EMBRYOLOGY

The development of the orofacial region is a complex process involving exact timing and multiple interactions between primordial structures. By the 4th week of intrauterine life (i.u.l), the primitive face consists of the five embryologic processes; frontonasal process above, a maxillary process on either side and two mandibular processes below, all of which are derived from the first pair of pharyngeal arches, and forms the boundary of the stomodeum. The frontonasal process shows olfactory pits which divide it into medial and lateral nasal processes. The rounded end of the medial nasal process (globular process) gives rise to the premaxilla, prolabium, columella and the apex of the nose. The lateral nasal processes forms the alae2,6.

At the 7th week of i.u.l, each maxillary process grows forward above the stomodeum and fuses with the lower edge of the lateral nasal process. It then extends across the lower margin of the olfactory pit to reach and unite with the medial nasal fold. The fusion of the maxillary processes with the nasal processes eventually forms a continuous ridge above the stomodeum from which the upper lip develops6.

Two maxillary extensions of mesoderm grow medially beneath the olfactory pits, joining in the midline and forming the primary palate. On each side of the face, the maxillary mesoderm gives a medially directed shelf-like projection called the palatal process, which extends as a free edge2,6. The two palatal process fuse at about the 9th week and fusion occurs in the soft palate area by the 11th week of i.u.l. These processes are first directed vertically downward on either side of the tongue, with the tongue projecting posteriorly between them. The palatal processes take a horizontal position as the tongue descends which eventually lead of fusion. The palate behind the incisive foramen, which is formed by the fusion of the two palatal shelves, is referred to as the secondary palate.

The embryologic basis for the formation of CL/P rests on the failure of the mesenchymal masses derived from any of the five facial prominences to meet and eventually fused6.

A cleft of the lip and palate (CL/P) is one of the commonest major birth defects1,2. The unexpected birth of a baby with CL/P is shocking and traumatic experience, generating anxiety for parents as well as the attendant health care team. In addition to the obvious physical defect, infants with CL/P are at risk for many other health problems, including nutritional problems, chronic ear infections, speech disorders, hearing defects, dental problems and psychosocial maladjustment1,3. Furthermore, approximately 50% of patients with CL/P have other associated malforma-

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tions, including many complex syndromes. Management of patients with CL/P is best done by a multidisciplinary team of caregivers who see sufficient numbers of patients to maintain clinical expertise in this area.

In Sub-Saharan Africa, between 60% and 80% of child deliveries take place at primary health care facilities manned by primary health care workers (PHCWs) including nurse-midwives, nursing assistants, auxiliary midwives, community health officers, and trained traditional birth attendants and in few cases general medical practitioners. For a child with CL/P, the newborn period is one of the most important and challenging times for the baby and the parents. In the care of patients with CL/P in Sub-Saharan Africa, PHCWs roles may be invaluable when dealing with issues of feeding, recurrent infections, psychosocial problems and development delays. By providing appropriate initial health care needs for infants and children with CL/P and referral to a cleft team, the PHCW could minimize morbidity in children with this birth defect and favour surgical scheduling at the right time.

CLASSIFICATION

CL/P varies in severity, from small notches in the lip to clefts that extend through the alveolar ridge in the mouth and involves the floor of the nostrils/palate. There is no entirely satisfactory system of classification for orofacial clefts and this is reflected in the wide variety of presentations. It is however, generally agreed that cleft lip with or without cleft palate represents varying degree of the same embryologic defect while isolated cleft represents a separate entity.

Kernahan and Stark proposed a classification using the incisive foramen as the dividing line between the primary and secondary palate:

- Clefts of the primary palate may involve only the lip or the lip and the alveolar process as far back as the incisive foramen.
- Clefts of the secondary palate may involve the soft palate only or the soft palate and hard palate as far forwards as the incisive foramen.
- Clefts involving both the primary and secondary palate.

When CL/P is described by the extent of the tissue involvement, they are unilateral or bilateral, and incomplete or complete. CL/P involving only one side of the face is unilateral and when both sides are involved they are bilateral. If the cleft is unilateral, which side is involved is noted. Incomplete CL/P is defined as involving the lip and the anterior part of the maxilla. Complete CL/P includes the lip, anterior part of the maxilla, and the hard and soft palate. Clefts of the lip and palate are more common on the left side and when the cleft is bilateral, the left side is usually the more severely affected. Submucous cleft palate occurs when muscular union across the velum is incomplete, despite an intact mucosal surface. CL/P may occur in isolation (nonsyndromic clefts) or in association with other anomalies (syndromic clefts).

EPIDEMIOLOGY

The incidence of CL/P is known to vary according to parental race/ethnicity and geographic origin, the sex of the embryo, and the family’s socioeconomic status. When racial differences were considered, the incidence of CL/P has been found to vary from 2.1/1000 in Asians, 1.0/1000 in Caucasians and 0.41/1000 in Black. Abyholm reported that the incidence of CL/P is highest in Mongoloids, low in black people and intermediate in Caucasians. Isolated cleft palate however rarely shows any coherent pattern of variation among races. Actual rate differences for cleft are usually attributable to underlying variation in the populations from which the different samples have been drawn. Such variations include variation in genetic susceptibility, basic differences in facial width among races and variation in environmental exposure.

Studies of the incidence and prevalence of CL/P also shows large variations in pattern among countries. The figures obtained vary from 18.2 per 10,000 live births in China, to 700 live births in the United States of America. In Malaysia, an incidence of 1.24 per 1,000 live births was reported. Iregbulum reported an incidence figure of 0.34 per 1,000 for cleft lip with or without cleft palate and 0.05 per 1,000 for isolated cleft palate in a study in Nigeria. Several reasons have been suggested for the possible increase in the incidence of CL/P recorded by some authors in recent time, among which include:

1. Better general and specific treatment for clefts, resulting in better social acceptance and higher fertility and fecundity.
2. Intermarriage of cleft patients or carriers.
3. Decreased mortality among cleft patients.
4. Better diagnosis and registration of cases.
5. Increasing exposure to environmental factors (drugs, diseases, pollution, and others).

GENETICS AND TERATOGENS

Despite decades of intensive investigation into the cause of CL/P, the pathogenesis is still not clear. Multiple genetic loci as well as exposure to various environmental teratogens have been implicated. When
evaluating the inheritance of oro facial clefting, it is necessary to distinguish syndromic and non syndromic clefts. The genetics of a cleft that occurs as part of a syndrome cannot be evaluated apart from the syndrome itself. Parents of children with non-syndromic cleft and the affected child have approximately a 4% risk of cleft occurrence in each succeeding child. If more than one person in the family has a cleft, then the risk rises to 10% to 12%. The unaffected siblings of a child with a cleft have an approximate 1% chance of having a baby with a cleft. If more than one family member has a cleft, the risk of cleft for offspring of unaffected siblings rises to 5-6%. PHCWs may need to refer parents/families to certified genetic counsellors for diagnosis and counselling regarding recurrence risk and prognosis for patients with complex cleft syndromes.

Exposure to various environmental teratogens during the first trimester of pregnancy has been reported to interfere with lip and/or palate formation. Many teratogens including steroids, anticonvulsants (phenytoin), diazepam, ethanol, 13-cis retinoic acid, methotrexate have been shown to cause clefting. Infections such as rubella and toxoplasmosis during first trimester have also been associated with clefting. A positive association have been found between maternal smoking and the risk of having a baby with a cleft. If more than one family member has a cleft, the risk for offspring of unaffected siblings rises to 5-6%. PHCWs may need to refer parents/families to certified genetic counsellors for diagnosis and counselling regarding recurrence risk and prognosis for patients with complex cleft syndromes.

**DIAGNOSIS OF CLEFTS**

Diagnosis of cleft lip can be made in utero by ultrasound examination. With the use of transvaginal sonography, accurate detection of cleft lip can be made at 13 to 16 weeks gestation, however the exact severity of the deformity may be difficult to assess. Reports on the use of transabdominal sonography in detecting cleft malformation has not been consistently successful due to its inferior resolution and lesser frequency when compared with the transvaginal approach. Detection of isolated cleft palate in utero may be very difficult because of its posterior and mid cephalic location. The diagnosis of CL/P is most commonly made at birth as a result of the infant’s characteristics physical appearance. In a cleft lip, there is a separation of the two sides of the lip. The separation often includes the bones of the upper jaw and or gums. Infants with cleft lip also have a characteristics nasal deformity that involves partial collapse or flattening of the nose and flaring of the alar base on the side of the cleft. Shortening or absence of a strip of tissue (columella) that connects the tip of the nose to the upper lip and separates and nostrils is also a frequent occurrence. Cleft palate is an opening in the roof of mouth, the two sides of the palate did not fuse as the unborn baby is developing. When examining the palate, the entire palate back to the tip of the uvula should be visualized. Gentle palpation of the palate with a finger may detect a notch in the posterior border of the hard palate suggestive of a submucous cleft. Other signs of submucous cleft include a bifid uvula or a translucent central zone.

When CL/P in a newborn is diagnosed at primary health care facility, the PHCW can assist in determining whether the birth defect is an isolated CL/P or potentially syndromic. This is more important because many syndromes associated with clefts can involve life-threatening malformations. Anomalies most commonly associated with CL/P are heart defects, ear deformities, skeletal malformations and genitourinary problems. Parents of children with CL/P have often stated that one of the most stressful situations is to be isolated without interaction by hospital staff during the initial days following birth. The PHCW should therefore provide adequate information about the birth defect to the child’s parents. It should be emphasized that the child is not in pain because of the deformity and that the condition presents no immediate threat to the infants’ survival and it can be surgically treated. A cleft specialist or cleft should be identified by PHCWs for adequate referrals of cleft cases.

**SPECIAL ISSUES FOR THE PHCWs**

**Feeding**

Feeding difficulties are often the worst problems presented by infants with CL/P and are caused by insufficient suction, milk regurgitation through the nasal cavity, and low food intake. All of these factors aggravate the infant’s nutritional status, resulting in low weight gain. In some instances feeding difficulties has been associated with death of infants with CL/P in developing countries. The diet of an infant with CL/P has been associated with death of infants with CL/P in developing countries. The diet of an infant with CL/P is important to surgery scheduling. Too often feeding issues take priority over the parents’ need for support and proper information.

The PHCW should play a vital role in helping parents develop and maintain satisfactory feeding methods. Parents should be informed that the feeding problem is due to the anatomic defect. As a general neonatal orientation, all mothers are to be encouraged to try breast-feeding. A newborn with a cleft lip may be able to feed in ways common to all infants. The breast tissue in most instances fills the lip defect,
allowing the infant to develop the necessary negative intraoral pressure to feed efficiently. It is however, extremely challenging and unlikely that a child with cleft palate to have success feeding directly from the breast. Frequently excessive air intake occurs, leading to burping and nasal regurgitation. The use of digital pressure or breast pump to express breast milk in a cup and using a spoon to feed the child may be encouraged in some cases. Breast feeding is beneficial in promoting optimal growth, easy digestibility and greater mother and infant bonding.

Parents are advised to feed the infant slowly and in an upright position, allowing gravity to prevent milk coming through the baby’s nose. Mothers should take their time with feeding and burp the baby frequently. Infant with cleft palate tends to swallow a lot of air during feeding. Where breast-feeding is not possible, information about other resources of feeding should be given to parents. These may include the use of compressible feeding syringe, dropper and a tube. When using a compressible bottle, parents should not rush and should direct the flow of milk to the side or back of the child’s mouth. Infants with cleft palate, typically take a very long time to feed and still take a small amount of formula from the bottle. Well-informed parents have been shown to have less stress with feeding issues. PHCWs are expected to monitor child’s weight gain and parent-child bonding difficulties and intervening appropriately. Whatever feeding method is used, weight gain in infants with CL/P may lag behind established normal weights of infants without clefts in the first 1 to 2 months of life but is usually comparable by 12 to 18 months.

**Middle Ear Diseases**

The presence of conductive hearing loss in patients with cleft palate has been known for more than a century, with Eustachian tube dysfunction being the primary cause of middle ear disease. Otitis media is an almost universal finding in a child with cleft palate, and long-term permanent hearing loss incidence ranging from 0-90% has been reported. The underaeration of the middle ear mucosa results in sterile ear mucosa inflammation and accompanying middle ear effusion with superimposed infection. As in other children, infection originating in the upper respiratory tract may also play a significant role. Middle ear disease can cause linguistic problems because language learning can be influenced negatively. Screening audiology is usually performed before cleft palate repair to detect children who may benefit from the insertion of ventilating tubes.

A patient with cleft palate typically requires long-term monitoring of the ears, nose, and throat due to the high prevalence of ear disease. Physical examinations of the ears are to be carried out by PHCWs on a regular basis, beginning within the first six months of life. Many patients with cleft palate may continue to have recurrent infection, necessitating multiple courses of antibiotics. Prompt referral to a cleft specialist may be indicated in some difficult cases.

**Dental Care**

Patients with CLP will require dental speciality services as a direct result of the defect. Children with clefts are at increased risk for congenitally missing teeth, displaced teeth, impacted teeth, supernumerary teeth and hypoplastic teeth. A higher incidence of neonatal teeth has been reported in patients with bilateral clefts when compared with those with unilateral clefts. Higher caries prevalence in the primary and permanent dentition has been found in children with clefts when compared with those without clefts.

In patients where the cleft involved the alveolar ridge, the lateral incisor may be absent, displaced or replaced by supernumerary teeth. In addition patients with CL/P are at higher risk for gingivitis, crossbite and teeth crowding.

The treatment of dental problems in patients with CL/P should be an integral part of the rehabilitative process and efforts should be made to ensure that dental procedures are as less traumatic as possible. Orthodontic treatment may be needed in various stages of child’s life. In the immediate newborn period, the use of oral orthopaedic splints or obturators may be used, to aid feeding and limit abnormal tissue displacement. As the primary dentition erupts, dental examination and referrals to appropriate providers for caries control, preventive measures, and space management be done. Dental and skeletal components are usually evaluated often with the aid radiograph, to determine if malocclusion is present or developing before primary dentition is completed. Depending on the age at which patient is first seen and also the goals to be accomplished, orthodontic management of malocclusion may be performed in the primary, mixed or permanent dentition. Alveolar bone grafting using cancellous bone from the iliac crest to correct alveolar cleft is often performed between 9 and 12 years of age when the canine root is 25% to 30% formed. This is to restore normal alveolar architecture to allow teeth to erupt normally and subsequently be moved orthodontically into the cleft site.

PHCWs as a matter of routine be alert to the developing dentition of a child with CL/P. Early counselling regarding good oral hygiene and prevention of caries in the child should be provided to parents. Counselling regarding the prevention of nursing bottle caries is to be emphasized. The importance of balanced diet with minimum of refined carbohydrates should be...
stressed and fluoride supplements be prescribed as appropriate. Pathologist may provide regular evaluation over a long period to monitor the child progress.

Social and Psychological issues

Although it is well established that 3% to 4% of all pregnancies result in the birth of an infant with a major birth defect, the majority of couples often approach pregnancy anticipating the birth of a healthy, structurally normal child20. The initial shock in parents caused by the discovery of a cleft is usually followed by fear, anger, guilt, and sadness. This may be more difficult in Sub-Saharan Africa, where in most cultures, myth and superstitions commonly accompanied the birth of a physically defective child21. The parents are often blamed and the families are shamed21. Infanticide following the birth of a child with congenital anomalies has been reported in some communities in Africa21,22.

PHCWs should provide important information about the scientific causes of CL/P to the parents and family, this should however, be done in a culturally sensitive manner. PHCWs need to be aware that even highly educated parents may have an incorrect idea concerning the etiology of CL/P, many of which imply personal fault22. It will be critical for these parents to be told that the CL/P is not their fault.

The presence of a child with a cleft places an enormous emotional burden on the child’s parents and may be impaired by feeding difficulties and feeling of maternal guilt12,20,21. PHCWs must be alert to difficulties in child and family psychosocial adjustment, and must provide support and understanding and work to foster positive relationships between the parents and the child with a cleft. Facilitating meetings with other families with children with similar conditions has been reported to decrease the negative social effects20. The psychosocial effects of CL/P often extend over the infancy, childhood, and adolescence of individuals who are born with this defect. As the child grows, poor self-esteem and impaired socialization may affect peer relationship and may produce a prolonged dependence on adults. In most cases, it is not only the individual’s own perception of the condition that contributes to the effects, but the reactions of the people in his or her environment as well22. PHCWs should provide early intervention by counselling parents regarding the possible social effects and expectations for growth and development. Assessing patients for depression is essential and prompt referral to a specialist for further counselling when indicated is necessary.

Treatment

Surgical goals of CL/P repair are directed toward achieving a normal facial appearance, feeding, speech, and hearing without significantly affecting the ultimate facial and psychosocial development of the child22. This is best achieved under a multidisciplinary team usually composed of a plastic surgeon, oral surgeon, speech pathologist, orthodontist, social worker, audiologist, and nutritionists.

The timing for CL/P surgery is controversial. The cleft lip is usually repaired when the child’s general health permits the safe use of general anaesthesia A commonly quoted guide is the ‘rule of tens’ which refers to weight greater than 10 lb, haemoglobin greater than 10gm/dL, and age greater than 10 weeks6,23. In general lip repair is done between the ages of 3 months21,24. By this time, anatomical lip landmarks have become more apparent, there has been time to fully bond with the infant, and the parents have had time to adjust to the cleft and the operation and necessary rehabilitation that may follow23. Cleft palate surgery is directed to palatal closure with a technique and timing that produce optimal speech and minimize facial growth disturbances. Usually the cleft palate is repaired when the child is between 12 and 18 months of age. Early repair has been associated with disturbance of midfacial growth. Speech development is adversely affected if palate closure is delayed past 2 years of age24.

Patients routinely undergo blood investigations and radiographs before surgery and are usually admitted to the hospital few days before the day of the surgery. Repair of cleft of the lip and that of the palate requires general anaesthesia with endotracheal intubation and may last for about 1 hour. Postoperative feeding protocols vary widely, with some cleft team recommending an immediate return to the breast or the bottle and others using syringe or tube feeding23. After primary repair the patient may often need to undergo further staged reconstructions later in life. Palatal and nasolabial fistula occur in 5% to 20% of patients after palatal surgery and is usually corrected by surgery12. Pharyngeal flap surgeries following palatal repair are done to improve on speech defect and are usually done from age 3 to 7 years23. Lip scar revision, if necessary, is often done when the child approaches school age and appearance becomes important23. Alveolar bone grafting is generally performed at the age 9 to 12 years.

For appropriate surgery schedule, PHCWs are essential in ensuring routine paediatric care, such as immunizations, assessment and monitoring of nutritional intake, and weight gain. PHCWs should provide appropriate information to parents on surgical treatments of cleft and assist families on the emotional adjustments to care. PHCWs must not see treatment of clefts as a simple closure of a wound1,17,19, but as a birth defect with lifelong physical, social, and psychological effects.
CONCLUSION

In Sub-Saharan Africa where three out of every four deliveries take place at the primary level of health care, the PHWCs have a vital role for families of children with congenital anomalies such as CL/P. PHCWs should provide proper information at a time when parents have questions. They should address the physical and psychological issues that arise. PHCWs can be a helpful link between families and the cleft specialist or a multidisciplinary team.

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