CHAPTER 10

CLEFT LIP AND PALATE

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I. ANATOMY AND DEFINITIONS

A. Cleft Lip (CL) alone, Cleft Lip with Cleft Palate (CLP), and Cleft Palate (CP) alone
   1. CL alone and CLP are along the same spectrum of morphology
   2. CP alone is a different entity

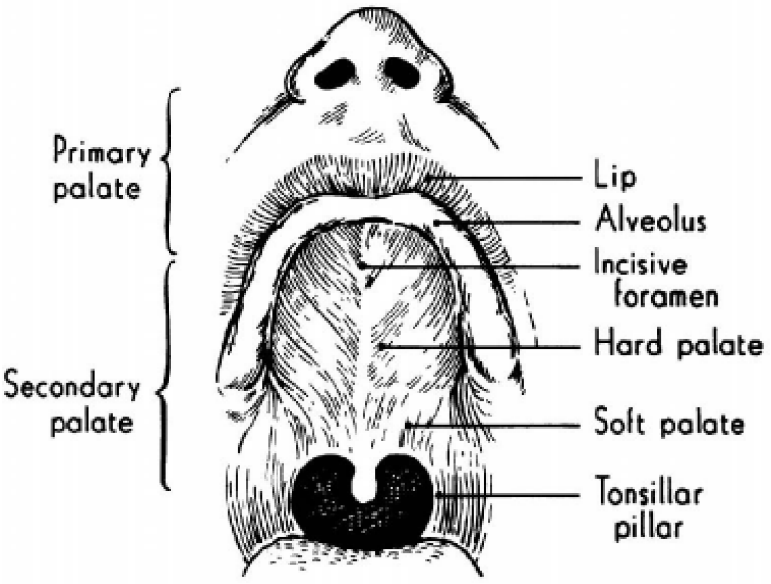

Figure 1. Basic anatomy of the palate

   3. CL occurs anterior to incisive foramen and can involve alveolus, due to failure of fusion of medial nasal processes and maxillary prominence at 4-6 weeks gestation
   4. CP divided into primary and secondary
      a. Primary CP is anterior to incisive foramen, due to failure of fusion of medial and lateral palatine processes
         i. Always involved in CLP
      b. Secondary CP is posterior to incisive foramen, due to failure of fusion of lateral palatine processes at 7-12 weeks gestation
         i. CP alone occurs in this region and soft palate, CLP can extend into secondary palate as well

B. Functional Deficits
   1. Cleft lip
      a. Cannot form fluid/air seal in eating/speech

b. Malocclusion of teeth
c. Cosmetic deformity with lack of continuity of muscle, skin, mucosa, nasal distortion

2. Cleft palate
   a. Cannot separate nose from mouth so air escapes during speech (velopharyngeal insufficiency)
   b. Cannot suck on breast/bottle well due to poor seal for intraoral negative pressure
   c. Middle ear disease/infections, often chronic

II. CLASSIFICATION

A. CL divided into unilateral/bilateral and incomplete/complete (Figure 2)

Figure 2. Classification of lip clefts

B. CP alone may be divided into incomplete/complete (Figure 3)
C. Complete CLP divided into unilateral/bilateral (Figure 3)

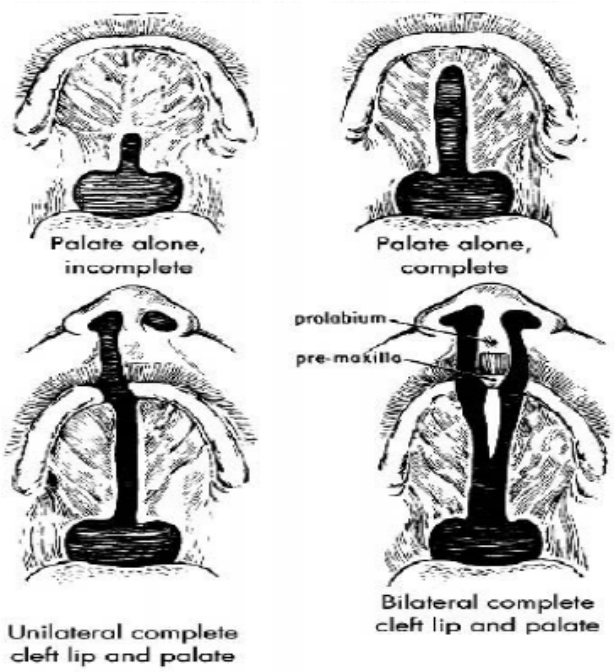

Figure 3. Classification of isolated cleft palates, and combination cleft lip and palates

III. DEMOGRAPHICS

A. Incidence/Demographics of CL alone/CLP
   1. 1:1000 in Caucasians
   2. 1:500 in Asians
   3. 1:2000 in those with African ancestry
   4. 2:1 males:females
B. Incidence/Demographics of CP alone
   1. 0.5:1000 without ethnic variation
   2. 1:2 males:females
C. Genetics
   1. Offspring occurrence risk (Table 1)
      a. Most cases of CL alone or CLP are sporadic, multifactorial, no isolated genetic cause, with only <15% syndromic
         i. Van der Woude's syndrome, most common syndrome associated with CL
         ii. Multifactorial risk factors include fetal exposure to drugs (phenytoin, EtOH, phenobarbital, diazepam), maternal smoking, advanced paternal age
      b. CP alone is often syndromic
         i. DiGeorge syndrome (most common) and Stickler syndrome

	Affected Relatives	**Predicted Outcomes**
CL +/- P		
	One sibling	= 4%
	One Parent	= 4%
	Sibling and a Parent	= 16%
CP		
	One sibling	= 2-4%
	One Parent	= 2-4%
	Sibling and a Parent	= 15%

Table 1. Risk of CL +/- P, or CP. Note if congenital lip pits are present, inheritance is autosomal dominant with variable penetrance (Van der Woude's Syndrome)

IV. TREATMENT

A. Multidisciplinary Care
    1. Plastic surgeon, dentist, orthodontist, audiologist, geneticist, social worker, speech/swallow therapist, nutritionist, otolaryngologist (middle ear issues), psychologist, pediatrician
B. Pre-operative techniques to bring cleft closer together
    1. Taping across lip segments
    2. Lip adhesion by suturing cleft margins together, with definitive repair later
    3. Nasoalveolar molding with pre-fabricated oral appliance and weekly adjustment to bring cleft segments closer together, can lengthen columella
C. Goals and Timing of Surgery
    1. CL repair – 2-3 months
        a. Repair skin, muscle and mucosa to restore continuity of lip, symmetrical length and function
        b. Simultaneous repair of both sides of a bilateral CL
        c. Can correct nasal deformity at time of CL repair
        d. Millard rotation-advancement repair is most commonly used unilateral CL repair in USA
    2. CP repair – 9-15 months
        a. One stage repair of both hard and soft palate
        b. Goal to separate oral and nasal cavities, lengthen palate, and reposition muscles in proper orientation (anomalous insertion of levator veli palatini (LVP) onto the posterior edge of the hard palate rather than decussating in midline)
        c. Furlow palatoplasty (double-opposing Z-plasty) allows for both oral and nasal closure, with lengthening of soft palate and realignment of LVP
    3. CL and nasal revisions – 2-4 years
    4. Alveolar cleft bone grafting– 6-8 years
        a. Performed at time of eruption of permanent maxillary canines, using most commonly iliac bone graft to fill alveolar defect
        b. Provides bony support to nasal base and incoming permanent teeth
    5. Orthognathic surgery to correct malocclusion of teeth – 15-18 years
        a. Lefort I osteotomy to advance maxilla to correct hypoplasia and reach normal occlusion, aided by orthodontics
    6. CL revisions and formal rhinoplasty – 14-18 years

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