Unilateral Cleft Lip Repair

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KEYWORDS

• Cleft lip • Cleft alveolus • Cleft nasal deformity • Nasoalveolar molding
• Presurgical infant orthopedics • Cheiloplasty • Gingivoperiosteoplasty • Primary cleft nasal surgery

KEY POINTS

- Modern cleft surgery requires four-dimensional and functional anatomic understanding of the cleft (and noncleft) lip, nose, and alveolus.
- Some techniques for nasolabial repair rely more on precise anatomic geometry, whereas others afford the surgeon a more flexible design.
- Consistent anthropometry enables accurate assessment and reporting of long-term outcomes; such reports are needed to guide perioperative care, delineate optimal repair principles, and resolve ongoing controversies.

INTRODUCTION

Since 390 BCE, the treatment of upper lip clefts has challenged generations of surgeons.[1] Cleft care is now concentrated in specialized, high-volume centers where multidisciplinary teams can best serve patients and their families. The American Cleft Palate–Craniofacial Association (ACPA) has established a framework for such centers that guides multidisciplinary team composition, interdisciplinary communication, cultural competence, psychosocial services, and outcomes assessment (www.acpa-cpf.org). Modern cleft reconstruction requires three-dimensional (3D) and functional anatomic understanding of the cleft (and noncleft) lip, nose, and alveolus. Accurate nomenclature defines shared challenges, improves classification schema, and builds consensus (where possible). Multiple repair techniques and modifications reflect the rich variety and continuous evolution of operative principles in cleft reconstruction. Diligent anthropometry enables accurate assessment of outcomes over time, the so-called fourth dimension.[2]

EMBRYOLOGY AND EPIDEMIOLOGY

Lip development occurs from the fourth to the seventh weeks of gestation.[3,4] The frontonasal prominence is formed from mesenchymal tissue ventral to the forebrain and leads to development of the medial and lateral nasal prominences.[5] The paired maxillary and mandibular prominences are formed from neural crest cells migrating from the first pharyngeal arch.[6] The primary palate, or premaxilla, similarly forms from the fusion of the medial palatine processes. A unilateral cleft lip occurs when there is failure of complete fusion between the advancing maxillary prominence and the fused medial nasal prominences on one side (**Fig. 1**). The cleft of the lip extends through the maxillary dentoalveolus to the incisive foramen when the medial palatine processes fail to fuse. The secondary palate, posterior to the incisive foramen, develops from the 6th to the 12th weeks of gestation as the lateral palatine processes fuse. A cleft palate occurs when there is incomplete fusion of the lateral palatine processes with the medial palatine process and/or nasal septum (**Fig. 2**). Various anomalous combinations in lip and palate development occur; it is more common to have cleft lip and palate (46%) than isolated cleft palate (33%) or isolated cleft lip (21%).[7] Additional failures of mesenchymal penetration result in an array of facial clefts.

Cleft lip (with or without cleft palate) occurs most commonly in boys and has a 6:3:1 ratio of

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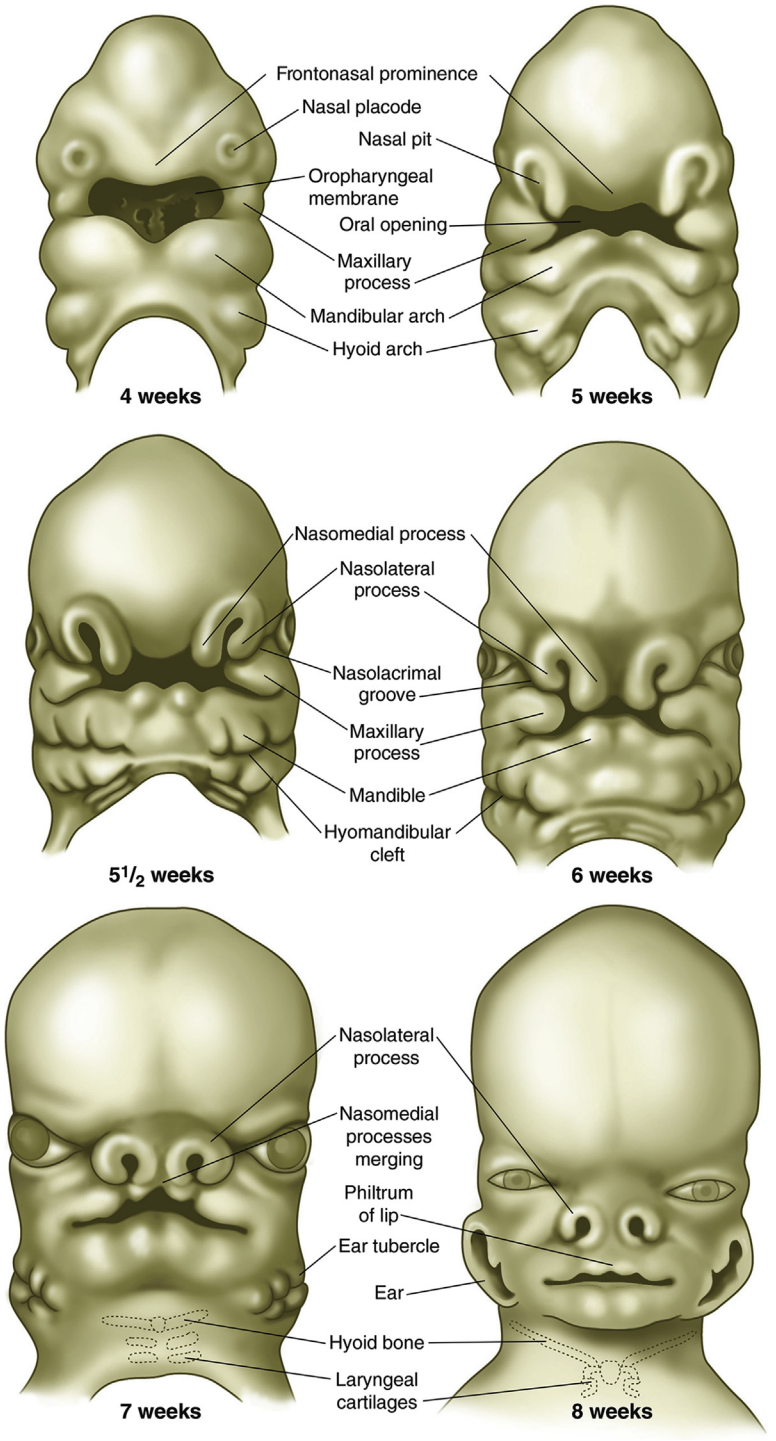

Fig. 1. Embryology of the normal lip. The face is initially made up of 5 prominences: a central frontonasal prominence, paired maxillary prominences, and paired mandibular prominences. These primordial structures appear during the fourth week of development and surround the primitive mouth. The maxillary prominence gives rise to the upper jaw, lip (lateral to the philtral column), orbital floor, and inferior portion of the lateral nasal wall. Rapid growth of the maxillary prominences, along with the medial nasal processes, during weeks 4 to 8 leads to a shift of the frontonasal process away from the stomodeum. The medial nasal processes fuse and give rise to the intermaxillary segment, which ultimately becomes the philtrum of the lip, the premaxilla, and the primary palate. (*From* Carlson BM. Head and neck. In: Carlson BM, editor. Human Embryology and Developmental Biology, Fifth Edition. Philadelphia: Elsevier Saunders, 2014; with permission.)

left/right/bilateral involvement.[7] If a child is born with a cleft lip, the risk of having another child with cleft lip is 4% and 9% after 2 affected children.[7] If a mother and child are affected, the risk to the second child is 15.3%.[7] The condition occurs in approximately 2 of 1000 Asians, 1 of 1000 white people, and 0.5 of 1000 African Americans.[8] Unlike isolated cleft palate, cleft lip is usually sporadic and associated with only a few syndromes: van der Woude (autosomal dominant, lip pits), 22q deletion