

Cleft Lip and Palate

CRITICAL ELEMENTS OF CARE

Produced by

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Children and Youth with Special Health Care Needs Program
and
Seattle Children's Hospital Craniofacial Center
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The *Critical Elements of Care* (CEC) considers care issues throughout the life span of the child. The intent of this document is to educate and support those caring for children with cleft lip and palate. The CEC is intended to assist the primary care provider and family members in the recognition of symptoms, diagnosis and care management related to a specific diagnosis. It provides a framework for a consistent approach to management of these children.

This document is available on the Seattle Children's Hospital website at:
<https://www.seattlechildrens.org>

DISCLAIMER: Individual variations in the condition of the patient, status of patient and family, and the response to treatment, as well as other circumstances, mean that the optimal treatment outcome for some patients may be obtained from practices other than those recommended in this document. This consensus-based document is not intended to replace sound clinical judgment or individualized consultation with the responsible provider regarding patient care needs.

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INTRODUCTION

The Washington State Department of Health, Division of Family and Community Services, Children with Special Health Care Needs Program has funded interdisciplinary work groups to identify Critical Elements of Care (CEC) for children with special health problems, including cleft lip and palate. This document was created by the cleft lip/palate consensus team, made up of primary care physicians, specialty providers, regional cleft lip/palate team coordinators, parents and third-party payers. The CEC draws upon a number of sources, especially the American Cleft Palate-Craniofacial Association (ACPA) Parameters of Care (2017) and Team Standards (2017) documents.¹⁻⁴

Central to these documents, which are summarized in the appendix, is the principle that **patients with cleft lip/palate are best cared for by an interdisciplinary team of specialists with experience in this field.** The CEC also draws on the literature of cleft lip and palate outcomes, as well as the experience of the CEC team members.

The goals of treatment for the child with a cleft lip/palate are:

- Repair the birth defect (lip, palate, alveolar ridge, nose)
- Achieve normal speech, language and hearing
- Achieve functional dental occlusion and good dental health
- Optimize psychosocial and developmental outcomes
- Minimize costs of treatment
- Facilitate ethically sound, family-centered, culturally sensitive care

Seven key themes are important for achieving these goals:

- Early assessment and intervention is imperative and should begin in the prenatal or newborn period with referral to a Cleft Lip/Palate Team.
- An interdisciplinary cleft lip/palate team is needed because cleft lip/palate outcomes are in surgical, speech, hearing, dental, psychosocial and cognitive domains.
- Providers with training and expertise in cleft lip/palate care are needed because of the complexity of treatment interventions and the expectation of outstanding outcomes.
- Continuity of care is essential because outcomes are measured throughout the child's life and team care is linked to improved outcomes.
- Proper timing of interventions is critical because of the interaction of facial growth, dental occlusion and speech.
- Coordination of care is necessary because of the complexity of the medical, surgical, dental and social factors that must be considered in treatment decisions.
- Proper early management leads to better outcomes, fewer surgeries and lower costs.

Organization of this Document

This document elaborates on the above goals and themes of treatment. Tables on pages 17 and 18 highlight key interventions by discipline and age group, respectively. These are explained more fully in the body of the document and in the sections that follow. A glossary of terms, description of cleft types, and resource guide with a listing of cleft lip/palate teams in Washington state are also included.

The following pages list problems and interventions for the child with a cleft lip/palate. Most of the interventions listed are provided by specialists on the cleft lip/palate teams. Others become the responsibility of the primary care provider (PCP). The division of these tasks will vary depending upon geographic location and the expertise and interest of the PCP. The services that result must be closely coordinated with the treatment plans of the patient's cleft lip/palate team. In addition, cleft lip/palate teams vary in both the disciplines participating and the interventions provided. For these reasons, specific provider disciplines are often not mentioned.

In many cases, the PCP will need to initiate a referral to the cleft lip/palate team and preauthorize visits with different specialists. It is the intent of this document to assist the PCP caring for these children by summarizing interventions for each age group. As the interventions listed are necessarily brief, appendices have been included to provide additional information in many of the key areas.

It is important to remember that children with cleft lip/palate may be eligible for Birth-To-Three services as mandated by the Individuals with Disabilities Education Act (IDEA). Referrals to these services can be facilitated by any Children with Special Health Care Needs Coordinator at local public health departments or by the cleft lip/palate team coordinator. No further mention of the IDEA or Birth-To-Three services will be made elsewhere in this document because other materials exist which describe these in detail.

NOTE: The interventions listed in this document are to be considered as guidelines only. All interventions may not be needed by every patient. Conversely, some patients may require interventions not mentioned in these recommendations. Each patient's care plan should be individualized considering medical needs, psychosocial and cultural variables, and resources available in each community. Communication between the community provider and the cleft lip/palate team members is essential for developing and implementing these care plans.

BACKGROUND

Types of Cleft Lip/Palate

Clefts of the lip and palate can vary considerably from one individual to the next. Some have both cleft lip and palate; some have only a cleft of the lip (also known as the primary palate); others have only a cleft of the palate (also known as the secondary palate). Clefts may be unilateral or bilateral.

Figure 1A is a drawing of a normal roof of the mouth. **Figure 1B** through **1F** are drawings of various types of clefts. With any kind of cleft lip, there may be extra, missing or poorly formed teeth in the area(s) of the cleft.

FIGURE 1A: NORMAL ROOF OF THE MOUTH

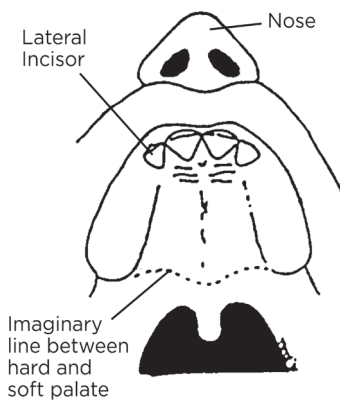
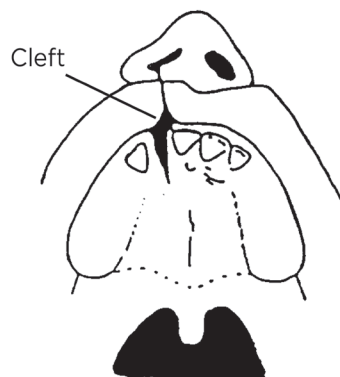


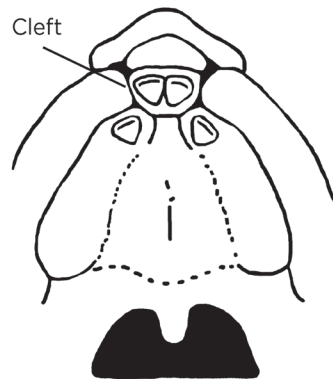
FIGURE 1B: UNILATERAL CLEFT

(EXTENDS COMPLETELY THROUGH THE LIP AND ALVEOLUS (GUMLINE))



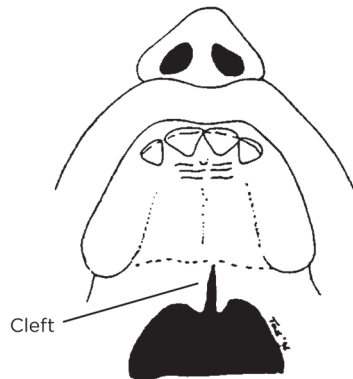
This causes the side of the nose to be collapsed and spread to the side.

FIGURE 1C: BILATERAL CLEFT LIP



The alveolus and the lip between the two cleft areas have been displaced from their normal alignment, and this allows this segment of the premaxilla to swing upward and protrude. In this situation, pre-surgical orthopedics may be desired.

FIGURE 1D: CLEFT OF THE SOFT PALATE



Even a "small" cleft of the soft palate can create an opening in the roof of the mouth, which an infant cannot close to create suction for nursing. As the child grows and before the palate is repaired, this opening also allows too much airflow through the nose, causing hypernasal speech.

FIGURE 1E: CLEFT WITH BOTH HARD AND SOFT PALATE

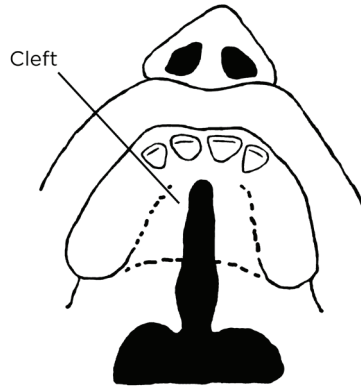


FIGURE 1F: UNILATERAL CLEFT OF THE LIP AND PALATE

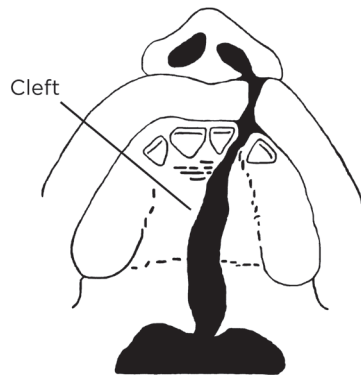
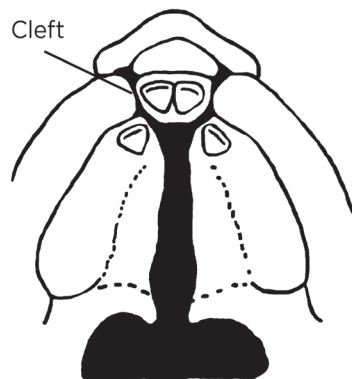


FIGURE 1G: BILATERAL CLEFT OF THE LIP AND PALATE



Illustrations of cleft types adapted with permission from *Looking Forward: A Guide for Parents of the Child with Cleft Lip and Palate*, (Mead Johnson, 1995). Illustrations adapted by Daryl Tong. Updated 2008. Available at: https://www.meadjohnson.com/pediatrics/us-en/sites/hcp-usa/files/MJN362-LF258-cleft-lip_o.pdf

Prenatal Diagnosis

In the past, prenatal diagnosis of a cleft lip was almost always made in association with other abnormalities in the fetus. With improvements in ultrasound technology, the prenatal diagnosis of isolated cleft lip is increasingly common. Detection rates for orofacial clefts using 2D ultrasound screening vary. A recent meta-analysis revealed that most studies reported detection rates between 9-50%, indicating a substantial proportion of diagnoses of cleft lip and palate are still missed.⁵ In the United Kingdom, routine views of the face and lips were added to antenatal ultrasound guidelines in 2000 and detection rates of cleft lip in low risk populations increased from 16-33% to 75% with 2D ultrasound between 18-23 weeks gestation.⁶ Conventional ultrasound can identify cleft lip and palate with standard views.⁷ The use of 3D ultrasound of the face improves detection rates significantly.⁷⁻⁹

Thus, if there is a family history of clefting or if there is a concern about a possible cleft on screening ultrasound, a referral should be made for a complete diagnostic ultrasound (including 3D images if possible) and genetic counseling. Ultrasounds obtained during 18-23 weeks gestation have been most accurate. However, if an expecting mother is being scanned early, for example, if amniocentesis is being considered (typically at 15-17 weeks), an additional later scan can be performed if there are concerns about a possible cleft.

Ultrasound can often establish whether a cleft lip is unilateral or bilateral. While ultrasound is a sensitive modality for detection of cleft lip, it is still difficult to make an antenatal diagnosis of a cleft palate, unless it is detected in association with a wide cleft lip. Detection rates for cleft palate without cleft lip vary from 0-22%.^{10,11} 3D ultrasound may also be more sensitive to diagnose palatal involvement. Multiple factors affect ultrasound quality: fetal position at the time of ultrasound, the mother's weight, the amount of amniotic fluid, the type of ultrasound machine, and experience of the sonographer.¹² These pitfalls are not a concern with other imaging modalities. Fetal MRI has been used to detect abnormalities including cleft palate, however, experience and availability is somewhat limited at this time.^{13,14}

Once a cleft lip and/or palate is identified, the family should be referred for genetic counseling to discuss options for additional testing, including amniocentesis. A complete pregnancy and family history should be performed. This should include information on any teratogenic exposures, maternal health issues, and the presence of family members with clefts or other congenital differences, developmental problems and genetic syndromes. If prenatal ultrasounds and tests reveal anomalies in addition to the cleft, the possibility of a syndrome or chromosome difference is more likely. Even if genetic tests are negative, parents should be informed that an accurate diagnosis and complete discussion of prognosis and recurrence risks can only take place after the baby is born.

When a cleft lip/palate is detected prenatally, the family should be referred to a cleft lip and palate team to learn about the care and management of children with clefts. The experience of learning about a cleft in an unborn child will have associated emotions and reactions that may be different for each individual. Initial feelings may include disbelief and confusion. With realization of the diagnosis may come a sense of loss and grief. Recognizing the loss and these feelings can help a family actively mourn. The process of recovery guides a family into a mode of problem solving, and opens up opportunities for learning about cleft care and their baby's needs.¹⁵

At the family's first visit with the cleft lip and palate team, feeding instructions, bottles, and a breast pump prescription should be provided, and a clear plan for the newborn period should be formulated. A description of the medical and surgical plans and timeline of care for a child with cleft lip and palate can be shared. This is an opportunity to formulate a feeding plan, learn about the future care their child will receive, and meet the providers involved in this care. The prenatal visit has the potential to increase a parent's sense of control and preparedness in the face of this unanticipated diagnosis. As the diagnosis of a birth difference often creates stress for a family, providing psychosocial and emotional support are also essential at this time. Supportive counseling and referral to community resources may be helpful. Appropriate resources and support will help families adjust to the unexpected news.

STANDARDS OF CARE

Overview of Standards of Care for Cleft Lip and Palate

General standards of care for children with cleft lip and palate and other craniofacial anomalies have been created by the American Cleft Palate-Craniofacial Association (ACPA). These standards are contained in two documents summarized below. Central to these documents is the principle that management of patients with cleft lip/palate is best provided by an interdisciplinary team of specialists with experience in this field. Both documents are available from the ACPA national office (see Resources, page 62).

ACPA Parameters of Care (Revised 2017)

This document is based on a national consensus conference funded by the Bureau of Maternal and Child Health, in conjunction with the ACPA. It draws on the 1987 Surgeon General's Report on children with special health care needs.

Fundamentals of care for children with cleft lip/palate (and other craniofacial anomalies):

1. Requires an interdisciplinary team of specialists with experience in cleft lip/palate.
2. Team must see sufficient numbers to maintain expertise.
3. Optimal time for team evaluation is in first few days or weeks of life.
4. Team should assist families in adjustment to the cleft.
5. Team should adhere to principles of informed consent, form partnership with parents, and allow participation of the child in decision-making.
6. Care is coordinated by the team, and is provided locally if appropriate team care is available.
7. Team should be sensitive to cultural, psychosocial and other contextual factors.
8. Team is responsible for monitoring short- and long-term outcomes, including quality management and revision of clinical practices, when appropriate.
9. Treatment outcomes include psychosocial well-being, and effects on growth, function and appearance.
10. Long-term care includes evaluation and treatment in the areas of audiology, dentistry/orthodontics, genetics/dysmorphology, nursing, oral and maxillofacial surgery, otolaryngology, pediatrics, plastic surgery, psychosocial services and speech-language pathology.

Standards for Approval of Cleft Palate and Craniofacial Teams (2017)

In March 2010 the ACPA transitioned from a self-reporting system to an active process with the development of the “Standards for Approval of Cleft Palate and Craniofacial Teams Commission on Approval of Teams” or CAT available at:

<http://acpa-cpf.org/wp-content/uploads/2017/06/standards.pdf>

Teams are to be evaluated and approved by the CAT, and then listed by the ACPA as Cleft Lip/Palate Teams if they meet the criteria outlined below. In addition, other teams may be listed which do not meet all these criteria, but are either new, provide only evaluation and treatment review, or serve low-population areas. In reporting professional services to the ACPA, teams may not include patients treated on missions to meet required standards.

Basic Criteria: Cleft lip/palate team must meet all eight:

1. Team meets face-to-face at least six times/year, with at least four disciplines present.
2. Team evaluates at least 50 new/return patients a year.
3. Team has central and shared files on each patient.
4. The team has at least one actively involved surgeon, orthodontist and speech-language pathologist. All patients are evaluated by these specialists and one other specialist.
5. Team assures that all children are evaluated by a primary care physician (pediatrician, family physician or general internist).
6. Evaluations of patients by this team include a screening hearing test and tympanogram (all patients with clefts are referred to an otolaryngologist for examination, consultation or treatment).
7. At least one surgeon on the team has operated on 10 or more patients for primary repairs of a cleft lip and/or palate in the past year.
8. Team refers patients requiring facial skeletal surgery (bone grafts, orthognathic surgery) to a surgeon with education, training experiencepreparing him/her for this surgery, and who also has performed 10 osteotomies or more in the past year.

Additional Criteria: Cleft lip/palate team must meet 30 of the following:

1. The team has a speech-language pathologist with education, training and experience in treatment of cleft lip/palate who attends team meetings.
2. The team has at least one speech-language pathologist who evaluates at least 10 patients/year with cleft lip/palate.
3. The team speech-language pathologist performs structured speech assessment during team evaluations.
4. Clinical speech instrumentation (e.g. videofluoroscopy, endoscopy, etc.) is used to assess velopharyngeal dysfunction (VPI).
5. The team has an orthodontist with education, training and experience in treatment of cleft lip/palate who attends team meetings.
6. The team has at least one orthodontist who provides care for at least 10 patients a year with cleft lip/palate.
7. Patients requiring orthognathic treatment (jaw surgery) are referred to an orthodontist with the education, training and experience for provision of orthodontic care as a part of orthognathic treatment.
8. Orthognathic surgical treatments are adequately documented with intraoral dental casts, facial and intraoral photographs, and appropriate radiographs.
9. Orthognathic surgical planning and outcomes are discussed at team meetings.
10. The team has or refers to a pediatric/general dentist/prosthodontist with education, training and experience in dental management of cleft lip/ palate.
11. The team has a surgeon who attends meetings with education, training and experience in treatment of cleft lip and palate.
12. The team has a psychologist, social worker or other mental health professional who evaluates all patients on a regular basis.
13. The team routinely tests or screens patients for learning disabilities, and developmental, psychological and language skills.
14. When indicated, the team collects school reports and other learning information.
15. The team has a nurse or other professional to provide supportive counseling and feeding information.
16. When requested by the family, the team refers to parent support groups in the community.
17. The team provides pre- and post-operative supportive counseling and instruction to parents and patients.
18. The team provides formal genetic counseling or clinical genetic evaluation.
19. Hearing is tested by an audiologist before the child is one year of age.
20. The team has an otolaryngologist with education, training and experience in treatment of cleft lip/palate.
21. The team evaluation includes an ear exam by an otolaryngologist on a routine basis beginning before one year of age.

22. After team evaluation, the patient and family have an opportunity to ask questions and discuss the treatment plan.
23. The team routinely prepares summary letters or reports containing the treatment plan to be sent to the family in a timely fashion.
24. Treatment plan reports are sent to the patient's care providers in the community in a timely fashion (with parental permission).
25. The team records include diagnosis.
26. The team records include complete medical history.
27. The team records include plan or treatment goals, which are reviewed regularly.
28. The team records include a social and psychological history.
29. The team records include dental and orthodontic findings and history.
30. When indicated, the team makes intraoral dental casts on patients.
31. The team takes facial photographs on patients in treatment or evaluation.
32. When indicated, the team takes appropriate radiographs including lateral cephalograms.
33. The team has an office and a coordinator.
34. The team supports, encourages or offers continuing medical education in cleft lip/palate care to members.
35. The team provides case management (follow-up, referral, coordination of care) and provides advocacy and assistance, as needed.

TABLE 1: SUMMARY OF CARE TEAM INTERVENTIONS BY SPECIALTY

Note: This table is only a summary and may not include all disciplines needed for a particular child. Team participants may also vary depending upon community and location.

SPECIALTY:	INTERVENTIONS:	REFER TO PAGE:
Nursing (team coordinator, public health nurse, feeding therapist)	<ul style="list-style-type: none"> • Coordinate care • Feeding counseling • Monitor psychosocial issues • Pre- and post-operative teaching 	24-26
Pediatrics/primary care/genetics	<ul style="list-style-type: none"> • Monitor medical issues • Assist with coordination of care and referrals • Monitor developmental and behavioral issues • Genetics/dysmorphology assessment 	27-30, 33-35
Social work and psychology	<ul style="list-style-type: none"> • Monitor psychosocial issues • Developmental/behavioral problems • Refer to community resources • Assist with coordination of care 	30-32
Surgery (plastic surgery, otolaryngology, oral and maxillofacial surgery)	<ul style="list-style-type: none"> • Lip and palate repair • Velopharyngeal surgery for VPI • Ear tubes • Rhinoplasty • Alveolar bone graft • Jaw surgery • Dental extractions • Airway assessment • Assist with VIP evaluation 	36-43, 51-53
Audiology	<ul style="list-style-type: none"> • Test and monitor hearing • Recommend preferential seating and amplification when appropriate 	40-42
Speech and language	<ul style="list-style-type: none"> • Monitor speech-language development • Assist with VPI evaluation • Communicate with school or outside therapists • Provide speech-language therapy; provide speech appliance (obturation) 	44-47
Orthodontics and dentistry	<ul style="list-style-type: none"> • Presurgical orthopedics as needed • Follow dental eruption, hygiene • Monitor facial and jaw growth • Move dental arches/teeth • Provide speech prosthesis, bridges, implants as needed 	48-50

TABLE 2: SUMMARY OF KEY INTERVENTIONS BY AGE

Note: This table is only a *summary* and does not contain every intervention that could be needed by a particular child at a certain age. For more details see pages referenced.

AGE RANGE:	INTERVENTIONS:	REFER TO TABLE:
Prenatal	<ul style="list-style-type: none"> • Refer to cleft lip/palate team • Review prenatal diagnostic studies • Genetic counseling • Address psychosocial issues • Provide feeding instructions • Make feeding plan 	3 (page 14)
Birth–1 Month	<ul style="list-style-type: none"> • Refer to cleft lip/palate team • Medical diagnosis and genetic counseling • Address psychosocial issues • Provide feeding instructions and monitor growth • Begin pre-surgical orthopedics if indicated 	4 (page 15)
1–6 Months	<ul style="list-style-type: none"> • Monitor feeding and growth • Repair cleft lip • Monitor ears and hearing • Begin/continue pre-surgical orthopedics if indicated 	5 (page 16)
5–15 Months	<ul style="list-style-type: none"> • Monitor feeding, growth, development • Monitor ears and hearing; consider ear tubes • Repair cleft palate • Instruct parents in oral hygiene 	6 (page 17)
16–24 Months	<ul style="list-style-type: none"> • Assess ears and hearing • Assess speech-language • Monitor development 	7 (page 18)
2–5 Years	<ul style="list-style-type: none"> • Assess speech-language; manage VPI* • Monitor ears and hearing • Consider lip/nose revision (if necessary) • Assess development and psychosocial adjustment 	8 (page 19)
6–11 Years	<ul style="list-style-type: none"> • Assess speech-language; manage VPI • Orthodontic interventions • Pre-surgical orthodontic interventions • Alveolar bone graft 	9 (page 20)
12–21 Years	<ul style="list-style-type: none"> • Jaw surgery rhinoplasty (if necessary) • Orthodontics; bridges, implants (if necessary) • Genetic counseling • Assess school/psychosocial adjustment 	10 (page 21)

*VPI = velopharyngeal insufficiency. See page.

TABLE 3: PRENATAL INTERVENTIONS

PROBLEMS:	INTERVENTIONS:
Family need for information on the care of a child with cleft lip/palate	<ul style="list-style-type: none"> • Refer family to cleft lip/palate team for information on cleft care, including feeding, speech, ear and dental problems, surgical management, etc.
Psychosocial stress in family	<ul style="list-style-type: none"> • Team nurse/social worker meet with family • Refer to community resources as needed
Anticipated closure of cleft lip/palate	<ul style="list-style-type: none"> • Team plastic surgeon meets with family if possible to discuss plans for closure
Need to anticipate feeding plan	<ul style="list-style-type: none"> • Team nurse coordinator or public health nurse counsels and provides feeding instructions and specialized bottles (see pages 25-26)
Team/family need for accurate medical/diagnostic information on cleft (and any other anticipated medical problems)¹⁶	<ul style="list-style-type: none"> • Team reviews ultrasound results and information from perinatologist, geneticist, primary care physician, etc. • If relevant, discusses implications for cleft care with family • Family discusses other issues with appropriate care providers (primary physician, OB, perinatologist, geneticist, etc.)

Summary of critical interventions at the time of prenatal diagnosis:

- Review prenatal diagnostic studies
- Genetic counseling
- Refer to cleft lip/palate team
- Make feeding plan
- Address psychosocial issues and concerns

TABLE 4: INTERVENTIONS FOR BIRTH THROUGH 1 MONTH

PROBLEMS:	INTERVENTIONS:
Need for interdisciplinary care of a child with a cleft	<ul style="list-style-type: none"> • Refer to cleft lip/palate team
Cleft lip/palate	<ul style="list-style-type: none"> • Team specialists assess the cleft and examine infant for dysmorphic features and other anomalies • Discuss diagnosis and treatment plan with the family • Address etiology and recurrence risks • Consider pre-surgical orthopedics, depending on the structure of the cleft (see page 49-50)
Feeding difficulty and high risk for poor weight gain	<ul style="list-style-type: none"> • Team nurse or public health nurse counsels family on appropriate feeding technique (see pages 25-27) • Assess weight weekly for first month to verify adequate gain: should regain birth weight by 2 weeks, and 5-7 oz. per week thereafter • Additional consultation with cleft feeding specialist or dietitian if needed • Provide psychological support if feeding plan involves loss of ability to breast-feed
Middle ear status, hearing, airway	<ul style="list-style-type: none"> • Assess middle ear status • Perform Brainstem Auditory Evoked Response (BAER) if newborn screen is abnormal • Rule out airway problems, especially if diagnosis is Robin Sequence or if the cleft is part of a syndrome (see page 27)
Family's need for information and psychosocial support	<ul style="list-style-type: none"> • Help family deal with guilt, loss and adjustment issues • Identify community resources and support groups • Address barriers to care: insurance issues, transportation needs, absence from work, language and cultural differences • Provide psychosocial support and assessment to optimize child and family adjustment

Summary of critical interventions for ages birth through 1 month:

- Referral to cleft lip/palate team
- Medical diagnosis and genetic counseling
- Feeding and growth interventions
- Address psychosocial issues
- Begin pre-surgical orthopedics if indicated

TABLE 5: INTERVENTIONS FOR 1 THROUGH 6 MONTHS

PROBLEMS:	INTERVENTIONS:
Need for interdisciplinary care of a child with a cleft	<ul style="list-style-type: none"> • Refer to cleft lip/palate team (if not already done) • Team coordinates care and needed surgeries
Cleft lip/palate	<ul style="list-style-type: none"> • Team specialists assess the cleft and examine infant for dysmorphic features and other anomalies that may become more apparent • Discuss diagnosis and treatment plan with the family • Address etiology and recurrence risks (if not already done) • Monitor pre-surgical orthopedics (if being used)
Feeding difficulties and growth problems	<ul style="list-style-type: none"> • Monitor feeding; provide instructions as needed • Verify adequate weight gain by plotting on growth grid • Refer to cleft feeding specialist/dietitian, as needed
Middle ear status, hearing, airway	<ul style="list-style-type: none"> • Monitor middle ear status (refer to otolaryngologist if needed) • Brainstem Auditory Evoked Response (BAER) if otoacoustic emissions abnormal (if not already done) • Monitor for airway problems if diagnosis is Robin Sequence or other syndrome (see page 27)
Cleft lip and nasal deformity	<ul style="list-style-type: none"> • Repair cleft lip, usually between 3-6 months
Family's need for specific pre- and post-operative lip repair information	<ul style="list-style-type: none"> • Teach pre- and post-operative care requirements (e.g. feeding plan, pain management, and, if recommended by your team, use of arm splints) • Assess family's understanding of these instructions and ability to follow through • Help family make the necessary medical and social arrangements
Family's ongoing need for information and psychosocial support	<ul style="list-style-type: none"> • Help family deal with guilt, loss and adjustment issues • Identify community resources and support groups • Address barriers to care: insurance issues, transportation needs, absence from work, language and cultural differences • Provide psychosocial support to optimize child and family adjustment

Summary of critical interventions for ages 1 through 6 months:

- Begin/continue pre-surgical orthopedics if indicated
- Monitor feeding and growth
- Repair cleft lip

TABLE 6: INTERVENTIONS FOR 7 THROUGH 15 MONTHS

PROBLEMS:	INTERVENTIONS:
Need for continued interdisciplinary care of a child with a cleft	<ul style="list-style-type: none"> • Cleft lip/palate team coordinates cleft care including surgeries
Middle ear status, hearing, airway	<ul style="list-style-type: none"> • Place/replace ear tubes if persistent or recurrent middle ear effusions > 3 months (coordinate with palate repair if possible) • Assess hearing at 8 months of age with behavioral audiogram and at 6-month intervals thereafter • Monitor for airway problems if diagnosis is Robin
Cleft palate	<ul style="list-style-type: none"> • Repair cleft palate, usually at 9-15 months (palatoplasty, see page 37)
Family's need for accurate genetic information	<ul style="list-style-type: none"> • Provide genetic counseling (if not already done) (see pages 33-35)
Family's need for specific pre- and post-operative palate repair information	<ul style="list-style-type: none"> • Teach pre- and post-operative care requirements (e.g. feeding plan, arm splints or hand mitts, pain management) • Assess the family's understanding of these instructions and ability to follow through • Help family make the necessary medical and social
Feeding, growth and development issues	<ul style="list-style-type: none"> • Advance to solid foods • Address feeding difficulties • Provide regular growth and development screening
Abnormal dental development	<ul style="list-style-type: none"> • Instruct parents in oral hygiene; monitor tooth eruption • Provide parents with information about expected dental development
Speech-language production	<ul style="list-style-type: none"> • Provide speech-language assessment and counseling to parents before the palate repair • Assess child's speech-language 3-6 months after palate repair
Family's ongoing need for information and psychosocial support	<ul style="list-style-type: none"> • Continue to review family/child adjustment issues • Identify community resources and support groups • Address barriers to care: insurance issues, transportation needs, absence from work, language and cultural differences • Provide psychological support to optimize child and family adjustment

Summary of critical interventions for ages 7 through 15 months:

- Monitor feeding, growth and development
- Consider ear tubes/assess hearing
- Repair cleft palate

TABLE 7: INTERVENTIONS FOR 16 THROUGH 24 MONTHS

PROBLEMS:	INTERVENTIONS:
Need for continued interdisciplinary care of a child with a cleft	<ul style="list-style-type: none"> • Cleft lip/palate team provides cleft care and coordination
Middle ear status, hearing, airway	<ul style="list-style-type: none"> • Monitor middle ear status every six months • Behavioral audiogram • Place/replace tubes if middle ear effusions persist for more than 3 months
Speech-language production	<ul style="list-style-type: none"> • Assess speech-language, especially velopharyngeal mechanism (see pages 44-45)
Feeding, growth and development issues	<ul style="list-style-type: none"> • Address feeding difficulties • Provide regular growth and development screening
Repaired cleft lip/palate	<ul style="list-style-type: none"> • Monitor integrity of the surgical repairs • Assess the palate for fistula(e)
Abnormal dental development	<ul style="list-style-type: none"> • Monitor tooth eruption and oral hygiene • Reassure parents regarding expected dental development
Family's ongoing need for information and psychosocial support	<ul style="list-style-type: none"> • Continue to review family and child adjustment issues • Identify community resources and support groups • Address barriers to care: insurance issues, transportation needs, absence from work, language and cultural differences • Provide psychological support to optimize child and family adjustment

Summary of critical interventions for ages 16 through 24 months

- Monitor ear tubes and hearing
- Assess speech-language and development
- Monitor development

TABLE 8: INTERVENTIONS FOR 2 THROUGH 5 YEARS

PROBLEMS:	INTERVENTIONS:
Need for continued interdisciplinary care of a child with a cleft	<ul style="list-style-type: none"> • Cleft lip/palate team provides cleft care and coordination
Middle ear status, hearing, airway	<ul style="list-style-type: none"> • Monitor middle ear status every 6 months • Place/replace ear tubes if middle ear effusions persist for > 3 months • Assess hearing at 6-month intervals until age 3, then every 6-12 months as indicated • Assess airway, sleep disturbances (workup if suspect obstructive sleep apnea that could be due to small jaw, large tonsils/adenoids, pharyngeal flap or sphincter pharyngoplasty) • Caution needed when considering adenoidectomy
Speech-language production	<ul style="list-style-type: none"> • Assess speech-language, monitor for velopharyngeal insufficiency (VPI) or other speech disorders. • Formal VPI workup, as indicated (see pages 38) • Consider treatment options (speech therapy, surgery, obturation)
Abnormal dental development	<ul style="list-style-type: none"> • Orthodontic exam and dental records at age 6 or 7 for bone graft timing and management of abnormal dentition • Dental extractions if needed
Child's overall developmental and behavioral adjustment	<ul style="list-style-type: none"> • Monitor for developmental/behavioral problems • Provide counseling or make referrals as needed
Family's ongoing need for information and psychosocial support	<ul style="list-style-type: none"> • Continue to review family adjustment issues • Identify community resources and support groups • Address barriers to care: insurance issues, transportation needs, absence from work, language and cultural differences • Provide psychological support to optimize child and family adjustment (school entry and peer comments may be sources of stress)

Summary of critical interventions for ages 2 through 5 years:

- Assess speech for VPI; consider interventions
- Monitor ear tubes and hearing
- Lip/nose revision (if necessary)
- Assess child's development, including language and psychological adjustment

TABLE 9: INTERVENTIONS FOR 6 THROUGH 11 YEARS

PROBLEMS:	INTERVENTIONS:
Need for continued interdisciplinary care of a child with a cleft	<ul style="list-style-type: none"> • Cleft lip/palate team provides cleft care and coordination
Middle ear status, hearing, airway	<ul style="list-style-type: none"> • Monitor middle ear status every 6-12 months • Place/replace ear tubes if middle ear effusions persist > 3 months • Perform audiogram every 6-12 months as indicated • Assess airway, sleep disturbances (workup if suspect obstructive sleep apnea that could be due to small jaw, large tonsils/adenoids, pharyngeal flap or sphincter pharyngoplasty) • Caution needed when considering adenoidectomy (see page 39)
Speech-language production	<ul style="list-style-type: none"> • Assess speech-language; monitor for velopharyngeal insufficiency (VPI) • Formal VPI workup as indicated (see pages 38) • Consider treatment options (speech therapy, surgery, obturation) • Communicate with school or outside therapist if the child is receiving speech therapy
Abnormal jaw growth and dental developmental	<ul style="list-style-type: none"> • Regular orthodontic exams and records to monitor jaw growth and readiness for alveolar bone graft • Orthodontics often needed before and after the bone graft • Dental extractions as needed; monitor dental hygiene
Persistent bony cleft of alveolus and oro-nasal fistula(e)	<ul style="list-style-type: none"> • Bone graft to the alveolar cleft(s) and closure of the oro-nasal fistula(e) (timing is critical)
Child's overall developmental and behavioral adjustment	<ul style="list-style-type: none"> • Monitor school performance, emotional and behavioral issues • Make referrals as necessary (see pages 30-32)
Child and family's ongoing need for information and psychosocial support	<ul style="list-style-type: none"> • Continue to review family adjustment issues • Identify community resources and support groups • Address barriers to care: insurance issues, transportation needs, absence from work or school, language and cultural differences • Provide psychological support to child and family • Involve child in decision-making process as age/abilities allow

Summary of critical interventions for ages 6 through 11 years:

- Assess speech for VPI; consider interventions
- Orthodontic interventions and alveolar bone grafting
- Monitor school performance and psychological adjustment
- Involve child in medical decision-making process

TABLE 10: INTERVENTIONS FOR 12 THROUGH 21 YEARS

PROBLEMS:	INTERVENTIONS:
Need for continued interdisciplinary care of teen/young adult with a cleft	<ul style="list-style-type: none"> • Cleft lip/palate team provides cleft care and coordination
Middle ear status, hearing, airway	<ul style="list-style-type: none"> • Monitor middle ear status every 6-12 months • Place/replace ear tubes if middle ear effusions persist for > 3 months <ul style="list-style-type: none"> • Audiometry every 6-12 months until ears are clear and hearing normal for 2 years • Assess airway, sleep disturbances (workup if suspect obstructive sleep apnea that could be due to small jaw, large tonsils/adenoids, pharyngeal flap or sphincter pharyngoplasty) • Caution needed when considering adenoidectomy (see page 39)
Speech-language production	<ul style="list-style-type: none"> • Assess speech, rule out velopharyngeal insufficiency (VPI) • Workup VPI if indicated (see pages 38) • Consider treatment options (therapy, surgery, obturation) • Communicate with school or outside speech clinician
Abnormal dental developmental	<ul style="list-style-type: none"> • Regular orthodontic exams and dental records to monitor bone graft and jaw growth • Final orthodontics when facial growth complete • Provide bridges or implants as needed
Maxillary/mid-face hypoplasia with malocclusion	<ul style="list-style-type: none"> • Orthodontic treatment and/or jaw surgery
Adolescent's and family's ongoing need for information and support	<ul style="list-style-type: none"> • Continue to review adolescent and family adjustment issues • Identify community resources and support groups • Address barriers to care: insurance issues, transportation needs, absence from work or school, language and cultural differences • Provide psychological support to optimize adolescent and family adjustment (peer teasing, adolescent self-esteem and school transitions are areas of focus) • Provide adolescent/family with appropriate genetic information, including risks for recurrence • Involve adolescent in medical decisions; respect preferences on elective procedures (see page 22-23)
Adolescent's overall developmental adjustment	<ul style="list-style-type: none"> • Review school performance, academic/vocational plans • Screen for behavioral/emotional problems; refer as needed

Summary of critical interventions for ages 12 through 21 years:

- Jaw surgery, rhinoplasty if needed
- Final orthodontics
- Genetic counseling

SPECIFIC CARE AND TREATMENT

Ethics and Children with Cleft Lip/Palate

Ethical issues arise whenever medical decisions are made for patients who legally cannot make their own decisions about medical care, due to their chronological age or concerns about intellectual disability or other compromising medical condition. These concerns take on special importance for young patients when the goal of medical therapies includes both objective and subjective outcomes. The key points in decision-making for these children are summarized below. In addition, changes in the health care system raise ethical issues relating to advocacy and access to care for this population. These will also be considered briefly.

Decision-making for Children

Key points in pediatric decision-making include the following:¹⁶

Incompetency of children

Patients under the age of 18 are presumed to be incompetent by virtue of age, and cannot give informed consent. The legal age of consent is an arbitrary designation, therefore when working with patients under 18 years of age, close attention must be paid towards their opinions.

Surrogate decision-makers, parental permission, “best interests” standards, and the harm principle

Surrogates, usually their parents, make medical decisions for young patients. These decisions must include several ethical components. They must reflect what is in the best interests of the young patient, they should not lead to harm,¹⁷ they should reflect the patient’s involvement in discussions about their ongoing treatment (assent), and they must take into consideration the surrounding values and right to autonomy related to family individuality.¹⁸ This must be done intentionally, as a determination of the appropriateness of any one decision can lead to identifying medical neglect or abuse. When considering a referral to local child abuse providers, one must consider not only whether the decision supports a child’s best interest, but also whether or not the decision poses some harm to the child. The analysis of the situation requires that a provider use the Harm Principle before taking steps to interfere in parental decision-making. (Diekema, 2011)

Informed consent

Health care providers are obligated to fully inform parents and patients of the risks and benefits of proposed care plans, including alternative therapies. Ideally, informed consent discussions should take place over time, not just at the point of surgical interventions, and with providers who have developed a positive relationship with the family. Discussions with parents and young patients should recognize cultural and ethnic differences, as these variables can affect the family's adjustment and understanding of their child's diagnosis and treatment.

Role of the young patient

Young patients need to be active participants in the consent process both for respect of their autonomy and for developing a foundation for future health (2004).¹⁹ By the developmental age of 7 years, most patients have the capacity to assent to interventions and procedures. When a young patient refuses treatment recommendations, their opinion should be honored whenever possible. The more serious or emergent the medical circumstances, the more difficult it can be to balance the autonomy of the patient, the family, and what is determined to be in the best interests of the patient. Any situation that leads to a decision to override the young patient's wishes must include a careful discussion about the importance of the therapies. Reasons for overriding the young patient's wishes should be explained carefully and the patient must be offered support during the interventions. The more elective the intervention, the more essential it is that the patient's wishes be honored.

Quality of Life as a Goal of Therapy*Deciding on quality of life interventions*

Many interventions for children with cleft lip/palate are intended to improve the quality of life, although most have a functional component as well. Interventions with an impact on the quality of life include correcting or minimizing facial deformity, improving dental appearance and function, optimizing psychosocial adjustment, improving developmental outcome, and normalizing speech and hearing. Because of the subjective nature of some of these therapy goals, it is important that patients demonstrate assent and participate in the decision-making process when appropriate. By adolescence, most patients are able to share fully in the decision-making process, and should have veto power for some procedures. There is considerable variability in the degree to which cleft lip/palate affects self-esteem and quality of life. It is important to elicit the patient's independent opinion about how any one intervention will impact their quality of life. Thus, some surgeries to correct these deformities are often postponed until the patient is old enough to share in the decision-making, unless the intervention must be performed earlier to be successful or safe. Making sure to get patient reported outcomes while also practicing shared decision-making will help to assure that the treatments done truly benefit the patient.

Changes in Health Care: Access and Advocacy

Changes in the health care system have potential impact for children with cleft lip/palate. Patients with relatively rare medical conditions such as these may have difficulty accessing care, because specialists with adequate experience and training in this field are relatively rare. There is risk that care will be parceled out in an uncoordinated fashion to a number of providers inside and outside of an insurance plan's network. In order to access team care it may be necessary for families, medical teams and insurance providers to work together to educate payers about the medical necessity of craniofacial treatments.

This collaborative advocacy with insurance companies needs to include language that focuses on how and why craniofacial treatments are best provided within a multidisciplinary team. An insurance company must be educated about why team care for a patient with cleft lip/palate represents a "basic level of health care". In addition to the problems with accessing care within established insurance networks, surgeries required for the treatment of cleft lip and/or palate are often viewed as optional or elective and as a result may not be paid for by the insurance company. The insurance company can initially assess surgical treatments as being "cosmetic". This, however, is incorrect as the surgeries are undertaken to bring the individual with a visible birth defect closer to the range of normal. The importance of such advocacy is described in the 2018 Parameters of Care for Evaluation and Treatment of Patients with Cleft Lip/Palate or Other Craniofacial Differences published by the American Cleft Palate and Craniofacial Association. In the preface to this document, the importance of "encouragement of adequate health care financing", is acknowledged. Another source of helpful definitions of the importance of funding for team care is the 1987 Surgeon General's Report on Children with Special Healthcare Needs.

Nursing and Coordination of Care

Specialty nurses, including cleft lip/palate team nurses and public health nurses associated with the Children with Special Health Care Needs Program (Department of Health), play an important role in the care of patients with cleft lip/palate. They assist with coordination of care, provide peri-operative counseling and help monitor psychosocial issues. When appropriate, they may also provide community outreach and refer to community resources. In addition, because of their knowledge of the medical issues at stake in the care of these children, they may be called upon to help with referrals.

One of the most important functions of nurses and other knowledgeable specialists is offering detailed feeding instructions and support for new parents of babies with cleft lip/palate. The importance of feeding issues in the care of these infants prompts this summary on the following pages:

Feeding the Infant with a Cleft Lip/Palate

I. For those infants with a cleft lip only:

Infants that have only a cleft lip can usually be fed by either breast or bottle. Some problem-solving may be needed to ensure that the infant can get a tight seal around the breast or nipple, particularly if the cleft is broad. Early referral to the infant-feeding specialists or nurses associated with cleft lip/palate teams can facilitate this problem-solving.

II. For those infants with a cleft palate, with or without a cleft lip:

The infant with a cleft palate will require specific bottles and a special feeding technique. Breast-feeding and use of a regular bottle are rarely possible. Lack of knowledge of this important fact can lead to poor weight gain or failure to thrive.

A. Why the infant with a cleft palate cannot breast-feed or use a regular bottle:

The purpose of the palate is to separate the mouth from the nose. Normally the soft palate at the back of the mouth moves up to close off the passage to the nose during feeding. This creates a closed system, and the sucking motions create negative pressure which pulls the milk out of the breast or bottle. A cleft palate prevents the infant from creating a closed system in his/her mouth, and makes it impossible for the milk to be pulled out. The infant will look like he/she is sucking, but he/she will be using up precious calories in a futile attempt to gain adequate nutrition.

B. How to feed the infant with a cleft palate:

The proper bottle is the key to a successful feeding plan. Currently, there are four options widely used. The first is the Dr. Brown specialty care feeder. This bottle utilizes a one-way valve inserted at the base of the nipple. In addition, different nipples which allow for flow options are available to customize to the specific needs of the infant. The nipple options include ultra-preemie, preemie, and #1-4. Other bottle choices include the Mead Johnson Cleft Palate Nurser, Medela® Special Needs Feeder, and the Pigeon Cleft Palate Nurser. The Mead Johnson Cleft Palate Nurser is a soft-sided bottle that is squeezed in coordination with the infant's sucking efforts, and thus milk is delivered into the mouth. The second is the Medela® Special Needs Feeder which has a large, compressible nipple with a one-way valve at its base that keeps the nipple full of milk. The nipple has variable flow rates depending on its position. The infant's effort to compress the soft nipple is often sufficient to dispense the milk into the infant's mouth, but this can also be assisted by squeezing the nipple to increase the flow. The Pigeon Cleft Palate Nurser, distributed by Phillips, also makes use of a one-way valve at the base of the nipple. In addition, the nipple is constructed with a thinner, more compressible underside so that the infant's tongue is effective in compressing the nipple to produce the flow. None of these bottles are available in stores, but all cleft teams can provide them to families and/or provide phone numbers or websites for

ordering. All four of these bottles work without the infant needing to create intraoral suction in order to pull milk out of the nipple. They all require parent training for proper use. For training, contact infant feeding therapist or nurse experienced in feeding infants with clefts.

Mothers and families need adequate psychosocial support to process the loss of the ability to breast-feed their infant. Pumping breast milk for use in a specialized bottle allows a mother to give her baby her own milk. The use of an appropriate, hospital grade, breast pump for long term use should be assessed and facilitated.

However, long-term pumping requires a considerable commitment of time and effort to maintain an adequate milk supply in the absence of normal infant sucking. The need for supplemental feedings with formula must be closely monitored.

C. Establishing feeding goals and monitoring weight gain for the infant with a cleft palate:

Even with a specialized bottle, close attention to weight gain is of great importance for the infant with a cleft palate. If the infant does not maintain appropriate growth, feeding re-evaluation and changes in technique may be needed. The nurse or feeding therapist experienced in feeding an infant with a cleft can evaluate the feeding process and make changes. Sometimes a consultation with a dietitian is needed to establish calorie goals and to provide recipes to increase the calories in the breast milk or formula. Infants with isolated cleft lip/palate should be able to maintain normal growth. There should be little tolerance for any failure to follow a normal growth curve in the first months of life.

These two feeding parameters must be observed to promote adequate weight gain:

1. The infant's intake over 24 hours should be 2.5 ounces of milk for each pound that he/she weighs.
2. No feeding session should take longer than 30 minutes. If it takes longer than this, the infant is working too hard and burning calories needed for growth. The measure of success of the feeding plan is adequate weight gain. During the first several weeks of life weekly weights and plotting the data on the growth curve are the proper way to evaluate this.

D. The introduction of solid foods:

The timing and strategy of introducing solid foods should be the same for the baby with a cleft palate as for any other child. Experiment with the consistency of the food to minimize regurgitation out of the nose while still allowing a smooth swallow. Some sneezing may occur because the exposed nasal passages but this does not impede the progression to solid food. Following each meal with swallows of milk or water is all that is needed to remove any remaining food in the mouth.

Partnering with Primary Care Providers

The primary care provider (PCP) is indispensable in the care of the patient with cleft lip and palate. Ideally, the PCP becomes an extended member of the cleft lip/palate team, following many of the same medical issues as the team specialists (such as recurrent ear infections, airway concerns, growth failure and developmental progress). In addition, the PCP may have the special task of advocating for the child in a particular health care system, and preauthorizing visits to the cleft lip/palate team providers. For these reasons it is essential that the PCP be familiar with the special aspects of cleft care.

The following are particularly important issues for the PCP:

- 1. Feeding.** Although many newborns have feeding problems, babies born with cleft palates are particularly at risk for significant failure to thrive. Infants with cleft palate are unable to generate sufficient suction to feed at the breast and must use specialized bottles made for infants with cleft palate. Without the appropriate bottle system, infants are at risk for inefficient, calorie-wasting attempts to suck, resulting in inadequate nutritional intake. In addition, some babies (e.g. with Robin Sequence, discussed below) have difficulty coordinating breathing, sucking and swallowing, which further impedes adequate intake. Growth parameters must be monitored very closely in the first few weeks to months of life. Infants with clefts involving the palate should be evaluated by nurses or feeding therapists with specialized training in feeding infants with clefts. The knowledgeable nurse in the newborn nursery can initiate proper feeding, but these infants should have long term monitoring, often with a PCP working closely with providers on a craniofacial team (see pages 51-53).
- 2. Robin Sequence.** Robin Sequence (RS) consists of mandibular hypoplasia (micrognathia), glossoptosis (posterior tongue position) and resultant upper airway obstruction (occluded or blocked airway). Most children with RS will also have a U-shaped cleft palate. This constellation of findings was first reported by the French stomatologist, Pierre Robin. If the baby appears to have this condition and is having difficulty breathing due to obstruction by the tongue, the baby should be placed in the prone position. Monitoring of the infants respiratory exam, oxygenation (oximeter) and ventilation (CO₂ levels) will help determine improvement of their airway obstruction. If the infant with RS continues to have evidence of airway obstruction at the base of tongue, escalation of care may include placement of a nasopharyngeal (NP) tube, temporary oropharyngeal tube, or intubation. Endotracheal intubation of an infant with RS can be difficult due to the abnormal anatomy. It is recommended to involve an experienced otolaryngologist if any of these interventions are needed. Some infants with RS will benefit from longer term use of the NP tube, early mandibular advancement through distraction osteogenesis, or tracheotomy to stabilize the airway.

Even when the baby with Robin Sequence appears to be comfortable at rest, he/she maybe so stressed during feeding that adequate weight gain does not take place. Therefore, close feeding, weight and growth monitoring is essential. Consideration of supplemental nasogastric tube feedings or gastrostomy tube feedings may be necessary for some infants.

Many factors may contribute to failure to thrive in babies with RS: difficulty coordinating suck/ swallow; inefficiency of feeding with the cleft palate; glossoptosis with increased work of breathing; gastroesophageal reflux; and caloric consumption. These issues can be difficult to resolve, and generally require the coordinated efforts of pediatrics/primary care, otolaryngology, nursing, infant feeding therapy and respiratory therapy. Serial CO₂ monitoring can help reveal hypoventilation. Babies with RS are at higher risk for having early obstructive sleep apnea, and a polysomnogram (sleep study) can be a useful tool to characterize sleep and breathing. A genetics evaluation is recommended, as over half of children with RS will have other anomalies or an underlying syndrome, identification of which will affect other screening and care recommendations. Monitoring and managing these important early issues should be with a cleft lip/palate team and in a hospital where there is access to pediatric anesthesia.

- 3. Middle Ear Effusions.** Infants with cleft palate are at high risk for recurrent and chronic middle-ear effusions or infections. Conductive hearing loss may result from middle-ear fluid, and 90-95% of infants with cleft palate will require ear tube placement, usually at the time of the palate repair. The craniofacial team will work with the PCP to make sure that the infant has completed newborn hearing screening and has ongoing evaluation by audiology and otolaryngology during early childhood (see pages 39-42).
- 4. Genetics/Dysmorphology.** Genes and environmental factors are both thought to contribute to facial development, and families often have questions about why their child was born with a cleft and whether this will happen in a subsequent pregnancy. A visit with a clinical geneticist and genetic counselor should be offered after the child is born, during adolescence, or whenever a family member has questions about etiology and recurrence risks. The possibility of a genetic syndrome should be considered if a patient has atypical facial features, developmental delays, or other anomalies. If the patient is followed by a cleft lip/palate team that is without a dysmorphologist or a geneticist, consider referral to an outside specialist. In some cases, the PCP may need to help with the referral for a genetics evaluation (see pages 33-35).

5. Dental Issues. Dental issues are of paramount importance in the management of patients with cleft lip/palate. PCPs may be asked to authorize visits to orthodontists, pediatric dentists or oral and maxillofacial surgeons, and must appreciate the integral medical role of these specialists in the care of the child with cleft lip/palate.

- First, if presurgical orthopedics are needed (e.g. tape or a nasoalveolar molding appliance to bring the lip/jaw segments closer together before surgery), an appropriate dental specialist (pediatric dentist, prosthodontist or orthodontist) must be involved in the initial assessment during the first few weeks of life.
- Second, good oral hygiene is essential for successful cleft habilitation. Thus once the teeth have erupted, preventive counseling should take place regarding baby bottle caries, proper tooth brushing, etc. The PCP has an important role to play reinforcing proper dental care and hygiene.
- Third, correct placement of the teeth and dental arches may be necessary before alveolar bone grafting can take place. Alveolar bone grafting is usually needed when clefts extend through the upper gum (alveolus). This procedure is generally performed between the ages of 7-10 years, depending upon dental development. The alveolar bone graft provides the foundation for the erupting teeth and support for the nasal base. Orthodontic evaluations and interventions are necessary before and after bone graft surgery.
- Orthodontic interventions are also needed in adolescence to bring teeth into final alignment and address malocclusion resulting from deficiencies in upper/lower jaw growth, a common problem in patients with cleft lip/palate. Some patients will also need jaw surgery because the deficiency is too great for orthodontic compensation alone. Orthodontists, oral and maxillofacial surgeons, and craniofacial surgeons affiliated with the cleft lip/palate teams generally evaluate and treat these issues. (see pages 48-50)

6. Development. Several studies have shown that children with orofacial clefts have an increased risk for developmental delays and poor academic outcomes when compared to unaffected peers. The potential reasons for these differences are numerous but may include hearing loss during infancy associated with the cleft palate, difficulty with speech production because of the anatomic differences, school absences for clinic visits or surgeries, syndromic forms of clefting that are associated with other anomalies or developmental delays, or psychosocial issues associated with facial or speech differences. Both PCPs and cleft lip/palate teams should monitor for problems with development, psychosocial issues, or problems in school, and refer for appropriate interventions and resources when needed (see pages 30-32).

7. General Medical Care. All children, including those with orofacial clefts, require ongoing well-child care and access to medical care when they are sick. While establishing a relationship with a PCP may be difficult initially because of the intense focus on cleft-related issues during early infancy, it is vital that the family partner with a PCP. The PCP will provide health supervision, anticipatory guidance for development and growth, provide immunizations, and treat acute medical problems when needed. The role of the PCP during adolescence is vital as the focus shifts towards educating and screening for issues related to sexual activity, substance abuse, depression and other health problems. Adolescence is a difficult time for most people, but can be especially difficult for those who look or sound different from their peers. Especially as the adolescent with a cleft begins to participate in decision-making around health care interventions, the PCP can provide support and help facilitate communication with the craniofacial team.

Psychosocial and Developmental Issues in Cleft Care

Psychosocial issues are a critical part of the assessment and management of the child with cleft lip/palate, and must be addressed from the onset of care. The birth of a child is always a time of great family adjustment, and it is especially stressful when the child is born with a birth defect such as cleft lip/palate.

Parents often experience feelings of sadness, guilt, anger and fear for their child's future social acceptance. Some parents feel the extent of their emotional turmoil is unwarranted with such a reparable birth defect, and experience guilt that a facial deformity is so disturbing to them. In addition, the feeding difficulties these infants experience can be threatening to new parents, who may doubt their own ability to feed and nurture an infant with such differences. The loss of the ability to breast-feed is especially traumatic for some mothers. In part, through good psychosocial support and proper instructions, most families are able to work through their own emotional turmoil and effectively master the skills needed to feed and nurture these babies. Other issues of concern for new parents relate to accessing professional and community services, securing adequate financial resources and coping with the stress of sending a child to surgery.

As the child grows, the family will have other concerns, often relating to teasing, peer acceptance, speech difficulties, learning and behavior problems. For many families, securing appropriate community and financial resources remain important issues. Children should have their evolving decision-making role acknowledged, and should be personally addressed during appointments. During adolescence there are new challenges, as the maturing teen strives for independence and copes with being different in a highly appearance-conscious culture. Adolescents and pre-teens should be given the opportunity to confidentially share feelings and concerns with a qualified professional. Older children and teens often require considerable support in preparing for major procedures such as alveolar bone grafting (usually performed between the ages of 6-10 years) and jaw surgery

(performed when growth is complete, in the late teen years). Psychosocial assessment and support may also become necessary when a high level of patient compliance and family commitment are required for certain interventions. When considering elective procedures such as lip scar revision or rhinoplasty to correct a facial difference, the maturing child's preferences should be respected. By age 9 or 10, typically developing children should be included in decision making for these elective surgeries.

Other important circumstances that are often addressed by a psychosocial professional include child abuse/neglect, substance abuse, domestic violence and other family dysfunction. It is not uncommon to see a child in a dysfunctional family become overly focused on "fixing my face" as a way for them to fix the dysfunction in the family. There is research to suggest that unless such emotional issues are addressed prior to surgery, such interventions alone are less likely to change self-image and improve quality of life.

A detailed and specific psychosocial assessment is appropriate for all families presenting to a cleft lip/palate team, regardless of socioeconomic status and perceived stability. In assessing children and families, their unique cultural and social characteristics must be taken into account, with a clear understanding of any implications for providing health care. Cultural differences as well as other unexplored parental worries and concerns often contribute to behavior perceived and labeled by health care providers as "noncompliant." Understanding cultural and psychosocial issues is essential for the delivery of good health care.

Learning Disorders and Behavioral Problems

Children with cleft lip/palate are at increased risk for learning disorders. Fluctuating hearing loss associated with middle ear disease may impair speech and language development. Some children with clefts may have learning difficulties associated with a syndromic diagnosis (e.g. 22q11.2 deletion syndrome, Opitz-Frias syndrome, fetal alcohol syndrome). However, children with isolated clefts (especially cleft palate), also appear to be at increased risk for learning problems. Children with cleft lip/palate may be at increased risk for behavioral disorders as well. Again, these disorders may be associated with a syndromic diagnosis, but can occur in children with isolated clefts as well. Symptoms may include social withdrawal, depression, conduct problems or school performance issues. Furthermore, social and educational circumstances, peer dynamics, problems in the child-parent relationship, and intrinsic characteristics of the child (including temperament and underlying cognitive problems) can combine to create a complex clinical picture. For all these reasons, children with cleft lip/palate should be monitored regularly for psychosocial, learning and behavioral problems. When such problems arise, relevant areas should be assessed, and the interaction of these variables recognized. Specialties suited to screen for these disorders include psychology, social work, nursing, developmental pediatrics, primary care, and speech-language pathology. However, all team members and primary care providers should be alerted to the potential for difficulties in these areas, so when problems arise, appropriate referrals can be made.

TABLE 11: KEY PSYCHOSOCIAL AND DEVELOPMENTAL INTERVENTIONS BY AGE

AGE:	INTERVENTIONS:
Birth to 1 Month	<ul style="list-style-type: none"> • Assessment of grief and loss issues • Identify and validate other concerns • Assess family functioning: recognize strengths, weaknesses, cultural differences • Assess family's understanding of medical information • Help incorporate family needs into treatment plan • Make appropriate community referrals
1–15 Months	<ul style="list-style-type: none"> • Follow-up on psychosocial needs of family • Check family arrangements for surgical stays (lip and palate repairs) • Review issues surrounding future pregnancies • Address family stresses surrounding surgery • Ensure family understands post-op care needs • Review financial issues
16–24 Months	<ul style="list-style-type: none"> • Review family's experiences with hospital and surgery • Explore how parents believe child is perceived by others because of appearance/ speech differences • Screen for developmental problems; make referrals if appropriate
2–5 Years	<ul style="list-style-type: none"> • Review family functioning • Review issues surrounding future pregnancies • At school entry, review concerns related to speech, appearance differences and peer acceptance • Screen for developmental/behavioral problems; refer if appropriate • Assess family's understanding of team treatment plan including management of speech problems • Talk directly with child to assess his/her concerns
6–11 Years	<ul style="list-style-type: none"> • Review family function and new stresses • Assess family need for community resources and help getting to medical appointments • Assess child's fears and concerns before surgeries and hospital stays, especially before bone graft • Assess child's concerns related to peer acceptance, speech and facial differences • Model/refer for social skills training, if needed • Screen for learning/behavioral disorders; refer as appropriate • Acknowledge child's evolving role in the decision-making process • Review plans requiring high patient/family compliance (e.g. orthodontic interventions, obturation), including financial issues and family and child's ability to follow through with treatment
12–21 Years	<ul style="list-style-type: none"> • Acknowledge teen's evolving role in the decision-making process • Assess teen's fears and concerns before surgeries/hospital stays • Check for unrealistic expectations of surgery • Assess teen's concerns related to peer acceptance, speech and facial differences • Model/refer for social skills training if needed • Screen for school problems; review academic/vocational plans • Assess psychosocial adjustment of teen and possibility of depression, substance abuse, etc.; make referrals as needed • Assess teen and family understanding of recurrence risks, need for additional genetic counseling

Genetics/Dysmorphology

A dysmorphology or genetics assessment is part of the complete evaluation of every child with a cleft. Cleft lip with or without cleft palate occurs in approximately 1:700 births worldwide, and 1:940 births in the United States. Isolated cleft palate occurs in approximately 1:1500 live births. Although the majority of patients with cleft lip and palate are otherwise healthy, approximately 25% have associated birth defects/chromosomal abnormality, or a genetic syndrome.²⁰⁻²³

Although there are more than 400 syndromes reported in association with cleft lip or cleft palate, the three syndromes that follow should receive special consideration. **(1) 22q11.2 deletion (a.k.a. velocardiofacial syndrome or DiGeorge syndrome)**, should be considered in children with velopharyngeal insufficiency, submucous cleft palate, or cleft palate; **(2) Van der Woude syndrome**, an autosomal dominant condition, should be considered in a child with either cleft lip/palate or cleft palate alone who has a family history of mixed clefting in which either the child or another family member has lower lip pits; **(3) Stickler syndrome**, an autosomal dominant disorder of collagen with variable congenital myopia, clefting, and arthropathy, should be considered in all infants with Robin sequence.

A complete medical history should be obtained on every child with a cleft, including a detailed prenatal history, teratogenic exposures, and a three-generation family history. This family history should include occurrences of clefting (and lower lip pits), hypodontia, other birth defects, developmental disabilities or known genetic syndromes. A complete physical examination by clinical geneticist or dysmorphologist should be pursued to identify dysmorphic features and/or associated birth defects or medical concerns. Additional studies, including ophthalmologic consultation, echocardiography, other radiographic studies, and genetic testing should be directed by the examination and family history to facilitate genetic diagnoses. While the majority of children with cleft lip and/or palate will not have a genetic syndrome, these conditions may have prognostic implications that must be taken into account to help guide medical decisions and counseling. It should be noted that some physical features that suggest a genetic syndrome may not develop until later in infancy, so continued vigilance is needed. Children who were thought to have nonsyndromic clefting that are later identified to have developmental delay should also be referred for genetic evaluation.

Parents typically have many questions about the etiology of clefts to be addressed by the cleft lip/palate team. There is considerable cultural and social variability in family attitudes towards birth defects and their causation. These issues should be explored and, when appropriate, correct information supplied recognizing that western medical information will not necessarily supplant other cultural and ethnic beliefs. Since genetic factors play a role in clefting conditions even in children with nonsyndromic clefting, information on causation and empirical recurrence risks should be provided to all families with clefts based upon the family history.

For parents with one affected child, the recurrence risk for future pregnancies is 2-5% for cleft lip/palate, and 1-2% for cleft palate alone. The risk is highest for bilateral cleft lip/palate (5-6%), compared to 2-3 % for cleft lip without cleft palate. This risk increases if there are additional family members with clefts, up to approximately 8-10% for nonsyndromic clefting. The recurrence risk for clefting associated with genetic syndromes varies with the mode of inheritance of the condition, and can range from 1-50%. Condition-specific recurrence risks and prenatal testing options should be provided to families of a child with syndromic clefting condition. A discussion regarding the potential preventative role of preconception/prenatal folate supplementation and avoidance of environmental risk factors (tobacco smoke, alcohol, and isotretinoin) should be considered.

Parents should be informed of the option of ultrasonography for future pregnancies. Routine mid-pregnancy ultrasound, done at 18-20 weeks gestation, has become a standard part of obstetrical care, and improving technology (including 3-D imaging) has led to increasing numbers of infants with cleft lip to be identified. Isolated cleft palate remains extremely difficult to identify on ultrasound due to technological limitations, so the majority of infants with cleft palate will be identified at birth. Infants suspected of multiple malformations may be referred for additional antenatal imaging, such as fetal magnetic resonance imaging or fetal echocardiography.

Ideally, a genetics evaluation should be considered at several points. After a prenatal diagnosis of cleft lip/palate, the family should be referred for a genetics evaluation and a complete diagnostic ultrasound. If appropriate, amniocentesis or other tests may be ordered, though the majority of infants with cleft lip and palate will have no other anomalies, and in this case no genetic testing may be appropriate. Preliminary genetics counseling should stress that diagnosis and risks of recurrence cannot be accurately discussed until after the baby is born and examined. At this time, families should also be referred to a cleft lip/palate team for discussion of management issues and formulation of a feeding plan.

If the diagnosis of a cleft lip/palate is made in the newborn period, a prenatal and family history should be taken, the infant examined for dysmorphic features and genetic counseling offered. Parents also can be informed of the possibility of ultrasonography for future pregnancies. If a formal genetics evaluation has not previously taken place, it should be offered now. Discussion of recurrence risk should occur in infancy, but the precise timing may be guided by the suspicion for an underlying syndrome, health of infant, and parental preference.

The possibility of a genetic condition should also be considered as the child matures, because facial morphology changes with growth. In addition, developmental problems and learning disorders may not surface until later. At adolescence, risks of recurrence should be revisited with both the patient and family.

Because of the rapid change in genetic information and technology, all families with adolescents should be offered the opportunity to have their concerns addressed in a formal genetics consultation. If a dysmorphologist or geneticist is not a member of the cleft lip/palate team, an outside consultation should be discussed, and a referral offered. Additional psychosocial support also may be needed at these times, as parents may have difficulty coping with the provided information.

TABLE 12: GENETIC AND DYSMORPHOLOGY INTERVENTIONS BY AGE

AGE:	INTERVENTIONS:
Prenatal	<ul style="list-style-type: none"> • Genetics consultation after ultrasound diagnosis, or parents have questions about recurrence risks
Birth to 1 Month	<ul style="list-style-type: none"> • Complete medical and family history • Dysmorphology/genetics assessment • Discuss prognosis and implications for treatment • Address etiology • Offer family additional counseling and resources when appropriate
2–15 Months	<ul style="list-style-type: none"> • Discuss recurrence risks, prenatal diagnosis for clefts (ultrasound) • Consider genetics consultation if patient has lip pits, hypotonia or other congenital anomalies
16–24 Months	<ul style="list-style-type: none"> • Consider genetics consultation if patient has lip pits, hypotonia developmental delay or other congenital anomalies
2–5 Years	<ul style="list-style-type: none"> • Consider genetic syndrome if developmental delays are present • Additional genetics workup as indicated
6–11 Years	<ul style="list-style-type: none"> • Consider genetic syndrome, especially if learning problems present • Additional genetics workup as indicated
12–21 Years	<ul style="list-style-type: none"> • Revisit recurrence risk issues and offer formal genetics consultation

Plastic Surgery

Implicit in the choice of a surgeon for the child born with cleft lip/palate is the understanding that the first surgeon to operate has the best opportunity for a good outcome. Once crucial tissues are surgically manipulated or lost, it may be difficult to achieve optimal results. With this information in mind, it is clear that qualifications and expertise are of paramount importance. They should include:

- Board certification or board eligibility in plastic surgery, otolaryngology, or oral and maxillofacial surgery with explicit documentation of training in cleft care.²⁴
- A surgical caseload that ensures regular experience in cleft lip/palate care and a low rate of surgical revisions.
- Affiliation with a cleft lip/palate team.
- Commitment to attend cleft lip/palate team meetings and discuss surgical planning and outcomes.
- Ongoing continuing medical education and expertise in cleft lip/palate care.

TABLE 13: KEY PLASTIC SURGERY INTERVENTIONS BY AGE

AGE:	INTERVENTIONS:
Prenatal	<ul style="list-style-type: none"> • Meet parents and child, outline plan
Birth to 1 Month	<ul style="list-style-type: none"> • Meet parents and child, outline plan • Consider pre-surgical molding in consultation with the appropriate dental specialist (see “a.” in About Surgical Interventions on page 37)
1–3 Months	<ul style="list-style-type: none"> • Monitor progress of pre-surgical molding with orthodontist
3–15 Months	<ul style="list-style-type: none"> • Repair cleft lip, usually at 3–6 months (see “b.” in About Surgical Interventions) • Repair cleft palate, usually at 9–15 mos (see “c.” in About Surgical Interventions)
16 Months–5 Years	<ul style="list-style-type: none"> • Monitor speech-language development with speech-language pathologist (refer for speech-language therapy as needed) • Monitor for symptomatic fistulae • Consider surgical management as needed for VPI (see “d.” in About Surgical Interventions, page 38) • Lip/nasal surgery as needed for residual deformity
6–11 Years	<ul style="list-style-type: none"> • Consider surgical management as needed for VPI (see “d.” in About Surgical Interventions, page 38) • Bone graft to alveolar cleft with closure of oronasal fistulae (see “e.” in About Surgical Interventions) • Lip/nasal surgery as needed for residual deformity
12–21 Years	<ul style="list-style-type: none"> • Rhinoplasty as needed (nasal revision) (see “g.” in About Surgical Interventions) • Lip surgery as needed • Orthognathic surgery (see “b.” in About Surgical Interventions and page 52)

About Surgical Interventions

- a. About pre-surgical orthopedics:** It may be difficult to obtain a good lip repair if the cleft of the lip and alveolus is very wide, or if there is a protruding premaxilla as in bilateral clefts. The lip and alveolar segments can be brought closer together or the premaxilla moved to a more normal position through an intervention called pre-surgical molding. This can involve the application of external taping across the cleft or a plastic orthopedic device taped in place (Nasoalveolar molding device).²⁵ The specifics regarding the timing and nature of the orthopedic device vary from center to center. Potential advantages and disadvantages of molding for a given child should be discussed with the cleft lip/palate team.
- b. About cleft lip repair:** If other medical factors are stable, cleft lip repair is usually done when the child is 3-6 months old. Closure involves meticulous repair of the skin, muscle, and mucosa of the lip. Correction of the cleft lip nasal deformity is usually done at the same time. In wide clefts, some surgeons first do a preliminary lip adhesion procedure (ie. a partial repair) to mold alveolar ridges, and the definitive repair is done several months later.

For the child who has had nasoalveolar molding, there is the additional possibility of primary closure of the alveolar cleft using the technique of gingivoperiosteoplasty (GPP). In a percentage of children undergoing this procedure, later alveolar bone grafting may not be needed.^{26, 27}

- c. About cleft palate repair:** The usual age for cleft palate repair is 9-15 months, which roughly corresponds to the emergence of early infant speech. Closure of the palate (palatoplasty) is complex and involves reorientation and closure of the layers of the soft palate, as well as tissues of the hard palate. The best chance of successful repair is at the initial repair. Failure of part or all of the repair to heal results in a fistula, which permits air or fluid to move between oral and nasal cavity. Inadequate function of the soft palate can result in nasal air leakage and a speech disorder known as velopharyngeal insufficiency (VPI). Pictures of typical clefts are provided on pages 3-5.

Some patients may have a submucous cleft palate, which is more difficult to diagnose. In a submucous cleft of the soft palate, there is continuity of the mucosa, but not of the underlying muscle.

A submucous cleft palate is classically diagnosed by the presence of a bifid (split) uvula, a tented central area in the soft palate, parasagittal bunching of the levator muscle, and a palpable notch at the back of the hard palate. Since most individuals with submucous cleft palate are asymptomatic, this type of palatal cleft is repaired only when there are significant symptoms (feeding problems, speech difficulties, and ear infections).

- d. About treatments for VPI:** Surgical intervention offers the possibility for long-term improvement in speech for the child with velopharyngeal insufficiency (VPI). Surgical options include palatal lengthening (e.g. Furlow technique) to achieve velopharyngeal closure. If this is not sufficient, a sphincter pharyngoplasty or pharyngeal flap may be considered. Disadvantages include: a risk for over-correction of the air leak leading to post-operative obstructive sleep apnea (OSA) and hyponasality. When these occur, additional surgical modifications may be needed. Tailoring the surgical intervention to match the size and characteristics of the velar gap as determined by the VPI workup after 3 years of age can lessen the likelihood of OSA. Speech prostheses (lifts or obturators) provide a non-surgical option for some patients, and may improve speech enough to minimize the need for future surgical intervention. However, they are labor-intensive and require family commitment and child cooperation. (For a more complete discussion of VPI and obturators, see pages 44-46.)
- e. About alveolar bone grafting:** Alveolar bone grafting is usually necessary to close the residual bony cleft in the maxilla and are usually performed between 6-9 years of age. These procedures are performed by an oral and maxillofacial surgeon or a plastic surgeon with special training/expertise in this area (see pages 51-53).
- f. About jaw surgery:** Oral-maxillofacial surgeons and craniofacial plastic surgeons may do orthognathic surgery. Refer to pages 52-53, for discussion of jaw surgery.
- g. About rhinoplasty:** Even though the nasal deformity is often addressed early in care, there can be residual deformities and difficulty breathing through the nasal passages. Definitive rhinoplasty can address both form and function and is typically performed at skeletal maturity.

Otolaryngology and Audiology

Participation of an experienced otolaryngologist is essential for good team care. The otolaryngologist must be familiar with the chronicity of the problems associated with clefts, the unique aspects of cleft care (such as the need to be cautious about adenoidectomy in patients with cleft palate), and the need for coordination with other surgical procedures. As with all cleft lip/palate team specialists, qualifications and experience of the otolaryngologist are important and should include:

- Board certification or board eligibility in otolaryngology
- A surgical caseload that ensures regular experience in cleft lip/palate care
- Affiliation with a cleft lip/palate team
- Commitment to attend cleft lip/palate team meetings, and to discuss surgical planning and outcomes
- Ongoing continuing medical education and expertise in cleft lip/palate care

On some teams, otolaryngologists repair the cleft lip and palate. Otolaryngologists also take part in the assessment and surgical management of velopharyngeal insufficiency in children with cleft lip/palate. They perform the instrumental speech evaluation (nasopharyngoscopy) with the speech-language pathologist to assess the velopharyngeal function, and together they recommend to the team the appropriate surgical or prosthetic intervention. Surgical options include a palate lengthening procedure (Furlow palatoplasty), pharyngeal flap, or sphincter pharyngoplasty to manage the VPI (page 38). Patients that have VPI in addition to abnormal sleep (obstructive sleep apnea) often require additional tests because treatment of VPI has a small risk of causing increased upper airway obstruction, and treatment of sleep apnea may impact speech, especially adenoidectomy. Polysomnography may be indicated to further characterize the severity of the sleep symptoms before or after speech surgery.

Due to the abnormal anatomy of the palate and/or oropharynx, the incidence of recurrent middle ear disease in children with cleft lip/palate is very high (90-95%). While middle ear status is frequently monitored by the primary care physician, it is essential that a qualified otolaryngologist also be involved because of the difficulty in diagnosing middle ear fluid reliably in infants and young children. Many of these children will require one or more sets of ear tubes in order maintain appropriate aerated middle ear spaces.

Infants with Robin sequence (glossoptosis, micrognathia, and often cleft palate) frequently have upper airway compromise. A comprehensive evaluation including awake fiberoptic upper airway endoscopy is recommended to guide treatment. Medical and respiratory supportive measures (i.e. prone positioning, oxygen supplementation) are the first line treatment to maintain adequate upper airway, ventilation, and weight gain. If those measures are insufficient to alleviate the respiratory distress, additional evaluation may

include operative airway evaluation including rigid bronchoscopy, CT scan to assess degree of micrognathia, and polysomnography. Depending on the results of those tests and based on the location of upper airway obstruction, the following treatments are discussed and offered to the family:

1. Placement of a nasopharyngeal airway tube (modified endotracheal tube) to stent open the airway.
2. Upper airway procedure that is directed to the individual level of obstruction, for example, supraglottoplasty (removal of obstructive tissue above larynx).
3. Mandibular distraction.
4. Tracheotomy.
5. Other procedures to relieve tongue base obstruction, but are less commonly performed include tongue lip adhesion and tongue suspension suture placement.

Audiology

The chronic middle ear effusions and infections experienced by the child with a cleft palate are often associated with hearing loss. For this reason, it is essential that hearing be monitored regularly by a qualified audiologist. Hearing loss secondary to middle ear disease is called a conductive hearing loss; sensorineural hearing loss occurs in a very small number of children with cleft lip/palate. Conductive hearing loss secondary to middle ear disease can vary in terms of degree and configuration, but most commonly it is a mild hearing loss. A persistent mild hearing loss can adversely influence speech and language, with potential consequences for cognitive development and psychological adjustment. Because of the unpredictable course of middle ear disease in young children, the early and routine audiologic monitoring of children with cleft palate is mandatory, and should include the use of tympanometry. A Brainstem Auditory Evoked Response (BAER) test (during nap or with sedation) may be required to fully evaluate the hearing function. The American Academy of Pediatrics recommends screening all newborns for hearing loss (with otoacoustic emissions/BAERs), and ongoing hearing assessments (as described above) in high-risk children including those with a cleft.^{28,29}

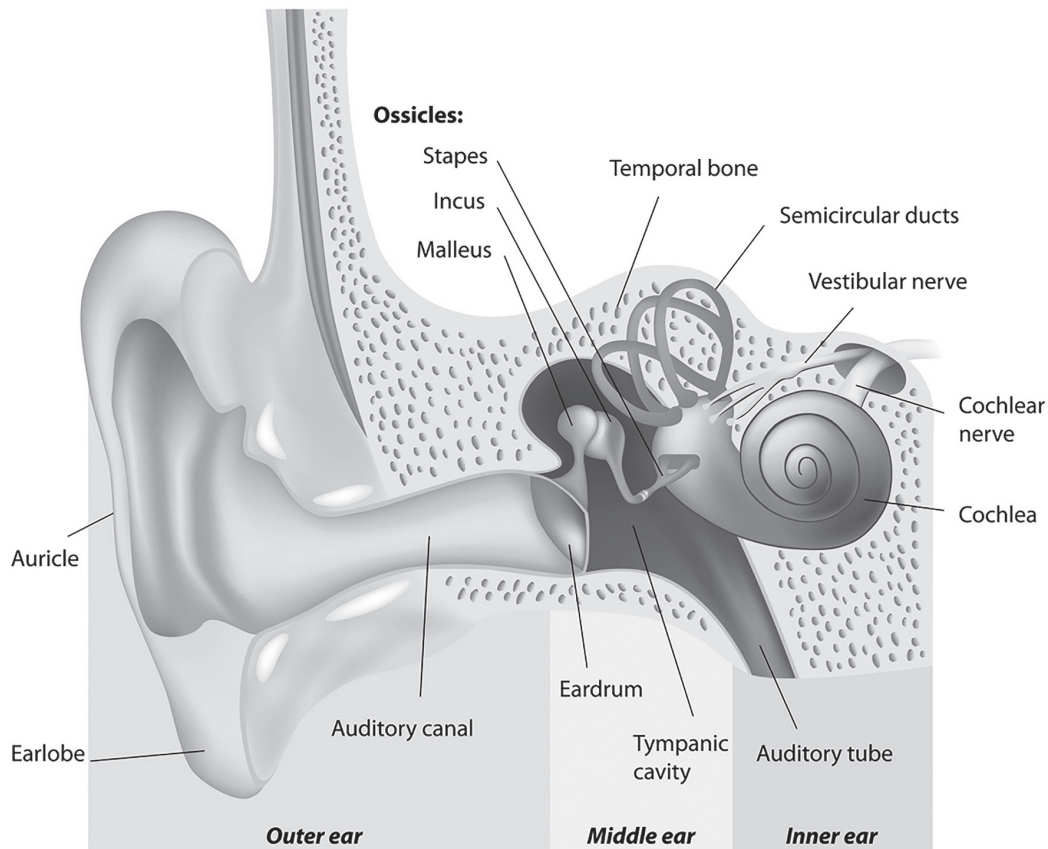
TABLE 14: KEY OTOLARYNGOLOGY AND AUDIOLOGY INTERVENTIONS BY AGE

AGE:	INTERVENTIONS:
Birth to 1 Month	<ul style="list-style-type: none"> • If Robin sequence or other syndrome, assess for airway problems and use positioning, nasopharyngeal tube, tracheotomy, or mandibular distraction as needed • Assess middle ear status (fluid or infection) • Assess hearing (BAER/otoacoustic emissions)
1–5 Months	<ul style="list-style-type: none"> • Monitor airway status, intervene as needed • Assess middle ear status • Assess hearing if not done already
5–15 Months	<ul style="list-style-type: none"> • Monitor airway after palate closure in Robin sequence/other syndromes • Place ear tubes with palate repair if middle ear fluid present > 3 months • Medical management for ear fluid or infections if tubes already present • Assess hearing with behavioral and impedance audiometry at 7-8 months of age, and monitor at 6-month intervals • Consider amplification when indicated
16–24 Months	<ul style="list-style-type: none"> • Assess airway status • Monitor middle ear status at least every 6 months; place/replace ear tubes as needed • Assess hearing every 6 months
2–5 Years	<ul style="list-style-type: none"> • Assess airway status (consider polysomnogram if clinical symptoms of obstructive sleep apnea reported in a child with small jaw, flat mid-face, large tonsils/adenoid tissue, sphincter pharyngoplasty or pharyngeal flap present) • Deviated septum may require repair • Use caution with adenoidectomy due to risk of post-operative VPI • Monitor middle ear status every 6-12 months • If ear tubes have been in place for 3 years or more, consider removal • Monitor hearing every 6 months to age 3 years, then every 6-12 months as needed • Nasal endoscopy with speech therapist to evaluate VPI (see page 38) • Consider surgical interventions for VPI (palate lengthening surgery, sphincter pharyngoplasty, or pharyngeal flap) usually by a speech surgeon (plastic surgeon or otolaryngologist)
6–21 Years	<ul style="list-style-type: none"> • Assess airway status (consider polysomnogram if small jaw, flat mid-face, large tonsils/adenoids, sphincter pharyngoplasty or pharyngeal flap present) • Deviated septum may require repair • Use caution with adenoidectomy due to risk of post-operative VPI • Monitor middle ear status every 6-12 months. Chronic tympanic membrane retraction can lead to cholesteatoma, which requires a surgical treatment; chronic problems may necessitate repeated ear tubes, tympanoplasty, mastoidectomy • Ear tube removal when appropriate • Nasal endoscopy with speech therapist to evaluate VPI • Consider surgical interventions for VPI (palate lengthening surgery, sphincter pharyngoplasty, or pharyngeal flap) usually by a speech surgeon (plastic surgeon or otolaryngologist) • Monitor hearing every 6-12 months until normal for 2 consecutive years

Anatomy of the Middle Ear

In children with cleft palate, the eustachian tube (auditory tube) often does not function properly, in part, because muscles responsible for opening the eustachian tube are involved in the cleft palate. If the eustachian tube does not open effectively, then negative pressure can develop in the middle ear space causing an accumulation of fluid. With fluid, the eardrum and the ossicles (middle ear bones) are unable to vibrate properly, hindering the transfer of sound energy from the air through these structures to the cochlea of the inner ear. If the eustachian tube remains blocked, or if the fluid persists long enough, the eardrum or the ossicles may be damaged. The presence of fluid is a risk factor for ear infections. The function of myringotomy tubes is to keep the middle ear space aerated, which normalizes the hearing. Ear tubes that are in place and functional should maintain an aerated space. If an infection develops with ear tubes in place, it can be treated using ototopical drops and rarely requires the use of systemic antibiotics.

FIGURE 2: ANATOMY OF THE EAR



Anatomy of the Roof of the Mouth

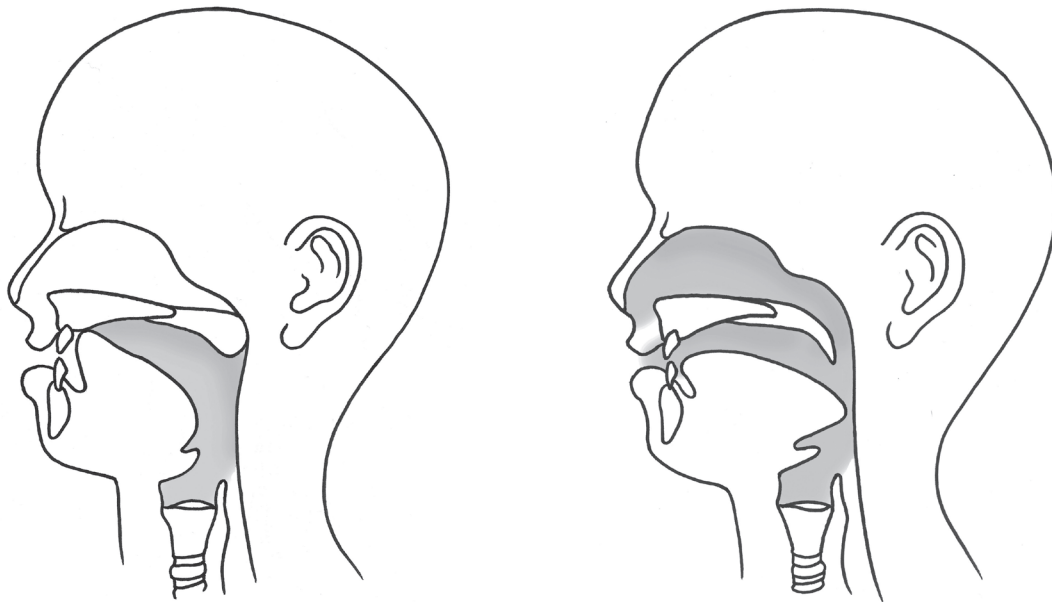
Hard Palate

The hard palate is the bony roof of the mouth.

Soft Palate

The soft palate is the muscular extension of the hard palate located at the back of the mouth. Movement of the muscles of the soft palate is essential for normal speech and eustachian tube function. In infancy, closure of the velopharyngeal space by palatal musculature is necessary for generating suction during feeding. Without adequate closure of this space, air and food escape through the nose.

FIGURE 3: ANATOMY OF THE PALATE



The soft palate is closed against the back of the throat during the swallow. It also allows pressure to build up in the mouth for speech sounds such as “p” and “b”.

The soft palate is open for breathing and for making speech sounds such as “m” and “n”.

Cleft Related Speech Considerations

The soft palate is critical for normal speech development. The palate elevates and touches the back of the throat to separate the nose from the mouth and allow pressure to build up in the mouth for certain speech sounds (i.e. 'p', 'b', 'sh')(see Figure 3, page 43). Children with an unrepaired cleft palate are not able to separate the nose from the mouth. Even after surgical closure of the palate, some children remain unable to create adequate intraoral pressure for normal speech. Any problem with a child's ability to separate the nose from the mouth during speech is called velopharyngeal dysfunction (VPD). Some types of VPD can be treated with speech therapy alone, and other types of VPD require a surgery or prosthesis. The three types of VPD are velopharyngeal insufficiency (VPI), velopharyngeal mislearning (VPM), and velopharyngeal incompetence. A problem with the structure of the palate or throat results in what is called VPI. VPM refers to speech habits that prevent the child from building up pressure in the mouth. Velopharyngeal incompetence refers to weakness or discoordination of the palate. Regular monitoring by a team speech-language pathologist will ensure that your child receives timely evaluations and appropriate intervention.

Speech and language assessments should take place regularly as outlined below by a certified speech-language pathologist with expertise in cleft lip/palate care. These regular assessments determine the type of VPD your child may have and what the best course of management may be. Regular assessments also allow the team speech-language pathologist to monitor progress in speech therapy and collaborate with community-based speech-language pathologists.

Often, more information is needed to determine the best plan, so an instrumental VPD evaluation will be recommended. This can take place when the child is old enough to cooperate (usually 3-5 years old). This evaluation will include a speech evaluation, nasopharyngoscopy and, if needed, a videofluoroscopic speech study. It is generally conducted by a team made up of a speech-language pathologist and an otolaryngologist. When the results of the VPD evaluation are available, recommendations for surgery, speech prosthesis or speech therapy can be made by the team.

TABLE 15: KEY SPEECH-LANGUAGE INTERVENTIONS BY AGE

AGE:	INTERVENTIONS:
6–9 Months	<ul style="list-style-type: none"> • Speech-language consultation to provide counseling about potential speech issues
9–24 Months	<ul style="list-style-type: none"> • Speech-language evaluation (three-six months after palate repair) • Refer for early intervention speech-language therapy as needed.
2–5 Years	<ul style="list-style-type: none"> • Annual speech-language evaluation • VPD evaluation and interventions as needed • Proceed with surgical or prosthetic management, if indicated • Refer for speech- language therapy, as needed • Communicate with community based speech-language pathologist to monitor progress of therapy
6–11 Years	<ul style="list-style-type: none"> • Annual speech-language evaluation until involution of adenoids • VPD evaluation and interventions, if needed • Refer for speech- language therapy as needed • Communicate with community based speech-language pathologist to monitor progress of therapy
12–21 Years	<ul style="list-style-type: none"> • Speech-language evaluation every 2-3 years or as needed • Speech-language therapy as needed • Communicate with community based speech-language pathologist to monitor progress of therapy • VPD evaluation and intervention as needed • Speech-language evaluation before and after orthognathic surgery

About Speech Prostheses

A speech prosthesis is a removable appliance that is attached to the teeth with wire clasps and is fabricated by a dental specialist in conjunction with the speech-language pathologist. There are two types of speech prostheses: lifts and obturators. A palatal lift appliance lifts the palate in order to close the velopharyngeal gap. An obturator closes the velopharyngeal gap with an acrylic bulb that matches the gap's size and shape. The design of the prosthesis is determined by the instrumental VPD evaluation, and its fit is modified and fine-tuned as needed. Due to the significant improvements in surgical approaches for the treatment of VPI, many teams no longer use obturation as a treatment modality.

Advantages:

- In the young child, it allows the development of correct speech sounds, improves intelligibility and may diminish behavior problems due to communication difficulties
- The speech bulb is infinitely adjustable in size and shape, and requires minimal movement of the velopharyngeal structures to work well
- It is removed at night so it does not cause obstructive sleep apnea
- Prosthetic treatment provides a reversible trial for controlling VPI before committing the child to a surgical procedure
- Prosthetic treatment makes control of VPI available to patients for whom surgery is not advised

Disadvantages:

- Requires parent commitment, patient compliance and a professional skilled at working with children
- Initial desensitization needed to reduce gag reflex
- Requires modification as the child grows
- Requires many office visits for development and adjustment
- Long-term presence of any removable dental prosthesis makes oral hygiene difficult and increases risk of cavities and periodontal problems
- Can be lost or broken

About Speech Surgery

Refer to page 38 for a discussion of the surgical management of VPI. Surgery provides a permanent treatment for VPI by reconfiguring the tissues of the velopharyngeal space so that the VP gap can be reliably closed for certain speech sounds while remaining open for other speech sounds and breathing. The advantage of surgery is the potential permanent resolution of VPI symptoms. The disadvantage is the potential for over-correction, creating obstructive sleep apnea and/or hyponasal speech (i.e. not enough air flow through the nose during speech).

About the Decision to Treat VPI

The decision to treat a child's VPI should be made through consultation among the child, parents, and members of the cleft lip/palate team. Recommendations for surgery, speech therapy, a speech appliance, or some combination of these should be based on the needs of the individual child.

Orthodontics and Dental Medicine

Orthodontics and dental care are integral parts of the habilitation of the child with a cleft lip/palate. The interventions of the orthodontist are particularly critical, and it is important that they have experience in cleft lip/palate care. Qualifications of the orthodontist include:

- Board certification or board eligibility in orthodontics
- An orthodontic caseload that ensures regular experience in cleft lip/palate care
- Affiliation with a cleft lip/palate team
- Commitment to attend cleft lip/palate team meetings and to discuss treatment plans and outcomes
- Ongoing continuing education in cleft lip/palate care

About orthodontics and dental care

Children with cleft lip/palate have both the usual childhood dental needs and special problems arising from the clefts. Good dental care is essential. These children have an increased need for preventive and restorative dental care due to underlying dental anomalies and the use of braces and other orthodontic appliances. Dental hygiene must be closely monitored by either a pediatric dentist or general dentist. Unhealthy teeth and gums compromise later orthodontic and surgical interventions, and may contribute to low self-esteem.

Early involvement of the orthodontist is necessary in the newborn period if pre-surgical orthopedics (external taping, internal appliance, or molding device) are used. Later, the orthodontist monitors the development and eruption of teeth. There may be missing, rotated, incorrectly shaped, extra or displaced teeth. Occasionally, extractions are needed.

The orthodontist also monitors facial growth and obtains important dental records (X-rays, models and photographs) needed for planning and timing interventions. Much of what the orthodontist does is orthopedic – positioning bony segments to provide the underlying framework for the soft tissue. This is especially true before important surgical procedures such as the alveolar bone graft or jaw advancement (see pages 52-53). Cleft repair without orthodontic intervention at the proper stages can produce unstable, inferior results, with subsequent tooth loss and inadequate chewing ability/jaw function.

Proper orthodontic care enhances soft tissue repair, speech production, oral function and self-image. Orthodontic treatment should be performed in discreet phases with specific, limited objectives. Continuous active treatment from early years through permanent dentition should be avoided. Neither the teeth nor the child can tolerate such lengthy treatment. Sometimes orthodontic interventions are carried out by community orthodontists in consultation with the cleft lip/palate team orthodontist. This facilitates community-based care while ensuring coordination with surgical, speech and other treatments planned by the team.

TABLE 16: KEY ORTHODONTIC/DENTAL INTERVENTIONS FOR THE CHILD WITH CLEFT LIP/PALATE

AGE:	INTERVENTIONS:
Birth – 5 Months	<ul style="list-style-type: none"> • Pre-surgical orthopedics (See About Pre-surgical Orthopedics on page 50)
5–24 Months	<ul style="list-style-type: none"> • Parent teaching regarding oral hygiene, dental development and future treatment plans • Monitor eruption of teeth and dental hygiene – pediatric dentistry
2–6 Years	<ul style="list-style-type: none"> • Orthodontic dental records (X-rays, photos) at 5-6 years of age in preparation for evaluation of jaw relationships, teeth and cleft size • Monitor dental hygiene; provide appropriate preventive and restorative care through ongoing follow-up with pediatric dentist • Orthodontic records as needed to determine timing of bone graft • Assist with speech prosthesis b, as needed (See About Speech Prostheses on page 50)
6–11 Years	<ul style="list-style-type: none"> • Positioning of maxillary segments in preparation for alveolar bone graft • Recommend extractions as needed • Following bone graft, orthodontist may align teeth in upper arch • If indicated, orthodontist may work with oral surgeon to help guide teeth which are not developing in correct position to the right place • Monitor growth; maxillary protraction as needed • Monitor dental hygiene; provide appropriate preventive and restorative care through ongoing follow-up with pediatric dentist • Assist with speech prosthesis as needed
12–21 Years	<ul style="list-style-type: none"> • Dental records to monitor jaw growth, dental development and bone graft (if not yet performed) • Braces for dental alignment as needed • If retrusion is severe, combination of jaw surgery and braces is needed – if underbite is severe, distraction osteogenesis may be considered once all permanent teeth are erupted. Otherwise, jaw surgery is indicated once jaw growth is complete (~16-19 years) • Prosthetic replacement of missing teeth as needed (See About Dental Prostheses on page 50) • Monitor dental hygiene; provide appropriate preventive and restorative care through ongoing follow-up with pediatric dentist • Prepare surgical splints for orthognathic surgery as needed

About Pre-surgical Orthopedics

If the cleft lip and alveolus is wide, or there is a protruding or prominent premaxilla (as in bilateral clefts), the segments can be brought closer together and the premaxilla moved to a more favorable position by use of external taping across the cleft or nasoalveolar molding device may be indicated. Due to the success of non-surgical nasoalveolar molding, most teams no longer use surgically placed intraoral devices. To be effective, these interventions, called presurgical orthopedics, must occur in the first weeks of life. Potential advantages and disadvantages for a given child should be discussed with the cleft lip/palate team at the time treatment is recommended.

About Speech Prostheses

A speech prosthesis is a removable appliance used to treat velopharyngeal insufficiency (VPI). Usually referred to as an obturator, it is fabricated by a dental specialist in consultation with the speech-language pathologist and is attached to the teeth with wire clasps. Refer to pages 46-47 for a discussion of VPI and obturators.

About Dental Prostheses

The disruption of anatomy caused by a cleft lip may result in congenitally missing or severely malformed (and non-usable) teeth. Dental bridges may be used to support and retain prosthetic teeth. Surgically placed dental implants are the treatment of choice for many patients.

Oral and Maxillofacial Surgery

An oral and maxillofacial surgeon on the cleft lip/palate team is responsible for several major surgical procedures important to the successful habilitation of cleft patients. The two most common of these are outlined in detail below. As with all cleft lip/palate care, the qualifications and expertise of the specialty providers is very important. The following criteria are offered as indicators of such expertise in oral and maxillofacial surgery:

- Board certification or board eligibility in oral and maxillofacial surgery or plastic surgery
- A significant surgical caseload of these procedures
- Participation in or affiliation with a cleft lip/palate team
- Commitment to attend cleft lip/palate team meetings and to discuss surgical planning and outcomes
- Ongoing continuing education and training in cleft lip/palate care
- For many teams, other surgical subspecialists may perform alveolar bone grafting and orthognathic (jaw) surgery. In these cases, the particular education, training and experience of the surgeon qualifying them to perform the surgery should be established

TABLE 17: ORAL AND MAXILLOFACIAL SURGERY INTERVENTIONS BY AGE

AGE:	INTERVENTIONS:
6–11 Years	<ul style="list-style-type: none"> • Bone graft of alveolar cleft and closure of oronasal fistulae (See About Alveolar Bone Grafting on page 52) • Selective tooth extractions as needed
12–21 Years	<ul style="list-style-type: none"> • If needed, orthognathic (jaw) surgery in consultation with the orthodontist (see About Orthognathic (Jaw) Surgery on page 52)

About Alveolar Bone Grafting

After the repair of the cleft lip, there typically remains a bony cleft in the maxilla and often an opening running from the nose to the mouth (under the upper lip) called an oronasal fistula.

When teeth erupt into the cleft, they are unsupported by bone and will likely be lost. Patients with oronasal fistulae may experience nasal regurgitation of food and drink into their nose when eating. Bone grafting of the cleft(s) and repairing oronasal communications is essential. Doing so joins the cleft segments of the maxilla, provides a bony base for erupting adult dentition, repairs the opening between the nose and mouth, and constructs the floor of the nose, providing support for the nasal alar base. For this procedure, cancellous bone is best, and is usually taken from the hip, though bone from the skull, lower jaw, or tibia may also be used. This procedure is usually performed by an oral/maxillofacial surgeon or plastic surgeon with special training in this area. Timing for this procedure is critical and requires close cooperation between the orthodontist and surgeon. In cases when a child has had nasoalveolar molding in infancy and a gingivoperiosteoplasty was done at the lip repair, an alveolar bone graft may not be needed.

These surgical procedures cannot take place unless the teeth and gums are healthy and the maxillary alveolar ridges have been properly positioned through orthodontic intervention. Proper dental and orthodontic care are essential to the successful habilitation of the child with cleft lip and palate.

About Orthognathic (Jaw) Surgery

The upper jaw (maxilla) is usually fully developed by age 15 years. In the child with a cleft, the maxilla may have intrinsic growth deficiency or may be impacted by scars from palatoplasty.

A size discrepancy between the upper and lower jaws results in a concave facial profile.

If the discrepancy between the jaws is slight, it can be managed by orthodontics alone. If maxillo-mandibular discrepancy is more severe, then jaw (orthognathic) surgery in conjunction with orthodontics is required for correction of the maxillo-mandibular relationship.

Orthognathic surgery is complex and requires the combined efforts of the orthodontist and surgeon. Pre-operative orthodontic treatment is necessary to position the teeth in the upper and lower jaws so they will match well when the jaws are repositioned. Surgical planning involves the use of photos, plaster dental models, cephalometric X-rays, and frequently 3D CT models of the patient's facial bones and teeth.

A long established surgical procedure, called a maxillary advancement (Le Fort I osteotomy), is sometimes done with a bone graft to increase the size of the upper jaw. An alternative technique being used by some surgeons involves similar cuts of the bones of the jaw, but instead of a bone graft and single stage advancement, new bone growth is stimulated and directed by a process called distraction osteogenesis. In this technique, pins are placed on each side of the cuts in the bone. These pins are then attached to an external frame called a distraction device or halo. Screws on the device are turned daily and gradually advance the healing bones until the desired lengthening has been achieved.

Sometimes, surgery on both the upper and lower jaws is required to correct the maxillomandibular relationship.

Potential advantages and disadvantages of these procedures for a given child should be discussed by the team at the time the surgery is being planned. This surgery can be performed by an oral/maxillofacial or craniofacial plastic surgeon with training in this area. Following surgical management of the jaws, the final phase of the orthodontic treatment is begun. During this phase, which usually lasts about one year, the occlusion between upper and lower teeth is optimized. Pre- and post-operative speech evaluation is required, because velopharyngeal insufficiency (VPI, see page 44) can result from the advancement of the upper jaw.

GLOSSARY

Alveolar ridge: the bony arches of the maxilla (upper jaw) and mandible (lower jaw) that contain teeth.

Alveolus/alveolar process: the bony area that supports the teeth.

Appliance, dental: a device worn in the mouth to provide a dental benefit.

Audiology: the study of hearing and hearing disorders.

Bifid uvula: uvula muscle divided into two parts.

Bilateral: having two sides, or pertaining to both sides.

Bone graft: a transplant or movement of bone from one site to another.

Brainstem Auditory Evoked Response (BAER): an electrophysiologic measurement of activity in auditory nerve and brainstem pathways.

Cheiloplasty: surgical repair of cleft lip.

Cleft: split or divided; refers to muscle, skin, bone.

Cleft lip: congenital deformity of the upper lip that varies from a notching to a complete division of the lip; any degree of clefting can exist (also known as a primary palate cleft).

Cleft palate: a congenital split of the palate that may extend through the uvula, soft palate, and into the hard palate; the lip may or may not be involved in the cleft of the palate (also known as a secondary palate cleft).

Cleft Palate-Craniofacial Team: group of professionals involved in the care and treatment of patients having cleft lip/palate and other craniofacial malformations; consists of representatives from some of the following specialties: audiology, genetics, nursing, oral surgery, orthodontics, otolaryngology, pathology, pediatrics, pedodontics, plastic surgery, prosthodontics, psychiatry, psychology, radiology, social work, and speech-language.

Craniofacial: pertaining to the cranium (the part of the skull that encloses the brain) and the face.

Dental arch: curved structure of the upper and lower jaws formed by the teeth in their normal position on the alveolar ridge.

Dentofacial orthopedics: orthodontics

Distraction osteogenesis: a technique that uses a device to slowly lengthen a bone(s) without requiring a bone graft.

Eardrum: tympanic membrane; it vibrates and transmits sounds from the air to the middle ear.

Effusion: accumulation of fluid in the middle ear.

Eustachian tube: the duct that connects the nasopharynx (located in the back of the throat and above the hard palate) with the middle ear; it is usually closed at one end, but opens with yawning and swallowing; it allows ventilation of the middle ear cavity and equalization of pressure on both sides of the eardrum.

Evoked otoacoustic emission (oAe): a screening test that specifically measures the cochlear response to presentation of a stimulus.

Fistula: abnormal opening from the mouth to the nasal cavity remaining after surgical closure of the original cleft.

Hard palate: the bony portion of the roof of the mouth.

Heredity: characteristics and traits genetically derived from one's ancestors.

Hypernasality: excessive nasal resonance during speech due to an excess of air flow into the nasal chamber.

Hyponasality: lack of nasal resonance during speech due to an inadequate amount of air flow. (As heard when a person has nasal congestion or decreased nasal airway space, e.g., “man” would sound like “bad” and “maybe” like “baby.”)

Impedance audiometry: physiologic test used to measure air pressure in the middle ear cavity and the ability of the eardrum to function normally (tympanogram).

Incidence: frequency of occurrence.

Incisor: a tooth that is located in the front of the mouth between the cuspids/canines (eye teeth).

Inner ear: the internal portion of the ear that contains the sensory end organs used for hearing and balance.

Language disorder or impairment: difficulty with language comprehension or expression; an interference with the ability to communicate effectively.

Mandible: U-shaped bone forming the lower jaw.

Maxilla: the bones forming the upper jaw.

Maxillary orthopedics: the movement of palatal segments by the use of appliances (also called dentofacial orthopedics).

Micrognathia: a condition characterized by abnormal smallness of the jaw.

Middle ear: portion of the ear containing the three small bones of the ossicular chain that transfers sound from the eardrum to the inner ear; it is attached to the tympanic membrane on one end and the oval window at the other end.

Nasal emission or nasal escape: the flow of air through the nose during speech, usually indicative of an incomplete seal between the cavities of the mouth and the nose.

Obturator: a plastic (acrylic) appliance, usually removable, used to cover a cleft or a fistula in the hard palate, or to help achieve velopharyngeal closure to promote clear speech.

Occlusion: relationship between the upper and lower teeth when they are in contact; it refers to the alignment of teeth as well as the relationship of the dental arches.

Orthodontics: the specialty of dentistry concerned with the correction and prevention of irregularities and malocclusion of the teeth and jaws.

Orthognathic: dealing with the cause and treatment of malposition of the jaw bones.

Orthopedics: the movement of bone by means of appliances rather than surgery.

Otitis media: infection of the middle ear, where thick mucous fluid accumulates; this is a common problem for children with cleft palate.

Otolaryngologist: physician specializing in the diagnosis and treatment of diseases of the ear and larynx; commonly referred to as an ear, nose and throat (ENT) specialist.

Palate: the roof of the mouth, including the front portion or hard palate, and the rear portion or soft palate (velum).

Pediatrician: physician specializing in the health, disease and treatment of children and adolescents.

Pharyngeal flap: a flap of mucosa and muscle taken from the back of the throat and attached to the soft palate. It is designed to create velopharyngeal closure during speech in patients with velopharyngeal insufficiency.

Pharynx: the opening at the back of the throat.

Premaxilla: the front middle portion of the upper jaw containing the front teeth (the incisors).

Rhinoplasty: surgical repair of a deformed nose.

Sphincter pharyngoplasty: a procedure in which the surgeon moves tissue from the back of the throat closer to the back of the palate. It is designed to treat velopharyngeal insufficiency.

Submucous cleft palate: a cleft of the muscle layer of the soft palate with an intact layer of mucosa lying over the defect.

Teratogen: something in the environment of the embryo causing defects in structural or functional development.

Tympanic membrane: eardrum.

Unilateral: one-sided.

Uvula: muscle extension on the soft palate that can be seen as a fleshy lobe in the midline of the posterior palate.

Velopharyngeal: pertaining to the soft palate and pharynx.

Velopharyngeal insufficiency (VPI): inadequate velopharyngeal closure resulting in hypernasality (excessive flow of air through the nose); also called velopharyngeal incompetence.

Velum: the soft palate.

(Glossary adapted with permission from: Berkowitz, Samuel. The Cleft Palate Story. Quintessence Publishing Co., Inc. Chicago: 1994)

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509-574-3268

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Fax: 509-574-3210

RESOURCES

American Association of Oral and Maxillofacial Surgeons. (2017). *Standards of Care for Cleft Lip and Palate*. https://www.aaoms.org/docs/member-center/parcare/parcare_5_skeletal_deformities.pdf

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Organizations and Support Groups for Cleft Lip/Palate

Aboutface

123 Edward Street, Suite 1003

Toronto ON Canada M5G 1E2

Phone: 416-597-2229

Toll Free: 800-665-FACE

Fax: 416-597-8494

Email: info@aboutfaceinternational.org

A nonprofit support network for people with facial differences. Their chapters and resources include newsletters, videos, and publications.

The American Cleft Palate-Craniofacial Association (ACPA)

acpa-cpf.org

1504 East Franklin Street, Suite 102

Chapel Hill, NC 27514-2820 USA

Phone: 919-933-9044

Fax: 919-933-9604

Email: info@acpa-cpf.org

The national organization for specialists involved in the treatment of cleft and craniofacial conditions. ACPA is the treatment authority. ACPA is moving from the role of being primarily an educational organization for professionals to one that interacts with government agencies, legislatures, insurance carriers and organizations representing patients with clefts and other craniofacial anomalies. This group has developed and published “Parameters for Evaluation and Treatment of Patients with Cleft Lip/palate or Other Craniofacial Anomalies.”

Children’s Craniofacial Association

ccakids.com

A national, non-profit organization dedicated to improving the quality of life for people with facial differences and their families. The CCA addresses the medical, financial, psychosocial, emotional, and educational concerns relating to craniofacial conditions.

Cleft Advocate

cleftadvocate.com

A nonprofit organization that provides educational and support resources for families and children with a cleft. The website includes insurance and advocacy information and online family networking.

The Cleft Palate foundation

cleftline.org

Referral/Information: 800-24CLEFT (800-242-5338)

A non-membership organization affiliated with the American Cleft Palate-Craniofacial Association. It provides referrals to the local teams and to parent support groups, and publishes brochures, fact sheets and newsletters.

FACES

www.faces-cranio.org

Phone: 423-266-1632

Toll Free: 800-332-2373

Email: faces@faces-cranio.org

A non-profit organization that provides financial assistance for nonmedical expenses such as travel, lodging and food incurred when traveling to a craniofacial center for reconstructive surgery. Support is offered on the basis of financial and medical need. A quarterly newsletter, information about craniofacial disorders, support networks and applications for financial assistance are available.

Seattle Children's Craniofacial Center

www.seattlechildrens.org/clinics-programs/craniofacial/conditions-treated

Friendly Faces

friendlyfaces.org

Started by the mother of a child with Treacher Collins to provide information and networking to families with any craniofacial condition.

Foundation for Faces of Children

facesofchildren.org

A nonprofit foundation that provides clear, accurate information and other educational resources to individuals born with craniofacial conditions and to their families. Resources include a video for families of children with clefts.

Parent-to-Parent

arcwa.org/parent_to_parent.htm

Toll Free: 800-821-5297

An organization serving Washington State that provides emotional support and information about disabilities and community resources to parents.

Starting Point and Resource Directory for Children with Special Needs

<http://cshcn.org/sites/default/files/webfm/file/StartingPointGuide.pdf>

Phone: 866-987-2500, press 4 (Children's Resource Line)

Starting Point is a guide that provides a overview of resources for families of children with special health care needs in Washington State. An online resource directory is also available at <http://cshcn.org/resources-contacts/resources-directory>. The resource directory lets families search options by topic and includes resources in Washington, Alaska, Montana and Idaho.

Wide-Smiles

widesmiles2.org

Phone: 209-942-2812

A private, non-profit organization that provides information and networking among families of children with clefts. It publishes a magazine to educate and encourage families and to share research information. It also sponsors an on-line Internet discussion list called CLEFT-TALK that connects families all over the world.

About This Document

These guidelines were developed through a consensus process. The design team was multidisciplinary with state-wide representation involving primary and tertiary care providers, family members and a representative from a health plan.

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