INTERDISCIPLINARY CLEFT CARE
GLOBAL PERSPECTIVES

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**Contents**

**Preface** ix  
**Acknowledgments** x  
**About the Editors** xi  
**Contributors** xiii

**Section I. Early Care of the Infant With Cleft Lip and/or Palate**

1. Embryology, Anatomy, and Classification of Cleft Lip and Palate  
   Carolyn R. Rogers-Vizena and Anne M. Burrows 3

2. Assessment and Management of Feeding and Nutrition  
   Claire Kane Miller, Brenda Thompson, Jennifer Kobler, and Tamara Rhodes 15

3. Early Speech and Language Development in Children With Cleft Lip and/or Palate  
   Adriane L. Baylis and Caitlin E. Cummings 27

4. Audiological Care of the Infant With Cleft Palate  
   Briana Kelly Dornan 43

5. Genetic Counseling and Syndromic Considerations  
   Melissa D. Kanack, Neda Zadeh, Touran Zadeh, and Raj Vyas 51

**Section II. Presurgical Dentofacial Treatment**

6. Dynacleft®, Lip Taping, and Passive Molding Appliances  
   Daniela Y. S. Tanikawa 63

7. Nasoalveolar Molding  
   Barry H. Grayson, Pedro E. Santiago, Mohammad M. Ahmed, and Serena N. Kassam 81

8. Active Dentofacial Orthopedics With Latham-Type Appliances  
   Elizabeth Ross, Christopher Ma, Carolyn R. Rogers-Vizena, and James K. MacLaine 95

   Serena N. Kassam, Mohammed M. Ahmed, and Amr M. Moursi 105

**Section III. Anesthetic and Pediatric Perioperative Considerations**

10. Cleft Lip and Cleft Palate: Anesthetic Considerations  
    Dima Daaboul, Pete G. Kovatsis, and Navil F. Sethna 123

11. Perioperative Considerations in Pediatric Cleft Repair  
    Marie Nader, Suying Lam, Jennifer Co-Vu, and Jennifer Munoz 133

**Section IV. Unilateral Cleft Lip Repair**

12. Primary Cleft Lip and Nose: Rotation Advancement and V-Cheiloplasty in Unilateral  
    Cleft Lip Repair  
    Usama S. Hamdan, Rami S. Kantar, and Antonio Melhem 145
13. Rotation Advancement Repair: Mulliken Modification
   Raj M. Vyas

14. Extended Mohler Repair: Cutting Technique
   Kerry A. Morrison and Roberto L. Flores

15. Unilateral Cleft Lip Repair: Anatomic Subunit Approximation
   Raymond W. Tse, David K. Chong, and David M. Fisher

16. Cleft Lip Repair With Vomerine Flap Closure of the Hard Palate
   Brian Sommerlad

Section V. Bilateral Cleft Lip Repair

17. Synchronous Premaxillary Setback, Vomerine Ostectomy, and Bilateral Lip Repair
   Usama S. Hamdan, Adam B. Johnson, Rami S. Kantar, Elsa M. Chahine, and Omar S. Al Abyad

18. Mulliken Technique for Bilateral Cleft Lip Repair
   Carolyn R. Rogers-Vizena and Alexander C. Allori

   Cassio Eduardo Raposo-Amaral and Cesar Augusto Raposo-Amaral

20. Two-Stage Bilateral Cleft Lip and Palate Repair
   Brian Sommerlad

Section VI. Cleft Palate Repair

21. Cleft Palate Repair With Radical Muscle Dissection
   Brian Sommerlad

22. Modified Furlow Double-Opposing Z-Plasty With Tissue Augmentation Palatoplasty
   Miles J. Pfaff and Joseph E. Losee

23. Minimal Incision Palatoplasty Technique
   Marcia R. Perez Dosal, America Ayuso Arce, and Rafael Villaseñor Caloca

24. Hard Palate Repair With Relaxing Incisions: Two-Flap and Bipedicle Palatoplasty
   Carolyn R. Rogers-Vizena and John G. Meara

25. The Anatomic Cleft Restoration Philosophy With Buccal Flap Approach
   Robert J. Mann

26. Staged Palate Repair: Soft Palate First
   Hans Mark and Jan Lilja

Section VII. Revision Lip and Palate Surgery

27. Oronasal Fistula Repair Surgery
   Mahmoud I. Awad and Krishna G. Patel

28. Secondary Cleft Lip Deformity
   Usama S. Hamdan, Rami S. Kantar, and Omar S. Al Abyad

Section VIII. Secondary Management of Dentoalveolar and Orthognathic Concerns

29. Phase I Dental Orthopedic and Orthodontic Treatment for Cleft Lip and Palate
   Pedro E. Santiago, Daniel Levy, Tiago Turri de Castro, and Ryan Cody
30. Alveolar Bone Grafting and Long-Term Outcomes  
   Karen Z. Carver and Bonnie L. Padwa
   389

31. Phase II Dental and Presurgical Orthodontic Treatment  
   Douglas Olson and Pradip R. Shetye
   405

32. Determinant of Facial Growth in Cleft Lip and Palate  
   Elçin Esenlik and Laura Mancini
   413

33. Orthognatic Correction: Surgery First  
   Yu-Ray Chen, Yu-Fang Liao, Chuan-Fong Yao, and Tingchen Lu
   425

34. Orthognathic Correction: Surgery Last  
   Connor J. Peck, Joseph Lopez, Jakob Lattanzi, and Derek Steinbacher
   443

35. Prosthetic Rehabilitation of the Cleft Alveolar Gap  
   Stephanie J. Drew, Shelly Abramowicz, James Davis Jr., and Steve Roser
   455

Section IX. Cleft Nasal Deformity

36. Primary Rhinoplasty  
   Christopher Brooks, Usama S. Hamdan, and Omar S. Al Abyad
   465

37. Secondary Cleft Tip Rhinoplasty  
   Beatriz Berenguer, Jesse Taylor, and Anna R. Carlson
   481

38. Definitive Septorhinoplasty in the Cleft Lip Patient  
   Grace Lee Peng, Usama S. Hamdan, and Babak Azizzadeh
   499

Section X. Evaluation and Management of Cleft-Related Speech Disorders

39. Evaluation of Speech and Resonance Due to Velopharyngeal Insufficiency (VPI)  
   Ann W. Kummer and Mikaela M. Bow
   513

40. The Role of the Speech-Language Pathologist in Cleft Care  
   Ann W. Kummer and Mikaela M. Bow
   523

41. Measuring Speech Outcomes  
   Debbie Sell and Valerie Pereira
   531

Section XI. Surgical Management of Velopharyngeal Insufficiency (VPI)

42. Posterior Pharyngeal Flap  
   Oksana A. Jackson and David W. Low
   549

43. Sphincter Pharyngoplasty  
   David W. Low and Oksana A. Jackson
   559

44. Revision Palatoplasty With Intravelar Veloplasty (±) Buccinator Flaps  
   Brian C. Sommerlad
   567

45. Revision Palatoplasty With Furlow Double-Opposing Z-Plasty  
   Philip Kuo-Ting Chen and Vikram S. Pandit
   581

46. Palatopharyngeal Augmentation  
   Monica K. Rossi Meyer and Deborah S. F. Kacmarynski
   595
Section XII. Psychosocial Considerations From Early Childhood to Adulthood

47. Provisions of Psychosocial Care in Cleft Lip and/or Palate: A Global Context
   Martin J. Persson and Matthew Ridley

48. Promoting Resilience in People With Cleft Lip and/or Palate Around the World
   Matthew Ridley and Martin J. Persson

49. Patient and Family Community Resources
   Gareth Davies, Erin Stieber, Ruben Ayala, Hugh Brewster, Jamie Perry, Serena Kassam,
   Taylor Snodgrass, and Usama S. Hamdan

Section XIII. Special Considerations for Outreach Settings

50. Planning and Execution of Overseas Outreach Programs
    Charanya Vijayakumar, Elsa M. Chabine, Rami S. Kantar, Nikbil Shah, Antonio Melhem,
    Omar Al-Abyad, Daniel Jaffurs, Raj M. Vyas, and Usama S. Hamdan

51. Quality Assurance Guidelines for Outreach Cleft Programs
    Elsa M. Chabine, Elie P. Ramly, Alexander P. Marston, Raj M. Vyas, and Usama S. Hamdan

52. Safety and Emergency Preparedness During Overseas Outreach
    Elsa M. Chabine, Antonio Melhem, Raj M. Vyas, and Usama S. Hamdan

53. The Future of Cleft Outreach: In Situ Simulation, Task Training, Augmented Reality, and
    Artificial Intelligence
    Jennifer Munoz Pareja, Marie Nader, Antonio Melhem, Omar S. Al Abyad, Usama S. Hamdan,
    Rami S. Kantar, J. Peter W. Don Griot, Raj M. Vyas, and Corstiaan Breugem

54. Building a Cleft Team
    Gaurav Deshpande and Jordan W. Swanson

Section XIV. Rare Facial Clefts

55. Rare Facial Clefts
    Nivaldo Alonso and Cristiano Tonello

56. Macrostomia: Functional and Aesthetic Repair
    Usama S. Hamdan, Raj M. Vyas, Rami S. Kantar, and Antonio Melhem

Index
The quest for optimal patient outcomes is a driving force for health care professionals. This is especially true for those caring for children and adults born with cleft lip and palate. Significant advances in interdisciplinary cleft care, coupled with exponentially expanding information dissemination, facilitate knowledge sharing and quality improvement on a global level. A remaining aspiration of those involved in cleft care is to “level the playing field” and achieve ideal outcomes at all income and resource levels. That goal inspired the current work.

Through print, illustration, and videography, this book presents the knowledge, skill, and evidence-based practice of an interdisciplinary group of cleft experts from around the world. Lessons from individuals practicing in a variety of cultures and resource environments have been thoughtfully assembled. The resulting work provides the reader with practical and personally vetted solutions for all aspects of cleft care. Highly accurate illustrations by surgeon-artist Dr. David Low and technical videos accompanying this book also enhance the reader’s ability to understand and apply techniques used by the authors to achieve successful outcomes. A multidisciplinary team approach is essential for rendering the best possible cleft care, and we hope this book serves as a catalyst for development of comprehensive cleft programs worldwide.
The editors are grateful to all the children and adults born with cleft lip and palate who were the inspiration for this project and for the lessons they have taught us in perseverance and humility. We are also indebted to the Global Smile Foundation postdoctoral fellows whose efforts made this book possible: Dr. Elsa Chahine, Dr. Antonio Melhem, Dr. Omar Al Abyad, Dr. Mario Haddad, and Dr. Robert Younan. Their initiative, dedication, and perseverance created the momentum needed to transform the idea for this book into reality. We are also incredibly appreciative of our coeditor, Dr. David Low, whose illustrations are found throughout this book. The anatomic detail and surgical accuracy rendered by a practicing cleft surgeon is truly a unique highlight of this book. Finally, we are thankful to our colleague authors, volunteers, and supporters whose tireless energy and focus on providing safe and comprehensive cleft care throughout the world have been instrumental in this undertaking.
Usama S. Hamdan, MD, FICS, is president and cofounder of Global Smile Foundation, a 501(c)(3) Boston-based nonprofit foundation that provides comprehensive and integrated pro bono cleft care for underserved patients throughout the world. He has been involved with outreach cleft programs for over three decades. Dr. Hamdan is an otolaryngologist/facial plastic surgeon with former university appointments at Harvard Medical School, Tufts University School of Medicine, and Boston University School of Medicine. He is also the founder of the International Comprehensive Cleft Care Workshop based on his special interests in simulation-based cleft training, empowerment, and sustainability initiatives for providing comprehensive cleft care as well as quality assurance strategies. For his philanthropic service to the people of Ecuador, he was awarded the knighthood, “Al Merito Atahualpa” En El Grado De Caballero, by the president of Ecuador in March 2005. He received honorary professorship at Universidad de Especialidades Espíritu Santo, School of Medicine, in Ecuador on March 5, 2015, for his contributions in the field of cleft lip and palate.

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Raj M. Vyas, MD, earned his BS in biology with honors and distinction from Stanford University and completed his MD at UCLA, where he was awarded the Stafford L. Warren Medal for most outstanding medical student and the Longmire Medal for most outstanding student in surgery. He then completed integrated plastic surgery residency at Harvard and Craniofacial Fellowship at New York University. Dr. Vyas is a professor of plastic surgery at UC Irvine School of Medicine, where he serves as vice-chair for the Department of Plastic Surgery and chief of pediatric plastic surgery at CHOC Children’s Hospital. His clinical and research interests include characterizing neonatal sleep and breathing disturbances, enhancing recovery after cleft/craniofacial surgery, understanding patient-reported psychosocial outcomes, and using technology to facilitate knowledge and skill transfer. Aligned with his passion for enhancing global capacity for interdisciplinary cleft care, Dr. Vyas is codirector of Global Smile Foundation’s International Research Fellowship and codirector of Research for Plastic Surgery Foundation’s SHARE program (Surgeons in Humanitarian Alliance for Reconstruction Research and Education). Dr. Vyas actively serves on dozens of regional, national, and international cleft committees, hospital consortia, and journal editorial boards.

Brian C. Sommerlad, MBBS, FRCS, qualified in medicine in Sydney, Australia. He went to the United Kingdom in 1968 to further his training and stayed. He is an honorary consultant plastic surgeon at Great Ormond Street Hospital for Children, London. He has been caring for children with clefts for 45 years—in the United Kingdom and by regularly working with colleagues in many less privileged countries over the past 22 years. His ongoing research interests have centered on palate anatomy and function and attempting to improve the speech outcomes of palate repair. He was cofounder in 2007 and is chairman of the U.K. charity CLEFT, which funds research into the causes and treatment of clefts and supports cleft centers in several low-resource countries.

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**Introduction**

Shape and form of the human face is rooted in early embryonic development. Similarly, cleft lip and/or palate (CL/P) begins with malformation early in embryonic development that persists as tissues differentiate and development progresses. This chapter will provide the embryologic and anatomic foundation necessary to understand CL/P pathology. In addition, common classification systems used to describe and document the extent of anatomic findings in CL/P will be presented to foster consistent language for communicating CL/P phenotype.

**Embryology**

Knowledge of normal facial embryology is important for understanding the underlying basis of craniofacial anomalies such as CL/P. Basic morphology of the face is established between the 4th and 8th embryonic weeks, developing from five primordia surrounding a central stomodeum (primitive mouth). These primordia include a single midline frontonasal prominence (FNP), paired maxillary prominences, and paired mandibular prominences. These prominences undergo mostly symmetrical growth followed by subsequent fusion to form the face.

**Development of the Face**

During the 4th week, the stomodeum invaginates, and nasal pits, or placodes, form in the FNP. In the 5th week, medial and lateral nasal processes form around the nasal pits (Figure 1–1). The FNP gives rise to the nose, philtrum, primary palate, perpendicular plate of the ethmoid bone and vomer (nasal septum), cribriform plates, and forehead. The medial nasal processes go on to form the nasal tip, columella, philtrum, and premaxilla/primary palate (central portion of the alveolus and anterior hard palate, including the central and lateral incisors). The lateral nasal processes form the nasal alae. Concomitant with FNP development, the first pharyngeal arch forms paired maxillary prominences that enlarge and migrate ventrally. The maxillary prominences give rise to the upper cheek and most of the upper lip, maxilla, zygoma, and secondary palate. The medial nasal processes and maxillary prominences fuse to form a continuous upper lip and primary palate by the end of the 6th week (Sperber et al., 2001). Disrupted fusion of the medial nasal process and maxillary prominence results in cleft lip and alveolus (Larrabee et al., 2004).

**Development of the Palate**

The palate develops between the 6th and 12th weeks from a single midline nasal septum originating on the
FNP and paired lateral palatine processes originating on the maxillary prominences (see Figure 1–1). The palatine processes grow inferiorly, lateral to the developing tongue. As the maxilla and mandible develop and enlarge, the tongue moves caudally, permitting the palatine processes to elevate into a horizontal position (known as “palatal shelves”) during the 7th and 8th weeks. The palatal shelves fuse with the primary palate in a Y-shaped configuration, with the apex forming the incisive foramen in the 6th and 7th weeks. Posterior to the incisive foramen, the palatal shelves fuse with each other and with the nasal septum starting anteriorly and extending posteriorly beginning around the 8th week and ending by the 12th week. Ossification of the primary palate and anterior secondary palate forms the hard palate. The posterior secondary palate does not ossify and forms the soft palate and uvula (Sperber et al., 2001). Failure of the palatal shelves to elevate and fuse at the midline results in a cleft palate (Larrabee et al., 2004).

Normal Lip and Nasal Anatomy

Upper lip surface anatomy reflects its embryologic formation and underlying muscular anatomy. Understanding key surface landmarks is critical for properly designing a cleft lip repair (Figure 1–2). The columellar-labial junction is the intersection between the columnella and superior part of the upper lip. Inferior to this is the philtral dimple, a cutaneous indentation in the center of the upper lip. The philtral ridges are raised areas of skin flanking the philtral dimple on either side. They extend inferiorly to elevated areas of vermilion called the peaks of Cupid’s bow. Between these two elevated areas is a vermilion depression, the low point or nadir of Cupid’s bow. The white roll is a raised area of skin just superior to the vermilion. The red line represents the transition from dry,
1. Embryology, Anatomy, and Classification of Cleft Lip and Palate

The keratinized vermillion extends from the labial mucosa. Finally, the median tubercle is the prominence at the most inferior extent of the upper lip.

In a normal upper and lower lip, the orbicularis oris muscle (OOM) extends from modiolus to modiolus (the confluence of facial muscles lateral to the oral commissure), creating a muscular sphincter. There are two major OOM subdivisions in the upper lip, the pars marginalis (inferior) and the pars peripheralis (superior). Pars marginalis is a continuous band of muscle fibers while pars peripheralis fibers decussate at the midline with fibers from the contralateral side inserting deep to dermis of the contralateral philtral ridge (Figure 1–3). This forms the underlying muscular architecture of the philtral ridges and dimple but is not the only factor contributing to philtral shape. Thickened dermis and connective tissue are present at the ridges, and there is a paucity of connective tissue at the dimple. In cross section, the configuration of the OOM is a subtle “J” shape with the distal tip of the “J” lying deep to the white roll and contributing to its raised appearance (Rogers et al., 2014).

In the normal nose, three-dimensional shape is primarily supported by the paired lower and upper lateral cartilages as well as the single midline cartilaginous septum (see Figure 1–3). The arch-shaped lower lateral cartilages are conceptualized as three sections. Inferiorly, the medial crura are narrow portions of the arch beginning superficial to the anterior nasal spine and extending along the inferior border.
of the septum, before bending laterally. This bend, or genu, is called the middle crus and supports the nasal tip. The cartilages widen, extending superolaterally from the genua as the lateral crura, providing soft tissue support to the lateral nasal tip and alae. The upper lateral cartilages lie superior to the lower lateral cartilages and extend laterally from the septum supporting the middle vault of the nose. The midline nasal septum supports the dorsum of the nose and separates the two nasal vaults. It intersects posteriorly with the perpendicular plate of the ethmoid bone (superior) and the vomer (inferior), which merges with the hard palate (Fisher & Mann, 1998).

The major blood supply to the upper lip and nose arises from branches of the paired facial arteries (Figure 1–4). The upper lip is primarily supplied by the superior labial branches. Beyond the superior labial branch point, the facial artery continues as the lateral nasal artery to supply to the nasal tip. The superior labial arteries anastomose with each other at the midline and give off septal and columellar branches. The columellar branches anastomose with the dorsal and lateral nasal arteries. The clinical significance of this rich anastomotic network is that the lip and nose remain well perfused despite disruption of major vascular branches.

Sensory innervation of the lip and nose comes from branches of the trigeminal nerve (cranial nerve V), primarily the infraorbital branch of the maxillary nerve, but with contribution from the external nasal branch of the ophthalmic nerve. These branches are amenable to regional anesthetic block during cleft lip repair (see Chapter 12).

The skeletal structures most relevant to a cleft lip are portions of the maxilla (Figure 1–5). These form the inferior borders of the piriform aperture that encompasses the nasal cavity. Anteriorly, a midline outcropping called the anterior nasal spine supports the nasal tip, columella, and superior upper lip. The maxilla also comprises the tooth-bearing alveolus and anterior hard palate.

**Cleft Lip/Nasal Anatomy**

Cleft lip involves structures of the lip, nose, and primary palate. Nasolabial tissue is malformed, rather than deficient. In a unilateral cleft, the nasal tip and ala on the cleft side are displaced inferiorly and posterolaterally, corresponding to deformity of the underlying lower lateral cartilage and maxilla. The arch of the genu is widened and lateral crus inferiorly and posterolaterally positioned (see Figure 1–3). In more severe unilateral clefts, the lateral crus has a downward deflection or “recurvatum.” The anterior nasal spine and caudal septum deviate away from the cleft and the mid-septum bows toward the cleft (Fisher & Sommerlad, 2011). The lip is divided where the philtral ridge should be, extending through the peak of Cupid’s bow and vermillion. This division may be incomplete, including only the inferior margin of the lip, or complete, extending all the way
into the floor of the nose. On the labial surface, the vermilion narrows beyond where the peak of Cupid's bow should be, with the vermilion-cutaneous junction and vermilion-mucosal junction eventually converging along the margin of the cleft. In a complete cleft, the OOM is discontinuous, whereas there may be superior bridging fibers in an incomplete cleft. Orbicularis oris muscle fibers are oriented toward the alar base lateral to the cleft and columella medial to the cleft (see Figure 1–3). With increasing cleft severity, the philtral dimple effaces but remains present, in contrast to a bilateral cleft. The philtral ridge remains present on the noncleft side (Rogers et al., 2014). The superior labial artery is discontinuous, oriented along the cleft margin (see Figure 1–4). The underlying maxillary alveolus is protuberant on the noncleft side and may be collapsed on the cleft side (see Figure 1–5). The lateral incisor tooth is often absent on the cleft side, but particularly in the setting of an incomplete cleft, there may be an abnormal lateral incisor or supernumerary tooth.

In a bilateral cleft lip, the nasal tip is broad and flat, with both lower lateral cartilages having the abnormal configuration described for the cleft side in a unilateral cleft lip (see Figure 1–3). In a symmetric bilateral cleft lip, the septum and anterior nasal spine are midline, but in some cases, a bilateral cleft is asymmetric with corresponding deviation. The lip is divided at the area of the philtral ridge bilaterally. The central soft tissue element, the prolabium, has narrow vermilion and lacks a white roll in a complete cleft. The lateral lip vermilion deformity is analogous to the cleft side in a unilateral cleft lip. Orbicularis oris muscle fibers are oriented toward both alar bases laterally (see Figure 1–3). In a bilateral complete cleft lip, the prolabium lacks muscle tissue and the philtral dimple is completely effaced, whereas in a bilateral incomplete cleft lip, a small amount of OOM may bridge the prolabium and a subtle philtral dimple may be present. The superior labial arteries supply lateral lip elements, while the columellar and posterior septal arteries supply the prolabium (see Figure 1–4). Prolabial blood supply is critical to bear in mind when designing a bilateral cleft lip repair. When there is a complete alveolar cleft, the premaxilla is protuberant due to excessive anterior growth at the premaxillary-vomerine suture and the lateral alveolar segments collapsed (see Figure 1–5). Blood supply to the premaxilla comes primarily from branches of the posterior septal and anterior ethmoidal arteries,
an important consideration during premaxillary setback. The lateral incisors may be absent, abnormal, or supernumerary teeth may be present bilaterally.

**Normal Palatal Anatomy**

The anterior two thirds of the hard palate is formed by paired palatine processes of the maxilla. The posterior third, mesial to the tooth-bearing alveolus, is formed by the paired palatine bones. Superiorly, the hard palate and vomer articulate supporting the nasal floor. The incisive foramen is located at the midline, just posterior to the alveolus at the junction of the primary and secondary palate. Structures anterior to the incisive foramen are considered the primary palate, and structures posterior to it, the secondary palate.

The soft palate, or velum, is located posterior to the hard palate (Figure 1–6). It is a muscular sling formed by the paired levator veli palatini, palatopharyngeus, palatoglossus, and musculus uvuli, reinforced by the aponeurosis of the tensor veli palatini muscles. The levator in particular extends from the temporal bone into the velum and acts as the primary elevator of the soft palate during speech, whereas the palatoglossus and palatopharyngeus muscles elevate the posterior tongue and lateral pharyngeal walls while drawing the soft palate inferiorly to propel the food bolus during deglutition. Together with the superior constrictor muscle in the pharynx, these muscles form the velopharyngeal sphincter that provides dynamic separation of the oro- and nasopharynx during deglutition and speech. In addition, the tensor veli palatini acts as the principal dilator of the Eustachian tube, with further input from the levator veli palati, salpingopharyngeus, and tensor tympani muscles (Cho et al., 2013).

Palatal and velopharyngeal blood supply comes from branches of the paired maxillary arteries, ascending pharyngeal arteries, and ascending pala-

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**FIGURE 1–6.** Palatal anatomy—normal anatomy from intraoral (left) and sagittal (right) views. Deep to the glandular submucosa of the soft palate, the palatoglossus and palatopharyngeus muscles enter. The levator enters deep to these muscles but interdigitates with them, approaching the midline to form the velopharyngeal sling. The tensor tendon wraps around the pterygoid hamulus, spreading into a broad aponeurosis approaching the midline. The musculus uvulae is primarily located on the deep/nasal surface of the velopharyngeal sling, extending from the tensor aponeurosis to the uvula.
tine arteries. The soft palate has a rich blood supply from branches of these arteries. The hard palate mucoperiosteum is primarily supplied by the greater palatine branches of the maxillary artery, an important consideration during cleft palate repair.

Hard palate sensory innervation comes from the greater palatine and nasopalatine branches of the maxillary nerve while soft palate sensation is from the lesser palatine branch of the maxillary nerve. The maxillary nerve is a branch of the trigeminal nerve amenable to regional anesthetic block at the pterygopalatine fossa (see Chapter 10).

Cleft Palate Anatomy

Cleft palate involves structures of the secondary palate. When the hard palate is cleft, the maxillary and palatine bones are separated both from the contralateral side and from the vomer, resulting in communication between the oro- and nasopharynx. When the soft palate is cleft, there is a midline separation of the velum (Figure 1–7). The tensor veli palatini aponeuroses, levator veli palatini, palatoglossus, and palatopharyngeus muscles are discontinuous with each other and disoriented anteriorly. The tensor aponeuroses inserts on the posterolateral hard palate, while the palatoglossus and palatopharyngeus muscles insert on the posteromedial hard palate and cleft edge. The levator runs parallel to the other velar muscles but fails to reach the midline (Fisher & Sommerlad, 2011). Thus, these muscles fail to interdigitate and form the dynamic sling needed to control oronasal airflow and facilitate Eustachian tube function.

Submucous cleft palate, a lesser form of cleft palate, involves muscular diastasis of the soft palate with intact mucosa. The diastasis may be visualized as a bluish “zona pellucida,” or furrow, in the soft palate. Other findings may include bifid uvula and/or notch in the posterior hard palate (Calnan, 1954). This type of cleft is often diagnosed during evaluation of abnormal speech.

Phenotypic Spectrum and Classification

The wide phenotypic variation of CL/P has inspired numerous systems for describing the extent of the defect. The classifications described here are not all-inclusive but are some of the most commonly used ways to document clefting.
Cleft Lip

Cleft lip is broadly classified as unilateral (one-sided) or bilateral (two-sided) and further categorized by the extent of the discontinuity. Complete cleft lip involves a separation extending from the lip margin through the nasal floor. In an incomplete cleft lip, there is separation at the free margin of the lip with a superior connection, ranging from a thin skin bridge or band to a substantial musculocutaneous connection. Lesser forms of incomplete cleft lip involve smaller degrees of disruption of the vermilion-cutaneous junction at the peak of Cupid’s bow: mini-microform (notch of the vermilion-cutaneous junction without elevation of the peak of Cupid's bow), microform (disruption of the free margin of the lip with <3 mm elevation of the peak of Cupid’s bow), and minor form (disruption of the free margin of the lip with 3 to 5 mm elevation of the peak of Cupid’s bow). Classifying a labial cleft according to these descriptive terms is useful for both verbally communicating the degree of clefting and determining extent of the lip repair (Yuzuriha & Mulliken, 2008).

Cleft Palate

Cleft palate is often described using the Veau classification (Figure 1–8), allowing clinicians to communicate the anatomic nature of the defect. A Veau I cleft involves the soft palate only, a Veau II cleft includes the hard and soft palate, a Veau III extends unilaterally along the junction of the primary and secondary palate (i.e., a unilateral complete cleft lip and palate),