
Cleft Lip and Palate

CRITICAL ELEMENTS OF CARE

Produced by

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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 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 Center for Children with Special Needs website at: www.cshcn.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.*

TABLE OF CONTENTS

Cleft Lip and Palate CRITICAL ELEMENTS OF CARE

INTRODUCTION	4
I. KEY INTERVENTIONS FOR CLEFT LIP/PALATE	
Overview: Summary of Key Interventions by Age	6
Prenatal.....	7
Birth through 1 Month.....	8
1 through 4 Months	9
5 through 15 Months	10
16 through 24 Months	11
2 through 5 Years.....	12
6 through 11 Years	13
12 through 21 Years	14
Overview: Summary of Key Interventions by Specialty	15
II. APPENDICES	
I. Standards of Care for Cleft Lip and Palate.....	17
II. Prenatal Diagnosis	20
III. Nursing, Coordination of Care and Feeding Issues	22
IV. Primary Care	24
V. Psychosocial and Developmental Issues in Cleft Care.....	27
VI. Genetics/Dysmorphology.....	30
VII. Plastic Surgery	33
VIII. Otolaryngology/Audiology.....	36
IX. Speech Problems in Children with Clefts	39
X. Orthodontics/Dental Medicine.....	42
XI. Oral and Maxillofacial Surgery	44
XII. Ethics and Children with Cleft Lip/Palate	46
XIII. Types of Cleft Lip/Palate	48
XIV. Glossary	50
XV. Washington State Cleft Lip/Palate Teams	52
XVI. References and Resources.....	53

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 (2004) and Team Standards (1996) documents^{1, 2, 3, 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,⁵ 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, 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 newborn period with referral to a Cleft Lip/Palate Team. When cleft lip/palate is diagnosed prenatally referral to a team should be offered.

- 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.
- 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.
- Better 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. Overviews on pages 6 and 15 highlight key interventions by age group and discipline, respectively. These are explained more fully in the body of the document and in the appendices 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.

Acknowledgments

We gratefully acknowledge the Mead Johnson Company for permission to use several illustrations and wording from their book, *Looking Forward*; Samuel Berkowitz and the Quintessence Publishing Company for permission to adapt the glossary from *The Cleft Palate Story*; and Daryl Tong for the medical illustrations.

- 1 *Surgeon General's Report: Children with Special Healthcare Needs*. Office of Maternal and Child Health. U.S. Department of Health and Human Services, June 1987.
- 2 "Parameters for the Evaluation and Treatment of Patients with Cleft Lip/Palate or Other Craniofacial Anomalies." American Cleft Palate-Craniofacial Association, 2004
- 3 *New Standards for Cleft Palate and Craniofacial Teams*. American Cleft Palate-Craniofacial Association, March 31, 2010 www.acpa-cpf.org/Standards/Standards_2010.pdf
- 4 Shaw, William C., et al. "A Six-Center International Study of Treatment Outcome in Patients with Clefts of the Lip and Palate: Parts 1-5." *Cleft Palate-Craniofacial Journal* 1992; 29(2): 393-418.

I. KEY INTERVENTIONS FOR CLEFT/LIP PALATE

Overview: 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	INTERVENTION	REFER TO PAGE
Prenatal	<ul style="list-style-type: none">• Refer to cleft lip/palate team• Medical diagnosis and genetic counseling• Address psychosocial issues• Provide feeding instructions• Make feeding plan	7
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 presurgical orthopedics if indicated	8
1-4 months	<ul style="list-style-type: none">• Monitor feeding and growth• Repair cleft lip• Monitor ears and hearing• Begin/continue presurgical orthopedics if indicated	9
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	10
16-24 months	<ul style="list-style-type: none">• Assess ears and hearing• Assess speech-language• Monitor development	11
2-5 years	<ul style="list-style-type: none">• Assess speech-language; manage VPI*• Monitor ears and hearing• Consider lip/nose revision before school• Assess development and psychosocial adjustment	12
6-11 years	<ul style="list-style-type: none">• Assess speech-language; manage VPI• Orthodontic interventions• Alveolar bone graft• Assess school/psychosocial adjustment	13
12-21 years	<ul style="list-style-type: none">• Jaw surgery, rhinoplasty (as needed)• Orthodontics; bridges, implants as needed• Genetic counseling• Assess school/psychosocial adjustment	14

*VPI = velopharyngeal insufficiency. See Appendix IX, pg. 39.

PRENATAL

PROBLEM	INTERVENTION
Family need for information on care of 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 crisis in family	<ul style="list-style-type: none"> • Team nurse/psychosocial 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 Appendix III, pg. 22)
Team/family need for accurate medical/diagnostic information on cleft (and any other anticipated medical problems) ⁵	<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:

- Medical diagnosis and genetic counseling
- Refer to cleft lip/palate team
- Make feeding plan
- Address psychosocial crisis

⁵ Previously, when the antenatal diagnosis of a cleft was made, it was often in the context of other anomalies and medical problems. However, with improvements in ultrasonography, isolated clefts now can be diagnosed more readily. Nonetheless, it is prudent to caution families that definite plans will be formulated after the baby is born and has been carefully examined. (See Appendix II, pg. 20.)

BIRTH THROUGH 1 MONTH

PROBLEM	INTERVENTION
Need for interdisciplinary care of the 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 presurgical orthopedics, depending on the structure of the cleft (see Appendix X, pg. 42)
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 Appendix III, pg. 22) • 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 • Repeat 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 Appendix VIII, pg. 36)
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 presurgical orthopedics if indicated

1 THROUGH 4 MONTHS

PROBLEM	INTERVENTION
Need for interdisciplinary care of the 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 (if not already done) • Discuss diagnosis and treatment plan with the family • Address etiology and recurrence risks (if not already done) • Monitor presurgical orthopedics (if being used)
Feeding 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) • BAER and evoked otoacoustic emissions (if not already done) • Monitor for airway problems if diagnosis is Robin Sequence or other syndrome (see Appendix VIII, pg. 36)
Cleft lip and nasal deformity	<ul style="list-style-type: none"> • Repair cleft lip, usually at 3-5 months (cheiloplasty, see Appendix VII, pg. 33) • May include primary nasal reconstruction (rhinoplasty)
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, use of arm splints, pain management) • 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 psychological support to optimize child and family adjustment

Summary of critical interventions for ages 1 through 5 months:

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

5 THROUGH 15 MONTHS

PROBLEM	INTERVENTION
Need for continued interdisciplinary care of the 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 7 months of age and at 6-month intervals thereafter • Monitor for airway problems if diagnosis is Robin Sequence or other syndrome (see Appendix VIII, pg. 36)
Cleft palate	<ul style="list-style-type: none"> • Repair cleft palate, usually at 9-15 months (palatoplasty, see Appendix VII, pg. 33)
Family's need for accurate genetic information	<ul style="list-style-type: none"> • Provide genetic counseling (if not already done) (see Appendix VI, pg. 30)
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, pain management) • Assess the family's understanding of these instructions and ability to follow through • Help family make the necessary medical and social arrangements
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 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 5 through 15 months

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

16 THROUGH 24 MONTHS

PROBLEM	INTERVENTION
Need for continued interdisciplinary care of the 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 • Place/replace tubes if middle ear effusions persist > 3 months • Assess hearing every 6 months
Speech-language production	<ul style="list-style-type: none"> • Assess speech-language, especially velopharyngeal mechanism (see Appendix IX, pg. 39)
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

2 THROUGH 5 YEARS

PROBLEM	INTERVENTION
Need for continued interdisciplinary care of the cleft	<ul style="list-style-type: none"> • Cleft lip/palate team coordinates 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 (see Appendix VIII, pg. 36)
Speech-language production	<ul style="list-style-type: none"> • Assess speech-language, monitor for velopharyngeal insufficiency (VPI) • Formal VPI workup, as indicated (see Appendix IX, pg. 39) • Consider treatment options (speech therapy, surgery, obturation)
Abnormal dental development and alveolar defect	<ul style="list-style-type: none"> • Orthodontic exam and dental records at age 4 or 5 for bone graft timing and management of abnormal dentition • Dental extractions if needed
Repaired cleft lip/palate	<ul style="list-style-type: none"> • Consider lip/nose revision before school entry • Close palatal fistula(e) if indicated
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
- Revise lip/nose before school if needed
- Assess child's development, including language and psychological adjustment

6 THROUGH 11 YEARS

PROBLEM	INTERVENTION
Need for continued interdisciplinary care of the 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 • Assess hearing 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 Appendix VIII, pg. 36)
Speech-language production	<ul style="list-style-type: none"> • Assess speech-language; monitor for velopharyngeal insufficiency (VPI) • Formal VPI workup as indicated (see Appendix IX, pg. 39) • 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 development	<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)
Repaired cleft lip/palate	<ul style="list-style-type: none"> • Close palatal fistula(e) if indicated • Consider lip/nose revision as needed
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 Appendix V, pg. 27)
Child’s 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

12 THROUGH 21 YEARS

PROBLEM	INTERVENTION
Need for continued interdisciplinary care of the 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 • Assess hearing 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 Appendix VIII, pg. 36)
Speech-language production	<ul style="list-style-type: none"> • Assess speech, rule out velopharyngeal insufficiency (VPI) • Workup VPI if indicated (see Appendix IX, pg. 39) • Consider treatment options (therapy, surgery, obturation) • Communicate with school or outside speech clinician
Abnormal dental development	<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
Repaired cleft lip/palate	<ul style="list-style-type: none"> • Consider lip/nose revision • Monitor palate for fistula(e)
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 Appendix V, pg. 27)
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
- Assess overall psychological adjustment
- Review school issues/vocational plans

Overview: Summary of Key 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	INTERVENTION	APPENDIX
Cleft lip/palate team	<ul style="list-style-type: none"> • Coordinate care • Provide experienced specialists • Monitor medical and social issues 	I
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 	III, V
Pediatrics/primary care/genetics	<ul style="list-style-type: none"> • Monitor general medical issues • Assist with coordination of care and referrals • Monitor developmental and behavioral issues • Genetics/dysmorphology assessment 	II, IV, VI
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 	V
Surgery (plastic surgery, otolaryngology, oral and maxillofacial surgery)	<ul style="list-style-type: none"> • Lip and palate repair; lip scar revision • Velopharyngeal surgery for VPI • Ear tubes • Rhinoplasty • Alveolar bone graft • Jaw surgery; dental extractions • Airway assessment 	VII, VIII, XI
Audiology	<ul style="list-style-type: none"> • Monitor hearing • Recommend preferential seating and amplification when appropriate 	VIII
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) 	IX
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 	X

II *APPENDICES*

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 Appendix XVI, pg. 53).

1. ACPA Parameters of Care (Revised 2007)

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 birth defect.
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 possible and appropriate.
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.

2. Standards for Approval of Cleft Palate and Craniofacial Teams (2010)

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” available at: http://acpa-cpf.org/Standards/Standards_2010.pdf.

Teams are to be evaluated by self-assessment, 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 overseas 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 an 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) on or off team.

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 experience preparing 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.



APPENDIX II

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. However, it is easy to miss cleft lip on diagnostic ultrasounds, particularly those performed for routine indications in the physician's office as the American Institute of Ultrasound in Medicine does not even require views of the lip on screening ultrasounds.⁶

A wide range of fetal cleft lip detection rates (between 13-63%) have been reported with routine antenatal 2D ultrasounds.⁷ 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 week 18-23 gestation.⁸ The use of 3D ultrasound of the face improves detection rate significantly.^{9,10,11}

Thus, if there is a family history of clefting or if there is a concern about a possible cleft for other reasons, a referral should be made for a complete diagnostic ultrasound (including 3D images if possible) and genetic counseling. Ultrasounds obtained during 18-24 weeks gestation have been most accurate.

However, if patients are scanned earlier, especially if amniocentesis is being considered (typically performed at 15-17 weeks); a 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. It is still very difficult to make an antenatal diagnosis of a cleft palate, unless it is detected in association with a large cleft lip. Reported detection rates for cleft palate only vary from 0-22%.^{12,13}

Recently, fetal MRI has been used to detect fetal abnormalities including cleft palate. Experience and availability of fetal MRI, however, is extremely limited at this time.^{14,15}

Once a cleft lip/palate is identified, the family should be referred for genetic counseling to discuss other testing, including amniocentesis. During the genetic counseling session, a complete pregnancy and family history should be performed. This should include information on any teratogenic exposures, and the presence of family members with clefts or other birth defects, developmental problems and genetic syndromes. 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/palate team to learn about the care and management of children with clefts. The diagnosis of this birth defect creates a crisis for a family, so attention to psychosocial and emotional issues is essential at this time. Most families experience a grief reaction, although many feel anger and/or guilt as well. Relationships may be strained and there may be blaming of various family members.

Supportive counseling and referral to community resources may be needed. If appropriate resources are provided, most families can adjust adequately to this unexpected news.

At the family's first visit with the cleft lip/palate team, feeding instructions should be provided, and a clear plan for the newborn period should be formulated. Additional medical information provided at this visit should include a general description of the types of problems the baby may encounter. This opportunity to formulate a feeding plan, learn about the future care their child will receive, and meet the providers involved in this care can greatly increase a parent's sense of control and preparedness in the face of this unanticipated diagnosis.

⁶ American Institute of Ultrasound in Medicine Guidelines, 2003.

- 7 Johnson, N. & Sandy, J. (2003). Prenatal diagnosis of cleft lip and palate. *Cleft Palate-Craniofacial Journal*, 40, 186-189.
- 8 Wayne, C., Cook, K., Sairam, S., Hollis, B., Thilaganathan, B. (2002). Sensitivity and accuracy of routine antenatal ultrasound screening for isolated facial clefts. *British Journal of Radiology*, 75, 584-589.
- 9 Chmait, R., Pretorius, D., Jones, M., Hull, A. James, G., Nelson, T., Moore, T. (2002), Prenatal evaluation of facial clefts with two-dimensional and adjunctive three-dimensional ultrasonography: a prospective trial. *American Journal of Obstetrics & Gynecology*, 187, 946-949.
- 10 Tonni, G. Centini, G., Rosignoli, L. (2005). Prenatal screening for fetal face and clefting in a prospective study on low-risk population: can 3- and 4- dimensional ultrasound enhance visualization and detection rate? *Oral Surgery Oral Medicine Oral Pathology Oral Radiology Endodontics*, 100,420-426.
- 11 Maarse W, Bergé SJ, Pistorius L, van Barneveld T, Kon M, Breugem C, Mink van der Molen AB. Diagnostic accuracy of transabdominal ultrasound in detecting prenatal cleft lip and palate: a systematic review. *Ultrasound Obstet Gynecol*. 2010 Apr;35(4):495-502. PubMed PMID: 20235140.
- 12 Cash, C., Set, P., Coleman, N. (2001). The accuracy of antenatal ultrasound in the detection of facial clefts in a low-risk screening population. *Ultrasound in Obstetrics and Gynecology*, 18, 432-436.
- 13 Hanikeri, M., Savundra, J., Gillett, D., Walters, M., McBain, W. (2006). Antenatal transabdominal ultrasound detection of cleft lip and palate in Western Australia from 1996 to 2003. *Cleft Palate-Craniofacial Journal*, 43, 61-65.
- 14 Ghi, T., Tani, G., Savelli, L., Colleoni, G., Pilu, G., Bovicelli, L. (2003). Prenatal imaging of facial clefts by magnetic resonance imaging with emphasis on the posterior palate. *Prenatal Diagnostics*, 23, 970-975.
- 15 Kazan-Tannus, J., Levine, D., McKenzie, C., Lim, K.-H., Cohen, B., Farrar, N., Busse, R., Mulliken, J. (2005). Real-time magnetic resonance imaging aids prenatal diagnosis of isolated cleft palate. *Journal of Ultrasound Medicine*, 24, 1533-1540.

APPENDIX III

Nursing, Coordination and Feeding Issues

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:

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. 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 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. There are three options currently widely used. The first is the Cleft Palate Nurser made by the Mead Johnson Company.

It 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 Haberman™ feeder available from the Medela Company. This feeder consists of a large, compressible nipple with a one-way valve at its base that keeps the nipple full of milk. 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 third option is the Pigeon Cleft Palate Nurser distributed by Children's Medical Ventures. This system 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 or provide phone numbers for ordering. All three 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 the cleft lip/palate team for referral to the

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 the specialized bottle allows the mother who wishes to breast-feed the ability to give her baby her own milk.

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 an upward climb on the growth curve, feeding re-evaluation and changes in technique may be needed. The nurse or 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 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 can be irritated by food. Following each meal with swallows of milk or water is all that is needed to remove any remaining food in the mouth.

APPENDIX IV

Primary Care

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 otitis media, 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. One reason for this is the difficulty they have creating suction with the cleft palate, leading to 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. Thus, growth parameters must be monitored very closely in the first few weeks of life. Adequate feeding is possible with special bottles and techniques; both available from cleft feeding specialists (usually nurses or feeding therapists) associated with cleft lip/palate teams. The knowledgeable nurse in the newborn nursery can initiate proper feeding, but it is essential that these babies be monitored over the long-term (see Appendix III, pg. 22).
2. **Robin Sequence.** Robin Sequence consists of mandibular hypoplasia (micrognathia), glossoptosis, and a posterior U-shaped cleft palate which results in a posterior tongue position that can interfere with breathing. 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 (glossoptosis), the baby should be placed in the prone position. If this does not relieve the infant's distress and allow for normal oxygenation (as monitored by an oximeter), then placement of a nasopharyngeal (NP) tube or temporary oropharyngeal tube is indicated. It is desirable to involve an experienced otolaryngologist if any of these interventions are needed.

Some of these babies may require prolonged use of the NP tube, a tracheotomy, or early mandibular distraction.

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 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 these babies: 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, occupational therapy and respiratory therapy. Occasionally, a polysomnogram (sleep study) with CO₂ monitoring may be necessary to determine if ventilation is adequate. These issues are best addressed with a cleft lip/palate team and in a hospital where there is access to pediatric anesthesia. In the event of a respiratory emergency, these babies can be very difficult to intubate because of their abnormal anatomy.

3. **Middle Ear Effusions.** Infants with cleft palate are at high risk for recurrent and chronic middle-ear disease (90-95%). Many of these infants will require ear tube placement. The hearing loss which may result from these effusions can be significant, and may interfere

with speech and language development. Due to the difficulty of reliably diagnosing middle ear effusions in infants, it is recommended that an otolaryngologist periodically evaluate these children (see Appendix VIII, pg. 36).

4. **Genetics/Dysmorphology.** Genetic counseling is necessary to provide patients and families within formation on recurrence risks, and should be offered after the child is born, at adolescence or whenever family questions about etiology and recurrence risks. Because a significant number of children with cleft lip/palate have genetic syndromes (especially those with a cleft palate), this possibility should be considered if a patient has atypical facial features, developmental delays, learning problems 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. Research suggests that pre-conceptual and prenatal dietary supplementation with folic acid may decrease the risk of cleft lip/palate, although the optimum dosing is not known. In any case, prospective mothers should take a minimum of 0.4 mg of folic acid—the amount in most prenatal vitamins—starting three months prior to conception. This is the general recommended dose for prevention of neural tube defects. The dose may be higher to prevent recurrences of cleft lip and palate (or neural tube defects) in families with a positive history. (See Appendix VI, pg. 30.)

5. **Dental Issues.** Dental issues are of paramount importance in the management of patients with cleft lip/palate. Unfortunately, dental care is often viewed as optional and not included in typical definitions of medical necessity, which is particularly deleterious for patients with cleft lip/palate. The PCP is often asked to authorize visits to orthodontists and 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 is 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 6-10 years, depending upon dental development. The alveolar bone graft provides the foundation for the erupting teeth and support for the nasal base. Orthodontic interventions are necessary before and after this bone graft.
- 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. A certain number of these 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 monitor these issues. (See Appendix X, pg. 42; Appendix XI, pg. 44.)

6. **Development.** Development should be monitored in all children. However, children with cleft lip/palate are at increased risk for developmental and behavioral problems. First, speech production problems can result from the anatomical differences associated with the cleft palate. Second, speech and language delays can result from intermittent hearing loss accompanying recurrent or persistent middle ear effusions. Third, a significant number of patients with cleft lip/palate (especially those with isolated cleft palate) will have a syndrome with developmental implications. Finally, psychosocial issues stemming from the cleft can affect the child's emotional well-being, school performance and overall developmental adjustment. Specialty help is available in these areas to assist children and families with these issues. Both PCPs and cleft lip/palate teams should monitor developmental, behavioral and psychosocial issues. Interventions and resources should be recommended as appropriate (see Appendix V, pg. 27).
7. **General Medical Care.** These children, like all others, require ongoing well-child care. It may be difficult to accomplish this when the focus of the first months is on cleft-related issues. However, it is important that the PCP continue regular health maintenance, including administration of immunizations, attention to any other health problems and provision of anticipatory guidance in other areas of health and development. During adolescence, health issues should be monitored appropriately, and the PCP should include 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.



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 repairable 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 mid- to late-teens). 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 facial disfigurement, the maturing child's preferences should be respected. By age 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 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 appear to be 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. velocardiofacial 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 (velocardiofacial syndrome, fetal alcohol syndrome), but can occur in children with isolated clefts as well. Symptoms may include social withdrawal, depression, conduct problems or school failure. 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 1 on the following page lists key psychosocial and developmental interventions by age.



TABLE 1: Key Psychosocial and Developmental Interventions by Age

AGE	INTERVENTION
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) • 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, including the availability of genetic counseling and prenatal ultrasound, and pre-conceptual folic acid supplementation • 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

APPENDIX VI

Genetics/Dysmorphology

A dysmorphology or genetics assessment is part of the complete evaluation of every child with a cleft. Cleft lip and palate affects approximately 1:1,000 Caucasian, 1:500 Asians, and 1:2,000 African Americans in this country. 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.^{16, 23}

Although there are more than 400 syndromes reported in association with cleft lip or cleft palate¹⁷ the following three syndromes should receive special consideration. Velocardiofacial syndrome, due to a deletion of chromosome 22q11.2, should be considered in children with velopharyngeal insufficiency, submucous cleft palate, or cleft palate. Van der Woude syndrome, an autosomal dominant condition, should be considered in a child with either cleft lip/palate or cleft palate who has a family history of mixed clefting in which either the child or another family member has lower lip pits. 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. Children who have a family history of mixed cleft types (both cleft lip and cleft palate in different family members) should be evaluated for hypodontia, lip pits, and anosmia. Additional studies, including ophthalmologic consultation, echocardiography, or other radiographic studies, and laboratory studies

(such as chromosome analysis) should be directed by the examination and family history to facilitate syndrome/chromosomal diagnoses. These conditions may have prognostic implications that must be taken into account to help guide medical decisions.

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 the nonsyndromic child, 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%. This risk increases if there are additional family members with clefts. Condition-specific recurrence risks and prenatal testing options should be provided to families of a child with syndromic clefting condition.

Parents should be informed of the option of ultrasonography for future pregnancies. A discussion regarding the sensitivity of prenatal ultrasound to detect clefts should be considered given that only 22% of cleft palates, 67% of cleft lip without cleft palate, and 93% of cleft lip with cleft palate can be detected on antenatal ultrasound studies between 18-24 weeks gestation if appropriate facial views are obtained.¹¹¹⁸ Similarly, 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.

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 maybe

ordered. 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. When the initial crisis has subsided (generally after six months), it is appropriate to bring up risks of recurrence again.

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.

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 2 on the following page is a summary of genetic and dysmorphology interventions by age.

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- 16 Wyszynski, D. (2002). *Cleft Lip and Palate: From Origin to Treatment*. New York: Oxford University Press.
 - 17 Online Mendelian Inheritance in Man (OMIM) - database of syndromes associated with cleft palate. www.ncbi.nlm.nih.gov/entrez/query.fcgi?CMD=search&DB=omim (Search "syndromes with cleft palate or cleft lip")
 - 18 Cash, C., Set, P., Coleman, N. (2001) The accuracy of antenatal ultrasound in the detection of facial clefts in a low-risk screening population. *Ultrasound in Obstetrics and Gynecology*, 18, 432-436.

TABLE 2: Genetic and Dysmorphology Interventions by Age

AGE	INTERVENTION
Prenatal	<ul style="list-style-type: none"> • Genetics consultation if ultrasound is abnormal, 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)
16-24 months	<ul style="list-style-type: none"> • Consider genetic syndrome if developmental delays or other atypical features are present • Additional genetics workup as indicated
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

APPENDIX VII

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.¹⁹
- 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 discuss surgical planning and outcomes.
- Ongoing continuing medical education and expertise in cleft lip/palate care.

Table 3 lists key plastic surgery interventions by age.

¹⁹ Depending upon locale, surgeons from these subspecialties may perform plastic surgery procedures on children with cleft lip/palate. In any case, the particular education, training and experience of the surgeon which qualifies him/her to perform these repairs must be established. This should include documented evidence of residency training (as an operating surgeon, not as an assistant) in lip, palate and nasal procedures. This cannot include patients treated on overseas missions or treated for craniofacial trauma. (ACPA Team Standards Self-Assessment Instrument, 1996)

TABLE 3: Key Plastic Surgery Interventions by Age

AGE	INTERVENTION
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 presurgical orthopedics in consultation with the appropriate dental specialist^a
1-3 months	<ul style="list-style-type: none"> • Monitor progress of presurgical orthopedics with orthodontist
3-15 months	<ul style="list-style-type: none"> • Repair cleft lip, usually at 3-5 months^b • Repair cleft palate, usually at 9-15 months^c
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^d • Lip/nasal surgery as needed for residual deformity
6-11 years	<ul style="list-style-type: none"> • Consider surgical management as needed for VPI^d • Bone graft to alveolar cleft with closure of oro-nasal fistulae^e • Lip/nasal surgery as needed for residual deformity
12-21 years	<ul style="list-style-type: none"> • Rhinoplasty as needed (nasal revision) • Lip/nasal surgery as needed for residual deformity • Orthognathic surgery (see Appendix XI, pg 44,^b)

a About presurgical orthopedics

It is difficult to obtain a good lip repair if the cleft in 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 presurgical maxillary orthopedics. This can involve the application of external taping across the cleft, a surgically applied internal device (Latham device)²⁰, or a plastic molding device taped in place (Nasoalveolar molding device)^{21,22}. The specifics regarding the timing and nature of the orthopedic device vary from center to center.

Potential advantages and disadvantages for a given child should be discussed with the cleft lip/palate team at the time treatment is recommended.

b About cleft lip repair

If other medical factors are stable, cleft lip repair is usually done when the child is between 3-5 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 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 certain percentage of children undergoing this procedure, later alveolar bone grafting may not be needed.

c About cleft palate repair

The ideal time for palatoplasty is less clear. Theoretically, optimal speech is best served by earlier repair, and optimal facial growth by later repair. Today 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 often involves reorientation and closure of the layers of the soft palate, as well as tissues of the hard palate. This helps to minimize nasal air leakage and velopharyngeal insufficiency. Occasionally, the palate is closed in

two stages; however, there may be a higher risk of fistulae and speech problems with this approach. The usual practice is to repair the palate completely the first time. Pictures of typical clefts are provided in Appendix XIII, pg. 48-49.

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 to achieve VP closure. If this is not sufficient, a sphincter pharyngoplasty or pharyngeal flap may be considered. Disadvantages include: a significant risk for over-correction of the air leak leading to post-operative obstructive sleep apnea (OSA) and hyponasalality. 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 VP I workup 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 Appendix IX, pg. 39.)

e About alveolar bone grafting

Alveolar bone grafting is usually necessary to close the residual bony cleft in the maxilla. These procedures are performed by an oral and maxillofacial surgeon or a plastic surgeon with special training/expertise in this area. See Appendix XI, pg. 44.

f About jaw surgery

Oral-maxillofacial surgeons and craniofacial plastic surgeons may do orthognathic surgery. Refer to Appendix XI, pg. 44, for discussion of jaw surgery.

²⁰ Millard, D. & Latham, R. (1990). Improved primary surgical and dental treatment of clefts. *Plastic and Reconstructive Surgery*, 86, 856-871.

²¹ Grayson, B. & Cutting, C. (2001). Presurgical nasoalveolar orthopedic molding in primary correction of the nose, lip, and alveolus of infants born with unilateral and bilateral clefts. *Cleft Palate-Craniofacial Journal*, 37, 193-198.

²² Santiago, P., Grayson, B., Cutting, C. (1998). Reduced need for alveolar bone grafting by presurgical orthopedics and primary gingivoperiosteoplasty. *Cleft Palate-Craniofacial Journal*, 35, 77-80.27

²³ Losee, J & Kirschner, R (2008) *Comprehensive Cleft Care*. McGraw-Hill Professional.

APPENDIX VIII

Otolaryngology

Infants with Robin sequence (cleft palate, micrognathia, and glossoptosis) frequently have upper airway compromise. If placing these infants in a prone position is not sufficient to alleviate the respiratory distress, the placement of a nasopharyngeal tube, tongue-lip adhesion, or tracheotomy may be necessary. Mandibular distraction is increasingly used for infants with significant airway compromise to avoid tracheotomy. In such cases, additional studies including: 1) nasendoscopy/laryngoscopy to assess airway, or 2) polysomnography (sleep study) to determine severity may be indicated.

On some teams, otolaryngologists repair the cleft palate (Appendix VII, pg. 33). Otolaryngologists also take part in the assessment and surgical management of velopharyngeal insufficiency in children with cleft lip/palate. They perform the nasopharyngoscopy with the speech-language pathologist to assess the velopharyngeal gap, and together they recommend to the team the appropriate surgical or prosthetic intervention. Depending on the otolaryngologist's surgical expertise, they may perform a palate lengthening procedure, pharyngeal flap, or sphincter pharyngoplasty to manage the VPI (Appendix VII, pg. 33). Some children develop obstructed breathing following pharyngeal flap or sphincter pharyngoplasty. In such cases, polysomnography may be indicated.

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.

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

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. A persistent conductive hearing loss can adversely influence speech and language, with 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 impedance audiometry (tympanograms). In addition, 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.^{24,25}

• *Table 4 on the following page lists key otolaryngology and audiology interventions by age.*

TABLE 4: Key Otolaryngology and Audiology Interventions by Age

AGE	INTERVENTION
Birth-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: ear tubes may be placed with lip repair if indicated • 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 repair²⁵ 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 6-7 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/adenoids, sphincter pharyngoplasty or pharyngeal flap present) • Deviated septum may require repair • Use caution with adenoidectomy²⁶ • Monitor middle ear status every 6 months • If ears clear > 18 months, consider removing ear tubes • 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 Appendix IX, pg. 39) • Consider surgical interventions for VPI (palate lengthening surgery, sphincter pharyngoplasty or pharyngeal flap) usually by 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 adenoidectomy¹² • Monitor middle ear status every 6 months; 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 to improve velopharyngeal function (palate lengthening surgery, sphincter pharyngoplasty or pharyngeal flap) • Monitor hearing every 6-12 months until normal for 2 consecutive years

²⁴ Cunningham, M. & Cox, E. (2003). Hearing assessment in infants and children: recommendations beyond neonatal screening. *Pediatrics*, 111, 436-440.

²⁵ Joint Committee on Infant Hearing. (2000). Year 2000 position statement: principles and guidelines for early hearing detection and intervention. *Pediatrics*, 106, 798-817.

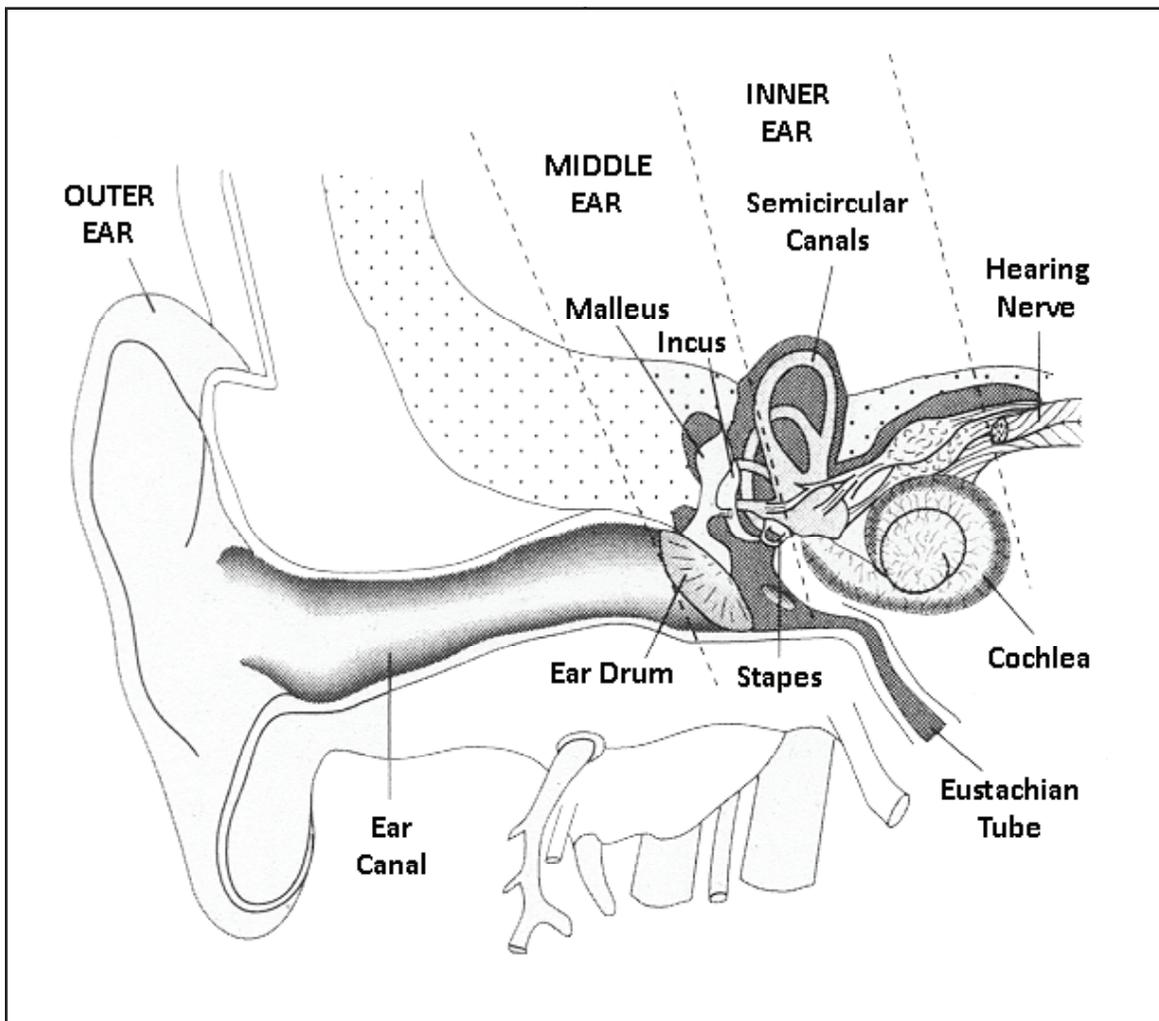
²⁶ Cleft palate repairs are usually performed by plastic surgeons, but in some centers otolaryngologists or oral and maxillofacial surgeons with special training perform these surgeries.

²⁷ Adenoidectomy should be approached cautiously in the patient with a cleft palate as adenoids often play a role in velopharyngeal competency, particularly the posterior and inferior portions of the adenoids. The cleft lip/palate team should review these issues before making a final decision on adenoidectomy.

Anatomy of the Middle Ear

In children with cleft palate, the Eustachian tube often does not function properly, in part, because muscles responsible for opening the eustachian tube are involved in the cleft. If the eustachian tube does not open effectively, then pressure changes occur and fluid may fill the middle ear space, which often leads to an ear infection. With fluid or infection, the eardrum and the ossicle bones of the middle ear no longer vibrate properly, hindering the transfer of sound from the air through these structures to the nerves of the inner ear. If the eustachian tube remains blocked or if the fluid persists long enough, the eardrum or the ossicles may be permanently damaged. The function of myringotomy tubes is to keep the middle ear aerated, normalize hearing, and prevent fluid from accumulating.

FIGURE 1: THE ANATOMY OF THE EAR



APPENDIX IX

Speech Problems in Children with Clefts

A cleft palate poses serious threats to speech development. In spite of surgical closure of the palate, many children remain unable to create adequate intraoral pressure for normal speech. This is called velopharyngeal insufficiency (VPI), and can usually be eliminated with surgical or prosthetic treatment to obturate (close) the site of air leakage. Speech and language assessments should take place regularly as outlined below by a certified speech-language pathologist with expertise in cleft lip/palate care. The team speech-language pathologist should also communicate with school and outside clinicians, and monitor therapy taking place with professionals who are not part of the team.

When VPI is significant and interferes with a child's communication, plans for a full "VPI workup" can be made. This can take place when the child is old enough to cooperate (usually 2-5 years old). This workup should include an oral exam, analysis of speech articulation, videofluoroscopic speech study and naso-endoscopy, and is generally conducted by a team speech-language pathologist, otolaryngologist and radiologist. Occasionally, nasometric studies and aerodynamic measures are also used. When the results of the VPI workup are available, recommendations for surgery, speech prosthesis or speech therapy can be made by the team.

Some children with cleft lip/palate also have an articulation and/or language disorder with or without VPI. Regular monitoring by a speech-language pathologist will ensure that other speech and language disorders receive timely assessment and intervention.

Table 5 lists key speech-language interventions by age.

TABLE 5: Key Speech-Language Interventions

AGE	INTERVENTION
6-9 months	<ul style="list-style-type: none"> • Speech-language consultation to address abnormal speech patterns
9-24 months	<ul style="list-style-type: none"> • Speech-language evaluation (>three months after palate repair)
2-5 years	<ul style="list-style-type: none"> • Annual speech-language evaluation • VPI workup if needed • Proceed with prosthetical or surgical management ^b (if child cannot cooperate, interventions may be recommended in the absence of a full workup) • If indicated, provide or refer for articulation and/or language therapy • Communicate with school and/or outside speech-language pathologist
6-11 years	<ul style="list-style-type: none"> • Annual speech-language evaluation until involution of adenoids • VPI workup and interventions if needed • Provide or refer for speech-language therapy if needed • Communicate with school and/or outside speech-language pathologist, and 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 if needed • Communicate with school and/or outside speech-language pathologist • Interventions for VPI if needed

^a About speech prostheses to treat VPI

A speech prosthesis is a removable appliance that is attached to the teeth with wire clasps, and is fabricated by a dental specialist in consultation 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 a plastic bulb that matches the gap's size and shape. The design of the prosthesis determined by the pattern of air leakage shown in the VPI workup, 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:

- May decrease the development of VPI-related misarticulations in young children, thereby allowing for normal speech development and decreasing the need for long-term speech therapy
- 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
- A speech bulb may make the child a better surgical candidate by decreasing the variability and/or size of the VP gap
- Some patients will not need permanent management of VPI (either prosthetic or surgical) following a course of prosthetic management
- 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 periodontal problems
- Can be lost or broken

^b About surgery to treat VPI

Refer to Appendix VII, pg. 33, 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 speech sounds requiring closure, but remain open for other speech sounds and for breathing during sleep. The advantage of surgery is the potential permanent resolution of VPI symptoms.

The disadvantage is the potential for over-correction, creating post-operative 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 and parents, and the relevant 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.

Anatomy of the Roof of the Mouth

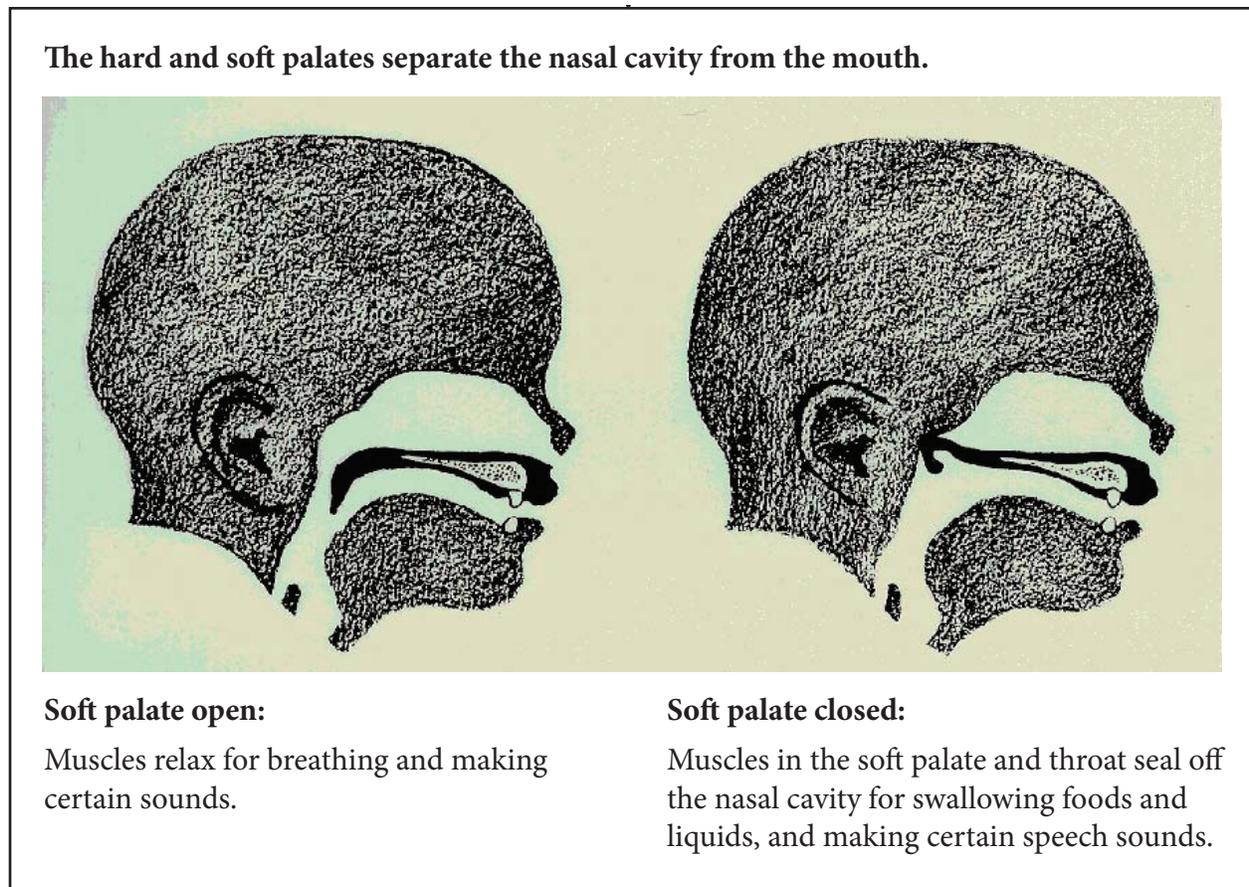
Hard Palate

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 2: ANATOMY OF THE PALATE



Adapted with permission from Looking Forward: A Guide for Parents of the Child with Cleft Lip and Palate (Mead Johnson, 2000).

Orthodontics/Dental Medicine

Orthodontics (also known as dentofacial orthopedics) 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 he/she 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. 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 presurgical 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 Appendix XI, pg. 44). Cleft repair without orthodontic intervention at the proper stages will 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 6 on the following page lists key orthodontic and dental interventions by age.

TABLE 6: Key Orthodontic/Dental Interventions for the Child with Cleft Lip/Palate

AGE	INTERVENTION
Birth-5 months	<ul style="list-style-type: none"> • Presurgical orthopedics ^a
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
2-5 years	<ul style="list-style-type: none"> • Orthodontic dental records (X-rays, photos) at 4-5 years of age in preparation for evaluation of teeth and cleft size • Monitor dental hygiene; provide appropriate preventive and restorative care • Orthodontic records as needed to determine timing of bone graft • Assist with speech prosthesis ^b, as needed
6-11 years	<ul style="list-style-type: none"> • Positioning of maxillary segments in preparation for alveolar bone graft • Recommend extractions as needed • Monitor dental hygiene; provide appropriate preventive and restorative care • Assist with speech prosthesis as needed • Monitor growth; maxillary protraction as needed
12-21 years	<ul style="list-style-type: none"> • Dental records to monitor jaw growth, dental development and bone graft • Braces for dental alignment as needed • If retrusion is severe, combination of jaw surgery and braces is needed • Prosthetic replacement of missing teeth as needed ^c • Monitor dental hygiene; provide appropriate preventive and restorative care • Prepare surgical splints for orthognathic surgery as needed

^a About presurgical orthopedics

If the cleft lip is very 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 normal position by use of external taping across the cleft, a surgically-placed intraoral appliance, or nasoalveolar molding device taped into place. 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.

^b 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 Appendix IX, pg. 39, for a discussion of VPI and obturators.

^c 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 false teeth. An option that is increasingly available is the use of dental implants (artificial teeth permanently placed in the alveolar ridge).

APPENDIX XI

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
- In some locales, 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 him/her to perform the surgery should be established

Table 7 lists key oral and maxillofacial surgery interventions by age.

TABLE 7: Oral and Maxillofacial Surgery Interventions

AGE	INTERVENTION
6-11 years	<ul style="list-style-type: none">• Bone graft of alveolar cleft and closure of oro-nasal fistulae ^a• Selective tooth extraction as needed
12-21 years	<ul style="list-style-type: none">• If needed, orthognathic (jaw) surgery in consultation with the orthodontist ^b

^a About alveolar bone grafting

Even after the repair of the cleft lip and/or palate, there typically remains a bony cleft in the maxilla and an opening running from the nose to the mouth (under the upper lip) called the oro-nasal fistula.

When teeth erupt into this cleft, they are unsupported by bone and will likely be lost. Bone grafting of the cleft(s) is essential. It joins the cleft segments of the maxilla, provides a bony base for erupting adult dentition 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 iliac crest, though bone from the skull or tibia may also be used. This procedure is usually performed by an oral/maxillofacial surgeon, or a 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 nasoalveolar molding in infancy and aging ivoperiosteoplasty 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.

^b About orthognathic (jaw) surgery

The mid-face (maxilla) is usually fully developed by age 15 years. In the child with a cleft, this bone may be hampered in its growth and development by scars from previous soft tissue surgical procedures.

A size discrepancy between the upper and lower jaws results producing a concave appearance.

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 dentoskeletal normalization.

Orthognathic surgery is complex and requires the combined efforts of the orthodontist and surgeon. It requires pre-operative orthodontic treatment 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 in some cases, 3D CT models of the patient's facial bones and teeth.

A long established surgical procedure is a maxillary advancement, and this 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, 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. Screws on the device are turned daily and pull the healing bones slowly apart until enough lengthening has been achieved. Potential advantages and disadvantages of these two 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 surgeon, or by a plastic-craniofacial surgeon with training in this area.

Following this surgical management of the maxilla, the final phase of the orthodontic treatment is begun. During this phase, which usually lasts about one year, the final occlusion between upper and lower teeth is established. Pre-and post-operative speech evaluation is required, because VPI (see Appendix IX, pg. 39) can result from the advancement of the upper jaw.

In the child with a cleft, orthognathic surgery is complicated by the distorted anatomy and residual scar tissue from the cleft palate repair. As a result, moving and stabilizing the maxilla may be very difficult. In some cases, it is necessary to move the maxilla forward and the mandible back to achieve the proper occlusion.



Ethics and Children with Cleft Lip/Palate

Ethical issues arise whenever medical decisions are made for children who cannot participate fully in the decision-making process. These concerns take on special importance for children with cleft lip/palate, because the goal of medical therapies for these children is to improve the quality of life, a subjective outcome. 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:

Incompetency of children. Children are presumed incompetent by virtue of age, and cannot give informed consent.

Surrogate decision-makers and the “best interests standards.” Medical decisions are made for children by surrogates, usually the parents, who must act in the best interests of the child. Parents cannot put the child’s health in jeopardy by requesting extreme therapies or neglecting medical needs. This is in contrast to medical decision-making for the competent adult, who has the right to request or refuse a range of available interventions. Although parents are given considerable latitude in defining the “best interests” of their child, their choices must fall within a range of reasonable responses. Suspected cases of medical neglect or abuse must be reported to the appropriate authorities. In the rare case where parents are not deemed appropriate decision-makers for their child, other surrogates are appointed by the courts.

Informed consent. Health care providers are obligated to fully inform parents 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 should recognize cultural and ethnic differences, which can impact the family’s adjustment to the visible birth defect and their attitude towards medical services. Such conversations often provide valuable insights into the beliefs and attitudes of families, and create opportunities for education around causality, etc.

Role of the child. As the child matures, it is appropriate to bring him/her into the decision-making process. By the developmental age of 7 years, most children have the capacity to assent to interventions and procedures, and therapies should be explained carefully; refusals should be honored when possible. The more serious or emergent the medical circumstances, the more parents and professionals are justified in ignoring the child’s preferences in order to administer needed therapies. Under such circumstances, the importance of the therapies and the reasons for overriding the child’s wishes should be carefully explained, and the child should be offered support during the interventions. The more elective the intervention, the more essential it is that the child’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 elective and 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 and developmental outcome, and normalizing speech and hearing. Because of the subjective nature of these therapy goals, it is important that children participate in the decision-making process as their age and abilities allow. By adolescence, most youngsters are able to share fully in the decision-making process, and should have veto power for elective procedures relating primarily to their quality of life. There is considerable variability in the degree to which the dentofacial disfigurement of this birth defect impacts

self-esteem and quality of life, and it is difficult to presume the impact for another individual. Thus elective surgeries to correct these deformities are often postponed until the child is old enough to share in the decision-making, unless the interventions must be performed earlier to be successful or safe.

Paying for quality of life interventions. Such surgeries are often viewed as optional by third-party payers, who have been reluctant to pay for quality of life interventions. Most of these surgeries are not “cosmetic” (improving upon the normal), but are undertaken to bring the individual with a visible birth defect closer to the range of normal. Oral health interventions also impact the quality of life, and are omitted from definitions of medical necessity. The recent commissioning of a *Surgeon General’s Report on Oral Health* provides an important opportunity to underscore the relationship of oral health to overall health, and its impact on the quality of life. Hopefully this report will ameliorate the lack of attention given to the oral health area, just as the earlier *Surgeon General’s Report on Children with Special Healthcare Needs* (1987) underscored the need for comprehensive and coordinated team care for children with complex medical problems, including cleft lip and palate.

Changes in Health Care: Access and Advocacy

Changes in the health care system have potential impact for children with cleft lip/palate. Children with relatively rare medical conditions such as these may have difficulty accessing care in such systems, because specialists with adequate experience and training in this field are relatively rare. Few managed care systems have the specialized staff needed to provide multidisciplinary team care for these children, or the volume of cases needed to monitor outcomes. Yet, managed care systems may be reluctant to go out of contract for full team care. The risk is that care is parceled out in an uncoordinated and piecemeal fashion to a number of providers inside and outside of the managed care plan. Although health care systems have an obligation to clearly disclose benefits and limitations of care, families should take responsibility for inquiring about coverage for needed services. Advocacy efforts

within such systems can include referencing and providing copies of the *ACPA Parameters of Care* (2004), *ACPA Team Standards Assessment Instrument* (2007) [www.acpa-cpf.org/teamcare/Parameters07rev.pdf], and this document, the *Critical Elements of Care for Cleft Lip/Palate* for Washington state (2010).

On the positive side, Medicaid managed care programs are now enrolling some children who previously had no access to health care. Thus there is the potential for an overall positive impact of managed care for children with cleft lip/palate because such systems often stress prevention and health maintenance. The preventive aspects of multidisciplinary team care for clefts should be emphasized with these providers and systems. The advantages of having experienced and skilled surgeons perform initial surgeries, psychosocial professionals to identify and target families in need of support, and dental professionals to monitor craniofacial growth and dental hygiene and plan optimal timing for interventions, should be clearly explained. Better early care means fewer complications and less need for surgical revisions; better outcomes for the child and family; and less cost to the health care system. Multidisciplinary team care for children with cleft lip/palate is essential health maintenance for children in need of complex care, and should be considered part of a “basic level of health care.” Special advocacy is needed by parents, patients and providers to make third-party payers and health policy makers aware of the importance of these issues.

APPENDIX XIII

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 3A is a drawing of a normal roof of the mouth. Figure 3B through 3F 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 3A: NORMAL ROOF OF THE MOUTH

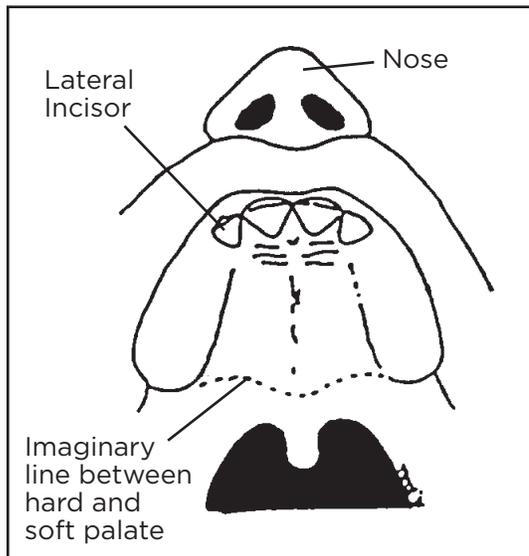
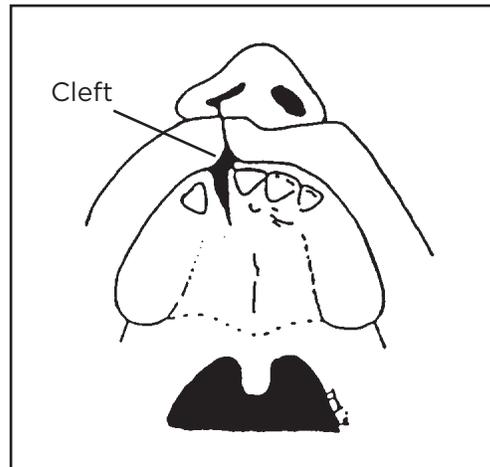
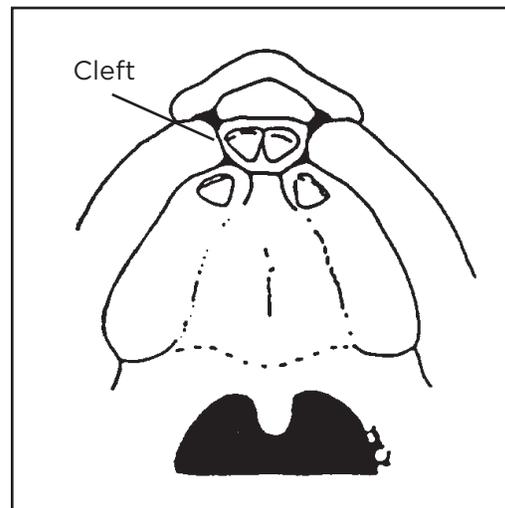


FIGURE 3B: UNILATERAL CLEFT (EXTENDS COMPLETELY THROUGH THE LIP AND ALVEOLUS (GUMLINE))



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

FIGURE 3C: BILATERAL CLEFT LIP

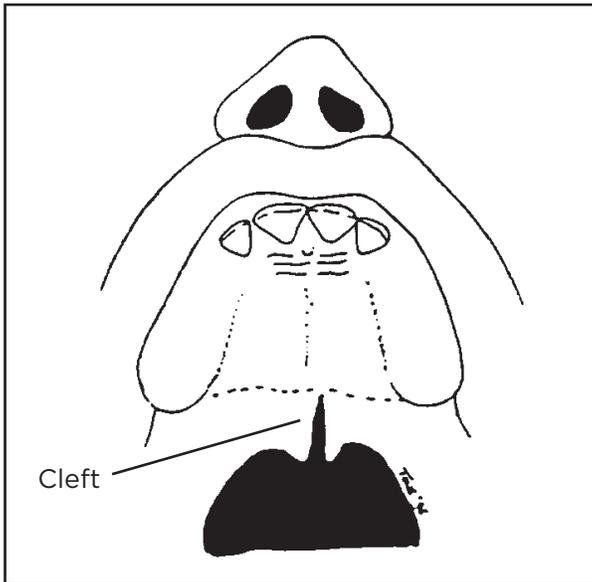


The alveolus and the lip between the two cleft areas have been displaced from their normal alignment, and this allows this segment of the premaxilla to swing upward and protrude. In this situation, presurgical orthopedics may be desired.

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.

Figure 3D is a drawing of a cleft of the soft palate. Figure 3E shows a cleft involving both the hard and the soft palates. Figure 3F shows a cleft involving the lip and entire palate.

FIGURE 3D: CLEFT OF THE SOFT PALATE



Even a “small” cleft of the soft palate creates 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 3E: CLEFT WITH BOTH HARD AND SOFT PALATE

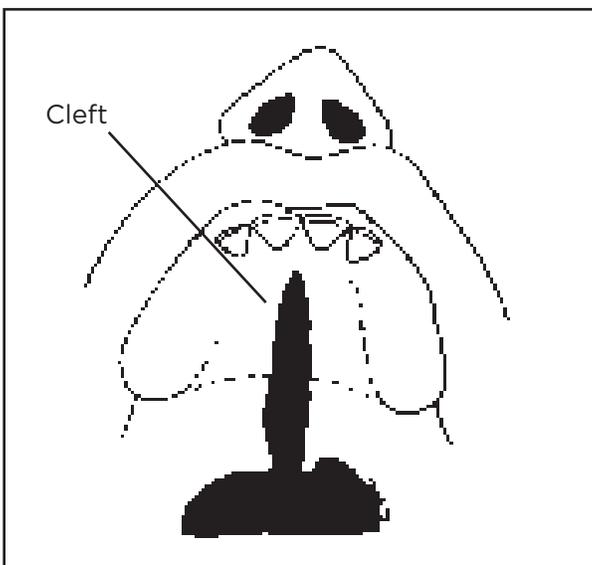
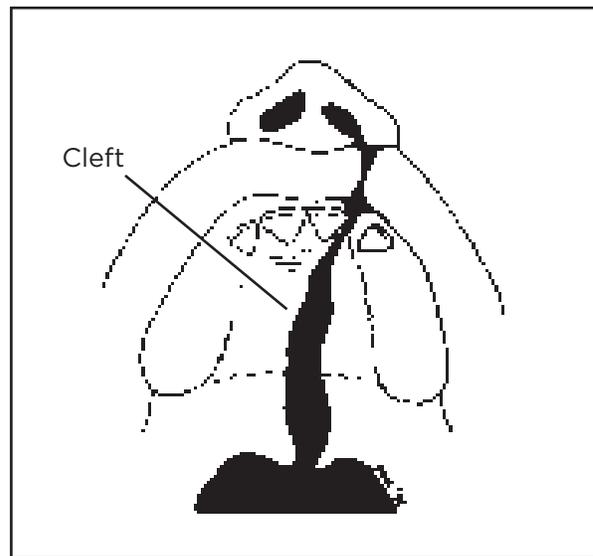


FIGURE 3F: CLEFT INVOLVING THE LIP AND THE ENTIRE PALATE



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.

APPENDIX XIV

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: pediatrics, plastic surgery, otolaryngology, audiology, speech-language pathology, pedodontics, psychiatry, orthodontics, prosthodontics, psychology, social service, nursing, radiology, genetics and oral surgery.

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: inflammation of the middle ear, where thick mucous fluid accumulates; this is a common problem for children with cleft palates.

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 pediatrics, the area of medicine dealing with the health and disease 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.

Adapted with permission from: Berkowitz, Samuel. The Cleft Palate Story. Quintessence Publishing Co., Inc. Chicago: 1994.

APPENDIX XV

Washington State Cleft Lip/Palate Teams

SEATTLE

**Craniofacial Center
Seattle Children's Hospital**

4800 Sand Point Way NE
P.O. Box 5371, MS: M2-8
Seattle, WA 98105

Medical Director: Michael L. Cunningham, MD, PhD
206-987-2528

Surgical Director: Richard Hopper, MD
206-987-3256

Contact: Marsha Ose, RN, BSN, MS

Toll Free: 866-987-2000, ext. 72188

Fax: 206-987-3824

SPOKANE

**Greater Inland Empire Maxillofacial Team
Children with Special Health Care Needs**

West 1101 College Avenue
Spokane, WA 99201-2095

Director: Lisa M. Ross, RN, BSN
Program Supervisor
509-324-1657

Contact: Laurel F. Vessey, RN, BSN
Team Coordinator
509-324-1652

Fax: 509-324-1699

TACOMA

**Southwest Washington Maxillofacial Review Board
Mary Bridge Children's Health Center**

PO Box 5299 MS: 316L-1-CHN
311 South L Street
Tacoma, WA 98415-0299

Phone: 253-403-1559

Toll Free: 800-552-1419, ext. 1559

Fax: 253-403-4700

Contact: Margie Bryerton, RN, BSN, CCRN
margie.bryerton@multicare.org
253-403-1559

YAKIMA

**Central Washington Cleft Palate Team
Children's Village**

3801 Kern Road
Yakima, WA 98902

Team Facilitator: Kerry Harthcock, MD

Contact: Cathy Buchanan, MSW
Team Coordinator
or

Jill Hilmes, MA
509-574-3200

Toll Free: 800-745-1077

Fax: 509-574-3210

APPENDIX XVI

References and Resources

GENERAL REFERENCES

- American Association of Oral and Maxillofacial Surgeons. (1995). *Standards of Care for Cleft Lip and Palate*. Berkowitz, S. (1994). *The Cleft Palate Story*. Chicago: Quintessence Publishing Co., Inc.
- American Cleft Palate-Craniofacial Association. (1993) Parameters for the evaluation and treatment of patients with cleft lip/palate or other craniofacial anomalies. *Cleft Palate-Craniofacial Journal*, 30, (Suppl. 1).
- American Cleft Palate-Craniofacial Association. (2010). *Standards for Approval of Cleft Palate and Craniofacial Teams-Commission on Approval of Teams*. www.acpa-cpf.org/Standards/Standards_2010.pdf
- American Cleft Palate-Craniofacial Association. (1996). *Team Standards Self-Assessment Instrument*.
- Berkowitz, S. (1994). *The Cleft Palate Story*. Chicago: Quintessence Publishing Co., Inc.
- Gorlin, R.G. (2001). *Syndromes of the Head and Neck*. [4th edition.] New York: Oxford University Press.
- Journal of Ultrasound Medicine*, 10, 577. (1991)
- Losee, J & Kirschner, R. (2008). *Comprehensive Cleft Care*. McGraw-Hill Professional.
- Moller K.T. & Starr, C.D. (Eds.). (1993). *Cleft Palate: Interdisciplinary Issues and Treatment*. Austin, TX: Pro-ed.
- Office of Maternal and Child Health. U.S. Department of Health and Human Services. (1987). *Surgeon General's Report: Children with Special Health Care Needs*.
- Shaw, William C., et al. (1992). A six-center international study of treatment outcome in patients with clefts of the lip and palate: parts 1-5. *Cleft Palate-Craniofacial Journal*, 29, 393-418.
- Sphrintzen, R.J. & Bardach, J. (1995). *Cleft Palate Speech Management: A Multidisciplinary Approach*. St. Louis, MO: Mosby.
- U.S. Department of Health and Human Services. *Oral Health in America: A Report of the Surgeon General*.

- Rockville, MD: U.S. Department of Health and Human Services, National Institute of Dental and Craniofacial Research, National Institutes of Health, 2000.
- Wyszynski D. (2002). *Cleft Lip and Palate: From Origin to Treatment*. New York: Oxford University Press.

PARENT GUIDES AND PAMPHLETS

- Cleft Palate Foundation. *Cleft Lip and Cleft Palate: The First Four Years*, 2001. (Also available in Spanish).
- Please note that the English version of this booklet is no longer available by mail. It is in the process of being split into two separate publications, *Your Baby's First Year* and *Toddlers and Preschoolers* which will be available online and by mail.
- Cleft Palate Foundation. *Cleft Lip and Palate: The School-Aged Child*, 2008. Available to download at www.cleftline.org/docx/Booklets/SAC-01.pdf or can be purchased from the foundation. (Also available in Spanish at www.cleftline.org/docs/Booklets/SAC-02.pdf.)
- Cleft Palate Foundation. *As You Get Older: Information for Teens Born with Cleft Lip and Palate*, 2008. www.cleftline.org/docs/Booklets/TEN-01.pdf.
- Cleft Palate Foundation. *Feeding an Infant with a Cleft*, 2009. Available to download at www.cleftline.org/docs/Booklets/FDG-01.pdf. (Also available in Spanish at www.cleftline.org/docs/Booklets/FDG-02.pdf.)
- Cleft Palate Foundation. *Genetics and You*, 2008. www.cleftline.org/docs/Booklets/GEN-01.pdf.
- Moller, K.T., Star, C.D. (1990). *A Parent's Guide to Cleft Lip Palate*. Minneapolis: University of Minnesota Press.
- Nackashi, J., et al. (2000). *Looking Forward: A Guide for Parents of the Child with Cleft Lip and Palate*. Evansville, IN: Mead Johnson.

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)

www.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

www.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

www.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

www.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/medical-conditions/chromosomal-genetic-conditions/cleft-lip-palate

Friendly Faces

www.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

www.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.

Let's Face It

www.nas.com/~letsfaceit/

Lists booklets, websites, and other resources for a wide variety of facial differences.

Parent-to-Parent

www.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

www.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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Center for Children
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