

Cleft Lip and Palate

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A 12-month-old male presents for a primary cleft palate repair. The patient was recently adopted, and no significant medical history has been documented.

In the preoperative holding area, the patient is being held and appears calm. The patient's pre-operative vital signs are: weight 10 kg, T 36.7°C, pulse 110 bpm, BP 90/54, RR 28, SpO₂ 99% on room air. On physical exam, the patient is noted to have a right-sided cleft palate. The patient appears to have slight micrognathia. The remainder of the physical exam is within normal limits.

When Does Development of the Face and Lip Occur? When Does Development of the Palate Occur?

The development of the lip occurs during weeks four to eight of gestation. The primary palate (palate that is anterior to the incisive foramen) forms during week six of gestation with the secondary palate formed during weeks six to twelve. Cleft lip and alveolus results from failure of fusion of the nasal and maxillary prominences, whereas cleft palate (secondary palate) results from failure of fusion of the palatal shelves.

What Are the Predisposing Risk Factors Related to Cleft Lip and Palate?

Causes of cleft lip and palate are multifactorial and reported etiologies include genetics, maternal exposures and risk factors (smoking, alcohol, gestational diabetes, folate deficiency), as well as teratogens (retinoic acid, valproic acid, phenytoin).

What Is the Incidence of Cleft Lip and Palate?

The incidence of orofacial clefts is 1 in 700 live births. Cleft lip with or without cleft palate is more common in

males (2:1) and certain ethnic populations (Asian, Native American). Nonsyndromic forms of cleft lip with or without cleft palate account for 70% of all cases.

How Are Different Types of Cleft Lip and Palate Classified?

The classification of craniofacial clefts was described by Dr. Tessier in 1976. Orofacial clefts are the most common craniofacial cleft deformity, and multiple classification systems for orofacial clefts exist. Cleft lip and palate can occur in combination or in isolation. Cleft lip is commonly described as unilateral or bilateral, and complete or incomplete. The clinical severity of cleft palate ranges from a submucosal, often undetectable, cleft to a complete bilateral cleft of the primary and secondary palate.

What Physiological Problems Are Common for Patients with Cleft Palate?

In cleft palate patients, improper closure between the palate and nasopharynx can lead to physiological problems of feeding (unable to generate negative pressure for swallowing and sucking) and difficulties with speech and language development. Eustachian tube dysfunction and middle ear disease related to abnormal insertion of the tensor veli palatini can result in conductive hearing loss. Inability to oppose maxillary and mandibular teeth leads to the inability to properly chew food.

When Is a Cleft Lip and/or Palate Typically Repaired?

Primary cleft lip repair is typically performed at around age 10–12 weeks. Historically, the “rule of 10s” described the preoperative parameters of weight over 10 lbs, hemoglobin over 10 g/dL, and age over 10 weeks. Primary cleft palate repair is typically

performed around age 9–12 months in order to optimize speech and language development.

What Other Procedures Are Common for These Patients?

Eustachian tube dysfunction, middle ear disease, and conductive hearing loss may be associated with cleft palate; these patients often require placement of myringotomy tubes. In the long term, these patients may require multiple procedures including lip and nasal revisions, additional palate surgery for velopharyngeal insufficiency, alveolar bone grafting, dental procedures, maxillary advancement, and cosmetic septorhinoplasty. Orthodontic intervention is usually required following palate repair to improve maxillary hypoplasia and crowding of the teeth.

What Is Velopharyngeal Insufficiency, and How Is It Treated?

Velopharyngeal insufficiency (VPI) is the insufficient separation of the nasopharynx from the oropharynx due to a shortened palate. During speech, escape of air through the nose can lead to several speech disorders including hypernasal speech and inability to produce specific speech sounds. Surgical treatment for VPI involves lengthening of the palate by palatal revision, pharyngeal flap, or pharyngoplasty (bringing the posterior pharyngeal wall forward with an artificial implant or fat injection).

What Are the Important Preoperative Considerations for Cleft Lip or Palate Repair?

Preoperative evaluation of all pediatric patients includes a detailed history of birth and family history, review of systems, and recent illnesses. Cleft patients may have chronic congestion that should be differentiated from an acute illness. A thorough physical examination should be performed, with a focus on the head and neck examination, as well as possible associated malformations. Attention should be placed on facial asymmetry, external ear anomalies, and crowding of maxillary teeth in teenage patients. If a particular syndrome or genetic diagnosis is suspected, further preoperative workup may be warranted. Laboratory studies are typically not warranted unless clinically indicated.

What Are the Important Components of a Pediatric Airway Exam?

Physical exam findings that might indicate a possible difficult airway include micrognathia, limited mouth opening, limited neck extension, and facial asymmetry. Micrognathia is associated with posterior regression of the tongue and a small hyomental space, making laryngoscopy difficult. Certain ophthalmic, auricular, and facial anomalies can occur in specific syndromes associated with difficulty with ventilation or tracheal intubation. Cleft alveolus, protruding premaxilla, and high-vaulted arch deformities contribute to difficult intubation in pediatric patients. Intubation may be challenging in patients with combined bilateral cleft lip and alveolus.

What Syndromes Are Commonly Associated with Cleft Lip/Palate?

Many syndromes are known to be associated with orofacial clefts. They include Trisomy 21 (Down syndrome), Nager syndrome, Treacher Collins syndrome, Stickler syndrome, hemifacial microsomia, 22q deletion syndromes, Goldenhar syndrome, and Pierre Robin sequence.

Clinical features of Pierre Robin sequence include micrognathia, glossoptosis, downward displacement of the base of tongue, and airway obstruction. Clinical features of Treacher Collins syndrome include mandibular hypoplasia, microtia, and eye colobomas.

Does This Child Need Premedication?

Stranger anxiety typically occurs around six to eight months of age. Premedication may be of benefit in older children, as these patients often require multiple surgical procedures throughout their lifetime and may have anxiety related to perioperative experiences. Although there are several choices of anxiolytic pre-medications, midazolam is most often chosen because of its relative safety, rapid onset with oral or nasal administration (5–10 minutes), and rapid offset (within 30 minutes). Premedication should be used with caution in patients at risk for airway obstruction as well as those with anticipated difficult airway.

How Would You Induce General Anesthesia?

Although most children's hospitals would use inhalational induction with sevoflurane in oxygen, with or without nitrous oxide, IV induction is an acceptable alternative and is used in some centers. Administration of a neuromuscular blocker to facilitate tracheal intubation is optional, though only after the proven ability to provide positive pressure ventilation. Many centers use an oral Ring-Adair-Elwyn (RAE) preformed tracheal tube that sits along the midline of the chin. It avoids distortion of the upper lip and angles of the mouth, and can accommodate the Dingman mouth gag used by the surgeon to facilitate access to the palate during surgery. Bilateral breath sounds should be confirmed in both flexion and extension positions to ensure adequate depth of the endotracheal tube (ETT) below the vocal cords with changes in positioning.

What Are the Most Important Intraoperative Considerations?

Intraoperative airway complications during cleft lip and palate repair include accidental extubation, and tracheal tube obstruction from kinking or from compression by the Dingman mouth gag. An abrupt change in the end tidal carbon dioxide tracing should prompt assessment of the tracheal tube and adequacy of ventilation. Communication with the surgical team is important as the surgical field and the airway occupy the same surgical space and are often accessed with difficulty during the procedure.

What Is Your Plan for Intraoperative Analgesia? What Nerve Blocks Are Available for This Procedure?

Because of the young age of these patients and their propensity toward upper airway obstruction, intraoperative and postoperative opioids should be used with caution. These patients are at increased risk of unplanned emergency tracheal intubation in the postoperative period as a result of respiratory depression from the use of opioids. Therefore, many centers will use an infraorbital nerve block with local anesthesia. This nerve is easily accessible as it exits the infraorbital foramen.

How Would You Prepare This Patient for Extubation? Awake or Deep Extubation? How Do You Protect the Airway Postoperatively?

Since these patients are at risk for postoperative airway complications, an awake extubation is preferred. Airway obstruction can occur secondary to tissue edema, or tongue swelling from the Dingman mouth gag. Increased oral secretions and airway manipulation can increase the risk of laryngospasm following extubation. Oropharyngeal airways should be avoided in these patients as placement could disrupt surgical suture lines and irritate mucosal flaps. Aggressive suctioning of the mouth and nose should be avoided for similar reasons and preferably done under direct visualization by the surgeon at the end of the procedure. If used, removal of mouth gag should be confirmed. In patients with a history of difficult mask ventilation or intubation, or concern for postoperative airway obstruction, the surgeon can place a traction suture through the tongue that when pulled can decrease airway obstruction and improve ventilation. In addition, the surgeon can place and secure a nasopharyngeal airway at the end of surgery under direct visualization.

What Postoperative Preparations Should Be Made? Can This Patient Be Discharged after PACU Discharge Criteria Are Met?

Postoperatively, younger patients can be placed in elbow immobilizers to avoid disruption of surgical sutures. The patient can be placed in prone or lateral positioning (recovery position) to improve ventilation and enable blood and secretions to pool into the cheek or out of the mouth. If there are concerns for postoperative airway edema or obstruction (pharyngeal flap, pharyngoplasty), admission to an intensive care unit and continued pulse oximetry is warranted.

Acknowledgments

We wish to acknowledge the image contributions to this chapter of Laura Monson, MD, and John Wirthlin, DDS, MSD.

Suggested Reading

- Arosarena OA. Cleft lip and palate. *Otolaryngol Clin North Am.* 2007;40(1):27–60. PMID: 17346560.
- Nargozian C, Ririe DG, Bennun RD, et al. Hemifacial microsomia: anatomical prediction of difficult intubation. *Paediatr Anaesth.* 1999;9(5):393–8. PMID: 10447900.
- Sheeran PW, Walsh BK, Finley AM, et al. Management of difficult airway patients and the use of a difficult airway registry at a tertiary care pediatric hospital. *Paediatr Anaesth.* 2014;24(8):819–24. PMID: 24471869.
- Shkoukani MA, Chen M, Vong A. Cleft lip: a comprehensive review. *Front Pediatr.* 2013;1:53. PMID: 24400297.
- Xue FS, Zhang GH, Li P, et al. The clinical observation of difficult laryngoscopy and difficult intubation in infants with cleft lip and palate. *Paediatr Anaesth.* 2006;16 (3):283–9. PMID: 16490092.