abnormalities; exotropia (29% Crouzon or Apert syndromes); lagophthalmus; exorbitism

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Anesthetic Considerations

Preoperative

Patients may have craniofacial anomalies—particularly Apert and Crouzon syndromes—which are associated with midface hypoplasia, obstructive sleep apnea and in some cases difficult intubation. Hence, detailed preop airway evaluation is necessary. Children with single-suture craniosynostosis are usually healthy. Surgery is often performed between 3–6 months of age, preferably when the infant weighs > 5 kg.

Respiratory	Patients with long-standing upper-airway obstruction due to choanal atresia, mandibular and maxillary hypoplasia, or other causes, may have chronic hypoventilation and hypoxia and may experience episodes of apnea. If the patient has Sx of acute URI, delay elective surgery at least 2 wk. The presence of fever, cough, and abnormal chest auscultation necessitates radiographic evaluation and pediatric consultation. **Tests:** As indicated from H&P.
Airway	Be aware of other congenital anomalies affecting the patient's airway, such as Apert, Klippel-Feil, Goldenhar, Pierre-Robin, Treacher-Collins, or Crouzon syndromes. Review any previous anesthetic records for patient to gain insights into appropriate airway management (e.g., FOI may be necessary). Consider elective tracheostomy under local anesthesia in patients with severe airway abnormalities.
Cardiovascular	Consider the coexistence of congenital cardiopulmonary anomalies, particularly in patients with Apert syndrome (autosomal dominant trait, craniosynostosis, syndactyly of hands and feet). Preop evaluation of a patient with a known or suspected heart defect should include thorough H&P, ECG, Hct, baseline O_2 sat, and CXR. For children with Sx of cardiac dysfunction or those requiring cardiac medication, it is advisable to consult with a pediatric cardiologist to optimize the patient's condition prior to surgery. **Tests:** Preop EKG indicated for patients with CHD; others as indicated from H&P.
Neurological	If only the sagittal suture is involved, ICP is usually normal. If more than one suture is involved, brain growth will be impaired, the patient will be developmentally delayed, and intracranial HTN

	may be present.
Hematologic	Surgery in early infancy (< 9 mo) is common; thus, allowable blood loss is small; blood transfusion usually is required.
Laboratory	**Tests:** Hct; PT; PTT; T&C blood. Other tests as indicated from H&P.
Premedication	Patients >9–10 mo old may benefit from premedication with midazolam (0.5–0.75 mg/kg po). Antibiotic prophylaxis for CHD (e.g., ampicillin 25 mg/kg + gentamicin 2.5 mg/kg iv) may be warranted (see AHA guidelines).

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Intraoperative

Anesthetic technique: GETA. Anticipate possible difficult airway. Heat OR to 78–80°. Forced air warmer. Heat all fluids to body temperature.

Induction	Surgery for craniectomies is extradural. Either mask induction with N_2O and inhalational agent or iv induction is suitable for the infant with a normal airway. If ICP is suspected to be elevated, consider mild hyperventilation after loss of consciousness. For a difficult airway, intubation may be facilitated by using an FOI while patient is awake or anesthetized and spontaneously ventilating. In rare situations, tracheostomy, under sedation and local anesthesia, may be necessary. Consider suturing ETT to prevent accidental extubation. In prone cases, unsutured endotracheal tubes can slip through tape due to pooling of oral or nasal secretions.
Maintenance	Maintenance anesthesia with inhalational agent or balanced (iv) anesthesia and long-acting muscle relaxant, should be adequate. Surgery may be prolonged. Control of ICP may be necessary (see below). A remifentanil infusion 100–300 ng/kg/min provides profound intraop analgesia and rapid awakening.
Emergence	Prompt awakening to allow neurological evaluation is an important goal. Excessive facial edema, particularly in patients with preoperative sleep apnea may require the patient to remain intubated postoperatively.
Blood and fluid requirements	Anticipate large blood loss. IV× 2 (as large as possible LR @: 4 mL/kg/h – 0–10 kg + 2 mL/kg/h – 11–20 kg + 1 mL/kg/h – > 20 kg (e.g., 25 kg = 65 mL/h) Warm all fluids. Humidify gases. Have 1–2 U PRBC or whole blood available in the room. Use LR for replacing deficit, maintenance, and 3rd-space fluid loss. Replace blood loss with colloid and PRBC mL for mL. Significant blood loss begins with scalp incision; allowable blood loss is small, so that it is important to begin transfusion early before hypovolemia occurs. EBV for an infant in this age group is 75 mL/kg. A good rule is to infuse a volume of blood equal to 10% of EBV prior to incision in the healthy infant after discussion with the attending surgeon regarding expected blood loss. Avoid NS (acidosis, ↑ bleeding) in children < 5 yr. Beware of ↑ K^+ and ↓ Ca^{++} associated with massive transfusion.
Control of ICP	Hyperventilation Osmotic diuretic Loop diuretic In some cases, it may be desirable to ↓ ICP. This can be accomplished by ↑ ventilation ($PaCO_2$ = 25–30 mmHg), diuretics (furosemide 1 mg/kg iv).
	Arterial cannulation for BP, monitoring of ABG, Hct, electrolytes, etc. ↑ K^+ and ↓ Ca^{++} are most common following transfusion with whole blood or FFP. VAE has been reported during craniectomies in infants;

		hence, a precordial Doppler and CVP line will be helpful. CVP may be particularly helpful in the infant with marginal cardiovascular status for volume assessment and drug administration. Close attention to $ETCO_2$ and, if available, ETN_2 monitoring is useful. If VAE is suspected the surgical field should be flooded and the head lowered immediately. CVP may be particularly helpful in the infant with marginal cardiovascular status for volume assessment and drug administration and if peripheral iv access is limited. Precordial or esophageal stethoscope is useful in cases where the patient is turned 180° or in cases of marginal pulmonary function.
Monitoring	Standard monitors (Appendix D-1). ± Arterial line ± CVP line ± Precordial Doppler ± Urinary catheter	
Positioning	and pad pressure points. eyes.	Positioning depends on surgical approach; most are performed with patient prone; however, use of the head-up position is not uncommon.
Complications	Oculocardiac reflex (OCR) → ↓↓ HR and ↓↓ BP	Notify surgeons and Rx with atropine 0.02 mg/kg iv.
	VAE	Be prepared to make prompt Dx of VAE (↓ $ETCO_2$ change in Doppler sounds, ↑ ETN_2 ↓ O_2sat, ↓ BP, ↑ HR) and Rx: notify surgeons, flood wound, ± head down, aspirate CVP, ± vasopressors.

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Postoperative

Complications	Hypovolemia with ↓ BP Hypothermia	Inadequate volume replacement may result in ↓ BP and acidosis. Hct to establish need for further fluid or blood therapy.
	Airway Complications	Excessive facial edema, history of a marginal airway preoperatively, or severe obstructive sleep apnea with sensitivity to opioids may necessitate maintenance of a secure airway postoperatively
Pain management	Parenteral opioids (see p. E-4).	
Tests	Followup Hct postop	Transfuse to keep Hct ≥ 30%.

Suggested Readings

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Major Secondary Craniofacial Surgical Procedures

Surgical Considerations

Description: These procedures usually are performed on children ≥ 5 years. There are two basic approaches. The first involves advancement of the upper face and frontal bone, frequently described as a **monobloc** (Fig. 12.8-3) or **frontofacial advancement.** The second variation, called **facial bipartition** or **periorbital osteotomy,** is for correction of telorbitism (widely spaced eyes), usually accomplished by a combined extra- and intracranial approach, using both plastic and neurosurgery teams.

Variant procedure or approaches: Many different variations of the above-named procedures can be performed; however, from an anesthetic standpoint, they are not significantly different. The use of **cranial bone grafts** and **rigid fixation** have shortened these somewhat lengthy procedures. Other bone grafts, however, from ribs and iliac crest, are occasionally required. These procedures frequently last ≥ 6 hours and blood loss can be very heavy. Reconstruction of the forehead and orbital area following a tumor excision, for example, uses a similar approach, but requires additional bone grafts.

Usual preop diagnosis: Craniofacial malformations; craniofacial deformities; telorbitism or hypertelorism; craniofacial dysostosis

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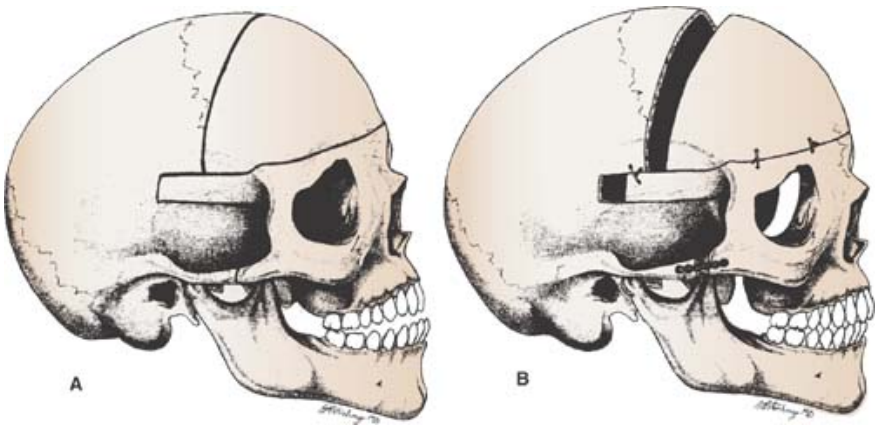

Figure 12.8-3. 3. Monobloc advancement. **(A)** Lines of osteotomy for monobloc osteotomy. **(B)** Advancement of midface, orbits, and frontal bone, and stabilization with bone grafts and miniplates. (Reproduced with permission from Aston SJ, Beasley RW, Thorne CH, eds: *Grabb and Smith's Plastic Surgery,* 5th edition. Lippincott-Raven, Philadelphia: 1997.)

Summary of Procedures

	Monobloc/Frontofacial Advancement	Facial Bipartition, Periorbital Osteotomies
Position	Supine	
Incision	Bicoronal, oral	Bicoronal, infraorbital
Special instrumentation	Horseshoe headrest, usually pediatric; disimpaction forceps; Midas Rex craniotome; resorbable plates and screws; mini/micro titanium plates and screws; ± distraction device to supplant bone grafting and rigid fixation.	
Unique considerations	Control of ICP: spinal drain may be placed, hyperventilation. Blood in room for anticipated transfusion.	
Antibiotics	Cefazolin 25 mg/kg q 8 h or vancomycin 10–15 mg/kg q 6 h, and cefotaxime 25–50 mg/kg q 6 h for oropharyngeal contamination and following dural tears	
Surgical time	4–10 h	
Closing considerations	Blood loss with Rainey clip removal for suturing. Full head-wrap dressing → head/neck movement with awakening.	
EBL	400–800 mL; may be formidable	
Postop care	ICU: 1–2 d; Monitor Hct.	
Mortality	0.6–1.6%	
Morbidity	Major complications: 14.3% Bone infection: 3–7% Meningitis Infection rates higher if: Adults rather than children (10 ×) Longer OR times and longer hospital stay Tracheostomy Foreign body (plates/screws/other alloplast) Anterior fossa entered Large dead space (e.g., in adult nongrowing brain) CSF leak: 4.5% ↑ ICP Air embolus: < 1% Blindness: < 1% Massive bleeding: < 1% Venous thrombosis Neurological injury: Rare	
Pain score	6	6

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Patient Population Characteristics

Age range	3–20 yr

Male:Female	1:1
Incidence	Crouzon syndrome: 1/25,000 Apert syndrome: 1/100,000
Etiology	Congenital (80%); occasionally trauma or tumor (20%)
Associated conditions	Depends greatly on the syndrome or disease entity. See Cranio synostosis, p. 1398.

Anesthetic Considerations

Preoperative

Craniofacial syndromes often are associated with maxillofacial deformities, mandibular abnormalities and challenging airway management.

Respiratory	Patients with long-standing upper airway obstruction due to choanal atresia, mandibular and maxillary hypoplasia, etc., may have chronic hypoventilation and hypoxia, and may have apnea episodes. If Sx of acute URI, delay elective surgery at least 2 wk. The presence of fever, cough, and abnormal chest auscultation necessitates radiographic evaluation and pediatric consultation. **Tests:** As indicated from H&P.
Airway	Be aware of other congenital anomalies affecting the airway, such as Apert, Goldenhar, Klippel-Feil, Pierre Robin, Treacher Collins, or Crouzon syndromes. Review any previous anesthetic records for insights into airway management (e.g., need for FOI). Consider elective tracheostomy under local anesthesia in patients with severe airway abnormalities.
Cardiovascular	Frequency of CHD is increased in patients with craniofacial abnormalities. Preop evaluation of patient with known or suspected heart defect should include H&P, ECG, Hct, baseline O_2sat, and CXR. For children with Sx of cardiac dysfunction or those requiring cardiac medication, it is advisable to consult with a pediatric cardiologist to optimize patient's condition prior to surgery. **Tests:** Preop ECG indicated for patients with CHD; others as indicated from H&P.
Neurological	Neurologic deficits, if any, should be documented preop.
Laboratory	Hb/Hct; therapeutic drug levels for patients taking anticonvulsants.
Premedication	Premedication is helpful for patients > 1 yr – oral midazolam 0.5–0.75 mg/kg or oral ketamine 6 mg/kg about 30–60 min before induction.

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Intraoperative

Anesthetic technique: GETA, with special consideration given to associated CHD, pulmonary, and airway problems.

Induction	In an otherwise healthy patient, inhalational induction with subsequent placement of iv lines is appropriate. Muscle relaxants facilitate intubation but should be used only when adequate mask ventilation can be assured. An oral RAE ETT is useful for this procedure and should be secured carefully in place (often by suturing). Intubation in a patient with airway abnormalities may be facilitated by using an FOI with patient awake or lightly anesthetized and spontaneously ventilating. In rare situations, tracheostomy, under

	sedation and local anesthesia, may be necessary.	
Maintenance	Standard pediatric maintenance (see p. D-2). Consider use of remifentanil infusion (100–300 ng/ kg/min) for supplemental analgesia and to facilitate rapid awakening.	
Emergence	Extubate trachea when patient is awake and protective airway reflexes have returned. Patients with reactive airway disease may require deep extubation.	
Blood and fluid requirements	Anticipate large blood loss. IV: 18 ga × 1–2 NS/LR @: 4 mL/kg/h – 0–10 kg + 2 mL/kg/h – 11–20 kg + 1 mL/kg/h –> 20 kg (e.g., 25 kg = 65 mL/h) Warm fluids. Humidify gases.	The goal of intraop fluid therapy is to replace preop deficits, intraop fluid, electrolyte, and blood losses, while providing maintenance fluids. Half of the calculated deficit (hours fasting × hourly maintenance fluid requirement) generally is replaced during the 1st h of anesthesia and the balance over the next 1–2 h. Surgical manipulation of tissue will cause 3rd-space fluid loss proportional to the degree of surgical trauma and tissue exposure. It may range from 0–10 mL/kg/h.
Control of blood loss	Deliberate ↓ BP	Deliberate ↓ BP can be accomplished by use of SNP, esmolol, or potent inhalational agents titrated to effect (MAP 50–60 mmHg).
Monitoring	Standard monitors (see p. D-1). Arterial line ± CVP line	Arterial line is essential for monitoring BP during deliberate ↓ BP and for ABGs and blood chemistries.
Control of ICP	Hyperventilation Mannitol Loop diuretics CSF drainage (> 1 yr)	For some procedures, it is essential to reduce intracranial volume to facilitate surgical access. If prolonged brain retraction is required, postop cerebral edema may ensue.
Positioning	and pad pressure points. eyes.	Positioning head above the heart facilitates venous drainage, but also increases the incidence of VAE. Do not hyperextend or hyperflex the head and neck. Flexion of the neck will move the ETT downward (mainstem intubation); extension will move the ETT upward (cuff leak).
Complications	Displacement of ETT Oculocardiac reflex (OCR)→ ↓↓ HR ↓ BP	Suture ETT to alveolar ridge. Notify surgeon. Rx: atropine 0.02 mg/kg.
	VAE Major blood loss	VAE should be suspected if sudden ↑ ETN_2, ↓ $ETCO_2$ ↓ O_2sat, ↓ BP, ↑ HR. Notify surgeon, flood surgical field with NS, support patient hemodynamically and D/C N_2O.

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Postoperative

Complications	↑ ADH secretion Diabetes insipidus (see Appendix D-1).	SIADH or DI may follow brain manipulation and may require pharmacologic intervention for Rx.
	Cerebral edema Pneumothorax Bleeding	Cerebral edema may →↑ ICP (headache, N/V, ↓ mental status, etc.) Pneumothorax (Sx = ↑ respirations, wheezing, ↓ BP, ↓ CO, ↓ O_2sat) may occur

	2° rib resection for bone graft. CXR.
Pain management	PCA (see p. E-4).
Tests	Hct

Suggested Readings

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Cleft Lip Repair—Unilateral/Bilateral

Surgical Considerations

Description: Cleft lip may be either unilateral or bilateral, associated frequently with clefts of the alveolus and palate. Surgical repair involves the design and execution of geometric flaps on the medial and lateral *(Print pagebreak 1408)* sides of the cleft and primary repair of the cleft nasal deformity. The most common unilateral technique is the **rotation advancement flap of Millard** (Fig. 12.8-4). Multiple bilateral lip repairs have been described, some repairing both sides simultaneously and some one side at a time (Fig. 12.8-5). Technique depends on the amount of prolabial and lateral element tissue available. Recently, **primary nasal repair** has been coupled with these bilateral procedures. These nasal repairs involve extensive mobilization of the alar cartilages and transfer of tissue up into the cleft nasal vestibule and floor, with nasal stents often placed. All of these factors can decrease or occlude nasal airway breathing. Although only a minority of neonates has been found to be true nasal obligatory breathers, this should be kept in mind for those postop patients with respiratory distress.

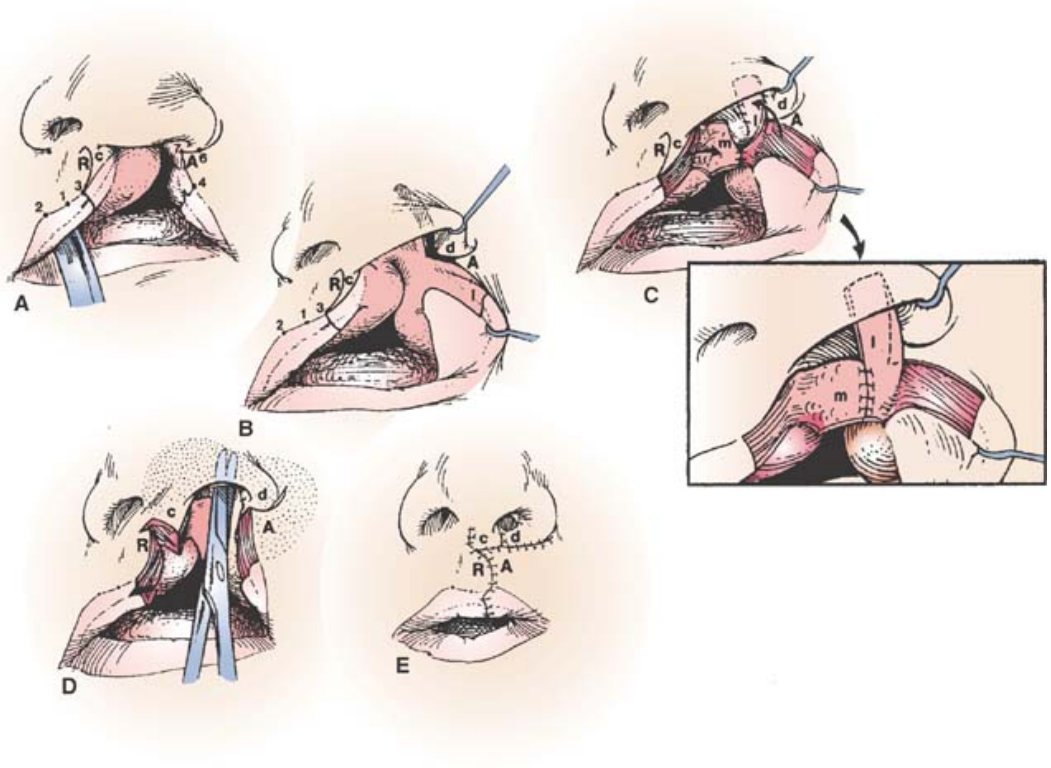

Figure 12.8-4. 4. Step-by-step unilateral complete cleft rotation advancement: m = medial mucosal flap; l = lateral mucosal flap. (Reproduced with permission from Aston SJ, Beasley RW, Thorne CH, eds: *Grabb and Smith's Plastic Surgery,* 5th edition. Lippincott-Raven, Philadelphia: 1997.)

Variant procedure or approaches: Other approaches commonly performed are those of the **Davies-** or **Tennison** -type (Z-plasty) lip repairs (Fig. 12.8-6). In large clefts, a lip adhesion may be performed as an initial stage several months before the actual definitive correction of the cleft lip. This procedure basically involves creating a wound on either side and suturing the muscles, mucosa, and skin together. The procedure itself is very short (45 min). **Presurgical orthopedic devices** may be placed and manipulated, instead of a lip adhesion, to bring a wide, bony cleft into better opposition for a tension-free complete repair. These are custom-fitted and may be fixed with pins to the palate. They are removed in the OR at time of repair.

Usual preop diagnosis: Cleft lip/palate

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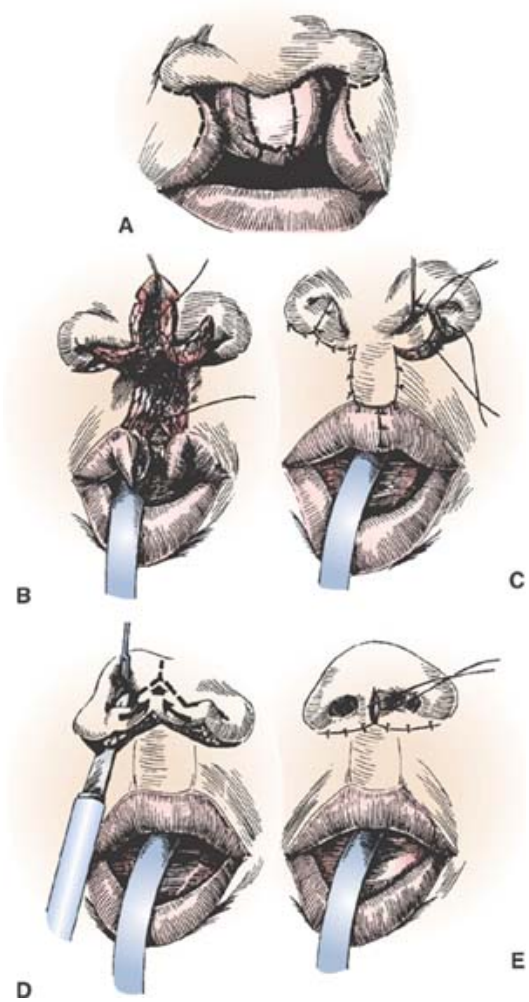

Figure 12.8-5. 5. Banked fork-flap procedure. In the first stage **(A-C)**, bilateral straight line repairs are done after the prolabium is divided vertically into three forks. The central limb is used to construct the center of the lip. The lateral forks are 'banked' at the alar bases for later use in constructing a columella. At the second stage **(D-E)**, bipedicle flaps from the nasal floor, which include the banked forks, are used in combination with a membranous septum incision to elongate the columella and increase tip projection. (Reproduced with permission from Aston SJ, Beasley RW, Thorne CH, eds: *Grabh and Smith's Plastic Surgery,* 5th edition. Lippincott-Raven, Philadelphia: 1997. After Millard DR Jr: Closure of bilateral cleft lip and elongation of columella by two operations at infancy. *Plast Reconstr Surg* 1971; 47:324–31.)

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Figure 12.8-6. 6. Z-plasty closure of lip. (Reproduced with permission from McCarthy JG, ed: *Plastic Surgery*. WB Saunders, Philadelphia: 1990).

Summary of Procedures

Position	Supine; table rotated either 90° or 180°, with oral RAE or anode tube toward chin; patient's head at the edge of head of bed; shoulder roll; neck extended; entire face exposed; scleral shields
Incision	Medial and lateral cleft margins into the nose and in the maxillary vestibule on the cleft side
Special instrumentation	* Throat pack (**NB:** 3 removal before extubation); oral RAE or anode tube.
Unique considerations	Local anesthesia with epinephrine injected **after** lip and nose markings complete. Pediatric patients should wake up in an unagitated state, because undue crying may place excessive tension on the repair. Immediate elbow restraints for children × 2 wk.
Antibiotics	Cefazolin 25 mg/kg iv
Surgical time	1.5 h (bilateral usually ½ h longer) Lip adhesion: 45 min
Closing considerations	Nasal stent; swelling and ointment may occlude nasal airway; smooth emergence important.
EBL	5–10 mL (higher with palatoplasty)
Postop care	Elbow restraints × 2 wk; PACU → room overnight → home POD 1.
Mortality	Minimal
Morbidity	Infection Wound breakdown Hypertrophic scars
Pain score	4

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Patient Population Characteristics

Age range	1 wk–6 mo
Male:Female	2:1—cleft lip and palate. Isolated cleft palate more common in females.
Incidence	1/750 for Caucasians; more common in Asians; less in African Americans. Left cleft more common than right; both more common than bilateral, in the ratio of 6:3:1.
Etiology	Multifactorial, including both genetic and environmental aspects
Associated conditions	Associated anomalies are seen in 29% of cleft lip cases, and may include major chromosomal deletions and/or duplications, along with possible severe mental retardation and CHD.

Anesthetic Considerations

See Anesthetic Considerations for Lip and Nose Surgery, (p. 1421).

Suggested Readings

1. Arosarena OA: Cleft lip and palate. *Otolaryngol Clin North Am* 2007;40(1):27–60.

2. Grayson BH, Cutting CB: Presurgical nasoalveolar orthopedic molding in primary correction of the nose, lip, and alveolus of infants born with unilateral and bilateral clefts. *Cleft Palate Craniofac J* 2001;38(3):193–8.

3. Gundlach KKH: Etiology, prevalence, growth and trends in cleft lip, alveolus and palate. In *Maxillofacial Surgery*. Booth PW, Schendel SA, Hausamen J-E, eds. Churchill Livingstone, Edinburgh: 1999, 991–1003.

4. Mommaerts Y: The traditional 'Millard' approach to lip and palate repair. In *Maxillofacial Surgery*. Booth PW, Schendel SA, Hausamen J-E, eds. Churchill Livingstone, Edinburgh: 1999, 1029–45.

5. Mulliken JB: Primary repair of bilateral cleft lip and nasal deformity. *Plast Reconstr Surg* 2001;108(1):181–94.

6. Schendel SA: Unilateral cleft lip repair–state of the art. *Cleft Palate-Craniofac J* 2000;37(4):335–41.

7. Sullivan WG: Respiratory distress following cleft lip repair: the role of obligatory nasal breathing in the infant. *Ann Plast Surg* 1988;20(6):590–2.

8. Winters JC, Hurwitz DJ: Presurgical orthopedics in the surgical management of unilateral cleft lip and palate. *Plast Reconstruct Surg J* 1995;95(4):755–64.

Palatoplasty

Surgical Considerations

Description: Cleft palate can be seen as either an isolated condition or in conjunction with clefting of the lip. The mildest form of cleft palate is the submucous, or occult cleft, in which there is no visible cleft but, rather, a nonunion of the soft-palate muscles. This is followed by the incomplete soft-palate cleft and, finally, the complete cleft, which includes soft and hard palates and may extend through the alveolar portion of the maxilla. Repair involves mobilizing the lateral soft tissue and moving it toward the midline to close the cleft and elongate the palate, if necessary. The most important goal of cleft-palate repair is the attainment of normal speech. Children with unrepaired or inadequately repaired clefts develop nasal-sounding speech patterns termed *rhinolalia*. Cleft-palate repair, therefore, usually is done when the child is 9–18 months old, before consequential speech development. In addition to closing the cleft itself, an important goal of palate repair is normal anatomic approximation of the levator palati muscles, which are responsible for oronasal valving in speech and swallowing. The cleft palate is closed by elevating the mucoperiosteum from the underlying bones and approximating it in the midline (**von Langenbeck technique**) (Fig. 12.8-7) or using a V-Y type of retrodisplacement and closure (**Wardill-Kilner technique**). In either method, the levator muscles are specifically dissected and the levator sling is reconstructed. A layered closure usually is accomplished, including repositioning of the uvular muscles.

There are several different approaches to the muscle reconstruction in the soft palate, generally termed **intravelarveloplasties.** Z-plasty of the soft palate, also called a **Furlow procedure,** (Fig. 12.8-8) has been used to lengthen the palate and reorient the palatal muscles across the cleft. The other procedures basically involve direct closure of the muscles and a push-back to lengthen the palate.

Usual preop diagnosis: Cleft palate

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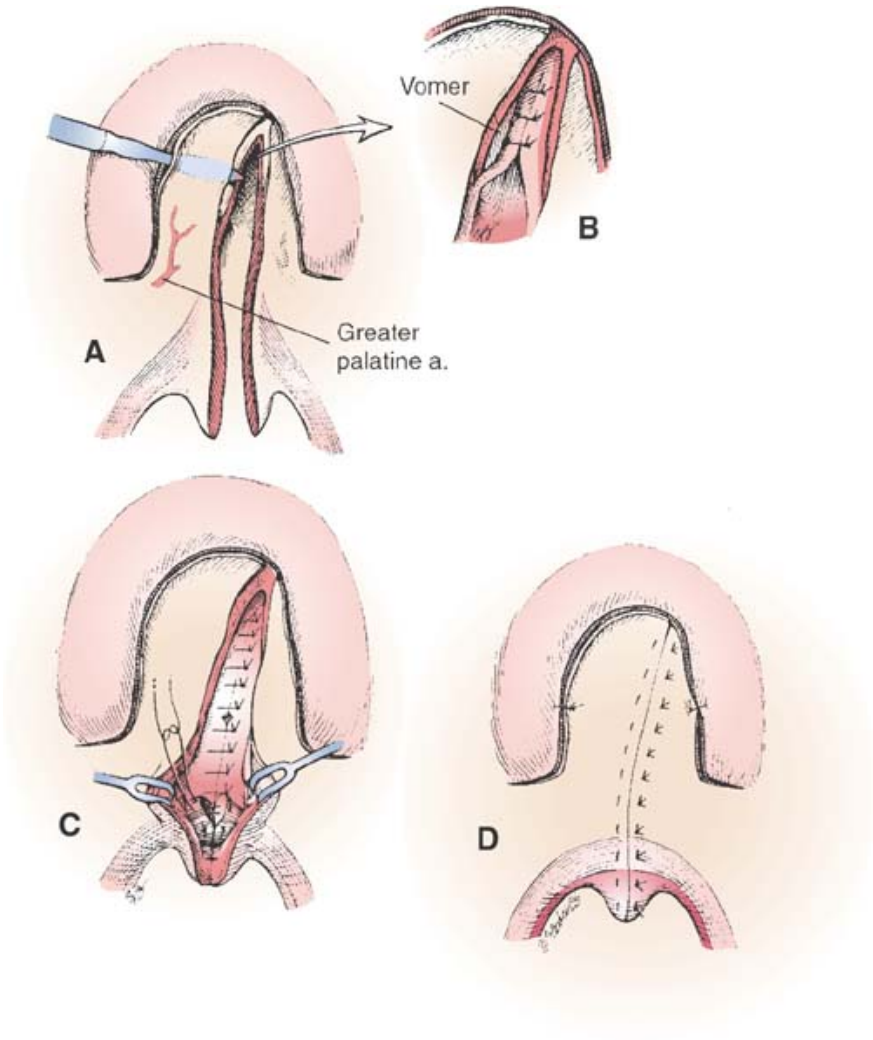

Figure 12.8-7. 7. Palatoplasty technique. **(A)** Cleft palate closure after healing of gingivoperiosteoplasty at 11–12 mo of age. Bilateral, unipedicled mucoperiosteal flaps, based on the greater palatine arteries, are elevated. **(B)** Anteriorly, the nasal floor is repaired by suturing the vomerine mucosa to the nasal mucosa on the cleft side. **(C)** The levator muscles are dissected free from the oral and nasal mucosa and released from the posterior edge of the hard palate. The levator muscles are approximated to each other in the midline. **(D)** The oral mucosa is reapproximated in the midline with interrupted horizontal mattress sutures. (Reproduced with permission from Aston SJ, Beasley RW, Thorne CH, eds: *Grabb and Smith's Plastic Surgery,* 5th edition. Lippincott-Raven, Philadelphia: 1997.)

Summary of Procedures

Position	Supine; table rotated 90°–180° with oral RAE tube extending down the midline of the lower jaw and taped to the chin.
Incision	Edges of the cleft palate and, possibly, the alveolar and pterygomandibular raphe areas.
Special instrumentation	Dingman mouth gag (when setting the gag, communication between surgeon and anesthetist allows ETT compression to be noted early); usually, a headlight; oropharyngeal pack
Unique considerations	The gag may be released intermittently to allow reperfusion of the tongue; each manipulation may affect the ETT. Minimal-to-moderate amount of blood in the oropharynx at end of procedure—should be carefully suctioned. Also, there may be some respiratory difficulties on emergence. Traction with a tongue suture often proves helpful in restoring patient's airway (maintained 24 h). Usually, oral or nasopharyngeal airways should not be placed in children.

Antibiotics	Cefazolin 25 mg/kg q 5–6 h (up to 1 g) × 5 d
Surgical time	1–1.5 h
Closing considerations	Child should not wake up crying and hypertensive. Tongue suture may be placed prior to extubation. **NB:** Dingman gag will stick to the ETT; therefore, holding the tube with a forceps deep in the oropharynx and careful removal of the gag avoids accidental extubation.
EBL	50 mL
Postop care	Elbow restraints; PACU → room; → home next day. OG tube placed, suctioned, and removed.
Mortality	Rare
Morbidity	Recurrent bleeding (requiring early return to OR): Rare Hematoma under palate Dehiscence of palate
Pain score	4

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Patient Population Characteristics

Age range	6–18 mo
Male:Female	1:3 (isolated cleft palate)
Incidence	1/1,000
Etiology	Failure of fusion of the palatal shelves from anterior to posterior. (Can be due to a persistent high-tongue position in utero, increased facial width, reduced facial mesenchyme and/or drugs such as steroids, anticonvulsants, and benzodiazepines, or infection.)
Associated conditions	Multiple associated conditions. Most common is the Pierre Robin syndrome, in which cleft palate is found in association with glossoptosis and a micrognathic retruded mandible. These children frequently have airway obstruction and, even at an older age, may have sleep apnea. Other associations include Klippel-Feil syndrome, Treacher Collins syndrome, CHD, chronic URI, chronic otitis media, subglottic stenosis.

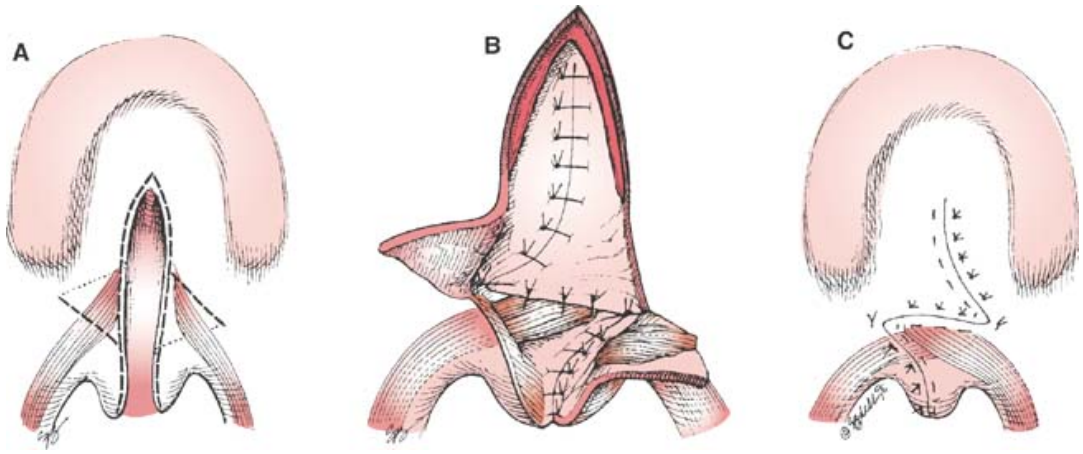

Figure 12.8-8. 8. Double opposing Z-plasty closure of an isolated cleft palate. **(A)** Design of the incisions. **(B)** Muscle included in the posteriorly based flap. **(C)** Final result with recreation of the levator sling. (Reproduced with permission from Aston SJ, Beasley RW, Thorne CH, eds: *Grabb and Smith's Plastic Surgery,* 5th edition. Lippincott-Raven, Philadelphia: 1997.)

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Anesthetic Considerations

See Anesthetic Considerations for Lip and Nose Surgery, p. 1421.

Suggested Readings

1. Arosarena OA: Cleft lip and palate. *Otolaryngol Clin North Am* 2007;40(1):27–60.

2. Gundlach KKH: Etiology, prevalence, growth and trends in cleft lip, alveolus and palate. In *Maxillofacial Surgery*. Booth PW, Schendel SA, Hausamen J-E, eds. Churchill Livingstone, Edinburgh: 1999, 991–1003.

3. Kirschner RE, Wang P, Jawad AF, et al: Cleft-palate repair by modified Furlow double-opposing Z-Plasty: *The Children's Hospital of Philadelphia Experience, Plastic and Reconstructive Surgery* 1999;104(7):1998–2014.

4. Mommaerts Y: The traditional 'Millard' approach to lip and palate repair. In *Maxillofacial Surgery*. Booth PW, Schendel SA, Hausamen, J-E, eds. Churchill Livingstone, Edinburgh: 1999, 1029–45.

Pharyngoplasty

Surgical Considerations

Description: Following the initial repair of palatal clefts, some children or young adults demonstrate continued hypernasal speech patterns, a condition called "velopharyngeal incompetence." This can be 2° a short soft palate, a large nasopharynx, or a soft palate that has inadequate movement either 2° scarring or due to neurogenic problems. The typical repair would be a superiorly based **pharyngeal flap** (Fig. 12.8-9) to the soft palate.

Variant procedure or approaches: The **Jackson modification of the Orticochea flap** uses the posterior tonsillar pillars, which consist of the palatopharyngeus muscle and overlying mucosa, to create a competent oronasal *(Print pagebreak 1415)* sphincter (Fig. 12.8-10). The flaps are based superiorly and repositioned horizontally to meet above and behind the soft palate. Although they act to augment the posterior pharyngeal wall, they also are intended to maintain their innervation and, therefore, augment sphincter activity. A posterior pharyngeal wall implant also may be placed.

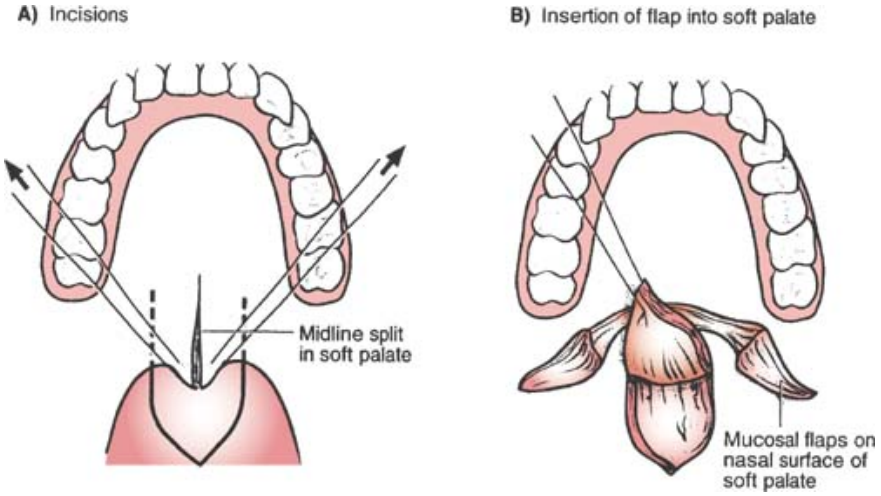

Figure 12.8-9. 9. Pharyngeal flap, superiorly based. (Reproduced with permission from Booth PW, Schendel SA, Hausamen J-E, eds: *Maxillofacial Surgery*. Churchill Livingstone, Edinburgh: 1999.)

Usual preop diagnosis: Velopharyngeal incompetence

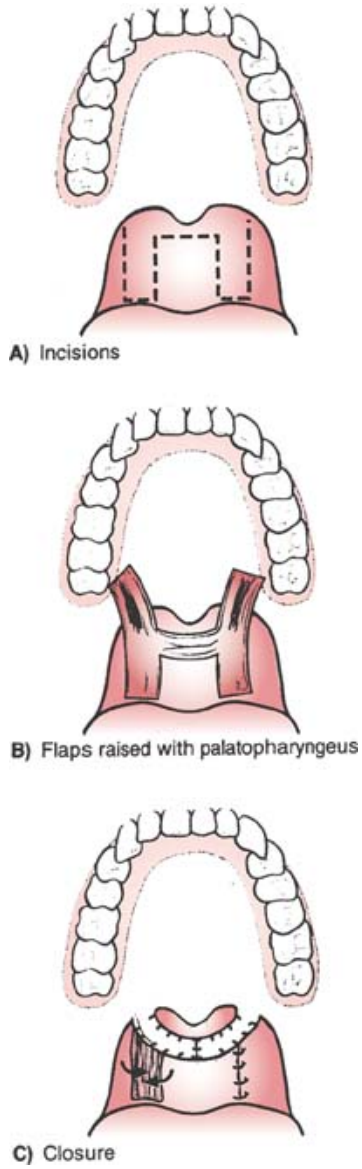

Figure 12.8-10. 10. Sphincter pharyngoplasty. (Reproduced with permission from Booth PW, Schendel SA, Hausamen J-E, eds: *Maxillofacial Surgery*. Churchill Livingstone, Edinburgh: 1999.)

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Summary of Procedures

Position	Supine, table rotated 90° or 180°; oral RAE tube extending down the midline chin and taped in position.
Incision	Involves incisions in soft and hard palates and in the posterior pharyngeal wall.
Special Instrumentation	Dingman mouth gag (caution with removal and manipulation re ETT); headlight
Unique considerations	Avoid oral or nasopharyngeal airways or nasal suctioning.
Antibiotics+ other meds	Cefazolin 25 mg/kg q 6–8 h (up to 1 g) × 3 d; periop steroids, depending on preop airway.
Surgical time	1–1.5 h
Closing considerations	Pediatric patients should not become hypertensive (↑ bleeding). There will be some nasopharyngeal drainage; thorough oral suctioning is important.
EBL	50–100 mL

Postop care	Avoid postop oral or nasopharyngeal airways or nasal suctioning. Be aware of possible occlusion of nasopharynx with flap and bleeding. Tongue suture can be placed for 24 h.
Mortality	Rare
Morbidity	Recurrent bleeding Hematoma under palate Dehiscence of palate Nasopharyngeal obstruction Secondary sleep apnea
Pain score	4

Patient Population Characteristics

Age range	3–11 yr most common
Male:Female	1:1
Incidence	15% of children undergoing cleft palate repair will need some type of secondary palatal lengthening procedure after 3 yr.
Etiology	Short and scarred palate; neurogenic palate; palate-to-pharyngeal ratio that is too small.
Associated conditions	Sleep apnea; Pierre Robin sequence, with gloscoptosis and micrognathia. Treacher Collins syndrome; microtia with craniofacial malformation; subglottic stenosis; CHD. Of special interest: some patients with cleft palate may have **velocardiofacial syndrome.** These children may have **medially displaced internal carotid arteries,** placing these major arteries in harm's way during dissection along the posterior pharyngeal wall.

Anesthetic Considerations

See Anesthetic Considerations for Lip and Nose Surgery, (p 1421).

Suggested Readings

1. Boorman JG, Bharathwaj S: Secondary palatal surgery and pharyngoplasty. In *Maxillofacial Surgery*. Booth PW, Schendel SA, Hausamen J-E, eds. Churchill Livingstone, Edinburgh: 1999, 1083–99.

2. Jackson IT: Sphincter pharyngoplasty. *Clin Plastic Surg* 1985;12(4):711–17.

(Print pagebreak 1417)
3. Markus AF, Precious DS: Secondary surgery for cleft lip and palate. In *Maxillofacial Surgery*. Booth PW, Schendel SA, Hausamen J-E, eds. Churchill Livingstone, Edinburgh: 1999, 1057–72.

4. Sloan GM: Posterior pharyngeal flap and sphincter pharyngoplasty: the state of the art. *Cleft Palate Craniofac J* 2000; 37 (2):112–22.

Also see Suggested Readings for Cleft Lip, (p 1421).

Alveolar Cleft Repair With Bone Graft

Surgical Considerations

Description: Alveolar cleft occurs as both bony and soft-tissue defects in the alveolar portion of the maxilla in the position of the lateral incisor tooth; thus, an oral/nasal fistula exists with this deformity. The size of the cleft is variable; it may be unilateral or bilateral and is associated with cleft lip and palate. The alveolar segments are often collapsed such that orthodontic expansion is required before bone graft and repair. These devices are maintained to stabilize the graft in situ for a 3-month healing period. The surgical procedure involves raising mucosal-gingival-periosteal flaps, advancing them, and performing a layered closure, starting with the nasal floor and working toward the oral cavity. A bone graft is placed in between these two layers to consolidate the upper arch. Cancellous bone usually is taken from the iliac crest or corticocancellous bone from the outer table of the skull.

Most commonly, the bone is harvested from the ilium. This can be accomplished via limited access and a trephine or via an open technique, depending on the amount of bone required. This portion of the procedure, especially by open technique, can add 50–100 mL of blood loss. Most nasal and lip revision surgery should be put off until the alveolus is reconstructed, since this is the base on which the lip and nose sit.

Variant procedure or approaches: In young children, the alveolar cleft procedure may be performed without the use of bone grafts at the time of lip or hard palate closure (**gingivoalveoloplasty,** Fig. 12.8-11) with the hope that preop alignment of the clefted alveolus and periosteal creation of new bone will fill the bony defect and allow subsequent normal tooth eruption. This is not always complete, and some of these children will need later bone grafting at age 7–8 years, before eruption of the permanent canine teeth.

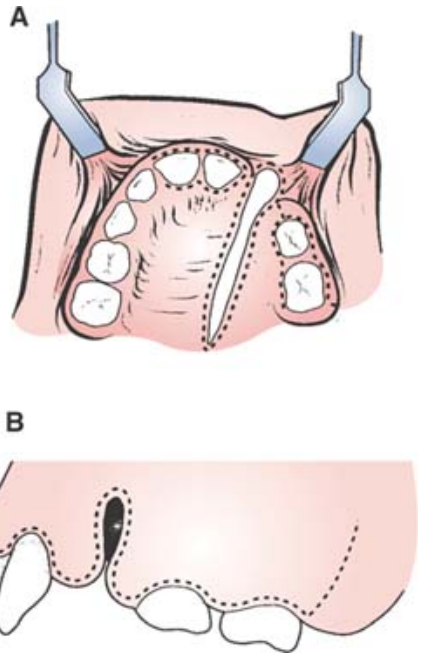

Figure 12.8-11. 11. Gingivoalveoloplasty (GAP). Gingival and mucosal incisions are shown on the palate **(A)** and vestibular **(B)** surfaces, extending along the cleft borders. (Reproduced with permission from Booth PW, Schendel SA, Hausamen J-E, eds: *Maxillofacial Surgery*. Churchill Livingstone, Edinburgh: 1999.)

Usual preop diagnosis: Congenital alveolar cleft

(Print pagebreak 1418)

Summary of Procedures

Position	Supine; table rotated 90°–180° ± roll under hip (bone harvest)
Incision	Oral, with the addition of iliac crest incision or scalp incision, either parasagittal or coronal
Special instrumentation	Throat pack; Dingman mouth gag; headlight. Two instrument setups used, to prevent cross-contamination from oral to iliac surgical sites.

Unique considerations	Important to ensure that the hip iliac crest bone graft site is on the opposite side from the anesthesiologist if the table is rotated only 90°. Midline oral RAE tube to chin. Care when manipulating Dingman, as it sticks to ETT.
Antibiotics	Cefazolin 25 mg/kg (up to 1 g) iv preop
Surgical time	1.5–2.5 h
Closing considerations	***NB:** Ensure that throat pack has been removed. Pediatric patients should not wake up in agitated state. Noncleft-side oral mouth gag and gentle oral suctioning permissible; avoid nasal suctioning, especially from the cleft side.
EBL	100–200 ml
Postop care	PACU → ward; walking POD 1 or 2 post-iliac graft.
Mortality	Rare
Morbidity	Bone graft loss: 2–10% Infection: 2–10% Refistulization: 2–10% Prolonged hip discomfort (bone graft donor site) Bleeding—oral or at bone donor site
Pain score	6

Patient Population Characteristics

Age range	8–12 yr
Male:Female	2:1
Incidence	Unknown
Etiology	Multifactorial, including both genetic and environmental aspects
Associated conditions	Associated anomalies are seen in 29% of cleft lip cases; and may include major chromosomal deletions and/or duplications, with the possibility of severe developmental delay, CHD.

Anesthetic Considerations

See Anesthetic Considerations for Lip and Nose Surgery, (p. 1421).

Suggested Readings

1. Brusati R, Mannucci N: Primary repair of the lip and palate using the Delaire philosophy. In *Maxillofacial Surgery*. Booth PW, Schendel SA, Hausamen J-E, eds. Churchill Livingstone, Edinburgh: 1999, 1005–28.

2. Posnick JC: *Craniofacial and Maxillofacial Surgery in Children and Young Adults*. WB Saunders, Philadelphia: 2000, 785–980.

3. Stassen LFA: Alveolar bone grafting–how I do it. In *Maxillofacial Surgery*. Booth PW, Schendel SA, Hausamen J-E, eds. Churchill Livingstone, Edinburgh: 1999, 1047–55.

4. Wolfe SA, Kawamoto HK: Taking the iliac bone graft: a new technique. *J Bone Joint Surg* 1978;60-A(3):411.

Also see Suggested Readings for Cleft Lip. p. 1421.

Secondary Cleft Lip/Nasal Surgery

Surgical Considerations

Description: Secondary deformities of the nose and lip develop, following the initial repair of either bilateral or unilateral cleft lip deformities. These subsequent deformities depend on the extent of the initial congenital anomaly, *(Print pagebreak 1419)* the quality of the surgical repair, and resulting oral/facial function. Revision can vary from a minimal scar revision (Fig. 12.8-12), to a complete opening and reconstruction of the lip and nose, with or without ancillary procedures such as **septorhinoplasty ± cartilage grafting, forked flaps** (Fig. 12.8-13), **fascial lip augmentation, or Abbe-Estlander flap** (lip-switch flap) (Fig. 12.8-14).

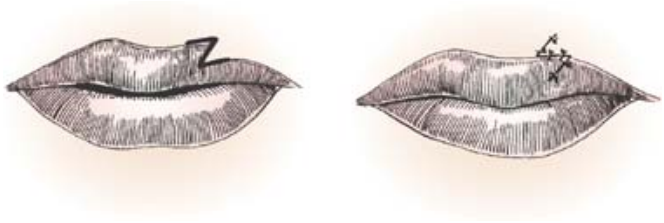

Figure 12.8-12. 12. Z-plasty to correct notching of the vermillion border. (Reproduced with permission from Aston SJ, Beasley RW, Thorne CH, eds: *Grabb and Smith's Plastic Surgery,* 5th edition. Lippincott-Raven, Philadelphia: 1997. Originally from Millard DR Jr: *Cleft Craft: The Evolution of Its Surgery*. Little & Brown, 1976.)

Occasionally, the individual born with a cleft lip and palate is severely deficient in tissue of the upper lip. This occurs most frequently in the bilateral condition. Correction involves switching tissue from the midline of the lower lip to the central portion of the upper lip, maintaining a pedicle of soft tissue between the lips, which usually contains the labial artery on one side. This pedicle normally is cut between 7–11 days. The redundant tissue in the mid portion of the upper lip is transferred to the columellar portion of the nose at the same time, which elongates this section (Fig. 12.8-14). To avoid disruption of the flap, the older child should be cautioned to avoid wide mouth opening in the postop period.

Usual preop diagnosis: Secondary cleft deformity

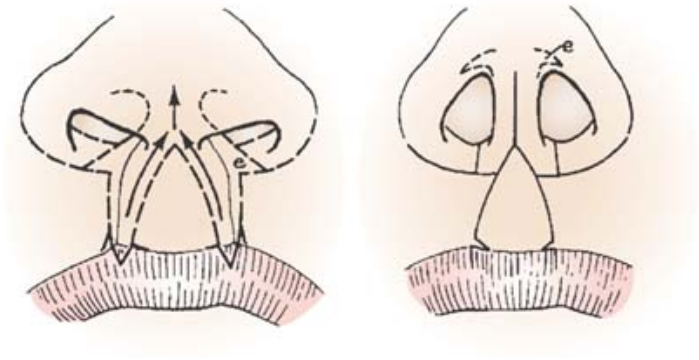

Figure 12.8-13. 13. Short columella associated with the bilateral cleft nose, elongated by forked flaps. (Reproduced with permission from Aston SJ, Beasley RW, Thorne CH, eds: *Grabb and Smith's Plastic Surgery,* 5th edition. Lippincott-Raven, Philadelphia: 1997.)

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Summary of Procedures

Position	Supine, table rotated 90°–180°; oral RAE; nasal RAE tube when working on lips only (especially if lips are held closed by a flap pedicle, making extubation more difficult).
Incision	Variable, in lip or nasal areas. (See Figs 12.8-12, 12.8-13, 12.8-14.)
Special instrumentation	Throat pack; possibly rhinoplasty instruments; oral RAE tube; head light; loupe magnification.

Unique considerations	Procurement of a cartilage graft is usually from the nasal septum or ear; thus, the head may need to be turned to one side and this area also prepped. Elbow restraints for children. A switch flap leaves the lips connected at their midportion by a thin, easily damaged, soft-tissue pedicle; therefore, the patient should wake up unagitated.
Antibiotics	Cefazolin 25 mg/kg (up to l g) iv preop
Surgical time	Variable, depending on the extent of revision: 0.5–3 h
Closing considerations	***NB:** Remove throat pack.
EBL	Minimal–75 mL
Postop care	May be obligatory mouth breathing post-rhinoplasty, or lips may be held mostly closed by the pedicle and swelling of a lip-switch flap; consider oral-pharyngeal airway; avoid wide mouth opening. PACU → room; elbow restraints in young children.
Mortality	Rare
Morbidity	Infection Wound breakdown Flap necrosis: < 1% (with lip switch) Bleeding
Pain score	3–5 (depending on extent of procedure)

Patient Population Characteristics

Age range	2–50 yr
Male:Female	1:1
Incidence	20–60% of patients with primary clefts
Etiology	Unsatisfactory outcome of previous lip/nose surgery
Associated conditions	Associated anomalies are seen in 29% of the clefts and may include major chromosomal deletions or duplications, with a possibility of severe mental retardation.

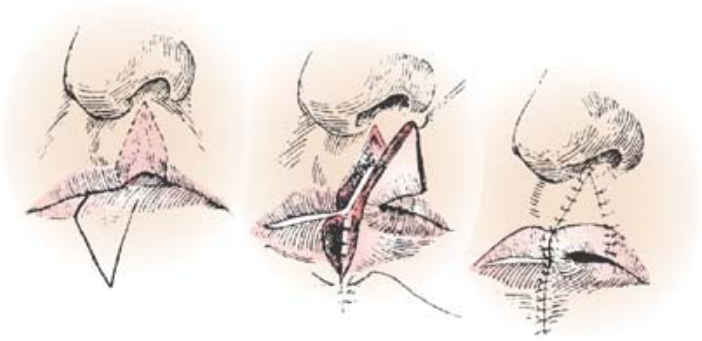

Figure 12.8-14. 14. Abbe-Estlander flap. Note lips sutured together. (Reproduced with permission from Converse JM, ed: *Reconstructive Plastic Surgery,* Vol 3, 2nd edition. WB Saunders, Philadelphia: 1977.)

(Print pagebreak 1421)

Anesthetic Considerations

See Anesthetic Considerations for Lip and Nose Surgery, p. 1421.

Suggested Readings

1. Palmisano BW: Anesthesia for plastic surgery. In: *Pediatric Anesthesia,* 3rd edition. Gregory GA, ed. Churchill Livingstone, New York: 1994, 699–741.

2. Sadove AM, Eppley BL: Correction of secondary cleft lip and nasal deformities. *Clin Plastic Surg* 1993;20(4):793–801.

3. Talmant JC: Cleft rhinoplasty. In *Maxillofacial Surgery*. Booth PW, Schendel SA, Hausamen J-E, eds. Churchill Livingstone, Edinburgh: 1999, 1133–71.

Anesthetic Considerations for LIP and Nose Surgery

(Procedures covered: cleft lip repair; palatoplasty; pharyngoplasty; alveolar cleft repair with bone graft; secondary cleft lip/nasal surgery)

Preoperative

The anesthesiologist should be aware of the parents' feelings about their child with a congenital malformation. The whole family needs to be treated with sensitivity and compassion. Cleft lip closure may be carried out as early as the first week of life in the healthy neonate; however, many surgeons and anesthesiologists find the ‘rule of ten’ helpful: the child should have an Hb >10 g, be 10 wk old, and weigh 10 lbs. The hard palate usually is closed between the ages of 1–5 yr; however, the soft palate should be closed prior to speech development (12–15 mo). **Palatoplasty** and **pharyngoplasty** usually are carried out from 1–15 years. Patients with these midline facial defects are most likely to have other associated anomalies, including CHD, subglottic stenosis, and Pierre-Robin or Treacher-Collins syndromes. In patients with severe OSA, post-op edema may impair ventilation. The possibility of remaining instubated post-op should be discussed with the surgical team and the family.

Respiratory	Careful assessment is necessary as associated anomalies may affect airway or lungs. Chronic otitis 2° eustachian tube dysfunction is common. Treat with antibiotics before surgery. Postpone surgery (2 wk) if Sx of acute URI present (e.g., runny nose, fever, sore throat, cough). Chronic aspiration may be associated with cleft lip/palate. **Tests:** as indicated from H&P.
Airway	Be aware of other congenital anomalies affecting the airway, such as Apert, Goldenhar, Klippel-Feil, Pierre Robin, or Treacher Collins syndromes. Review any previous anesthetic records for insights into airway management. Consider fiber optic intubation (FOI) in patients with suspected difficult airway. Also consider elective tracheostomy under local anesthesia in patients with severe airway abnormalities. Patients with severe subglottic stenosis may require preop tracheostomy. Patients with difficult airways or severe obstructive sleep apnea undergoing palatal surgery should be evaluated for ICU monitoring postoperatively.
Cardiovascular	CHD is frequently associated with cleft palate. Preop evaluation of a patient with a known or suspected heart defect should include thorough H&P, EKG, Hct, baseline O_2sat, and CXR. For children with Sx of cardiac dysfunction or those requiring cardiac medication, it is advisable to consult with a pediatric cardiologist to optimize the patient's condition prior to surgery. **Tests:** Preop ECG indicated for patients with CHD; others as indicated from H&P.
Nutritional	Infants with cleft lip/palate may have problems with oral feeding. Assess nutritional status from physical exam and by comparison to expected growth for age. **NB:** NPO after midnight for solids. Patients should continue to have clear liquids up until 2 h preop.

Neurological	Delayed development of speech is common in the older child with cleft palate. Some of these children may be hearing impaired. Preop preparation and discussion is important to minimize the impact of these communication problems.
Psychological	Many patients with orofacial congenital malformations require multiple procedures; emotional support and psychological assessment of these patients are essential. Preoperative visits and consultation with play-therapists can be helpful, particularly if the child with a difficult airway will need a preoperative IV.
Hematologic	High incidence of iron deficiency anemia; T&C for 1 U PRBC (cleft palate).
Laboratory	**Tests:** HCT Other tests as indicated from H&P.
Premedication	< 9 mo old rarely needs premedication; > 9 mo old, may benefit form midazolam (0.5–0.75 mg/kg po); alternatively ketamine (5 mg/kg po) 30 min preop may be used.

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Intraoperative

Anesthetic technique: GETA; warm room to 75–80°F; forced air warmer.

Induction	Typically, an inhalational induction (sevoflurane ± N_2O/O_2) while patient is breathing spontaneously. Airway obstruction is best treated with an oral airway. Anticipate difficult laryngoscopy if large, prepalatal cleft present. Intubate with oral RAE tube and secure in midline of lower lip. In patients with difficult airways, preoperative IV and FOI is the technique of choice. IV glycopyrollate and nebulized local anesthetics may facilitate FOI. Avoid muscle relaxants for difficult intubations until ETT is placed. For children with difficult airways and a small posterior pharynx, consider preop dexamethasone 0.5 mg/kg IV to decrease palatal edema. After induction, consider morphine 0.1 mg/kg, ketamine 0.1 mg/kg for analgesia.
Maintenance	Standard pediatric maintenance (see p. D-3) ± muscle relaxant. Airway is shared with the surgeons. The Dingman mouth gag is used for surgical exposure and may inadvertently compress the ETT or cause an endobronchial intubation. Monitor PIP before and after placement of Dingman. Flexion of the neck also may cause endobronchial intubation. Extension of the neck may cause complete or partial extubation. Adequacy of ventilation should be checked after every position change. Bilateral breath sounds should be equal after final positioning. In cases with the table turned 180°, a precordial stethoscope placed over the left chest can help elucidate endobronchial tube migration. ETT should be sutured to the alveolar ridge. In palatoplasty, the palate is infiltrated with epinephrine (in lidocaine usually) →↓ blood loss + ↑ dysrhythmias (halothane > sevoflurane). ↑ $PaCO_2$↑↑ dysrhythmias.
Emergence	*Pharyngeal (throat) packs are usually placed to prevent aspiration of blood. **NB:** Packs must be removed before extubating the trachea. Consider laryngoscopy to inspect airway and remove blood and clots before extubation. A tongue stitch is useful postop following cleft palate surgery. It may be used to pull the tongue forward to relieve postop respiratory obstruction. Extubation in the lateral (tonsillar) position is useful in promoting drainage of blood and secretions.
Blood and fluid requirements	IV: 18–20 ga × 1 NS/LR @: 4 mL/kg/h – 0–10 kg + 2 mL/kg/h –11–20 kg + 1 mL/kg/h – > 20 kg (e.g., 25 kg = 65 mL/h) Blood loss replaced by 3:1 crystalloid or 1:1 colloid (e.g., 5% albumin or 6% hetastarch). Rarely, a blood transfusion may be indicated for hemorrhage.
Monitoring	Standard monitors (see p. D-1)

Positioning		and pad pressure points. eyes.
Complications	Obstructed ETT→↑ PIP Mucous plugging Hemorrhage	ETT to see that it is not partially or completely obstructed by mouth gag. bilateral breath sounds.

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Postoperative

Complications	Retained throat pack Airway edema → croup Hemorrhage Obstructive sleep apnea	for retained throat pack if there are Sx of airway obstruction in immediate postop period. Rx of postintubation croup consists of cool, humidified, 100% O_2 mask, or nebulization 2.25% racemic epinephrine (0.5 ml in 3 mL NS). Racemic epinephrine is given for its vasoconstrictor, rather than its bronchodilator, effect. If posterior pharyngeal edema is present, consider dexamethasone 0.5 mg/kg iv.
Pain management	Acetaminophen (10–15 mg/kg po or 20–30 mg/kg pr) q6H Fentanyl 1 mcg/kg iv; Morphine 0.05–0.1 mg/kg iv q 2–3 h prn	Avoid oversedation in patients with Abbe-Estlander repair 2° airway obstruction. Aim for child to be mildly sedated (decreases bleeding), but not obstructing; intraop morphine, ketamine helpful to ensure smooth awakening.
Tests	Hct, if indicated.	Others as indicated.

Otoplasty

Surgical Considerations

Description: There are a number of congenital ear malformations. The two most frequently encountered in the OR are **prominent ear** and **microtia.** Both conditions can be unilateral or bilateral.

Prominent ears are usually an isolated finding. The ear is examined in thirds to determine where the prominence lies, and the surgery is tailored to correct the specific excesses. The antihelical fold is usually flattened and requires reshaping. The prominence of the ear, as measured by its projection from the mastoid process, is decreased accordingly. This usually involves an elliptical skin incision in the posterior ear area, dissection over the mastoid, and one, or a combination of three techniques—mattress sutures, cartilage scoring, and/or resection.

Variant procedure or approaches: All procedures are similar, with minor differences in suturing and amount of resected tissue. In addition to the posterior incisions, an anterior incision can be used in some approaches.

Microtia is within the congenital anomaly spectrum of hemifacial microsomia, and the associated facial malformation may include a small asymmetric jaw, creating a difficult intubation. Reconstruction is most often accomplished with autologous rib graft as a multistaged procedure. This donor site comes with the attendant risks of pneumothorax and hemothorax (Fig. 12.8-15). Stage one is the creation of a cartilaginous framework, with placement into a cutaneous pocket symmetric with the normal ear, if present (Fig. 12.8-16). Stage one is accomplished once the rib cartilage has grown to sufficient size—usually, 6–7 years of age. Stage two requires transposition of the lobule 3 months after stage one. Stage three is the elevation with skin graft of the framework from the head posteriorly. Stage four is the creation of a tragus and conchal excavation.

Variant procedure or approaches: Recently, **porous polyethylene implants** have been used to avoid donor-site morbidity, and **temporoparietal fascial flaps** provide coverage to avoid alloplastic extrusion.

Usual preop diagnosis: Ear malformation; prominent ears; microtia; anotia

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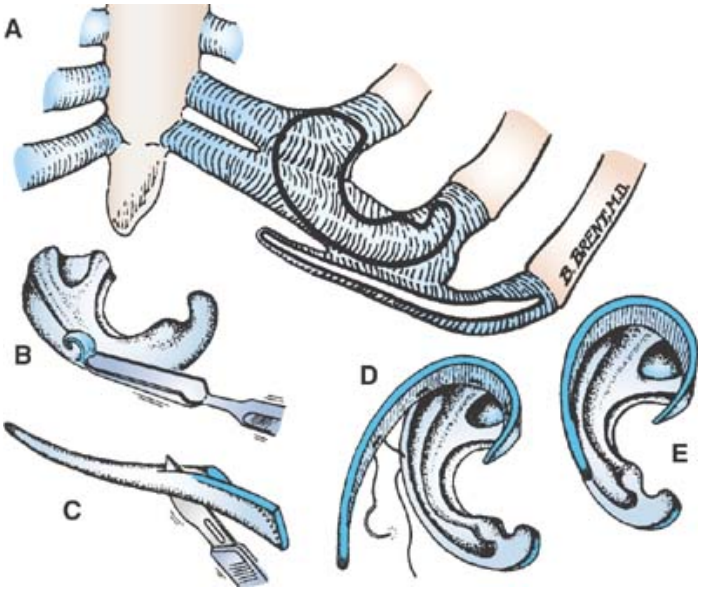

Figure 12.8-15. 15. Fabricating an ear framework from costal cartilage. **(A)** Donor site: the contralateral thorax. The helical rim is obtained from a 'floating' rib cartilage, the main pattern from the synchondrosis of two cartilages. **(B)** Sculpting the main block. **(C)** Thinning the 'floating' rib cartilage to produce a delicate helical rim. **(D)** Affixing the rim to the main framework block. **(E)** Completed framework. (Reproduced with permission from Aston SJ, Beasley RW, Thorne CH, eds: *Grabb and Smith's Plastic Surgery,* 5th edition. Lippincott-Raven, Philadelphia: 1997.)

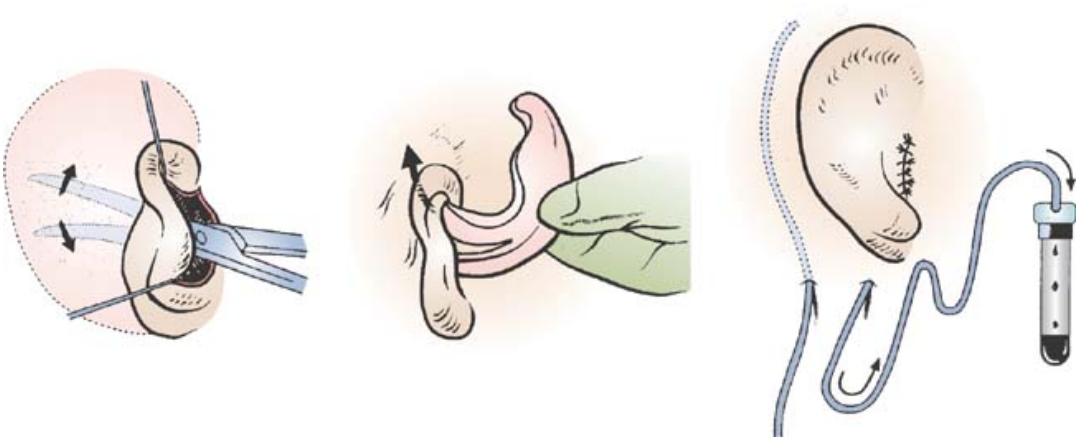

Figure 12.8-16. 16. The cutaneous "pocket." The vestigial native cartilage is excised; then a skin pocket is created. To provide tension-free accommodation of the framework, the dissection is carried out well beyond the proposed auricular position. Using two silicone catheters, the skin is coapted to the framework by means of vacuum tube suction. (Reproduced with permission from Booth PW, Schendel SA, Hausamen J-E, eds: *Maxillofacial Surgery*. Churchill Livingstone, Edinburgh: 1999.)

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Summary of Procedures

Position	Supine; table rotated either 90° or 180°; oral intubation
Incision	Posterior ear; occasionally anterior ear; microtia, stage-dependent; and chest wall
Unique considerations	Head turned from side-to-side during operation.
Antibiotics	Cefazolin 25 mg/kg (up to 1 g) iv preop
Surgical time	2–4 h (depends on unilateral vs bilateral); 1st stage microtia much longer

Closing considerations	Ear dressing requires 5–10 min at end of procedure. Delicate vacuum test-tube drainage system in 1st stage microtia repair fixed to head dressing.
EBL	10–100 mL
Postop care	PACU → room
Mortality	Rare
Morbidity	**Protruding ears:** Hematoma formation: < 1% Infection: < 1 % Asymmetrical ear reduction: < 1% Suture extrusion < 1% **Microtia:** Pneumothorax or hemothorax Hematoma, infection, and skin loss: 1.6% (total) Overgrowth: 41%
Pain score	4

Patient Population Characteristics

Age range	6+ yr
Male:Female	2:1
Incidence	Unknown
Etiology	**Microtia:** 9.2% bilateral; 32% left vs 58% right; 4.9% recurrence within immediate family **Protrusion:** Also seen among family members. Most are bilateral.
Associated conditions	**Protruding ears:** none known **Microtia:** Brachial arch deformities: bone/soft tissue deficit, 36.5%; facial nerve weakness, 15.2%; cleft lip ± palate, 4.3%; macrostomia, 2.5%; urogenital defects, 4%; cardiovascular malformations, 2.5%; other, 1.6%

Anesthetic Considerations

See Anesthetic Considerations for Ear Surgery, Chapter 3.0 Otolaryngology, p 229.

Suggested Readings

1. Brent B: Ear reconstruction. In *Maxillofacial Surgery*. Booth PW, Schendel SA, Hausamen J-E, eds. Churchill Livingstone, Edinburgh: 1999, 1419–28.

2. Brent B: Technical advances in ear reconstruction with autogenous rib cartilage grafts: personal experience with 1200 cases. *Plastic Reconstruct Surg J* 1999;104(2):319–38.