Orofacial Clefts

Goals & Objectives

Course Description
"Orofacial Clefts" is an online continuing education course for occupational therapists and occupational therapist assistants that presents current information concerning congenital orofacial clefts including epidemiology, embryology, etiology, classification, associated syndromes, diagnosis, and interdisciplinary care.

Course Rationale
The purpose of this course is to present contemporary information regarding orofacial clefts to therapists and assistants so that they may have an improved understanding of the condition and provide optimal care to individuals affected by this disorder.

Goals & Objectives
At the end of this course, the participants will be able to:
1. define the epidemiological occurrence and societal impact of orofacial clefts
2. identify the embryological sequence that results in orofacial clefts
3. identify etiology and risk factors for orofacial clefts
4. describe orofacial clefts utilizing accepted classification systems
5. identify syndromes that have associated orofacial clefts
6. recognize current mechanisms used to diagnosis orofacial clefts
7. list and define the roles of cleft palate and craniofacial teams
8. define the standards of care and treatment goals for individuals with orofacial clefts
9. identify common feeding issues associated with orofacial clefts
10. differentiate between the various surgical techniques used to repair orofacial clefts
11. recognize the psychosocial issues experienced by individuals with an orofacial cleft
12. identify audiology issues and interventions associated with orofacial clefts
13. identify speech-language issues and interventions associated with orofacial clefts
14. identify dental issues and interventions associated with orofacial clefts

Course Provider – Innovative Educational Services

Course Instructor - Michael Niss, DPT

Target Audience - Occupational therapists and occupational therapist assistants

Course Educational Level - This course is applicable for introductory learners.

Course Prerequisites - None

Method of Instruction/Availability – Online text-based course available continuously.

Criteria for Issuance of CE Credits - A score of 70% or greater on the course post-test.

Continuing Education Credits - Three (3) hours of continuing education credit
AOTA - .3 AOTA CEU, Cat 1: Domain of OT – Client Factors, Context
NBCOT – 3.75 PDUs

Innovative Educational Services
To take the post-test for CE credit, go to: WWW.CHEAPCEUS.COM
**Course Outline**

<table>
<thead>
<tr>
<th>Topic</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Course Goals &amp; Objectives</strong></td>
<td>1</td>
</tr>
<tr>
<td><strong>Course Outline</strong></td>
<td>2</td>
</tr>
<tr>
<td><strong>Overview Orofacial Clefts</strong></td>
<td>3</td>
</tr>
<tr>
<td><strong>Epidemiology</strong></td>
<td>3</td>
</tr>
<tr>
<td><strong>Embryology</strong></td>
<td>3-4</td>
</tr>
<tr>
<td><strong>Etiology</strong></td>
<td>5-7</td>
</tr>
<tr>
<td><strong>Classification</strong></td>
<td>7-10</td>
</tr>
<tr>
<td>- Tessier Classification</td>
<td>7-8</td>
</tr>
<tr>
<td>- Van der Meulen Classification</td>
<td>8-9</td>
</tr>
<tr>
<td>- LAHSAL Classification</td>
<td>9-10</td>
</tr>
<tr>
<td>** Syndromes with Associated Orofacial Clefts**</td>
<td>10-14</td>
</tr>
<tr>
<td>- Van der Woude Syndrome</td>
<td>11</td>
</tr>
<tr>
<td>- Pierre Robin Sequence</td>
<td>11-12</td>
</tr>
<tr>
<td>- Treacher Collins Syndrome</td>
<td>12</td>
</tr>
<tr>
<td>- Apert Syndrome</td>
<td>12-13</td>
</tr>
<tr>
<td>- Crouzon Syndrome</td>
<td>13</td>
</tr>
<tr>
<td>- Stickler Syndrome</td>
<td>13-14</td>
</tr>
<tr>
<td><strong>Diagnosis</strong></td>
<td>14</td>
</tr>
<tr>
<td><strong>The Health Care Team</strong></td>
<td>14-16</td>
</tr>
<tr>
<td>- Cleft Palate Team</td>
<td>15</td>
</tr>
<tr>
<td>- Craniofacial Team</td>
<td>15-16</td>
</tr>
<tr>
<td><strong>Standards of Care</strong></td>
<td>16-19</td>
</tr>
<tr>
<td>- ACPA Parameters of Care</td>
<td>16-17</td>
</tr>
<tr>
<td>- Standards for Team Approval</td>
<td>17-19</td>
</tr>
<tr>
<td><strong>Treatment Goals</strong></td>
<td>19-20</td>
</tr>
<tr>
<td><strong>Nursing and Feeding</strong></td>
<td>20-22</td>
</tr>
<tr>
<td>- Feeding Issues</td>
<td>20</td>
</tr>
<tr>
<td>- Bottle Feeding</td>
<td>20-21</td>
</tr>
<tr>
<td>- Breast Feeding</td>
<td>21-22</td>
</tr>
<tr>
<td><strong>Nasoalveolar Molding (NAM)</strong></td>
<td>22</td>
</tr>
<tr>
<td><strong>Surgical Interventions</strong></td>
<td>22</td>
</tr>
<tr>
<td>- Schweckendiek’s Technique</td>
<td>22-23</td>
</tr>
<tr>
<td>- Von Langenbeck Palatoplasty</td>
<td>23-24</td>
</tr>
<tr>
<td>- Modified Von Langenbeck Palatoplasty</td>
<td>24</td>
</tr>
<tr>
<td>- The Furlow Z-plasty</td>
<td>24-25</td>
</tr>
<tr>
<td>- Palatal Lengthening – V-Y Pushback</td>
<td>25</td>
</tr>
<tr>
<td>- Alveolar Bone Grafting</td>
<td>25-26</td>
</tr>
<tr>
<td>- Orthognathic (Jaw) Surgery</td>
<td>26-27</td>
</tr>
<tr>
<td><strong>Psychosocial Issues</strong></td>
<td>27-28</td>
</tr>
<tr>
<td><strong>Otolaryngology and Audiology</strong></td>
<td>28-29</td>
</tr>
<tr>
<td><strong>Speech and Language</strong></td>
<td>29-34</td>
</tr>
<tr>
<td>- Communication Disorders</td>
<td>29-31</td>
</tr>
<tr>
<td>- Abnormal Nasal Resonance</td>
<td>31</td>
</tr>
<tr>
<td>- Velopharyngeal Insufficiency (VPI)</td>
<td>32-33</td>
</tr>
<tr>
<td>- Speech Therapy</td>
<td>33</td>
</tr>
<tr>
<td>- Timeline for Speech Therapy Intervention</td>
<td>34</td>
</tr>
<tr>
<td><strong>Dental and Orthodontic Care</strong></td>
<td>35-36</td>
</tr>
<tr>
<td>- The Deciduous Dentition Period</td>
<td>35</td>
</tr>
<tr>
<td>- Mixed Dentition Period</td>
<td>35-36</td>
</tr>
<tr>
<td>- The Permanent Dentition Period</td>
<td>36</td>
</tr>
<tr>
<td><strong>Timeline for All Health Care Interventions</strong></td>
<td>36-37</td>
</tr>
<tr>
<td><strong>Resources</strong></td>
<td>38</td>
</tr>
<tr>
<td><strong>References</strong></td>
<td>39-40</td>
</tr>
<tr>
<td><strong>Post-Test</strong></td>
<td>41-42</td>
</tr>
</tbody>
</table>
Overview Orofacial Clefts

Orofacial clefts are among the most common congenital malformations worldwide. Typically, they require complex multidisciplinary treatment throughout childhood and can have lifelong medical and psychosocial implications for affected individuals. The two main types of oral clefts are cleft lip and cleft palate. Cleft lip is the congenital failure of the maxillary and median nasal processes to fuse, forming a groove or fissure in the lip. Cleft palate is the congenital failure of the palate to fuse properly, forming a grooved depression or fissure in the roof of the mouth. Clefts of the lip and palate can occur individually, together, or in conjunction with other congenital malformations.

Epidemiology

Epidemiologic studies of isolated (i.e., without other malformations or syndromes) cleft lip and/or cleft palate have been conducted worldwide, often resulting in varying prevalence rates. Differences in geographic and ethnic distributions may account for some but not all of the variations. Other factors contributing to the diverse figures are the inclusion criteria used to group cleft types (i.e., CL ± P versus CP) or define the cleft population (i.e., all cases of cleft including other birth defects versus cases of isolated cleft).

Cleft of the lip, palate, or both is one of the most common congenital abnormalities. The average prevalence of cleft lip with or without cleft palate is 7.75 per 10,000 live births in the United States and 7.94 per 10,000 live births internationally. (Tanaka, 2012)

The Centers for Disease Control and Prevention (CDC) recently estimated that each year 2,651 babies in the United States are born with a cleft palate and 4,437 babies are born with a cleft lip with or without a cleft palate. Cleft lip is more common than cleft palate. About 70% of all orofacial clefts are isolated clefts.

Cleft lip with or without cleft palate is observed more frequently in males, while isolated cleft palate is more typically seen in females (Mossey & Little, 2002). High rates of cleft lip with or without cleft palate are seen in Latin America, China, and Japan, but are relatively low in Israel, South Africa and southern Europe. Isolated cleft palate rates are high in Canada and parts of northern Europe, but low in Latin America and South Africa (Leck and Lancashire, 1995).

Embryology
Healthy structuring of the lip and palate requires a complex series of events to unfold between the 4th and 10th weeks of human embryonic development. The primitive oral cavity is surrounded by the frontonasal prominence, the paired maxillary processes, and the paired mandibular processes, all of which are dependent on the successful migration of the neural crest cells from the neural folds to the mesenchymal tissue of the early craniofacial region.

The 6th week of development results in the formation of the upper lip and the primary palate. It is important to note that at the same time, the lateral nasal process is vulnerable as it undergoes numerous cell divisions, and in fact may not properly close if growth is disturbed in any way. During the 7th week, the palatal shelves rise and fuse to form a midline epithelial seam. Following this fusion, is a differentiation of bony and muscular tissue, leading to the development of the hard and soft palates. By week 10, a separation of the oral and nasal cavities is completed, which will allow for simultaneous chewing and breathing (Sperber, 2002).

The development of the face is coordinated by complex morphogenetic events and rapid proliferative expansion, and is thus highly susceptible to environmental and genetic factors, rationalizing the high incidence of facial malformations. During the first six to eight weeks of pregnancy, the shape of the embryo’s head is formed. Five primitive tissue lobes grow:

- Frontonasal Prominence (lobe a) - one from the top of the head down towards the future upper lip;
- Maxillar Prominence (lobes b & c) - two from the cheeks, which meet the first lobe to form the upper lip;
- Mandibular Prominence (lobes d & e) - and just below, two additional lobes grow from each side, which form the chin and lower lip;

If these tissues fail to meet, a gap appears where the tissues should have joined (fused). This may happen in any single joining site, or simultaneously in several or all of them. The resulting birth defect reflects the locations and severity of individual fusion failures (e.g., from a small lip or palate fissure up to a completely malformed face). (Dudas, 2007)

The upper lip is formed earlier than the palate, from the first three lobes named a to c above. Formation of the palate is the last step in joining the five embryonic facial lobes, and involves the back portions of the lobes b and c. These back portions are called palatal shelves, which grow towards each other until they fuse in the middle.

This process is very vulnerable to multiple toxic substances, environmental pollutants, and nutritional imbalance. The biologic mechanisms of mutual recognition of the two cabinets, and the way they are glued together, are quite complex and obscure despite intensive scientific research. (Dudas, 2007)
Etiology

The excess or deficiency of certain nutrients has been linked to orofacial clefts in humans. These nutrients are integral in normal palatogenesis and play a role in the molecular-biological processes as substrates, cofactors, and ligands. Current research suggests that in fact a combination of genetics and environmental factors contribute to the expression of clefting of the lip and/or palate. Of particular interest is the nutritional environment of the developing embryo. This, of course, is dependent upon the mother’s nutritional intake, as well as the genes responsible for nutrient transfer and metabolism. Specifically, the embryo’s exposure and metabolism of folate and vitamin A has been studied in an effort to develop nutritional strategies for the prevention of orofacial clefting.

Folate has been identified as a one-carbon donor for DNA methylation, which allows cells to maintain control of gene expression. A folate deficiency can therefore promote aberrant gene expression, leading to the development of orofacial clefts.

Vitamin A, known as retinoic acid, is a lipid-soluble molecule that binds to specific intracellular or membrane-bound receptors. An excess of Vitamin A may saturate a cell’s receptors rendering them inactive, and resulting in cellular damage.

Although the specific manner in which palatogenesis is influenced remains unknown, several suggested pathways are implicated. These include: the homocysteine pathway, in which riboflavin, folate, pyridoxine, cobalamin, and zinc act as cofactors or substrates; the oxidative pathway, in which a balance must be met by oxidants (e.g. glucose and homocysteine) and antioxidants (e.g. ascorbic acid and glutathione); and hematopoiesis, in which iron, cobalamin, and folate play an important role. Additionally, consideration must be paid to the three ways in which nutrients can influence gene expression: directly by regulating the development of orofacial gene expression; indirectly through epigenetic events; or by affecting genomic stability.

Risk Factors

Medications
Maternal intake of vasoactive drugs, which include pseudoephedrine, aspirin, ibuprofen, amphetamine, cocaine, or ecstasy, as well as cigarette smoking, have been associated with higher risk for oral clefts (Lammer 2004). Anticonvulsant medications such as phenobarbital, trimethadione, valproate, and dilantin have been documented to increase incidence of cleft lip and/or cleft palate (Holmes 2004,). Isotretinoin (Accutane) has been identified as potential causative factors for oral clefts (Lammer 1985). An association between maternal intake of sulfasalazine, naproxen, and glucocorticoids during the first trimester has been
suggested (Kallen 2003). Aminopterin (a cancer drug) has also been linked to the development of oral clefts (Warkany 1978).

**Maternal Age**
Several studies have reported increased risk of oral clefts with increased maternal age (Shaw 1991). However, larger studies failed to identify advanced maternal age as a risk factor for oral clefts (Vallino-Napoli 2004). Conversely, another study found a greater risk for cleft lip among younger mothers (Reefhuis, 2004).

**Race**
There are racial/ethnic differences in risk for oral clefts. Asians have the highest risk (14:10,000 births), followed by whites (10:10,000 births) and African Americans (4:10,000 births) (Das 1995). Among Asians, the risk for oral clefts is higher among Far East Asians (Japanese, Chinese, Korean) and Filipinos than Pacific Islanders (Yoon 1997). Amerindian populations in South America have been found to have higher rates than other “mixed” populations (Vieira 2002).

**Genetics**
Genetic factors are believed to account for some defects, often in combination with one or more environmental factors. Several loci have been identified for cleft lip with or without cleft palate, and, in one case, a specific gene has also been found. In cleft palate alone, one gene has been identified, but many more are probably involved. There is evidence of two main types of cleft lip and palate in whites. The first type is controlled by a single gene, which may code for a transforming growth factor-alpha (TGF-alpha) variant. The second type is multifactorial in nature. There is also some evidence that maternal and/or infant gene variations in conjunction with maternal smoking may lead to oral clefts in the infant (Lammer 2004).

**Metabolism**
There is some evidence that indicates that a defect in the maternal metabolism of specific dietary elements may also be a contributing factor in producing an affected child (van Rooij 2003). In this instance, the presence of a gene identified as MTHFR 677TT in conjunction with a low folate diet may lead to increased orofacial clefting. There is also an indication that even with adequate folate intake, these clefts may still occur in some cases (Lammer 2004).

Other metabolic factors that may affect the presence of orofacial clefts include maternal ability to maintain red blood cell zinc concentrations and myo-inositol concentrations (a hexahydrocyclohexane sugar alcohol) (Krapels 2004). Maternal ability to maintain adequate levels of Vitamins B6 and B12 and fetal ability to utilize these nutrients are also seen as a factor in the development of oral clefts (van Rooij 2003). When these nutrients are not metabolized properly, errors in DNA synthesis and transcription may occur (van Rooj 2004).

**Sex**
Infant sex influences the risk for oral clefts. Males are more likely than females to have a cleft lip with or without cleft palate, while females are at slightly greater risk for cleft palate alone (Blanco-Davila 2003). One study indicated that family history of clefts, birth order, maternal age at birth, first-trimester maternal smoking, and alcohol consumption during pregnancy did not explain the sex difference (Abramowicz 2003).

**Classification**

There are several different systems for classifying orofacial clefts. Three of the most commonly used classifications systems are the Tessier classification, Van der Meulen classification, and the LAHSAL Classification.

**Tessier Classification**

Paul Tessier published a classification on facial clefts based on the anatomical position of the clefts. (Tessier, 1976) It is based on the consideration of the orbit, nose and mouth as key reference points through which craniofacial clefts constant flow through meridians. The cracks are numbered from 0 to 14, and the number 8 serves as the equator. Thus, the cracks from 0 to 7 represent lower hemisphere facial clefts, while those between the 9 and 14 of the top are the cranial fissures.

These 15 different types of clefts can be put into 4 groups, based on their position: midline clefts, paramedian clefts, orbital clefts and lateral clefts. The Tessier classification describes the clefts at soft tissue level as well as at bone.
level, because it appears that the soft tissue clefts can have a slightly different location on the face than the bony clefts.

**Midline clefts**
The midline clefts are Tessier number 0 and number 14. The clefts comes vertically through the midline of the face. Tessier number 0 comes through the maxilla and the nose, while Tessier number 14 comes between the nose and the frontal bone.

**Paramedian clefts**
Tessier number 1, 2, 12 and 13 are the paramedian clefts. These clefts are quite similar to the midline clefts, but they are further away from the midline of the face. Tessier number 1 and 2 both come through the maxilla and the nose, in which Tessier number 2 is further from the midline (lateral) than number 1. Tessier number 12 is in extent of number 2, positioned between nose and frontal bone, while Tessier number 13 is in extent of number 1, also running between nose and forehead. Both 12 and 13 run between the midline and the orbit.

**Orbital clefts**
Tessier number 3, 4, 5, 9, 10 and 11 are orbital clefts. These clefts all have the involvement of the orbit. Tessier number 3, 4, and 5 are positioned through the maxilla and the orbital floor. Tessier number 9, 10 and 11 are positioned between the upper side of the orbit and the forehead or between the upper side of the orbit and the temple of the head. Like the other clefts, Tessier number 11 is in extent to number 3, number 10 is in extent to number 4 and number 9 is in extent to number 5.

**Lateral clefts**
The lateral clefts are the clefts which are positioned horizontally on the face. These are Tessier number 6, 7 and 8. Tessier number 6 runs from the orbit to the cheek bone. Tessier number 7 is positioned on the line between the corner of the mouth and the ear. A possible lateral cleft comes from the corner of the mouth towards the ear, which gives the impression that the mouth is bigger. It’s also possible that the cleft begins at the ear and runs towards the mouth. Tessier number 8 runs from the outer corner of the eye towards the ear. The combination of a Tessier number 6-7-8 is seen in the Treacher Collins syndrome. Tessier number 7 is more related to hemifacial microsomia and number 8 is more related to Goldenhar syndrome.

**Van der Meulen Classification**
Van der Meulen classification divides different types of clefts based on where the development arrest occurs in the embryogenesis. (Van der Meulen, 1983) A primary cleft can occur in an early stage of the development of the face (17 mm length of the embryo). The developments arrests can be divided in four different location groups: internasal, nasal, nasalmaxillar and maxillar. The maxillar location can be subdivided in median and lateral clefts.
Internasal dysplasia
Internasal dysplasia is caused by a development arrest before the union of the both nasal halves. These clefts are characterized by a median cleft lip, a median notch of the cupid’s bow or a duplication of the labial frenulum. Besides the median cleft lip, hypertelorism can be seen in these clefts. Also sometimes there can be a underdevelopment of the premaxilla.

Nasal dysplasia
Nasal dysplasia or nasoschisis is caused by a development arrest of the lateral side of the nose, resulting in a cleft in one of the nasal halves. The nasal septum and cavity can be involved, though this is rare. Nasoschisis is also characterized by hypertelorism.

Nasomaxillary dysplasia
Nasomaxillary dysplasia is caused by a development arrest at the junction of the lateral side of the nose and the maxilla, which results in a complete or non-complete cleft between the nose and the orbital floor (nasoocular cleft) or between the mouth, nose and the orbital floor (oronasal-ocular cleft). The development of the lip is normal.

Maxillary dysplasia
Maxillary dysplasia can manifest itself on two different locations in the maxilla: in the medial or the lateral part of the maxilla.

• *Median maxillary dysplasia* is caused by a development failure of the medial part of the maxillary ossification centers. This results in secondary clefting of the lip, philtrum and palate. Clefting from the maxilla to the orbital floor has also been reported.

• *Lateral maxillary dysplasia* is caused by a development failure of the lateral part of the maxillary ossification centers, which also results in secondary clefting of the lip and palate. Clefting of the lateral part of the lower eyelid is typical for lateral maxillary dysplasia.

LAHSAL Classification
The LAHSAL code is based upon the striped Y diagrammatic classification (Kriens, 1991). The relevant parts of the mouth are subdivided into six parts:

• right lip
• right alveolus
• hard palate
• soft palate
• left alveolus
• left lip
Orofacial Clefts

The code is then written as if looking at the patient. The first character is for the patient’s right lip and the last for the left lip.

The LAHSAL code indicates a complete cleft with a capital letter, an incomplete cleft with a lower case letter and no cleft with a dot.

For example:
Bilateral complete cleft of lip and palate: LAHSAL
Right complete cleft lip: L.....
Left incomplete cleft lip and alveolus: ....al
Incomplete hard palate, complete soft palate cleft: ..hS..

Syndromes with Associated Orofacial Clefts

Clefts of the lip and palate often co-occur with other significant congenital anomalies, especially in cases of isolated cleft palate. The association between orofacial clefts and specific genetic syndromes is between 5%-7%. Cleft lip with or without cleft palate is a common feature of more than 200 known disorders, and isolated cleft palate is a feature of more than 400 known disorders.

Several of the syndromes most commonly associated with orofacial clefting include:

- Van Der Woude Syndrome
- Pierre Robin Sequence
- Treacher Collins Syndrome
- Apert Syndrome
- Crouzon Syndrome
- Stickler Syndrome
Van der Woude Syndrome

Van der Woude syndrome (VWS) is a rare autosomal dominantly inherited disorder. It occurs across both genders in approximately 1 in 100,000 births. It is most commonly linked to chromosome 1q32-q41, but a second link has been attributed to Ip34. There is a 50% chance of affected individuals passing along the syndrome to their children. IRF6 has been identified as the specific gene mutated in approximately 70% of Van der Woude patients.

Clinical characteristics of Van der Woude syndrome include clefts of the lip and/or palate, lip pits, hypodontia, missing premolars, bifid uvula, ankyloglossia, hypernasal voice, as well as systemic manifestations including syndactyly. Paramedian lip pits are the most common physical manifestation, occurring in approximately 87.5% of all cases. They result when the embryonic lateral sulci fail to completely fuse. The pits are observed as transverse or circular slits atop small elevations on the surface of the lip. In many cases, minor salivary ducts open in to these pits, resulting in visible salivary discharge. This condition tends to cause a great deal of discomfort and embarrassment to the individual (Reddy et al., 2011).

Absence of the mandibular secondary premolars is frequently observed with VWS. Absent teeth have direct clinical consequences. It is therefore important to determine which teeth are missing as early as possible so that orthodontic intervention can proceed accordingly (Oberoi & Vargervik, 2005).

Pierre Robin Sequence

Pierre Robin sequence (PRS) occurs in approximately 1 in every 8500 births across both genders. A genetic link with PRS has observed when the dysregulation of SOX9 and KCNJ2 occurs. Pierre Robin sequence is characterized by mandibular micrognathia (undersized lower jaw), U-shaped cleft palate, and glossoptosis (atypical downward or posterior placement of the tongue) (Rangeeth et al., 2011).

The expression of PRS occurs during the 4\textsuperscript{th} and 8\textsuperscript{th} weeks of embryological development. During this period, the mandibular prominence lies between the stomodeum (a precursor to the embryonic mouth) and the first branchial groove, which outline the caudal (posterior) limits of the face. During the 6\textsuperscript{th} week of development, the free ends of the mandibular arch grow and meet ventrally. This arch causes the posterior placement of the mandible, and keeps the tongue placed high in the nasopharynx. As a result, medial growth and fusion of the palatal shelves becomes impaired. Hypoplasia of the mandible occurring before the 9\textsuperscript{th} week of embryologic development may therefore be an initiating factor in PRS.
Due to mandibular deficiency, newborns with PRS experience varying degrees of airway obstruction and feeding difficulties. The micrognathia and glossoptosis seen in PRS often results in poor nutrition, failure to thrive, gastroesophageal reflux, hypoxia, hypercapnia (increased amounts of carbon dioxide), cor pulmonale (failure of the right side of the heart), neurologic impairment, and in some cases death (Gozu et al., 2010).

Additionally, many children with a diagnosis of PRS develop articulation difficulties secondary to the occurrence of micrognathia and glossoptosis. Specifically, the fricatives (‘f’, ‘s’, and ‘sh’) and the plosives (‘p’ and ‘t’) are observed to be the most affected speech sounds for this population. In these cases, a speech therapist can help implement a plan to assist in remediating the resulting articulation errors (Van den Elzen et al., 2001).

**Treacher Collins Syndrome**

Treacher Collins syndrome (TCS) is a rare autosomal dominant disorder affecting approximately 1 in every 50,000 babies. A correlation between TCS and a mutation of the treacle gene TCOF1 has been established. Affected individuals have a 50% chance of passing along this syndrome to their children.

TCS shows a high variation in expressive phenotype, with characteristics ranging from almost severe to almost unnoticeable. Physical abnormalities of TCS include downward slanting eyes with notched lower eyelids, sunken cheekbones, sunken jawbones, broad mouth, pointed nasal prominence, high arched palate, malformed auricular pinnae, conductive hearing loss, and cleft lip and/or palate (Shete et al., 2011). Additionally, some TCS patients may exhibit a retrognathic mandible and glossoptosis resulting in airway obstruction.

Physical anomalies of the ears are a common characteristic of TCS. Abnormalities of the shape, size, and position of the auricular pinnae (outer ears) are frequently observed in this population. Other malformations include atresia of the external auditory canals, as well as irregular or absent auditory ossicles. Specifically, radiographic analysis has revealed fusion between the rudiments of the malleus and incus, partial absence of the stapes and oval window, and in some cases the complete absence of the middle ear and epitympanic space. As a result of these anomalies, TCS patients will experience bilateral conductive hearing loss.

**Apert Syndrome**

Apert syndrome is an autosomal dominant condition occurring in approximately 1 out of every 65,000 births. Mutations on adjacent amino acids (Ser252Trp and Pro253Arg) of human fibroblast growth factor receptor-2 (FGFR2) account for 99% of all cases of Apert syndrome. Apert syndrome accounts for 4.5% of the
craniosyntosis syndromes, in which a premature fusion by ossification of one or more of the fibrous sutures in an infant’s skull occurs (Hansen et al., 2004).

Physical characteristics of Apert’s syndrome include craniosyntosis, midfacial malformations, syndactyly of the hands and feet, bifid uvula, and clefts of the soft palate (occurring in 76% of Apert’s patients). Additionally, individual’s with Apert’s syndrome frequently experience a conductive hearing loss, most likely due to malfunctioning eustachian tubes resulting in persistent otitis media with effusion (Rajenderkumar et al., 2005).

**Crouzon Syndrome**

Similar to Apert Syndrome, Crouzon’s Syndrome is also an autosomal dominant disorder. Occurrence within the United States is approximately 1 in 60,000 births. Crouzon’s Syndrome is caused as a result of a mutation of the human fibroblast growth factor receptor-2 (FGFR2). Approximately 4.8% of all cases of craniosyntosis are secondary to Crouzon’s Syndrome.

The primary physical characteristic of the disorder is a premature fusion of the coronal and sagittal sutures, beginning during the child’s first year of life. Other physical malformations include midfacial hypoplasia, maxillary hypoplasia, shallow orbits, mandibular prognathism, overcrowding of the upper teeth, V-shaped maxillary dental arch, bifid uvula, and cleft palate (Padmanabhan et al., 2011).

**Stickler Syndrome**

Stickler Syndrome is an autosomal dominant disorder of collagen connective tissue. Patients with Stickler’s syndrome are subclassified into type 1 or type 2, depending on the locus heterogeneity. Both groups demonstrate similar systemic features. Approximately 75% of patients with Stickler’s syndrome fall within the type 1 vitreous phenotype. Specifically, a distinct folded membrane is found within the retrolental space, and can be observed to be occupied with a vestigial vitreous gel.

Stickler patients falling within the type 2 vitreous phenotype demonstrate sparse and abnormally thickened fibrous bundles throughout the vitreous cavity. A diagnosis of Stickler’s syndrome should be carefully considered in cases of: infants born with spondyloepiphyseal dysplasia with myopia or deafness, infants/children demonstrating sporadic retinal detachment in association of joint hypermobility, clefting and/or deafness, as well as infants born with a family history of rhegmatogenous retinal detachment.

Stickler syndrome is most commonly expressed in ophthalmic, orofacial, auditory, and articular manifestations. Congenital nonprogressive myopia, as well as retinal detachment is common. Other phenotypic expressions include a flat
midface with depressed nasal bridge, short nose, anteverted nares, and micrognathia. Midline clefting is also common, ranging in severity from a cleft of the soft palate to the Pierre Robin Sequence. Stickler syndrome patients typically experience hearing loss in the higher frequency ranges and of such a minor degree that patients are often unaware of the deficit. Secondary to the presence of clefting, patients often suffer from recurrent serous otitis media resulting in a conductive hearing deficit.

Diagnosis

Clefts in unborn babies are often detected with an ultrasound examination during a routine antenatal appointment. This antenatal scan typically takes place at around 20 weeks. The accuracy of sonography for prenatal diagnosis of cleft lip and palate is highly variable and dependent on the experience of the sonographer and the type of cleft. Reported rates of detection for cleft lip and palate range from 16% to 93% (Shaikh, 2001). Isolated cleft palate is rarely identified prenatally. Furthermore, even when a cleft lip is visualized sonographically, it is difficult to determine whether the alveolus and secondary palate are also involved. (Stroustrup Smith, 2004)

MRI is used increasingly for evaluation of fetal abnormalities that are difficult to identify on sonography alone (Levine, 2003). Fetal MRI is less dependent than sonography on optimal amniotic fluid volume, fetal position, and maternal body habitus. Additionally, visualization of small structures on MRI is not limited by bone shadowing. (Stroustrup Smith, 2004)

If a cleft lip or palate is not picked up during an antenatal appointment, the cleft is nearly always diagnosed after the baby has been born. However, in some cases, for example a submucous cleft palate where the cleft is hidden in the lining of the mouth, a diagnosis may not be made for several months or even years, when speech problems develop.

The Health Care Team

Patients with orofacial clefts are best cared for by an interdisciplinary team of specialists with experience in this field. Generally, there are two variations in specialized teams that provide services to individuals with cleft lip and/or palate (Seattle Childrens Hospital, 2010). The Cleft Palate Team (CPT) provides coordinated and interdisciplinary evaluation and treatment to patients with cleft lip and/or cleft palate; while The Craniofacial Team (CFT) provides coordinated and interdisciplinary evaluation and treatment for patients with a wide range of craniofacial anomalies or syndromes.
Cleft Palate Teams (CPTs)

Minimally, The Cleft Palate Teams (CPTs) must include the following:

- Surgeon
- Orthodontist
- Speech-Language Pathologist

It is common in the United States for most Cleft Palate Teams to be comprised of more than four specialists.

Craniofacial Teams (CFTs)

Craniofacial Teams (CFTs) specialize in craniofacial anomalies or syndromes. These teams conduct surgery that involves an intracranial approach. A greater number and variety of specialists is needed to address the patient’s individual and complex needs. Professionals on the Craniofacial Team include:

- **Craniofacial Surgeon.** Commonly trained initially as a plastic surgeon, otolaryngologist or oral and maxillofacial surgeon, performs corrective/reconstructive surgery of the craniofacial complex.

- **Otolaryngologist (ENT).** Ear, Nose and Throat Specialist. Many malformations involve defects in the airway passage, inflammation of the middle ear and/or hearing and speech defects. Such complaints are treated by the ENT-Specialist. He/she is also responsible for the hearing tests.

- **Audiologist.** Identifies, treats and monitors any disorders of the auditory or vestibular system.

- **Pediatrician.** Provides diagnostic evaluations, management of medical problems, and coordinates team care. An important role of the team pediatrician is communication with the primary care provider to monitor the child’s overall health and development.

- **Pediatric dentist.** Assesses, identifies, and treats dental pathology.

- **Craniofacial orthodontist.** As member of the craniofacial team the craniofacial orthodontist takes care of the non-surgical treatment of the malposition of the jaws. He/she is responsible for the pre and post operative treatment of jaw surgery and monitors growth by means of X-rays and plaster casts.

- **Orthodontist.** Responsible for the design and fabrication of fixed and removable orthopaedic and orthodontic appliances for the cleft patient from birth through to adulthood. He/she also fabricates dental study models that are used to monitor growth.
• **Prosthodontist.** Plans and fabricates an obturator to close defects that surgery is not capable of closing. Many patients with congenital deformities are missing teeth or have poorly shaped teeth and require a denture prosthesis.

• **Speech-language pathologist.** Evaluates and monitors speech development to help determine if speech therapy, prosthetic devices, or surgery are needed to improve speech skills.

• **Psychologist.** Monitors the child’s development and teaches the child how to deal with the social aspects of a facial deformity. The psychologist also aids the parents when needed.

• **Clinical Genetist.** After thorough family research, advises on heredity with regard to a syndrome. Sometimes a final diagnosis can be defined only after genetic examination.

• **Social worker.** He/she is counsellor of the parents and family when there are problems resulting from the syndrome, treatment and/or hospitalisation. She/he acts as an advisor and is able to contact various official authorities, in and outside the hospital.

• **Nurse.** From hospitalization till discharge the nurse is responsible for the daily health care and nurture of the child. The nurse also addresses feeding difficulties due to cleft.

**Standards of Care**

The American Cleft Palate – Craniofacial Association (ACPA) established the general standards of care for children with cleft lip and palate and other craniofacial anomalies. These standards are presented in two documents: “ACPA Parameters of Care” and “Standards for Approval of Cleft Palate and Craniofacial Teams”

**ACPA Parameters of Care (Revised 2007)**

The ACPA Parameters of Care (2007) is based on a national consensus conference funded by the Bureau of Maternal and Child Health, in conjunction with the ACPA.

The fundamental principles of care for children with cleft lip/ palate and other craniofacial anomalies include all of the following:

1. An interdisciplinary team of specialists with experience in cleft lip/palate is required.
2. The team must evaluate and treat sufficient numbers to maintain its expertise.
3. The first few days or weeks of life is the optimal time for team evaluation.
4. The team should assist families in adjustment to the birth defect.
5. The team should adhere to the 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. The team should be sensitive to cultural, psychosocial and other contextual factors.
8. The team is responsible for monitoring short and long-term outcomes, including quality management and revision of clinical practices, when appropriate.
9. Treatment outcomes include the child's psychosocial wellbeing, and effects on growth, function and appearance.
10. Long-term care includes evaluation and treatment in the areas of audiology, dentistry/orthodontics, genetics/dysmorphology, nursing, oral and maxillofacial surgery, otolaryngology, pediatrics, plastic surgery, psychosocial services and speech language pathology.

**Standards for Approval of Cleft Palate and Craniofacial Teams**

In 2010, the American Cleft Palate-Craniofacial Association (ACPA) and Cleft Palate Foundation (CPF) established standards for care (available at: http://acpa-cpf.org/team_care/standards/) that identified the following six components as essential to the quality of care provided by interdisciplinary teams of health care specialists to patients with craniofacial anomalies, regardless of the specific type of disorder:

- Team Composition
- Team Management and Responsibilities
- Patient and Family/Caregiver Communication
- Cultural Competence
- Psychological and Social Services

The following ACPA and CPF standards are necessary conditions for approval of both cleft palate and craniofacial teams.

**Standard 1: Team Composition**

1.1 The Team includes a designated patient care coordinator to facilitate the function and efficiency of the Team, ensure the provision of coordinated care for patients and families/caregivers and assist them in understanding, coordinating, and implementing treatment plans.
1.2 The Team includes Speech-Language Pathology, Surgical, and Orthodontic specialties.

1.3 The Team includes members who are qualified by virtue of their education, experience, and credentials to provide appropriate care and to maintain currency with best practice.

1.4 The Team demonstrates access to professionals in the disciplines of psychology, social work, psychiatry, audiology, genetics, general and pediatric dentistry, otolaryngology, and pediatrics/primary care.

1.5 The Craniofacial Team must include a surgeon trained in cranio-maxillofacial surgery and access to a psychologist who does neuro-developmental and cognitive assessment. The results of the neuro-developmental and cognitive assessment must be part of the CFT team assessment record. The Team also must demonstrate access to refer to a neurosurgeon, ophthalmologist, radiologist, and geneticist. The participation of these individuals should be documented in each patient’s team report.

**Standard 2: Team Management and Responsibilities**

2.1 The Team has a mechanism for regular meetings among core Team members to provide coordination and collaboration on patient care.

2.2 The Team has a mechanism for referral and communication with other professionals.

2.3 Team care is provided in a coordinated manner. The sequencing of evaluations and treatments serves the patient’s overall developmental, medical, and psychological needs.

2.4 The Team re-evaluates patients based on Team recommendations.

2.5 The Team must have central and shared records.

**Standard 3: Patient and Family/Caregiver Communication**

3.1 The Team provides appropriate information to the patient and family/caregiver about evaluation and treatment procedures.

3.2 The Team encourages patient and family/caregiver participation in the treatment process.

3.3 The Team will assist families/caregivers in locating resources for financial assistance necessary to meet the needs of each patient.

**Standard 4: Cultural Competence**

4.1 The Team demonstrates sensitivity to individual differences that affect the dynamic relationship between the Team and the patient and family/caregiver.

4.2 The Team treats patients and families/caregivers in a non-discriminatory manner.
Standard 5: Psychological and Social Services

5.1 The Team has a mechanism to initially and periodically assess and treat, as appropriate, the psychological and social needs of patients and families/caregivers and to refer for further treatment as necessary.

5.2 The Team has a mechanism to assess cognitive development.

Standard 6: Outcomes Assessment

6.1 The Team has mechanisms to monitor its short-term and long-term treatment outcomes.

6.2 The Team has and implements a quality management system.

Treatment Goals

The goals of treatment for the child with a cleft lip/palate are (Seattle Children’s Hospital, 2010):

- 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

Keys 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.

Nursing and Feeding

Depending on the location and severity of the cleft, a newborn baby may have difficulties with sucking. A suckling baby uses its tongue to push the nipple against the roof of its mouth. The muscular motions of the jaw and soft palate at the back of the mouth allow suction to draw the milk. The cleft makes it hard to seal the mouth properly over the nipple, preventing the vacuum necessary to draw milk out of the breast or bottle.

Feeding Issues

Nasal regurgitation: Babies without a fully formed palate frequently leak liquids from their noses as they eat. Nasal regurgitation can pose problems over time and create nasal and sinus congestion.

Burping: Babies with cleft palate need to be burped often. Burping frequently during a feeding will make the baby more comfortable and may help to reduce spitting up.

Feeding times: Feeding should not exceed 20-30 minutes. Longer feedings are more work for the baby and more stress for parents and families.

Bottle Feeding

There are several special bottles and nipples available which help make feeding easier.

Mead Johnson Bottle
Mead Johnson Bottle is a soft squeeze bottle with a long cross cut nipple. It is designed to be “pulse” squeezed with the baby’s sucking and swallowing. The bottle has an extended nipple that allows milk to be directed past the cleft and cross cut allows increase flow with squeezing.

Haberman Feeder Bottle
The Haberman feeder’s design enables the feeder to be activated by tongue and gum pressure, imitating the mechanics involved in breastfeeding, rather than by sucking. A one-way valve separates the nipple from the bottle. Before starting the feeding, air is squeezed out of the nipple and is automatically replaced by breastmilk or formula through the valve. Milk cannot flow back into the bottle and...
is replenished continuously as the baby feeds. A slit valve opening near the tip of the nipple shuts between jaw compressions, preventing the baby from being overwhelmed with milk. Stopping or reducing the flow of milk is controlled by rotation of the nipple in the baby's mouth. Usually the nipple is marked with lines that indicate zero flow, moderate flow, and maximum flow. For infants who need assistance with their feeding efforts, the mother may apply a gentle pumping action to the body of the nipple.

**Pigeon Bottle**
The Pigeon bottle’s Cleft Palate Nipple is specially designed to appropriately feed cleft palate infants and infants with poor sucking strength. The latex-free, Y-cut nipple has a thick and thin side with a one-way valve that prevents excessive air intake and allows milk to flow only when sucked by the baby.

**Breastfeeding**

The following recommendations are made concerning breastfeeding based on reviewed evidence (Reilly, 2007):

1. As these infants are prone to otitis media, mothers should be encouraged to provide the protective benefits of breast milk. Evidence suggests that breastfeeding protects against otitis media in this population. (Aniansson et al., 2002)

2. Babies with a CL/P should be evaluated for breastfeeding on an individual basis. In particular, it is important to take into account the size and location of the baby’s CL and/or CP, as well as the mother's wishes, previous experience with breastfeeding, and supports.

3. Mothers who wish to breastfeed should be given immediate access to a lactation advisor to assist with positioning, management of milk supply, and expressing milk for supplemental feeds.

4. Mothers should be counseled about likely breastfeeding success. Where direct breastfeeding is unlikely to be the sole feeding method, the need for breast milk feeding and, when appropriate, possible delayed transitioning to breastfeeding should be discussed.

5. Breast milk feeding (via cup, spoon, bottle, etc.) should be promoted in preference to formula feeding. In these circumstances, assistance with hand expression/pumping breast milk should commence on day 1.

6. Monitoring of a baby’s hydration and weight gain may be important while a feeding method is being established. If inadequate, supplemental feeding should be implemented or increased.

7. Modification to breastfeeding positions may increase the efficiency and effectiveness of breastfeeding.

8. If a prosthesis is used for orthopedic alignment prior to surgery, caution should be used in advising parents to use such devices to facilitate breastfeeding, as
there is strong evidence that they do not significantly increase feeding efficiency or effectiveness. (Prahl, Kuijpers-Jagtman & van’t Hof, 2005)

9. Evidence suggests that breastfeeding can commence/recommence immediately following CL repair and 1 day after CP repair without complication to the wound. (Cohen, Marschall & Schafer, 1992)

10. Assessment of the potential for breastfeeding of infants with a CL/P as part of a syndrome/sequence should be made on a case-by-case basis, taking into account the additional features of the syndrome that may impact on breastfeeding success.

Nasoalveolar Molding (NAM)

Depending on the width of the cleft and the presence or absence of a cleft palate, a short period of reshaping the mouth and nose may be recommended. Nasoalveolar Molding (NAM) is a technique in which the alveolus (gum ridges) and/or nose are molded with an appliance similar to an orthodontic retainer. This is usually done by a specially trained orthodontist prior to surgery, in order to make surgery simpler. The baby wears the appliance 24 hours a day for a period of weeks or months. It does not interfere with feeding or breathing for the baby.

Surgical Interventions

Surgical intervention for the remediation of orofacial clefts varies greatly within and between countries. Which surgical technique is considered most effective continues to provoke controversy. (Ling, 2000)

The goals of surgical repair are:

- separation of the nasal and oral cavities
- construction of a watertight and airtight velopharyngeal valve
- preservation of facial growth
- development of aesthetic dentition and functional occlusion

Schweckendiek’s Technique

This surgery is used to close soft palate clefts only. (If a hard palate cleft also exists, its repair is deferred to a later date). Incisions are made in the soft palate, muscle bundles are dissected, a levator sling is reconstructed, and the soft palate is closed. It is usually performed at age 3 to 12 months.

Advantages of this surgery:

- construction of a velopharyngeal valve at an early age
- minimal disturbance in future facial growth
Disadvantages of this surgery:
- necessity of an additional operative procedure and hospitalization
- required dental prosthesis

(Ling, 2000)

**The Von Langenbeck Palatoplasty**

The Von Langenbeck palatoplasty is the oldest procedure still in use today. This surgical approach involves a simple closure technique without seeking to lengthen the palate. Anterior and posterior based bipedicle mucoperiosteal flaps of the hard and soft palate are advanced medially to close the palatal cleft.

The basic goals of the procedure are to close the abnormal opening between the nose and mouth, to help the patient develop normal speech, and to aid in swallowing, breathing and normal development of associated structures in the mouth. The procedure is usually performed on infants.

(Ling, 2000)
The ideal age for the patient is between six and twelve months of age. If the surgery is carried out much beyond three years of age, speech development may not be optimal. A common adverse condition secondary to cleft palate repair is the presence of oronasal fistulas. Oronasal fistulas typically result from an inadequate closure of the hard palate. They can occur anywhere along the line of the cleft, but are most commonly found at the junction of the hard and soft palates and at the anterior area of the cleft. Postoperative fistulas often produce hypernasal speech, nasal emission, and nasal food regurgitation.

The Von Langenbeck surgical technique has also been criticized for allowing for a high rate of velopharyngeal insufficiency (VPI). VPI is a condition in which inadequate closure exists between the soft palate and the posterior pharynx during speech. As a result of the insufficient closure, air leaks up into the nasopharynx producing a hypernasal quality of speech.

**Modified Von Langenbeck Palatoplasty**

A modified version of the Von Langenbeck palatoplasty was developed in an effort to significantly reduce the occurrence of post-operative oronasal fistulas. The modification of the original surgical technique involves easing unwanted tension at the anterior portion of the cleft. This is accomplished by creating and incorporating an anterior oromucosal triangular flap allowing for nasal side closure of the anterior section of the cleft.

**The Furlow Z-plasty**

The Furlow Z-plasty surgical technique uses opposing mirror image Z-plasties of both the oral and nasal mucosa. This procedure narrows the velopharyngeal space by medializing the tonsillar pillars. The Z-plasty also promotes the lengthening of the soft palate by preventing contraction of a longitudinal scar. Finally, the Furlow Z-plasty reassigns the muscle fibers of the levator veli palatini into a transverse orientation within the soft palate.

![Diagram of Furlow Z-plasty](image)

(Ling, 2000)
Orofacial Clefts

The Furlow Z-plasty technique results in fewer incidences of VPI; and subsequently, results in a higher patient quality of speech secondary to a decrease in hypernasality.

**Palatal Lengthening – V-Y Pushback**

This surgical approach is utilized to accomplish two goals: closure of of the cleft, and also lengthening of the palate.

The surgery creates two posterior unipedicle flaps and either one or two (depending on the extent of the cleft) anterior unipedicle palatal flaps. These anterior flaps are then advanced or rotated medially, and the posterior flaps are retrodisplaced with a V to Y technique.

![Diagram of Palatal Lengthening – V-Y Pushback](Ling, 200)

**Advantages:**
- lengthening of palate
- improved speech resulting in more extensive palatal clefts than those obtained by Von Langenbeck’s method

**Disadvantages:**
- failure to provide mucosal coverage of the retrodisplaced palate’s nasal surface which reduces the palatal lengthening obtained in the final result secondary to scar contracture of the raw nasal surface
- difficulty of closing the cleft’s alveolar portion
- fistulas in the thin mucoperiosteal membrane near the hard/soft palate junction

**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.

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 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.

Psychosocial Issues

Most children who have their clefts repaired early enough are able to have a happy youth and social life. Having a cleft palate/lip does not inevitably lead to a psychosocial problem. However, adolescents with cleft palate/lip are at an elevated risk for developing psychosocial problems especially those relating to self concept, peer relationships and appearance. Adolescents may face psychosocial challenges but can find professional help if problems arise. A cleft palate/lip may impact an individual’s self-esteem, social skills and behavior. There is research dedicated to the psychosocial development of individuals with cleft palate. Self-concept may be adversely affected by the presence of a cleft lip and or cleft palate, particularly among girls. (Leonard, 1991)

During the early preschool years (ages 3–5), children with cleft lip and or cleft palate tend to have a self-concept that is similar to their peers without a cleft. However, as they grow older and their social interactions increase, children with clefts tend to report more dissatisfaction with peer relationships and higher levels of social anxiety. Experts conclude that this is probably due to the associated stigma of visible deformities and possible speech impediments. Children who are judged as attractive tend to be perceived as more intelligent, exhibit more positive social behaviors, and are treated more positively than children with cleft lip and or cleft palate. (Tobiasen, 1984) Children with clefts tend to report feelings of anger, sadness, fear, and alienation from their peers, but these children were similar to their peers in regard to “how well they liked themselves.”

The relationship between parental attitudes and a child’s self-concept is crucial during the preschool years. It has been reported that elevated stress levels in
mothers correlated with reduced social skills in their children. (Pope, 1997)
Strong parent support networks may help to prevent the development of negative self-concept in children with cleft palate. In the later preschool and early elementary years, the development of social skills is no longer only impacted by parental attitudes but is beginning to be shaped by their peers. A cleft lip and or cleft palate may affect the behavior of preschoolers. Experts suggest that parents discuss with their children ways to handle negative social situations related to their cleft lip and or cleft palate. A child who is entering school should learn the proper (and age-appropriate) terms related to the cleft. The ability to confidently explain the condition to others may limit feelings of awkwardness and embarrassment and reduce negative social experiences. (CPF, 2007)

As children reach adolescence, the period of time between age 13 and 19, the dynamics of the parent-child relationship change as peer groups are now the focus of attention. An adolescent with cleft lip and or cleft palate will deal with the typical challenges faced by most of their peers including issues related to self esteem, dating and social acceptance. (Pope, 2005) Adolescents, however, view appearance as the most important characteristic above intelligence and humor. This being the case, adolescents are susceptible to additional problems because they cannot hide their facial differences from their peers. Adolescent boys typically deal with issues relating to withdrawal, attention, thought, and internalizing problems and may possibly develop anxiousness-depression and aggressive behaviors. (Pope, 2005) Adolescent girls are more likely to develop problems relating to self concept and appearance. Individuals with cleft lip and or cleft palate often deal with threats to their quality of life for multiple reasons including: unsuccessful social relationships, deviance in social appearance and multiple surgeries.

Otolaryngology and Audiology

Middle-ear disease is very common in infants with cleft palate and causes hearing loss that can last into childhood. The development of hearing is affected as children with cleft lip and palate and cleft palate universally present with otitis media with effusion (OME) and is often present within the first 6 months of life. Otitis media with effusion is a condition which presents with fluid in the middle ear and is not accompanied by signs or symptoms of an acute infection. The high prevalence of OME in children with orofacial clefts is due to Eustachian tube dysfunction.

The muscles responsible for the opening of the Eustachian tube include the tensor veli palatini and the levator veli palatini, which exhibit an abnormal point of insertion in children with CLP due to the palate not fusing during fetal development. This lateral point of insertion causes lack of anchorage which does not allow proper opening of the Eustachian tube. Therefore, the opening of the Eustachian tube is compromised and the middle ear cavity is not properly
ventilated. This lack of ventilation produces negative pressure and results in a retracted tympanic membrane and the secretion of mucous from the tissues through osmosis into the middle ear cavity. (Flynn, 2009) Middle ear pathology often leads to conductive hearing loss (CHL) in 50 to 93% of patients. This high incidence of OME and hearing loss is also found after surgical repair of the cleft leading to persistent otologic pathology.

Otologic goals for individuals with cleft palate are to provide adequate hearing, maintain ossicular continuity and adequate middle ear space, and prevent deterioration of the tympanic membrane. Patients with eustachian tube dysfunction are evaluated every 3-4 months until it resolves. Indications for myringotomy and tube insertion include a significant conductive hearing loss or persistent middle ear effusion, recurrent otitis media, or tympanic membrane retraction. (Deskin, 1998)

Speech and Language

Children with clefts and craniofacial abnormalities are also at a greater risk for resonance, articulation and expressive language problems. The Speech-Language Pathologist plays an integral part in the assessment and treatment of such disorders if they are present. Initially, it is within the SLP’s scope of practice to educate the child’s family on normal speech and language development, and also to provide an assessment of pre-linguistic speech-language abilities before the child turns 6 months old. If it is found that the speech and language skills are not developmentally appropriate or age appropriate for the child, arrangements should be made by the SLP to employ an early intervention strategy that would facilitate speech sound production and provide language stimulation. In addition to the initial testing and treatment plan, throughout the course of the child’s development, the SLP has widespread responsibilities:

- Speech-language evaluations and documentation should be conducted twice during the first two years of life and at least annually thereafter until the age of six.
- Speech-language screenings should take place annually until the child develops adenoids and every two years after until dental maturity.
- Conduct re-evaluations as deemed necessary by other CLP team members.
- Instrumental assessments for velopharyngeal function for patients with audible nasal air emissions or resonance disorders.

Communication Disorders

Children with cleft lip and palate often show delays in language development that
encompass both receptive (comprehension) and expressive (production) language. (Nagarajan, 2009)

The etiology of the speech disorder is often multifactorial and complex in nature with many structural and non-structural factors potentially interacting to cause speech problems. (Hodgkinson, 2005)

Etiological factors include (Grunwell, 2001):
1. abnormal oronasal structure and/or function e.g. velopharyngeal insufficiency, nasal airway deviations, hearing and ENT problems, residual clefts and oronasal fistulae.
2. abnormal oronasal structure and growth e.g. dental and occlusal anomalies.
3. abnormal neuromotor development e.g. abnormally learned neuromotor patterns, developmental learning deficits and neurological factors.
4. abnormal or disturbed psychosocial development e.g. impact of physical difference on parents and child.

Typically, the pressure consonants (stops, fricatives and affricates) are more affected than the other sounds. Various atypical consonant productions can be observed in individuals with cleft lip and palate. (Henningsson, 2008)

The common error patterns noticed in individuals with cleft lip and palate are (Nagarajan, 2009):
- Abnormal backing of oral targets to postuvular place (to pharyngeal or glottal)
- Abnormal backing of oral targets within the oral cavity (to uvular or velar or palatal)
- Nasal fricatives
- Nasal consonants for oral pressure consonants
- Nasalized voiced pressure consonants
- Weak oral pressure consonants
- Other misarticulations involving lateralized or palatalized productions of fricatives
- Developmental articulation/phonological error

Cleft type errors of speech sound production are broadly classified into two types: obligatory and compensatory. (Trost-Cardamone, 1997)

Obligatory errors are the result of the physical characteristics of the articulatory or velopharyngeal system. They include errors in production due to interference of structural abnormalities, such as malaligned tooth, residual clefts, oronasal
fistula, etc. These errors are unlikely to respond to therapy. They generally result in changes in the manner of articulation and almost always require surgical or prosthetic intervention.

Compensatory errors include errors that occur due to maladaptive articulatory placements learned by children during the developmental period. A child making compensatory errors is attempting to compensate for their inability to produce the correct sound by making the sound elsewhere in the mouth. Residual speech problems may persist after surgical correction of the anatomy, because the production has become part of the child's phonological system. For example, a stop consonant is produced as a stop, but with a posterior placement of the articulator (/p/ produced as a /k/ or /t/). (Nagarajan, 2009) These errors are amenable to speech therapy; and the prognosis and outcomes of therapy are better if the intervention is started early.

**Abnormal Nasal Resonance**

Another characteristic seen in a majority of individuals with cleft lip and palate is abnormal nasal resonance. For most English sounds, the majority of acoustic energy resonates within the oral cavity. For the nasal sounds (/m/, /n/ and /ng/), the majority of sound resonates within the nasal cavity. The resonance of speech is primarily influenced by the size and shape of the oral, nasal, and pharyngeal cavities; as well as the functioning of the velopharyngeal valve. Structural disturbances such as obstructions in the nasopharynx, large oronasal fistula, and velopharyngeal dysfunction often result in abnormal nasal resonance.

Hypernasality describes a perceptual judgment that there is excessive nasal resonance for oral sounds. Hypernasality can not occur on voiceless sounds, because these sounds do not have any acoustic energy. (Kummer, 2008) If voiceless sounds are perceived to be sounding nasal, then it is likely to be nasal air emission or there may be hypernasality on the vowel that precedes or follows the voiceless sound. Occasionally, hypernasality results due to velopharyngeal mislearning, wherein only certain sounds are perceived to be hypernasal. This is referred to as phoneme-specific hypernasality, which occurs due to the incorrect placement of oral structures for certain sounds (e.g., ng/l, ng/r). (Nagarajan, 2009)

Hyponasality indicates a decreased nasal resonance. It is most noticeable with nasal consonants, but it is also present on vowels. This is because during the production of vowels, especially high vowels, there is some transmission of resonance through the soft palate. Hyponasality causes the speaker to sound as if they have a cold.

A cleft lip and palate can create either hypernasality or hyponasality. In some individuals, hypernasality and hyponasality co-exist, resulting in a mixed nasality.
**Velopharyngeal Insufficiency (VPI)**

Located between the oral and nasal cavities, the velopharyngeal valve is a tridimensional muscular valve that consists of the lateral and posterior pharyngeal walls as well as the soft palate. The function of the velopharyngeal valve is to separate the oral and nasal cavities during speech and swallowing. Air is directed through the mouth for oral sounds and through the nose for nasal sounds during speech production. (Nagarajan, 2009)

When this mechanism is impaired in some way, the valve does not fully close, and a condition known as velopharyngeal insufficiency (VPI) can develop. (This disorder is also known as “palato-pharyngeal insufficiency”, “velopharyngeal inadequacy,” and “velopharyngeal dysfunction”.)

It is estimated that 10–20% of children undergoing primary palatoplasties before the age of 18 months have associated VPI. (Shprintzen, 2005) The occurrence of VPI may be much higher in children who undergo primary palate repair at later ages.

**VPI Evaluation**

If the clinician suspects that VPI is present and interfering with a child's communication, a complete VPI evaluation should be performed. (The child must be old enough to cooperate; usually 2-5 years old). This evaluation is generally conducted by a speech-language pathologist, otolaryngologist and radiologist. The evaluation often includes the following:

- oral exam
- analysis of speech articulation
- videofluoroscopic speech study
- naso-endoscopy

Occasionally, nasometric studies and aerodynamic measures are also used.

Assessment of VPI requires a thorough perceptual evaluation. Testing should include progressive speech/language challenges (including syllables, words, and sentences) that place a higher demand on the velopharyngeal functioning. The results of the evaluation document the velopharyngeal function and provide necessary information about whether additional instrumental evaluation (such as nasoendoscopy and videofluoroscopy) are needed.

Both the nasoendoscopy and videofluoroscopy may be utilized effectively to locate anatomical and physiological defects that can cause VPI. Nasoendoscopy is a minimally invasive procedure that provides a view of all the structures of the velopharyngeal mechanism and determines the location, size, and shape of the velopharyngeal opening. Compared to videofluoroscopy, nasoendoscopy helps
more in the visualization of even small openings in the velopharyngeal valve. However, videofluoroscopy gives a better picture of the length of the velum and its upward movement during speech. It also provides a view of the entire length of the posterior pharyngeal wall. The information from multiple views (frontal, basal, and lateral) has to be extrapolated to appreciate the functioning of the valve in all the three dimensions. Also, exposures to radiations in videofluoroscopy necessitate the procedure to be brief. Comparatively, videofluoroscopy is a less invasive procedure than nasoendoscopy. (Nagarajan, 2009)

Nasometry is another procedure that may be utilized for the assessment of resonance. It is a noninvasive procedure that measures the oral and nasal energy used during speech. The test does not allow the clinician to visualize the velopharyngeal port, so the resonance measurement is recorded indirectly. The instrument generates a numerical score that indicates the nasal acoustic energy of an individual’s speech. These values are then compared to the standardized norms for that particular language for interpretation. (Nagarajan, 2009)

Speech Therapy

Therapeutic speech intervention for individuals with cleft lip and palate often begin prior to surgical repair. The focus of therapy in very young infants, is on training the parent / caregiver to stimulate the child’s ability to understand and use language. There is evidence that early therapeutic intervention using parents helps reduce compensatory errors. (Scherer, 2008)

Children as young as three years old can be direct participants in therapy for the correction of misarticulations issues. The therapeutic objective is to correct the placement of the oral structures for speech sound production and direct the appropriate airflow. (However, misarticulation problems secondary to structural defects cannot be corrected through speech therapy unless the structural deformity is first surgically corrected.) If the structural correction is necessary, it should be followed with speech therapy to correct the functioning / production of speech sounds. Providing feedback using multiple auditory, visual, and tactile modalities is important in the management of articulation and resonance disorders.

Because individuals with cleft lip and palate have a structural deformity and not a muscle weakness, oromotor exercises (such as blowing, sucking, whistling, and electrical stimulation) are not useful in facilitating the correct production of speech sounds; and they should be avoided in this population. (Golding-Kushner, 2001)
Timeline for Speech and Language Therapy Intervention

Intervention can take place at any time but must be appropriate, timely and tailored to the individual. The aim of the Speech and Language Therapist is to facilitate normal development, prevent problems and to help the child with a orofacial cleft achieve their optimum communication potential.

The following outline of speech and language therapy management is cited as good practice (Russell, 2001):

**Birth to palate repair**
Support and information to parents/caregivers about the development of communication and how the cleft may affect speech development. Encouraging normal patterns of parent-child interaction.

**Palate repair to 18 months**
Monitoring general communication development and advising parents as appropriate, particularly in relation to emerging babble patterns. At 18 months children will have a routine assessment which looks at play skills, social development, interaction, receptive language, expressive language and consonant production.

**18 months to 3 years**
Continued monitoring of all aspects of communication with particular regard to velopharyngeal function and consonant production. At 3 years the child will have a more formal assessment of receptive and expressive language skills and an audio and video recorded sample of speech that is analyzed in detail.

**3 years to 4.5 years**
Monitoring and intervention as required, based on comprehensively analyzed speech sound data. Liaison with external agencies, particularly nurseries and education.

**5, 10, 15 and 20 year**
Collection and analysis of speech data using audio and video recordings.

**Other investigations**
The specialist Speech and Language Therapist with the cleft team will contribute to assessment, diagnosis and treatment planning for children requiring further surgery for velopharyngeal insufficiency affecting speech outcome and symptomatic palatal fistulae. In addition, assessment is required for older children and adults who require maxillary advancement and those needing prosthetic management of velopharyngeal insufficiency where surgery is contraindicated.
Children with clefts of the lip and palate frequently have dental problems in the area of the cleft. The issues correspond to three different developmental stages (Victorian Regional Group, 2008):

- Deciduous dentition period between 6 months & 7 years
- Mixed dentition period (where there are both deciduous and permanent teeth present, between 7 & 23 years)
- Permanent dentition period beyond 12 years (where no deciduous teeth are remaining).

The Deciduous Dentition Period

The upper deciduous (or first teeth), are not greatly affected, although the teeth just next to the cleft or clefts may be slightly displaced into the palate, or may be very slow in erupting or coming through the gums. Occasionally one or other of these teeth may not form. Behind the cleft of the gum pad, one or all of the teeth (which are the deciduous canine and two deciduous molars) may turn inwards towards the palate, and the bone, which holds them, may collapse. This is generally not a serious problem and may be readily connected by orthodontic treatment when the child is about 4-6 years.

It is difficult to prevent such collapse at an earlier stage. If there is only minor collapse, orthodontic treatment may be deferred until the child is 8 or 9 years old. A very important point to note is that the orthodontist mostly uses the deciduous teeth to hold appliances with which the upper dental arch is reshaped and repositioned. For this reason, special care must be taken of the teeth, with attention to diet, eating habits, and tooth-brushing. Otherwise no orthodontic treatment can be given, and the child's malocclusion will always become worse, never better.

If the front deciduous teeth are displaced because of the cleft, it is not always possible, or very helpful, to correct them, as this may not sufficiently help the position of the permanent front incisor teeth which appear later.

Mixed Dentition Period

It is during this stage that the permanent incisor teeth first appear, and they may not be as well position as the first teeth were. However, as long as the teeth continue to be well looked after, the orthodontist can use appliances to improve their positions. It must be understood that a plate or other dental device may have to be worn by the child to hold the corrections. Such devices are a source of worry because they must be kept scrupulously clean, otherwise the teeth against which they rest may decay and the gums may become unhealthy.
The Permanent Dentition Period

All the deciduous teeth are replaced by permanent teeth by about the age of 12 years, although sometimes the canine tooth near the cleft does not appear for quite some time after this. While these teeth are forming and appearing, the child's jaws are continuing to grow. The teeth move around and adjust their positions during this growth, although they do not improve their positions. This is why it is almost always necessary to carry out a final orthodontic correction between 12-14 years. This treatment is usually done with bands and arch wires which are fixed to the teeth for a period sometimes longer than 12 months. Sometimes one or two permanent teeth may not form in the child with a cleft lip or cleft palate. For this reason, after orthodontic treatment a partial denture is made to fill the empty space in the dental arch. When the child is grown beyond adolescence the denture might be replaced with a fixed dental bridge, or implants.

Timeline for All Health Care Interventions

The following timeline for health care interventions is based on the published recommendations of The American Cleft Palate-Craniofacial Association (ACPA) and Cleft Palate Foundation (CPF). (The Center for Children with Special Needs Seattle Children’s Hospital, 2010)

Infancy:
- Audiology screening
- Audiology follow-up
- Consultation with pediatrician (Peds Screening)
- Consultation with a geneticist
- Consultation/Visit with plastic surgeon
- Contact made with Orofacial Team Coordinator
- Contact made by Social Services
- Contact with other specialties as necessary
- Evaluation by a Feeding specialist
- Follow-up with pediatrician (Peds Screening)
- Orthodontics for palatal control device if required
- Otolaryngology screening
- Pediatrician / PCP Follow-up
- Repair cleft lip
- Repair cleft palate, if appropriate – age 9-12 months
- Social Service Follow-up

Age 12 to 18 months:
- Cleft palate repair, when determined necessary
- Contact with other specialties, as necessary
Orofacial Clefts

- Developmental Screening
- Pediatric dentist for oral examination, preventive education/procedures,
- PE Tube placement or evaluation
- Speech / language evaluation

Age 2 to 3 years:
- Cleft palate repair, when determined necessary
- Contact with other specialties, as necessary
- Developmental Screening
- Pediatric dentist for oral examination, preventive education/procedures,
- PE Tube placement or evaluation
- Speech / language evaluation

Age 4 to 6 years:
- Contact with other specialties, as necessary
- Evaluation by craniofacial team for future orthognathic surgery
- Evaluation for VPI (velopharyngeal incompetence)
- Orthodontia-dental/facial orthodontia treatment initiation
- Screening for developmental and special education needs
- Surgical procedures as needed, such as pharyngeal flap or palatal lift

Age 6 to 9 years:
- Contact with other specialties, as necessary
- Screening for self esteem, teasing issues and referral to psychology as needed
- Bone grafting of alveolar cleft

Age 9 to 11 years:
- Bone grafting of alveolar cleft
- Contact with other specialty as necessary
- Orthodontics

Age 11 to 18 years:
- Contact with other specialties, as necessary
- Lip revision / rhinoplasty
- Orthodontia (Alveolar ridge notch & Cleft palate patients only)
- Orthognathic surgical procedures with post-op evaluation of VP function and articulation by speech pathologist
- Prosthodontic Dentistry
Resources

About Face USA
P.O. Box 969 Batavia, IL 60510-0969
Phone: 888-486-1209
www.aboutfaceusa.org
e-mail: info@aboutfaceusa.org
This group provides support and guidance to people with facial differences. They also publish a newsletter and sponsor support groups.

American Cleft Palate-Craniofacial Association (ACPA)
1504 East Franklin St., Suite 102, Chapel Hill, NC 27514-2820
Phone: (919) 933-9044
www.acpa-cpf.org
An international non-profit medical society of health care professionals who treat and/or perform research on birth defects of the head and face.

AmeriFace
PO Box 751112
Las Vegas, NV 89136-1112
Phone: (888) 486-1209
http://www.ameriface.org
info@ameriface.org
Provides educational and emotional support to persons born with craniofacial anomalies and their families.

Children’s Craniofacial Association
13140 Coit Road, Suite 307 Dallas, TX 75240
Phone: 800-535-3643 Fax: 214-570-8811
www.ccakids.com
e-mail: contactCOA@ccakids.com
This group provides information, a newsletter, parents’ networks and an assistance fund for families who must travel for medical care.

Cleft Palate Foundation (CPF)
1504 East Franklin Street, Suite 102, Chapel Hill, NC 27514-2820 USA
Phone: (800) 242-5338
www.cleftline.org
Produces free publications and provides information about clefts and other craniofacial anomalies.

Craniofacial Foundation of America
975 E. Third Street, Chattanooga, TN 37403
Phone: 423-778-9176
www.craniofacialfoundation.org
Their extensive web site is a graphical consumer’s guide to many craniofacial surgical procedures.

FACES - The National Craniofacial Association
P.O.Box. 11082, Chattanooga, TN 37401
1-800-3FACES3
www.faces-cranio.org/
e-mail: faces@faces-cranio.org
The National Craniofacial Association is a non-profit organization serving children and adults throughout the United States with severe craniofacial deformities resulting from birth defects, injuries, or disease. There is never a charge for any service provided by FACES.

Foundation for Faces of Children
258 Harvard St., # 367, Brookline, MA 02446
Phone: (617) 355-8299
http://facesofchildren.org
info@facesofchildren.org
Provides patients and families with information about facial differences, and advocates for the best care possible for children with facial differences.

The Smile Train
41 Madison Ave., 28th Floor, New York, NY 10010
Phone: 1.800-932-9541
www.smiletrain.org
Email: info@smiletrain.org
Provides free cleft surgery and related treatment for children who would otherwise never receive it.
References


Krapels, IPC, Van Rooij, IALM, Weavers, R.A.; et al. MYO-inositol, glucose and zinc status as risk factors for non-syndromic cleft lip with or without cleft palate in offspring: a case-control study. BJOG 2004; 111: 661-668.


Orofacial Clefts


Shprintzen RJ. The velopharyngeal mechanism. In: Berkowitz S, editor. Cleft lip and palate Diagnosis and management. 2nd ed. Germany: Springer; 2005


Shprintzen RJ. The velopharyngeal mechanism. In: Berkowitz S, editor. Cleft lip and palate Diagnosis and management. 2nd ed. Germany: Springer; 2005


Orofacial Clefts

Post-Test

1. Which of the following statements regarding embryonic development of the face is FALSE?
   A. Formation of the upper lip and the primary palate occur in the 6th week.
   B. The maxillary prominence (lobes A & B) meet to form the upper lip.
   C. The separation of the oral and nasal cavities is completed by week 10.
   D. Formation of the palate is the last step in joining the five embryonic facial lobes.

2. Which of the following has been identified as a causative factor for orofacial clefts?
   A. Folate deficiency
   B. Vitamin A deficiency
   C. Acetaminophen overuse
   D. Presence of the GFT-Beta gene

3. Which of the following statements regarding orofacial cleft classification is TRUE?
   A. The Tessier Classification system is based on where the developmental arrest occurred during embryogenesis.
   B. The Van der Meulen Classification system numbers clefts 0 to 14 based on anatomical position.
   C. The LAHSAL Classification system subdivides the mouth into 5 parts.
   D. An incomplete hard palate and complete soft palate cleft is documented using the LAHSAL classification system as “...hS...”

4. ________ is a genetic syndrome characterized by mandibular micrognathia, U-shaped cleft palate, and glossoptosis.
   A. Van der Woude Syndrome
   B. Pierre Robin Sequence
   C. Treacher Collins Syndrome
   D. Apert Syndrome

5. Which of the following statements is INCORRECT regarding nursing and feeding of an infant with an orofacial cleft?
   A. It is common for infants with a cleft palate to experience nasal regurgitation.
   B. Feeding should last at least 30-40 minutes to allow the infant to receive adequate nutrition.
   C. The Pigeon bottle has a Y-cut nipple with a thick and thin side and a one-way valve.
   D. Breastfeeding can begin (resume) immediately following a cleft lip repair and 1 day after cleft palate repair.
6. The _________ surgical technique narrows the velopharyngeal space, reassigns the levator veli palatini muscle fibers, and results in decreased incidence of VPI.
   A. Von Langenbeck Palatoplasty
   B. Furlow Z-Plasty
   C. Palatal Lengthening – V-Y Pushback
   D. Alveolar Bone Grafting

7. Children with orofacial clefts have a high prevalence of _________________.
   A. otitis media secondary to Eustachian tube dysfunction
   B. deafness secondary to auditory ossicle malformation
   C. vertigo secondary to inner ear hyperplasia
   D. tinnitus secondary to dorsal cochlear nucleus occlusion.

8. Which of the following is NOT one of the common speech error patterns experienced by individuals with cleft lip and palate?
   A. Abnormal backing of oral targets to postuvular place.
   B. Nasal fricatives
   C. Hyper-pressurized oral consonants
   D. Developmental articulation/phonological errors

9. Which of the following therapeutic activities are recommended to facilitate correct speech sounds in individuals with cleft palate?
   A. Oromotor exercises
   B. Electrical stimulation
   C. Both A & B
   D. Neither A nor B

10. Which statement regarding dentition is TRUE?
    A. A deciduous canine tooth that turns inward towards the palate is a serious problem.
    B. Deciduous teeth must be corrected to improve the position of the permanent front incisor teeth.
    C. Deciduous teeth are used to hold appliances that reshape the upper dental arch.
    D. Final orthodontic correction should occur between 16-18 years of age.