3. Comprehensive Cleft Care

This chapter is largely based on the work entitled, "Parameters for the Evaluation and Treatment of Patients with Cleft Lip/ Palate or Other Craniofacial Anomalies," produced by the American Cleft Palate-Craniofacial Association and originally published in the Cleft Palate-Craniofacial Journal. This document can be found and downloaded from the ACPA website: www.acpa-cpf.org

TEAM CARE

Children born with cleft lip, cleft palate, or other deformities require comprehensive, coordinated care provided by health care providers that are readily accessible and responsive to the individual needs of the patients and their families.

This multidisciplinary care should include:

- An interdisciplinary approach to care
- Facilitation of parent/professional collaboration in the care of children
- Sharing of unbiased and complete information with patients and parents
- Sensitivity to cultural differences
- Encouragement of parent-to-parent support
- Incorporation of the developmental needs of infants, children, and adolescents into health care plans
- Assurance of the availability of comprehensive services including social, emotional, and cognitive aspects of health care
- Encouragement of community-based services
- Establishment of protocols to assess quality of care

FUNDAMENTAL PRINCIPLES

- Management of patients with craniofacial anomalies is best provided by an interdisciplinary team of specialists.
- Optimal care for patients with craniofacial anomalies is provided by teams that see sufficient numbers of patients each year to maintain clinical expertise in diagnosis and treatment.
- The optimal time for the first evaluation is within the first few weeks

of life and, whenever possible, within the first few days. However, referral for team evaluation and management is appropriate for patients at any age.

- From the time of the first contact with the child and family, every effort must be made to assist the family in adjusting to the birth of a child with a craniofacial anomaly and to the consequent demands and stress placed upon the family.
- Parents/caregivers must be given information about recommended treatment procedures, options, risk factors, benefits, and costs to assist them in (a) making informed decisions on the child's behalf, and (b) preparing the child and themselves for all recommended procedures. The team should actively solicit family participation and collaboration in treatment planning. When the child is mature enough to do so, he or she should participate in treatment decisions.
- Treatment plans should be developed and implemented on the basis of team recommendations.
- Care should be coordinated by the team but should be provided at the local level whenever possible; however, complex diagnostic and surgical procedures should be restricted to major centers with the appropriate facilities and experienced care providers.
- It is the responsibility of each team to be sensitive to linguistic, cultural, ethnic, psychosocial, economic, and physical factors that affect the dynamic relationship between the team and the patient and family.
- It is the responsibility of each team to monitor both short-term and long-term outcomes. Thus, longitudinal follow-up of patients, including appropriate documentation and record-keeping, is essential.
- Evaluation of treatment outcomes must take into account the satisfaction and psychosocial well-being of the patient as well as effects on growth, function, and appearance.
- The quality of care for patients with craniofacial anomalies and related disorders must be carefully monitored by the team providing diagnosis, treatment planning and treatment services. This requires (1) longitudinal assessments of the outcomes of treatment, (2) periodic team review of the clinical outcome data, and (3) team

adaptation of treatment procedures when clinical outcome assessments do not reach referenced criteria.

Interdisciplinary Cleft Care Team

- Cleft surgeon(s)
- Anesthesiologist(s)
- Pediatrician(s)
- Clinical Coordinator
- Operating Room Nurses
- Recovery Room Nurses
- Ward Nurses
- Nutrition Team
- Audiology
- Otolaryngology
- Dentist
- Orthodontist
- Medical Records Specialist
- Photo Imaging Technician
- Biomedical Technician
- Speech Therapist
- Child Life Specialist
- Patient Mobilization
- Patient Care Services
- Genetics

TEAM RESPONSIBILITIES

- Optimization of care requires regular team evaluations for assessment of treatment outcomes and updating of treatment plans. Continuity of care enhances the evaluation and treatment process for patients of any age.
- Integrated case management to assure quality and continuity of patient care and longitudinal follow-up.
- Each patient requires comprehensive, interdisciplinary treatment planning to achieve maximum habilitation with efficient use of parent and patient time and resources.
- Maintain centralized and comprehensive records on each patient, including histories, diagnoses, reports of evaluations, treatment plans, reports of treatment, and supporting documentation such as photographs, radiographs, dental models, and audiotaped speech recordings.
- Evaluate patients at regularly scheduled intervals, the frequency and specific content of those evaluations being determined by the condition and needs of the patient and family.
- Hold regularly scheduled face-to-face meetings for discussion of findings, treatment planning, and recommendations for each patient.

- Develop a longitudinal treatment plan for each patient that is modified as necessitated by craniofacial growth and development, treatment outcomes, and therapeutic advances.
- Weigh all treatment decisions against the expected outcomes and related factors such as facial growth, hearing, speech, dentition, and psychosocial impact on patient and family.
- Communicate the treatment recommendations to each patient and family in written form as well as in face-to-face discussion.
- Demonstrate sensitivity and flexibility in provision of care to accommodate linguistic, cultural, and ethnic diversity among patients and their families, ensuring that appropriate interpreters are available to assist in both verbal and written communication.
- Assist families in locating resources for financial assistance necessary to meet the needs of each patient.
- Communicate on a routine and ongoing basis with direct care.

CLEFT LIP AND CLEFT PALATE SURGERY

- In addition to primary surgical closure of the lip and palate, many patients will require secondary surgical procedures involving the lip, nose, palate, and jaws that usually are staged from infancy through adulthood. These procedures usually are staged over a period of several years.
- In all cases, surgical techniques should be individualized according to the needs and condition of the patient. Surgical procedures should be coordinated to minimize the number of anesthetic exposures and hospitalizations.
- Evaluation of complications (morbidity and mortality) of cleft lip and palate repairs should be completed on an annual basis and subjected to peer review.
- The major factor in the quality of surgical outcome is the skill, training, and experience of the cleft care team.

SURGICAL TREATMENT OF A CLEFT LIP AND AND PALATE BY AGE

AGE	TREATMENT	CLEFT TEAM MEMBERS
Prenatal	Prenatal imaging, diagnosis, and couseling.	Multidisciplinary
Newborn	Feeding assessment, medical assessment, genetic counseling, treatment information.	Multidisciplinary
0-3 months	Presurgical orthopedics.	Orthodontist, plastic surgeon.
3 months(or after presurgical orthopedics).	Primary cleft lip repair and tip rhinoplasty ± gingivoperiosteo-plasty.	Plastic surgeon.
12 months (delayed if airway or medical concerns)[a]	Primary cleft palate repair with intraveral veloplasty ± bilateral myringotomy and tubes .	Plastic surgeon, otolaryngologist.
Diagnosis of velopharyngeal insufficiency (3-4 years)	Secondary palate lengthening or pharyngoplasty, speech obturator.	Speech pathologist, plastic surgeon, otolaryngologist, orthodontist.
School-age years	Treatment of secondary lip and nasal deformities.	Plastic surgeon.
7-9 years (mixed dentition)[b]	Secondary alveolar bone graft.	Orthodontist, plastic surgeon, oral surgeon.
Postalveolar grafts[a]	Presurgical orthodontics.	Orthodontist.
Puberty	Definitive open rhinoplasty.	Plastic surgeon.
Skeletal maturity	LeFort I ± mandible orthognathic surgery	Plastic surgeon, oral surgeon.

Table 3-1. Timeline of Surgical Tretment for Cleft Lip and Cleft Palate.

Primary Cleft/Lip Palate Surgery:

- An anesthesiologist knowledgeable and experienced in pediatric anesthesia must be present for all surgical procedures involving children.
- Surgical repair of the cleft lip is usually initiated within the first 12 months of life and may be performed as early as is considered safe for the infant.
- Pre-surgical maxillary orthopedics to improve the position of the maxillary alveolar segments prior to surgical closure of the lip may be indicated for some infants.
- The nasal deformity is an integral part of the cleft lip. Primary nasal repair may be done at the time of the primary lip repair.
- The goal of lip repair is to restore the normal functional and anatomic features.
- In the normally-developing child, the palate should be closed by the age of 18 months and preferably earlier.
- The goal of palate surgery is to achieve normal function. Repair of the soft palate should involve levator veli palatini muscle reconstruction.
- Patients with submucous clefts should be monitored closely and their palates repaired only if there is evidence of feeding, otologic, or speech problems.

Secondary Cleft/Lip Palate Surgery:

- Although rhinoplasty and nasal septal surgery are usually advocated only after completion of nasal growth, earlier intervention for reasons of airway problems or nasal tip deformity may be indicated.
- The timing of nasal surgery should be discussed with the patient and parents so that the goals are understood and expectations are realistic.
- The patency of the nasal airway should be considered when planning either nasal reconstructive procedures or secondary velopharyngeal operations such as a pharyngeal flap or other type of pharyngoplasty.
- Secondary palatal and pharyngeal surgery for velopharyngeal inadequacy should be performed only after evaluation of the velo-

pharyngeal mechanism and review by the team.

- Surgical or prosthetic closure of palatal fistulae may be needed if the fistulae are symptomatic.
- Secondary cleft lip surgery is sometimes needed to optimize aesthetic results.
- Tonsillectomy and/or adenoidectomy may be indicated to permit safe performance of a pharyngeal flap or other type of pharyngoplasty. In some instances, enlarged tonsils may interfere with velopharyngeal function, and a tonsillectomy may be indicated.
- The timing of bone grafting of the alveolar cleft should be determined by the stage of dental development. The graft should be placed before the eruption of the permanent maxillary teeth in the region of the cleft, although in late-presenting patients bone grafting may necessarily take place after full eruption of the permanent teeth. The timing of the operative procedure should be determined in collaboration with the orthodontist. Autogenous bone should be used when tooth movement through the graft is anticipated. The occlusion should be stabilized before materials that have potential to impede tooth movement are used.
- Orthognathic surgery (and/or distraction osteogenesis) is indicated when orthodontic treatment cannot achieve functional and/or acceptable esthetic occlusion and facial harmony. Such surgery should be timed to minimize any adverse effect on possible subsequent growth, and the timing should be determined in consultation with the team. Whenever possible, orthognathic surgery should be delayed until physical maturation is essentially completed. Earlier surgery may be indicated when there are serious concerns regarding a compromised air way, jaw function, speech, or psychosocial adjustment. However, the patient and family must understand that additional procedures may be required to optimize the outcome.

DENTAL AND ORTHODONTIC CARE

Dentistry

- Patients with craniofacial anomalies require dental services as a direct result of the medical condition and as an integral part of the rehabilitative process. Due to the often complex nature, patients

are generally best treated by referral to a pediatric dentist.

- Provision of dental services for these patients includes not only primary care but routine maintenance throughout life. Dental care should occur within six months of the eruption of the first tooth and no later than twelve months of age and continues throughout life.
- Includes dental examinations, caries control, and preventive, restorative, and prosthetic dental treatment as needed. Patients should be closely monitored for periodontal disease, anomalies of the dentition and disturbed eruption. Prosthetic appliances in the form of an obturator to close fistulae or a speech appliance for velopharyngeal insufficiency may be indicated in some situations.

Orthodontic Services

- Patients with craniofacial anomalies require orthodontic services as a direct result of the medical condition and as an integral part of the habilitative process. Orthodontic care may include primary orthopedic treatment in infancy and can extend through adulthood.
- Treatment often takes place in phases which may include treatment in infancy, the primary dentition, the transitional dentition, and the permanent dentition. The skeletal and dental components should therefore be regularly evaluated to see if a malocclusion is present or developing.
- When indicated, orthodontic treatment prepares a patient for alveolar bone grafting of the cleft maxilla, for correcting malocclusions and for preparation of jaw surgery.
- Diagnosis and treatment planning requires a variety of diagnostic records as well as clinical examination. Records are obtained in a serial fashion to monitor dentofacial growth and development as well as the results of ongoing treatment.
- Dental films, cephalometric radiographs, photographs and computer imaging as indicated should be utilized to evaluate and monitor dental and facial growth and development
- For patients at risk for developing malocclusion or maxillary-mandibular discrepancy, diagnostic records including properly occluded dental study models should be collected at appropriate intervals.

- As the primary dentition erupts, each team evaluation should include a dental examination and referral to appropriate providers for caries control, preventive measures and restorative dental treatment, and space management if such services are not being provided.
- Before the primary dentition is completed, the skeletal and dental components should be evaluated to determine if a malocclusion is present or developing.
- Depending upon the goals to be accomplished, and also upon the age at which the patient is first seen, orthodontic management of malocclusion may be performed in the primary, mixed or permanent dentition. In some cases, orthodontic treatment may be necessary in all three stages.
- While continuous active orthodontic treatment from early mixed dentition to permanent dentition should be avoided, each stage of therapy may be followed by orthodontic retention and regular observation; the orthodontic retention period for the permanent dentition may extend into adulthood.
- For some patients with craniofacial anomalies, functional orthodontic appliances may be indicated.
- For patients with craniofacial anomalies, orthodontic treatment may be needed in conjunction with surgical correction (and/or distraction osteogenesis) of the facial deformity.
- Congenitally missing teeth may be replaced with a removable appliance, fixed restorative bridgework, or osseointegrated implants.
- Patients should be closely monitored for periodontal disease.
- Prosthetic obturation of palatal fistulae may be necessary in some patients.
- A prosthetic speech appliance may be used to treat velopharyngeal inadequacy in some patients.
- The concept of modern presurgical infant orthopedics uses an oral prosthesis similar to an obturator to approximate the cleft alveolar segments. A maxillary impression is taken of the newborn and an acrylic appliance is made from a plaster model that was cut and modified with the cleft gap slightly closed. By repeating this step and frequently modifying the appliance, orthodontists are able to

close not only the alveolar gap, but also the hard palatal cleft by influencing bone growth direction.

- Active appliances use a hard acrylic plate and controlled forces, sometimes from extraoral traction (bonnet with straps), to move the maxillary alveolar segments into approximation.
- The Lathum appliance is an active pinretained appliance designed to exert a forward force to the lesser posterior segment of the unilateral cleft maxilla. It consists of a two-piece maxillary splint that overlies the palatal shelves and is retained by short medial pins. An expansion screw connecting the two pieces can be moved to adjust the widths of the lateral palatal segments. An orthodontic elastic chain is used to retract the premaxilla. By adjustment of these independent controls, the premaxilla is brought back into its proper position in the arch before the primary repair. The Latham device requires a surgical procedure to introduce the device and another to remove it.
- Passive appliances generally consist of an alveolar molding plate made of a hard outer shell and a soft acrylic lining.
- By gradual alteration of the tissue surface of the acrylic plate, the alveolar segments are gently molded into the desired shape and position by direction of alveolar growth. The devices allow continued growth by a passive molding action without permitting medial movement of the buccal segments. Once the segments are in proper position, early lip repair and bone grafting could be performed.
- Presurgical nasal and alveolar molding (NAM) includes as its objectives the active molding and repositioning of the nasal cartilages and alveolar processes, and lengthening of the deficient columella. This modification of the traditional approach to presurgical molding plate therapy takes advantage of the plasticity of cartilage in the newborn infant during the first 6 weeks after birth. This technique has been demonstrated to have a positive influence on the outcome of the primary nasal, labial, and alveolar repair, and has been adopted by an increasing number of cleft teams.
- Shortly after birth, an impression of the intraoral cleft defect is made using an elastomeric material in an acrylic tray. A conventional molding plate is constructed on the maxillary study model

from clear orthodontic resin. The molding plate is applied to the palate and alveolar processes, and secured through the use of surgical adhesive tapes applied externally to the cheeks and to an extension from the oral plate that exits the horizontal labial fissure. The molding plate is modified at weekly intervals to gradually approximate the alveolar segments. This is achieved through the selective removal of acrylic from the region into which one desires the alveolar bone to grow ("negative sculpting"). At the same time, soft denture liner is added to line the appliance in the region from which one desires the bone to be moved. The ultimate goal of this sequential addition and selective removal of material from the inner walls of the molding plate is to align the alveolar segments and achieve closure of the alveolar gap. With NAM, alignment and approximation of the alveolar segments is controlled to create a natural arch form. The effectiveness of the molding plate is enhanced by adequately supporting the appliance against the palatal tissues, and by taping the left and right lip segments together between clinical visits.

- The nasal changes of NAM are achieved by the use of a nasal stent rising from the labial vestibular flange of the acrylic intraoral molding plate. The medio-lateral position of the nasal stent is adjusted as it lifts the nasal tip. The shape of the nostrils and alar rims is carefully molded to resemble the normal configuration of these structures through modifications gradually made to the nasal stents.

AUDIOLOGIC CARE

- Individuals with craniofacial anomalies may have congenital abnormalities of the auditory structures and are also subject to an increased incidence of ear disease.
- These children are at high risk for hearing disorders that may occur intermittently or become permanent, and that vary from mild to severe.
- Hearing loss can have a significantly adverse influence on speech and language development, educational and psychological status, and eventually on social and vocational status.
- Children with craniofacial anomalies require ongoing audiologic

surveillance.

- Each child should have an appropriate assessment of hearing sensitivity for each ear within the first three months of age.
- The timing of audiological follow-up examinations should be determined on the basis of the child's history of ear disease or hearing loss. Audiological follow-up examinations should continue through adolescence.
- Acoustic-immittance (tympanometric) measures should be obtained as a part of each audiological evaluation to monitor middle ear status.
- All children undergoing myringotomies and placement of ventilating tubes should be seen pre- and postoperatively for audiologic assessment.
- When a persistent hearing loss is identified, amplification (hearing aids, auditory training systems) should be considered.
- When hearing loss occurs in the presence of microtia or atresia, whether unilateral or bilateral, bone conduction amplification should be considered depending upon degree of loss; an implantable bone conduction aid may be a treatment option.
- Once amplification has been provided, a regular follow-up schedule is needed to monitor hearing thresholds and the function of the amplification system.
- For any child with a documented hearing loss, referral should be made to the child's school district for appropriate educational services as soon as the hearing loss is identified.
- In the absence of a positive history of otologic disease or hearing loss, audiologic examination or screening should still be carried out at least yearly through the age of six years to assure adequate monitoring of hearing.

GENETIC / DYSMORPHOLOGY SERVICES

- A comprehensive clinical genetic evaluation is a key component in the management of patients with congenital craniofacial anomalies and should include (1) diagnosis, (2) recurrence risk counseling, and (3) counseling regarding prognosis. Complex syndromes involving craniofacial anomalies may not fully express clinical

manifestations that can be recognized in the first year of life. Thus, genetic screening and follow-up evaluations will be necessary for some patients until puberty. Patients who are first seen by the team at later ages should also be evaluated.

- Prenatal ultrasound will often detect cleft lip with or without cleft palate. Isolated cleft palate is rarely identified by ultrasound. A timely referral to specialists experienced in diagnosis, management and treatment of children with these and related conditions should be made.
- Indications for referral for a complete genetic evaluation include, but are not limited to, (a) positive family history; (b) prenatal growth deficiency; (c) unexplained postnatal growth deficiency; (d) developmental delay or mental retardation; (e) associated major malformations and/or disorders; (f) associated minor malformations and/or disorders inconsistent with the genetic background; (g) family request; and (h) recognized genetic diagnosis.
- For families who live in areas which are geographically remote, the team should arrange for referral to an accessible source where genetic services can be provided, and should encourage the family to follow through on the referral.
- Personal genetic counseling should be offered to the maturing adolescent.

NURSING CARE

- Complex interdisciplinary management for individuals with congenital craniofacial anomalies requires a high level of ongoing coordination of services. The patient and family need appropriate information to understand the rationale for, and implications of, such management.
- Nursing assessment, interventions, and ongoing follow-up evaluations are integral to the long-term care needs of the child or individual with congenital craniofacial anomalies and family.
- Nurses may provide feeding assessments, interventional teaching, and follow-up of nutritional and growth assessments while serving as role models of acceptance and nurturance for the patient and family.

- Nurses prepare of patients and families for what to expect when either in-patient or out-patient surgical procedures are scheduled. Information should be provided on pre- and postoperative feeding, special positioning and equipment, wound care, skin management, activity restriction, anticipated outcome or general status immediately following surgery, and the necessity for, and timing of, postoperative examinations. Instructions must be given in a manner that assures understanding on the part of the family and other caregivers.
- Specialized clinical nurses are an integral to the optimal care in the operating room, post anesthesia care unit, and ward environments.

OTOLARYNGOLOGY

- Comprehensive care of children with craniofacial anomalies typically requires long-term monitoring of the ears, nose, and throat due to the prevalence of ear disease, ear malformations, and upper airway problems.
- Physical examination of the ears should be provided on a regular basis, beginning within the first six months of life and continue through adolescence.
- Treatment of middle ear disease may include use of antibiotics, myringotomies with insertion of tympanotomy tubes, tympanoplasties, gel foam patches, removal of cholesteatomas, mastoidectomies, and middle ear reconstruction.
- Any child with middle ear disease should be carefully followed during and after treatment to ensure a successful outcome and to be sure that tympanotomy tubes are removed if necessary.
- Although some otolaryngologic operative procedures such as myringotomies may be classified as minor procedures, the potential psychological impact on the child and family attendant upon any surgical procedure must be taken into consideration, particularly when the child is subjected to repeated procedures. All procedures should be preceded by appropriate exchange of information with the patient and family, and every attempt made to allay fears regarding effects of anesthesia, the immediate post-operative course, post-operative precautions, etc.

- Sequential airway assessments may be required to differentiate anatomical causes of airway difficulties from other causes, e.g., allergies. Such assessments may require endoscopy, radiologic studies, airflow studies, CT scans, MRI's, and polysomnography.
- Structural and functional laryngeal problems may exist in these patients, and may require medical as well as surgical treatment.
- An adenoidectomy and/or tonsillectomy as well as other oropharyngeal (tongue reductions, mandibular distraction, etc.) And laryngotracheal procedures may be indicated if the child is experiencing sleep apnea or other signs of airway obstruction. However, the team should provide a speech evaluation and assessment of the velopharyngeal mechanism prior to any decision to perform such a procedure.

PEDIATRIC CARE

- Pediatric care provided within the context of the team is fundamental in assuring that the health needs of the child with craniofacial anomalies are fully identified and appropriately treated.
- A primary care physician should be identified for each child, and should become an extended member of the team.
- Physical examinations should be provided on a regular basis within the context of the team.
- Parental questions regarding health issues should be addressed.
- Parental understanding of the child's health needs and of the team treatment plan should be monitored.
- When special needs are identified, referral to appropriate specialists should be made in cooperation with the primary physician. Family follow-through on these referrals should be monitored.
- Frequent monitoring is required for children who may be at risk for growth failure, delayed development, abuse and neglect, or any other significant problem.
- The child's health status should be evaluated prior to any surgical procedures that are planned.

PSYCHOLOGICAL AND SOCIAL SERVICES

- The accomplishment of the goals of treatment of the patient with craniofacial anomalies requires periodic assessment of the psychosocial needs of both the patient and the family. The psychosocial interviewer may come from the ranks of professionals such as those in social work, psychology, pediatrics, nursing, and psychiatry. However, psychological tests must be administered and interpreted under the supervision of a licensed psychologist, preferably a person familiar with craniofacial anomalies and related speech and hearing disorders.
- Psychosocial screening interviews should be conducted periodically to assess parental competence and nurturance, child management skills, parent-child relationships, and the emotional and behavioral adjustment of the child. The high rate of learning disorders in children with craniofacial conditions requires that each child be screened for potential learning disorders beginning in infancy until cognitive competence has been established. Each family should receive or be referred for psychological evaluation, and, as appropriate, intervention when possible problems are identified through the screening process.
- Parents should receive guidance regarding such problems as behavior management, teasing, rejection by other family members, public attitudes, fear of and expectations from surgical procedures, and emotional adaptation to treatment.
- Screening evaluations should be conducted to identify possible problems in the child's cognitive development, behavior, self concept, educational progress, and psychosocial development. The screening evaluations should be conducted periodically, beginning in infancy and continuing through adolescence. When problems are suspected or identified in these areas, the child should receive or be referred for formal developmental/cognitive evaluations, guidance, counseling, or other assistance as needed.
- When there is concern for possible developmental delay, repeated psychological evaluations are necessary to facilitate appropriate program planning and prevent inappropriate labeling of the child.
- Information about learning performance, through liaison with

schools, should be obtained periodically. When problems are suspected or identified, arrangements should be made for cognitive and educational assessments.

- Children who have craniofacial anomalies may benefit from contacts with other children who have similar conditions. The team should facilitate contact with other children and their families through support groups and networking.
- Social skills training programs should be available to help children and adolescents learn how to handle stressful social situations.
- As they mature, children should be given information about their craniofacial anomaly, and should be permitted and encouraged to become active participants in treatment planning. All care providers should be sensitive to how treatment discussions can be perceived by children, and should do everything possible to ensure that the child understands the treatment plan as much as possible. Towards this end, it is helpful for the team to ensure that each child has someone who will listen to his/her fears, concerns, and opinions regarding treatment.
- Referrals for vocational training and guidance should be provided for adolescents, as appropriate.

SPEECH-LANGUAGE PATHOLOGY

- Children who have craniofacial anomalies are at high risk for speech-language disorders. Evaluation of speech and language development provides information that is needed by the team in planning of treatment, particularly surgical and dental management. Further, information about the patient's speech and language is important in the assessment of the outcome of treatment. Speech-language evaluations should occur often enough to assure adequate documentation of each child's progress and to develop appropriate recommendations for intervention.
- Each child and family should be seen for discussion of normal speech and language development and assessment of pre-linguistic speech-language development before or by six months of age.
- When the development of speech and language skills is not at an

age-appropriate or developmentally appropriate level or when early speech productions are deviant, arrangements should be made for an early speech-language intervention program to facilitate speech sound development, provide language stimulation, and establish a home program to be carried out by parents and other care providers.

- With the possible exception of children with cleft lip only, speech-language evaluations with appropriate documentation should be conducted for each child at least twice during the first two years of life and at least annually thereafter until the age of six years.
- After the age of six years, even if speech-language development has been appropriate and no problems are noted, screenings should take place annually until after adenoid involution, and at least every two years thereafter until dental and skeletal maturity are reached.
- For children with problems in speech and language development and for those in whom the basis for continuing velopharyngeal inadequacy (VPI) cannot be definitively determined, re-evaluations should take place as often as deemed necessary by members of the interdisciplinary team in consultation with local care providers.
- Speech evaluations, conducted as part of a team visit should always include perceptual assessment of speech articulation, speech resonance, and voice.
- If articulation is deviant and characterized by maladaptive compensatory misarticulation errors associated with cleft palate, speech therapy may result in positive changes in velopharyngeal closure.
- Speech evaluations are necessary pre- and post-treatment to determine candidacy for, and outcomes of, surgical, behavioral and/or prosthetic management of the velopharyngeal system
- For patients with craniofacial anomalies who are candidates for orthognathic surgery, pre- and postoperative perceptual speech evaluations are necessary.
- Instrumental assessment of velopharyngeal function is required for all patients with resonance disorders, audible nasal air emission, or both. Procedures may include multiview videofluoroscopy, nasopharyngoscopy, and, at times, aerodynamic measures and na-

sometric studies, all of which should be conducted by or with the participation of the team speech-language pathologist.

- When voice is deviant, direct imaging of the larynx is necessary prior to initiating voice therapy.
- Remedial speech-language therapy will be needed for some patients with craniofacial anomalies.
- Biofeedback therapy may be useful in treating some patients with velopharyngeal dysfunction.
- Blowing exercises and oral-motor therapy are not useful in treating velopharyngeal dysfunction.

PATIENT MOBILIZATION

- Patients are the foundation of any cleft care team. A central step in provision of care is to make patients aware that opportunities for treatment exist and to facilitate access.
- This seemingly straightforward notion becomes exponentially more complex in developing regions and with a population that is concentrated in areas far away from billboards, televisions, newspapers, or other forms of public communication.
- In more rural and isolated regions it becomes exceedingly challenging to identify patients with cleft lip and cleft palate and to educate them about the treatment process.
- Differences in religion, ethnicity, language, and socioeconomic level can contribute to mistrust by patients.
- An ongoing cleft care team requires a robust patient recruitment team responsible for connecting people in need of surgical care to the cleft team for treatment.
- A manager of develops relationships and systems for increasing patient awareness and access, and a patient mobilization coordinator is tasked with strategy and implementation of recruitment efforts.
- A team of patient counselors is effective in rural areas to identify cleft patients and provide patient education on treatment of cleft lip and cleft palate, and services available.
- Understanding and utilizing existing infrastructure is essential to increase penetration into the rural regions. This includes the ad-

ministrative system of the local government, education systems, healthcare systems, professional and social affiliations. Engagement with officials at all levels helps to identify cleft patients in their individual areas.

- Community health activists throughout the region my be educated in large workshops on cleft lip and cleft palate, and dispersed into their villages to create awareness and mobilize their communities to identify patients.
- Local prescreening camps may be organized. Health activists bring patients from their communities to these camps, where the patient counselors assist patients with the process of accessing care.
- Once patients are identified, they are registered and provided with plans for travel to the cleft team for medical evaluation and treatment.
- The goal is to make this process as direct and simple as possible, so that patients may break through the barriers to obtaining care.

PATIENT CARE SERVICES

- In developing regions with very poor populations, a patient care team is very helpful to coordinate clinical services and hospitality to continuously large numbers of needy patients coming from distant regions.
- This team assumes responsibility for patients and their families upon arrival to the center, coordinating food, hygiene, and shelter.
- The patient care members coordinate extensive logistics needed for all patients to receive multidisciplinary clinical evaluations, and to synchronize investigations and care plans between professionals.
- Once scheduled for surgery, patients and attendants are admitted to the hospital and provided with amenities for a comfortable stay.
- The patient care team members also serve as liaisons between providers and patients to make the treatment process as straightforward, clear, and enjoyable as possible.
- Families in very poor regions face intense challenges to travel for surgery, including leaving necessary daily wages and additional children, and so every effort was made to screen patients and pro-

vide surgical operations within a single trip.
- After surgery, the patient care team assists patients with transportation back to their homes, and provides instructions and coordination for postoperative follow-up appointments.

NUTRITION SERVICES

- Poverty, poor maternal diet, and inadequate infant and young child feeding contribute to malnutrition in the state, and cleft children are especially vulnerable. Impaired ability to effectively breastfeed frequently causes life-threatening malnutrition in this patient population, as maternal milk is the principal source of nourishment.
- Previous studies have additionally suggested that malnourished children are at higher risk for surgical complications. A nutritional assessment by the Operation Smile in Northeast India team found that among children with cleft lip and cleft palate in Assam, 64% were malnourished by at least one standard anthropometric indicator, and 78% were anemic.
- A nutrition team is tasked with developing systems and protocols to identify malnourished patients, provide education and treatment, and monitor progress.
- Physicians, nurses, nutritionists, and social workers collaborate to rescue these children, with the goal of eliminating the malnutrition and preparing them for successful operations.
- Special protocols identify malnourished patients during screening and medical assessment by a pediatrician.
- Severely malnourished patients, as defined by World Health Organization standards, should be admitted to an inpatient malnutrition unit for immediate treatment.
- Patients with mild or moderate nutrition are referred to a patient manager to register them in the nutrition program and complete specialized forms to record demographic information, anthropometric measurements, medical and growth history, and feeding information.
- The treatment plan is prescribed by the pediatrician, including necessary laboratory work, medical therapy, and food/vitamin supplementation.

- Nurse educators specifically trained in nutrition conduct intensive inpatient education clinics with the patients and parents. Culturally appropriate counseling and education stressed proper feeding methods and diet to maximize parental understanding of the problem and necessary intervention.
- Once parents demonstrate the ability to provide adequate interventions at home, the patients are released and entered in a nutrition database. Regular clinic visits track progress and additional interventions are provided as necessary.
- Once patients are adequately nourished and candidates for surgery, they are referred to the surgeons for evaluation and scheduling.

PATIENT EDUCATION

- Simple and direct communication with patients in their native language is essential for quality surgical care.
- Providing proper patient education and discharge instructions in a postoperative setting has been associated with many benefits including decreased readmissions, decreased visits to other healthcare providers, improved postoperative pain control, decreased healthcare costs, and improved overall patient satisfaction.
- Significant challenges to effective patient-provider interactions include language, culture, and socioeconomic levels.
- This can be improved through implementation of standardized postoperative protocols and a culturally focused patient education program. Postoperative care is standardized among all surgeons and pediatricians, and the nurses are provided with specific education and training regarding postoperative care protocols.
- Nurses lead a targeted education program that includes individual and group teaching to patients and parents. They distribute discharge materials including pictographs and written in the local language and may show instructional videos on the postoperative ward.
- Patient understanding and outcomes improved remarkably, research has proven the utility of these programs in markedly decreasing postoperative complication rates

PATIENT FOLLOW UP

- Social, economic, and geographical challenges create intense challenges to patient travel to and from the cleft team in a developing regions, leading to traditionally low follow-up rates in these areas.
- Patients are often day laborers and cannot afford to take time off of work or pay the cost of transportation, which is often cumbersome and time-consuming.
- Also, postoperative patients with good outcomes often do not understand the need for follow-up visits, as they perceive no complication and therefore no need for physician intervention.
- Reliable communication is also very challenging and often difficult to maintain because of the sometimes transient nature of local telephone numbers.
- An extensive amount of organization is needed to effectively follow the patients and the goal of follow-up programs is to implement protocols that would lead to successful postoperative evaluations for as many patients as possible.
- Ultimately, this improves care through clinical examinations and advice to patients, scheduling future surgeries, and evaluating the quality of repairs being performed.
- Components of a successful follow up program may include block scheduling of follow-up appointments, improved discharge teaching, clear patient discharge records, repeated telephone reminders, and provision of transport costs to parents. Complimentary food and shelter may be considered for patients needing an overnight stay because of distance or timing of transportation.
- Patients returning to follow-up should be evaluated in a multidisciplinary fashion, and scheduled for future examinations, therapy, and procedures as indicated.
- Patients who are “no-shows” should be called weekly to provide a future appointment.
- District based outreach follow-up and speech therapy programs are innovative strategies to take plastic surgeons, speech therapists, and patient counselors back into the districts to improve access to services.

KEY READING

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