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All That An Obstetrician Should Know About Cleft Lip and Palate

Adenwalla *H5,* Narayanan *PV,* Rajshree *CJ*
Charles Pinto Centre for Cleft Lip and Palate, jubilee Mission Hospital, Trichur - 680005

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Introduction

There are many distressing moments that an obstetrician meets with in the dramatic situation that is associated with the birth of a child. One of these is to be faced with the predicament of explaining to a mother that her child has an unfortunate deformity of a cleft lip or a cleft palate or both. However, with the present state of the art these children can be restored to normal appearance and function. The moment calls for sensitivity and a great deal of precise information.

Once the parents recover from the shock of realization of this deformity, they have a host of questions to ask and there is no better person to answer these questions than the knowledgeable obstetrician who already has the parents' confidence. The pediatrician and the reconstructive surgeon come into the picture much later. It is also imperative that these three specialists should complement each other and should not contradict each other on basic information, so that they cause no conflict in the minds of already distressed parents. The obstetrician must exude confidence so that the mother takes her baby home optimistic and reassured that her child is going to be normal.

Incidence

Clefts of the lip and palate and other facial clefts constitute the commonest deformity of the face and the second commonest congenital deformity that one is faced with. One in 1000 to 1 in 1500 children born in this country manifest a cleft lip, a cleft palate or both. From the analysis of cases seen in our department (Table I) one can infer that

1) There is a predominance of males over females.
2) A predominance of left sided clefts over right sided cleft
3) The commonest cleft is a unilateral cleft of lip and palate
4) The rarest cleft is a cleft of the lower lip.

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Correspondence :
Dr. Adenwalla H.5.
Charles Pinto Centre for Cleft Lip and Palate,
Jubilee Mission Hospital, Trichur - 680 005.

Literature offers no logical explanation for these consistent findings in all the reported series that we have scrutinized.

Table I: Types of clefts encountered at our center (January 1957 to May 2003)

	Right	Left	Male	Female	Total
Unilateral cleft lip	723	1,089	1,027	786	1,813
Bilateral cleft lip			367	322	689
Cleft palate			785	727	1,512
Unilateral cleft lip and palate	1,330	1,982	1,850	1,466	3,316
Bilateral cleft lip and palate			876	700	1,576
Median cleft lip			47	44	91
Lateral cleft lip			73	66	139
Oro-ocular cleft			33	25	58
Cleft of lower lip			1	1	2
Total			5198	4001	9,196

Type of clefts

The obstetrician will commonly come across the classic defect of the lip or palate or both. In this the cleft goes through the floor of the nose and is commonly referred to as a "hare lip" which is really a misnomer as the hare has a cleft in the center. However clefting can take place along embryological fusion lines. These are the rarer clefts going into the eye, to the ear and the still rarer cleft of the upper lip in the midline, clefting the nose. The rarest cleft is a cleft of the lower lip (Table I).

Though various methods of classification of clefts exist the one most commonly followed is that of Kernahan[1]. The extent of clefting is depicted on a "Y". with the clefts of the primary palate being anterior to and those of the secondary palate posterior to the incisive foramen, which is at the junction of the limbs as shown in the (Fig. 1). It is imperative that the obstetrician and the pediatrician be familiar with this classification so that they and the reconstructive surgeon can communicate with one another perfectly when they encounter a child with a cleft.

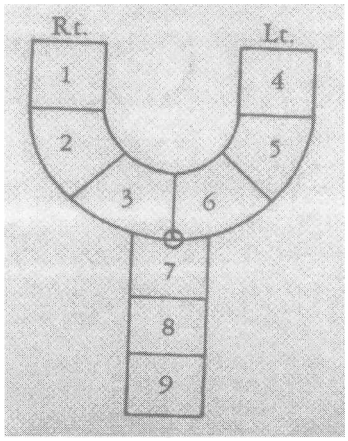

Figure 1: The right and left limbs of the "Y" are divided into three sections : the anterior portion = lip (1 and 4), the middle + alveolus (2 and 5) and the posterior = the area of the hard palate from the alveolus back to the incisive foramen (3 and 6). Posterior to the incisive foramen the vertical limb of the "Y" is divided into three sections. Hard palate (7 and 8) and soft palate (9).

A cleft of the primary or secondary palate is partial if it does not extend up to the incisive foramen and complete if it does.

The obstetrician must be aware of some of the not so typical conditions. First amongst these is the microform cleft lip, which manifests with one or all three of the following defects: (1) a nose with the classic cleft deformity (2) a scar on the body of the lip as if the child has been operated on (3) a notch on the vermillion. A child with a microform cleft lip requires surgical correction. A minor nasal deformity if it exists by itself may be ignored.

A submucous cleft palate has a greater chance of being missed unless the clinician is aware of the condition and looks for it before the child begins to speak. The three criteria for the diagnosis of a submucous cleft palate are: (1) a notch on the hard palate (2) a blue line running down the center of the soft palate (3) a bifid uvula. A child with such a palate has a deficiency of muscle and sometimes speaks with velopharyngeal incompetence. If he does so, he requires a formal repair with a good intravelar dissection and retroposition of the levator palati muscle.

An even more subtle condition is a submucous occult cleft palate. The palate looks completely normal on inspection through the mouth but on nasoendoscopy shows a furrow on the nasal aspect of the soft palate. If a child with such a palate has velo-pharyngeal incompetence he requires a formal repair like a submucous cleft palate. All the above three conditions are due to late intrauterine fusion of the facial elements.

It is nice for the clinician to know that one can get a child with velo-pharyngeal incompetence due to other causes. For example, paralysis of the soft palate and occasionally a capacious pharynx can cause a cleft like nasal speech.

Prenatal Diagnosis

Antenatal ultrasound is now performed routinely to gain information about fetal well-being and to detect congenital defects. Clefts of the lip and palate are diagnosed both incidentally on routine scans and also on specific referrals in at risk patients. Detection rates of clefts of lip alone, or of lip and palate together, are higher than that for cleft palate alone. However, with the use of a color Doppler ultrasound, detection rates of isolated cleft palates can also be improved", Prenatal ultrasound has been able to spot cleft in 22-33% of cases. Diagnosis is significantly improved if the scan is done after 20 weeks".

Counseling

Once a diagnosis of a cleft lip and palate is made on prenatal ultrasound, many questions will arise in the minds of the concerned parents and would naturally be directed first at the obstetrician.

The following are the common questions asked:

1) Why has the defect occurred?

Orofacial cleft may arise from an interaction of genetic and environment factors.

i) Genetic Factors

About 20 genes (called orofacial genes) have been suggested as causes of clefts. With advanced research, the gene list is getting longer and making the etiology more complex.

Family studies have shown that siblings of patients with cleft lip with or without cleft palate (CL/P) have an increased incidence of CL/P but the same is not true of isolated cleft palate (CP). Conversely siblings of patients with CP have an increased incidence of CP but not of *CLIP* [4].

About 3% of clefts are associated with other syndromes and in these the cause may be mutation of genes, chromosomal aberrations or genetic changes secondary to drug or alcohol ingestion.

ii) Environmental factors

The list of environmental factors involved in the formation of clefts is increasing gradually. Though they are less important than the genetic factors, these can be controlled, and hence form a means of

minimizing the occurrence of clefts. Examples of such environmental factors ares :

1. The anti-epileptic drug phenytoin
2. Exposure to carbon monoxide
3. Maternal hypoxia
4. Cigarette smoking
5. Deficiency of folic acid and excess of vitamin A. The anti-acne drug accutane (isotretinoic acid) can cause clefts.
6. Anecdotal reports exist of increased incidence of clefts in children born to mothers living at higher altitudes, presumably due to hypoxia.

Many of these agents act by inhibiting the electron transport chain and consequent ATP production.

2) Can there by other congenital anomalies along with the cleft ?

Various studies indicate that fetuses diagnosed with clefts may have other congenital anomalies",

Clefts may be associated with defects of the brain (median clefts), cardiovascular anomalies (velocardiofacial syndrome) etc. In all about 400 syndromes involving facial clefts have been described. In clinical practice, the number of cleft children seen who are syndromic is much less, presumably because of the mortality related to the more serious syndromes.

3) Should the pregnancy be terminated?

This decision rests entirely with the concerned parents. However, once the nature of the defect, and the fact that it is entirely treatable are explained to the parents, they could be expected to opt to continue the pregnancy. In a study by Mathews et al", of the nine families who returned the written survey none would consider abortion for an isolated cleft.

4) What is *the chance of a cleft in future pregnancies?*

The predicted recurrence in subsequent pregnancies based on reported data is given in Table II.

Table II : **Recurrence in future** Pregnancies.

	Predicted Recurrence	
	CLIP	CP
One sibling	4.4	2.5
One parent	3.2	6.8
One sibling and one parent	15.8	14.9

The risk to siblings born of unaffected parents rises from 4.4% to approximately 9% after two affected children are born", Armed with such data, the parents will be in a better position to make an informed decision regarding future pregnancies.

5. What are the problems that can arise due to the cleft and how can they be ameliorated ?

It has often been said that the untreated or badly treated cleft can affect every function of the face except sight, and even sight can be affected in an oro-occular cleft; not to mention the severe psychological trauma that it inflicts on both parent and child.

A cleft palate is associated with malnutrition, malocclusion, deficiency in maxillary growth, Eustachian tube malfunction, middle ear disease (going on to deafness) and repeated respiratory infection (sometimes of a serious nature). Problems can arise in speaking, alimentation, hearing, breathing, taste and smell. Besides there can be distortion of normal facial appearance and expression while speaking. Communication, a basic need of all mankind, becomes hesitant; lack of communication may lead to severe retardation of mental growth and the general ongoing process of learning which is so important for the normal development of a child.

i) Feeding

There has been an advance in feeding techniques, bottles and other feeding equipment in the West in recent years. One of them is a valvular pressure-controlled feeding bottle. None of these are cost effective and therefore, are not applicable to conditions in our country.

Mothers should be advised to breast feed their children. Babies with some of the smaller clefts can be effectively put to the breast and can suckle. If they can't, then the breast milk should be expressed under sterile conditions and given to the child. Breast milk should be given to the cleft baby as long as possible keeping a close watch on weight gain consistent with the age of the child. If the child does not gain weight as required the feeds must be supplemented with either infant feeding formula or full strength undiluted cow's milk (a good and cheaper alternative).

Regarding the mechanism of feeding, a bottle with a larger hole in the nipple is tried first; if this fails the child is fed with an indigenous baby feeding cup (Fig.2)

Spoon-feeding is messy and leads to a lot of spillage. Very rarely the infant may have to be tube-fed for a few days. Every mother should be provided with a feeding

chart and she and her pediatrician should watch and record the baby's weight gain. If the child does not gain weight in proportion to its age as given on the chart there must be something wrong with the child or the feeds administered. It is imperative that a healthy well-nourished child must be presented to the surgeon for reconstruction. An iron tonic is usually advised after 6 weeks of age and continued through surgery, as it is best to avoid blood transfusions prior to or during surgery due to the hazards involved.

All cleft children must be burped several times during the feed as they tend to swallow more air during a feed than a normal child. If this is neglected the child either rejects the feed or vomits due to a full stomach.

A cleft child is more prone to episodes of diarrhea and the mother must be advised to consult a pediatrician in such an eventuality without much delay.

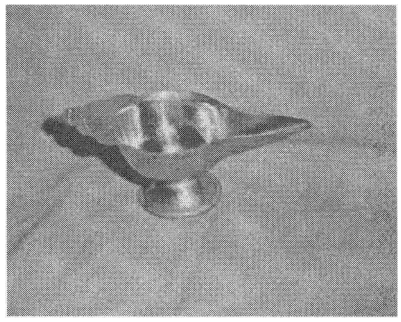

Figure 2 : Indigenous baby feeding cup.

ii) Respiratory infection

Cough, colds and respiratory tract infection are very common especially in cleft palate children and continue to be so till the palate is repaired. However, this is no reason for a pediatrician to insist on early surgery on an unfit child. It is hazardous to operate on a child within 10 days of a respiratory infection.

Respiratory distress of a serious nature, obstructive in etiology, may be seen in the early days of a Pierre Robin sequence which is usually associated with a wide high arched "U" shaped cleft palate. The condition is self-limiting and disappears as the child grows. In most cases it suffices to nurse the child in the lateral or prone position. During the last 44 years of cleft surgery we have had to perform a glassopexy just twice. In most cases the retrognathism also disappears and in only a few cases do these children require a genioplasty after the age of 16 years.

Another cause of breathing difficulty in a cleft child is laryngomalacia. These children are more comfortable in the propped up position. At times this distressing problem manifests in the immediate post-operative period. Other rarer causes of breathing difficulty in a cleft child can be an associated diaphragmatic hernia or an eventration of the diaphragm. A tracheo-esophageal fistula may be an even rarer cause.

iii) ENT problems

Cleft palate children prior to and sometimes for a short period after the surgical closure of the palate are prone to the formation of glue in the middle ear, which if neglected, can go into an acute or chronic otitis media (this complication takes place due to malfunction of the Eustachian tubes). The early insertion of grommets to prevent hearing loss cannot be overemphasized, remembering always that"a child who does not hear well will not speak well". The ENT surgeon must be advised to avoid removing the adenoids of a cleft palate child as this may worsen the child's velopharyngeal incompetence and in the process worsen his speech. He must also be guided that if a tonsillectomy is indicated it must be done with the utmost care to preserve the palato-pharyngeus muscle (the posterior pillar of the fauces) which is vital for performing a sphincter pharyngoplasty if needed for a particular child.

io) Mental Status

An obstetrician should be aware of the fact that a syndromic cleft child or a child with a median cleft, and rarely an ordinary cleft child may be mentally retarded.

All syndromic and median clefts of the face must have a CT scan of the brain. Median clefts with absence of pre-maxilla and hypotelorism usually have severe impairment of brain development and a very poor life expectancy" and therefore should not be operated upon. They are also prone to post-operative hyperpyrexia and convulsions.

v) Ophthalmic problems

In an oro-occular cleft the mother must be instructed about the care of the eye. It is the responsibility of the reconstructive surgeon to get the eye covered so that the child may not lose its sight due to exposure keratitis. This is a tragic and avoidable catastrophe.

vi) Care of the pre-maxilla and prolabium in *bilateral clefts*

A projecting pre-maxilla is deprived of salivary wash. Due to drying, cuts and rhagades form on the mucosal surface of the prolabium; these can get infected and cause undue delay in surgery The mother must, therefore, be instructed to keep the prolabial mucosa and premaxilla

moist with frequent application of potable water.

vii) When should treatment be commenced and what is *the current protocol of surgical management?*

There is no hard and fast rule as to when a cleft lip should be repaired, neither is there a universal consensus. While prenatal surgery is feasible, it is questionable. In our opinion as the state of the art exists today, the disadvantages far outweight the advantages. The American propagate the idea that to avoid psychological trauma to the mother, the baby should be operated upon in the neonatal period. The rule of lOs which calls for repair at the age of 10 weeks is universally accepted by western surgeons. We do not think that this rule applies to our Indian babies who are much smaller and not so well nourished.

For quite some time now we have been operating on our unilateral cleft lips at the age of 5 months. The lip and nose are corrected at the same stage. The anticipated result is such that no secondary soft tissue operation is necessary on these children either on the lip or the nose. In our opinion such a result can only be achieved if there is adequate tissue bulk to work on with comfort. Hence, we do not advise or perform surgery in the neonatal period.

It is said that a cleft palate should be repaired before the child begins to speak but the tendency now is to repair the palate earlier - even as early as 9 months. We in our department operate on a cleft palate at 1 year 5 months. This takes into consideration both speech and the damage done to maxillary growth by too early a palate repair.

One must be aware that the use of a good cleft-trained speech therapist is vital. He can, however, not compensate for a badly repaired palate. His role is to eliminate minor velopharyngeal incompetence, faulty pronunciation, glottal stops, vocal fricatives and grimacing. An intelligent mother must work in conjunction with a cleft-oriented speech therapist to give the child an optimum speech result.

A bilateral cleft lip can be repaired one side at a time, the first side at 4 months and the other side at about 8 months of age. We repair both sides in one stage at about 7 to 9 months of age. We believe that we achieve a better lip and nose symmetry if we operate on both sides at one stage. For a bilateral cleft lip a columella lengthening is performed at 3 years of age. All clefts of the alveolus, unilateral and bilateral, require alveolar bone grafting at 7 years.

If a secondary rhinoplasty is necessary it should be performed at 7 years. A well done cleft lip should not require a secondary rhinoplasty. If there is maxillary regression, an orthognathic procedure, a Lefort I osteotomy is indicated after the age of 16 years.

If the child's speech manifests velopharyngeal incompetence one of the pharyngoplasties is performed at 12 to 14 years of age. Every attempt is made to minimize the number of operative procedures; at the same time, there should be no compromise on an optimum result.

What can be done to prevent clefts?

While little is known yet on how to prevent the orofacial clefts, studies suggest that taking multivitamins containing folic acid (400 micrograms per day) before conception and during the first 3 months of pregnancy may help prevent cleft lip / palate and isolated cleft palate. Ordinary vitamin supplements can reduce the incidence of cleft by more than 20%. But as mentioned before, vitamin A should not be given in excess. Mothers at high risk should be prescribed a larger dose of folic acid (1 mg. per day).

Fetuses with certain predisposing genes may be at increased risk for clefts if their mothers smoke or drink. Drugs such as some antiepileptics (phenytoin) have been linked to an increased incidence of cleft lip / palate. Mothers taking such drugs have to consult the concerned doctors to alter the therapy to some other safer drug during the pregnancy. Isotretinoin, an anti-acne drug related to vitamin A, can cause very serious birth defects including cleft lip and palate if a woman becomes pregnant while taking the drug even in small doses for a short period.

Reync'd et al[8] have shown in a study of mice that a combination of folic acid and methionine have a synergistic action in prevention of clefts. All these precautions must be taken before the mother becomes pregnant as clefting takes place in the fetus during the first trimester of pregnancy. If precautions are taken after the pregnancy is confirmed it might be too late.

The sum total of information on clefts and craniofacial anomalies is ever-increasing. The state of the art has progressed beyond recognition in a man's life time. The obstetrician needs to be well informed to actively participate in the counseling and treatment of these unfortunate babies. The concerned obstetrician, the pediatrician and the reconstructive surgeon form the tripod of support and hope for families that have been devastated by this misfortune.

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