

## CLEFT LIP AND PALATE: REVIEW ARTICLE

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Article Received on 18/05/2018

Article Revised on 08/06/2018

Article Accepted on 29/06/2018

### ABSTRACT

Cleft lip and palate are the most common facial deformity. It may involve lip only, lip and palate and palate only. The main reasons of clefting in infants may be either environmental (such as smoking, alcohol, poor nutrition) or genetic factors (such as familial factors and chromosomes). A number of specialists involves in treatment of clefting and decides the best treatment plan depending on the site of defect and age of the infant.

**KEYWORDS:** Cleft Lip & cleft palate, etiology, treatment.

### DEFINITIONS

Cleft lip is defined as a congenital deformity that occurs in the primary palate which is located anteriorly to incisive foramen. Its occurrence may be unilateral, bilateral, complete or incomplete. Cleft palate is defined as a congenital abnormality that occurs in the secondary palate (soft and hard palate). Its occurrence may be unilateral, bilateral, complete or incomplete.<sup>[1,2]</sup>

### Epidemiology

Generally, its incidence is about 1 in 700 births and there are about 1000 new cases in UK annually. It is most common in the left side than right side. Unilateral clefts are the most common than bilateral clefts with ratio of 9:1 and its occurrence is twice on the left side in comparison to right side. On the other hand, bilateral clefts are the most common on the left side than right side with a ratio of 6:3:1. According to the race, male are affected by cleft lip and palate than females with a ratio of 2:1. On the other hand, females show the highest incidence of isolated cleft palate. The incidence of clefting differs according to the nature, geography and ethnicity. For example, the occurrence of clefting in African people showed about 0.3 per 1000 while Caucasian and Japanese populations demonstrated about 1 and 2.1 per 1000 respectively.<sup>[1,2,3]</sup>

### Classification

Several authors classified the cleft lip and palate.<sup>[4]</sup> Veau (1931) classified the clefts into four main groups:

- Clefts of soft palate.
- Clefts of hard palate.
- Unilateral clefts of the lip, alveolus and palate.
- Bilateral clefts of the lip, alveolus and palate.

According to Koch *et al.*, (1995), Kernahan (1971) suggested a new classification of cleft lip and palate and it gives the shape of Y letter and includes:

- 1 and 4 represent the right and left side of the nasal floor, respectively.
- 2 and 5 represent the right and left side of the lip, respectively.
- 3 and 6 represent the right and left side of the paired alveolar segment, respectively.
- 7 represent the primary palate.
- 8 and 9 represent the secondary palate (Figure 1).

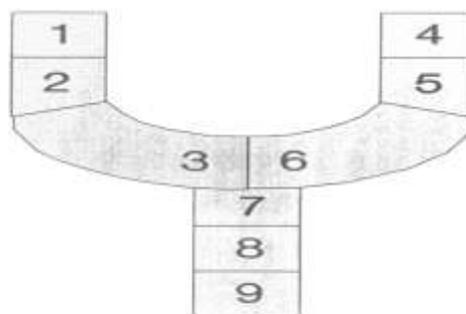


Figure 1: Kernahan classification of cleft lip and palate. (Koch *et al.*, 1995).

### Etiology

#### 1. Environmental factors

##### 1.1. Smoking and alcohol

Smoking and alcohol are considered as the main causes of clefts where high risk of developing a cleft is found in pregnancies with high alcohol abuse and smoking.<sup>[5,6]</sup>

#### 2. Nutritional factors

Nutritional status plays an essential role in developing cleft lip and palate. Vitamin B6 deficiency was the main

reasons of increased risk of clefts in the Netherlands and the Philippines.<sup>[1,6]</sup> In addition, folic acid deficiency was observed as the main reason of increasing clefts.<sup>[1,5]</sup> It was observed that the addition of folic supplement decreased the incidence of clefts in USA and North America.<sup>[6]</sup> Furthermore, zinc is considered as the main element in the fetal development. The deficiency of this nutrient cause cleft lip and palate. It was observed that mother with children with clefts had lower concentrations of zinc in comparison to mothers with children without clefts.<sup>[1,6]</sup>

### 3. Medications

Clefting of the lip and palate can result from some medications such as corticosteroid steroids in which some pregnancies take due to insomnia and anxieties. In addition, retinoid drugs are considered as one of the main reasons that cause clefts in infants because of exposing pregnant women to these drugs.<sup>[6]</sup>

### 4. Organic chemicals and solvents

Exposure to chemical and solvents can cause clefts of the lip, palate or both.<sup>[6]</sup>

### 2. Genetic factors

Family history is considered as one of the reasons may lead to cause cleft lip and palate. For example, the risk of transfer one parent having cleft lip and palate to their child is 9%. The risk of transfer unaffected parents having a child with a cleft lip and palate to their second child is 4%.<sup>[2]</sup> The occurrence of cleft lip and palate is either associated with many syndromes such as

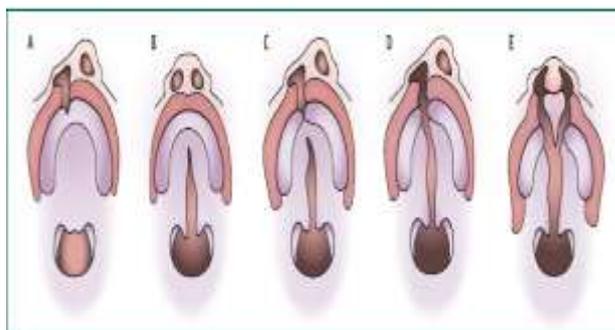
- Pierre Robin syndrome
- Sticklers syndrome
- Treacher Collins syndrome
- Hemifacial microsomia
- Ectodermal dysplasia

Velocardiofacial syndrome .So, in these cases, it called syndromes cleft palate.<sup>[2,3,5]</sup> On the other hand, clefts may occur without syndrome (non syndrome cleft palate).<sup>[2,5]</sup> In addition, other genetic factors which cause cleft lip and palate related to

- Trisomy 13
- Trisomy 18
- Trisomy 21 (1,2,3)

### Cleft types

The types of clefts are presented in the figure below.



**Figure 2: Types of clefts (Mossey *et al.*, 2009) A represents cleft lip and alveolus.**

B represents cleft palate.

C represents incomplete unilateral cleft lip and palate.

D represents complete unilateral cleft lip and alveolus.

E represents complete bilateral cleft lip and palate.

### Common problems associated with cleft lip and palate

The cleft lip and palate causes many problems:

#### 1. Speech problem

Patients with a cleft palate have speech problems which are result from velopharyngeal dysfunction. Inability of soft palate to move upward to provide a contact with nasal cavity results in a passing of air through the nose instead of oral cavity. This condition is known as hypernasality speech.<sup>[7]</sup> This case can be treated with a surgery to provide the velopharyngeal closure. Pharyngeal flap and sphincter pharyngoplasty are considered as the reliable surgeries for correcting the velopharyngeal deficiency in patients with a cleft lip and palate.<sup>[2]</sup>

#### 2. Hearing problem and ear infection

Otitis media is a condition where a fluid is accumulated in the middle ear and results in ear infection. This is due to the abnormal action of Eustachian tube opening by two muscles which are tensor veli palatine and levator veli palatine. This leads to the lack of ventilation to the middle ear cavity and accumulation of fluid inside the middle ear. This condition is presented in the child with cleft palate in the first six months of life.<sup>[5]</sup>

#### 3. Dental problems

Dental problems involve abnormalities in the size and shape of the teeth, For example, the permanent lateral incisor shows abnormalities in size and shape in the side of cleft, abnormalities in the position of teeth, delay of eruption of permanent teeth and delay of formation of permanent teeth.<sup>[5,8]</sup>

#### 4. Feeding and nutritional problems

Feeding problems in babies with cleft lip and palate occur because babies are incapable of sucking either their mother's nipple or from a bottle. Therefore, this affects the weight and growth of the baby because the amount of milk or food is not enough for growth. There are a variety of methods that enable the baby to feed and

gain a normal weight such as the use of disposable syringe, spoon and cup and prosthetic obturator device.<sup>[8,9]</sup>

### 5. Cosmetic problems

Patients with cleft lip have cosmetic problems and also cause problems for production of labial sounds. Babies with cleft lip face difficulty when they try to make a contact between upper and lower lips.<sup>[8,9]</sup>

### 6. Psychological problems

All above problems impair the psychological side of a patient with cleft lip and patient where they suffer from depression, anxiety and lack of esteem and they are incapable of communicating with their peers in the school. Furthermore, some patients feel anxiety due to the other people's reactions and worried about meeting people in social events.<sup>[5]</sup>

### Treatment

The aims of treating a child with a cleft lip and palate are to:

- Provide normal speech
- Provide normal feeding
- Improve psychological side
- Offer normal dentition and occlusion
- Normal hearing
- Achieve normal aesthetics.<sup>[8,9]</sup>

### Cleft lip and palate team

The treatment involves a team who consists of a number of specialists:

- Cleft lip and palate surgeons
- Orthodontist
- Nurse
- Dentist
- Speech and language therapist
- Paediatricians
- psychologist
- Ear, nose and throat surgeon
- Co-ordinator manager.<sup>[8,9]</sup>

### Treatment of cleft lip and palate

#### 1. Early treatment and evaluation

Initial assessment of a child with cleft lip is undertaken to know the extent of the cleft. The parents are informed of the cleft by a specialist nurse. The team often discusses with parents the treatment plan of their child. Parent feels worry of this defect and it is very important to show them previous cases of clefts which were treated to improve the psychological side.<sup>[8,9]</sup>

The most important thing is the feeding of the infant. A variety of feeding devices are available and this depends on the type of clefts. For example, for infants with isolated cleft lip, a bottle or breast can be used. On the other hand, infants with cleft lip and palate face challenges when feeding they are incapable of sucking

either their mother's nipple or from a bottle. For that reason, feeding devices such as nipples, cross cut nipples and longer nipple can be successfully assist the infant when feeding.<sup>[8,9]</sup>

The mother can try fed her child if she wishes. In case of cleft lip the mother can place her finger over the lip to provide a seal between her nipple breast and lip defect. In case of unilateral cleft palate, the mother can put her nipple breast on the non-affected side of the palate (Beumer *et al.*, 2011). In case of cleft palate, the mother can put their nipple breast on the side of the defect with supporting her breast by fingers.<sup>[1,8,9]</sup>

The child should be evaluated genetically by an assessment of the following:

- Prenatal medical history
- Any family medical history with clefts
- Examination of child for congenital heart disease, limb and ocular abnormalities
- Any birth abnormalities that may occur with clefts
- Evaluation of family members should be undertaken to assess any genetic factors.
- The child is referred for examination in many departments
- Diagnosis of cleft both syndromic and non syndromic.<sup>[1,8,9]</sup>

### Surgical treatment

#### Repair of the lip, nose, and anterior palate

#### Repair of the lip

This surgery is undertaken about 3 months of child life and it involves the mobilization of the tissue in the defect side of the lip and dissection of orbicularis oris muscle. This permits the closure of the lip (Figure 3).<sup>[1,2,5,9]</sup>

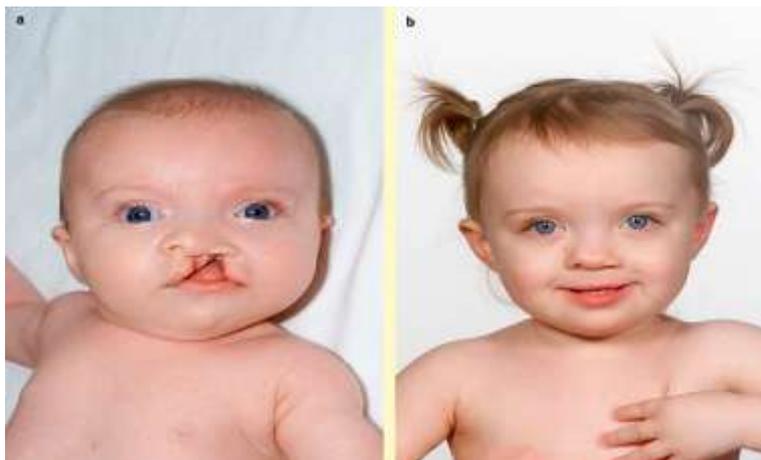


Figure 3a: Pre-operative view. Figure 3b: Post-operative view.

Figure 3: (a and b). Pre – operative view and post – operative view appearances of a left unilateral cleft lip and palate (Goodacre and Swan, 2008)

**Repair of anterior palate**

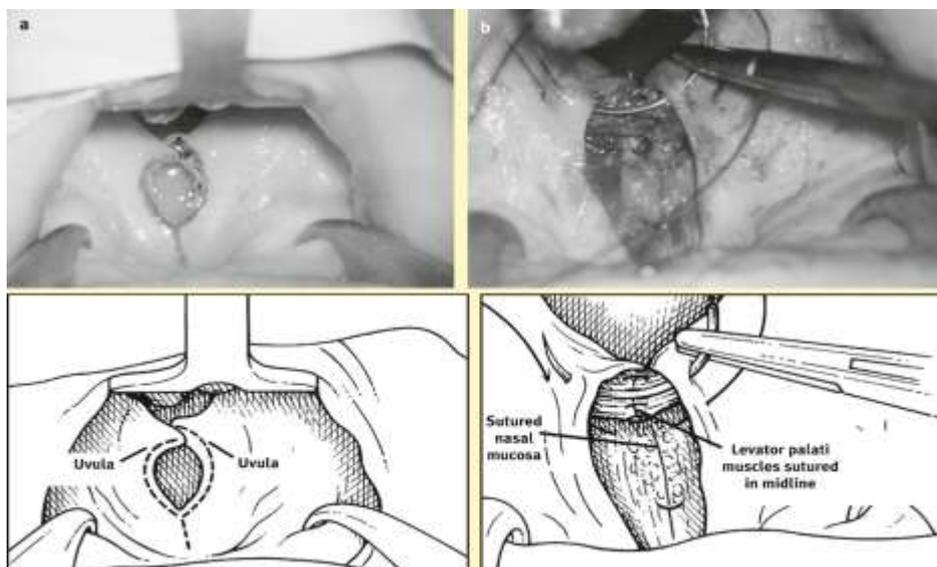
Anterior palate repair involves the use of vomerine flap which is taken from nasal septum to close the defect.<sup>[1,2,5,9]</sup>

**Repair of the nose**

Nasal repair involves the separation the adhesion between ala cartilage and overlying skin and underlying mucosa and then these structures are repositioned again using different methods of fixation.<sup>[5,9]</sup>

**Repair of the cleft palate**

Cleft palate surgery is performed about 6 month of child life and involves a mobilization of mucoperiosteal flaps of the hard palate. Soft palate muscles are then dissected from the bony edge of the hard palate and repositioned. An incision is then made in the lateral part of the palate to reduce the tension in the midline (Figure 4).<sup>[1,2,5,9]</sup>



a. Pre-operative view

b. Intra-operative view

Figure 4: (a and b). Pre-operative view and intra-operative view of palatal repair (Goodacre and Swan, 2008).

**Alveolar bone grafting**

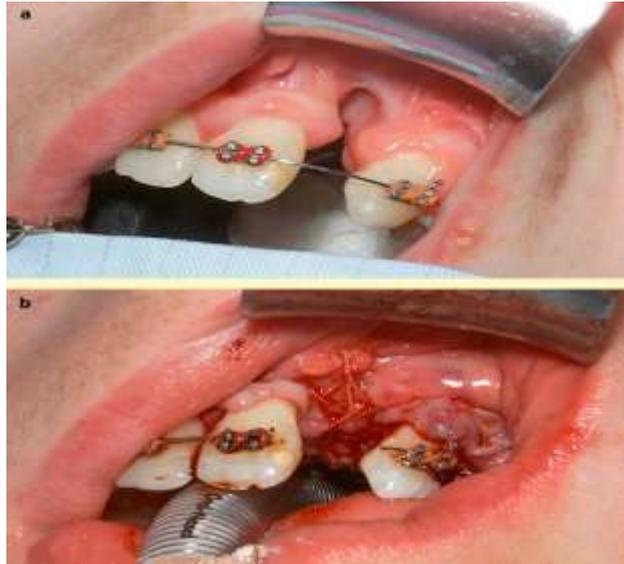
This surgery involves filling a bony defect with cancellous bone (from the iliac crest or tibial plateau) and it is performed at the time of mixed dentition (7-14 years) (Figure 5).

The aim of this treatment is to:

- Allow eruption of maxillary permanent teeth

(Especially canine incisor).

- Facilitate orthodontic treatment when necessary.
- Provide support for ala base.
- Provide a good base for prosthetic rehabilitation.
- Close the gap between oral and nasal cavity.<sup>[1,2,5,9]</sup>



**a. Pre-operative view b. Immediate post-operative view.**  
**Figure 5(a - b): Bone grafting of the cleft alveolus (Good acre and Swan, 2008).**

### Secondary palate surgery

In some cases palatal repair did not solve the speech problem. For this reason, the surgery is undertaken to provide velopharyngeal closure between soft palate and pharyngeal wall. The surgery involves the elevation of myomucosal flap (Pharyngeal flap) from the posterior pharyngeal wall and then this flap is attached to the posterior soft palate to achieve the closure between soft palate and pharyngeal wall.<sup>[1,2,5,9]</sup>

### Orthognathic surgery

Orthognathic surgery is indicated if orthodontic treatment is incapable of establishing an optimum occlusion and aesthetic requirements. This surgery is undertaken to reposition the upper and lower jaws in correct position. In case of maxillary retrognathia, maxillary jaw is repositioned forward with moving the mandible backward to establish class 1 relationship.<sup>[1,2,5,9]</sup>

### Clinical psychologists

Clinical psychologist has an important role in the treatment of cleft lip and palate patients. They assist to improve the psychological side of the patient in the society. Children with cleft lip and palate are incapable to communicate with their peers in the school and in society.<sup>[1,2,5,9]</sup>

### Possible appliances and their function

#### 1. Nasoalveolar moulding

Nasoalveolar moulding is presurgical prosthetic prosthesis used in both unilateral and bilateral cleft patients. They act to reduce the degree of clefts and facilitate the closure of both lip and palate. In addition they facilitate feeding of the patients because they obdurate the defect. They act to lengthen the columella.<sup>[1,2,5,9,10]</sup>

### Unilateral cleft lip and palate

#### Fabrication of nasal alveolar moulding

The construction of NAM devise involves taking several steps. Primary impression is recorded using silicone putty. The use of alginate and elastomeric materials is contraindicated because they may stick in the undercut and cannot be removed. The infant is positioned upside down with supporting head to prevent tongue from falling posteriorly (Figure 6A and B).<sup>[8,10]</sup> The impression is cast in dental stone (Figure 6C) and the cleft segments are blocked with wax and then duplicated. Clear acrylic resin is used to fabricate the plate which is fitted to create a normal palate (Figure a, b and c).<sup>[8,10]</sup>



a. During impression taking b. Impression of silicone putty c. Master cast.  
**Figure 6 (a - c): Fabrication of nasaoalveolar moulding in unilateral clefts (Beumer *et al.*, 2011).**

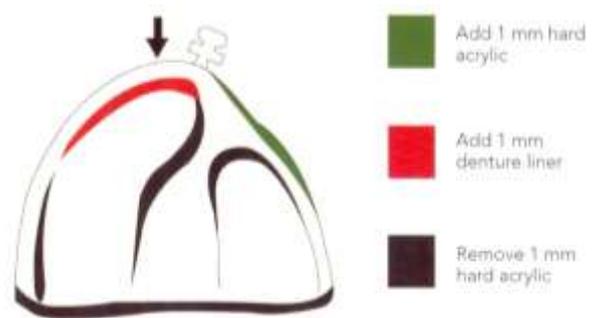


a. Master Cast b. Oral portion of NAM device c. Completed NAM device  
**Figure 7(a- c): Fabrication of nasaoalveolar moulding in unilateral clefts (Beumer *et al.*, 2011).**

The retention of the plate is done using elastic band and tape which are put on small button which is located anteriorly on the clefts.<sup>[8,10]</sup>

**Subtractions and additions to NAM**

The clefts either unilateral or bilateral are divided into two segments: greater alveolar and lesser segments. The process of moulding involves removing some of acrylic resin from some area and adding to other areas. This process continues weekly until two segments are located in close position. The aim of adding and removing acrylic resin is to allow the segments to move into relieved areas (Figure 8).<sup>[8,10]</sup>



**Figure 8: Subtractions and additions made to NAM device (Taylor, 2000).**

When the size of clefts is reduced to 0.5mm, the nasal portion is added to mould the nasal cartilage and reposition of columella. The current prosthesis is extended superiorly and contacts the nasal tip and dome posteriorly (Figure 9A-C). The adjustments to nasal portion continue weekly to correct the position of nasal dome and columella.<sup>[8]</sup>



A. Nasaoalveolar moulding. B. nasal stent positioned in nostril. C. lip tapping across the cleft.  
**Figure 9(A-C): Nasaoalveolar moulding with Nasal stent in its position with lip tapping across the cleft. (Taylor, 2000).**

**Bilateral cleft lip and palate**

The process of fabrication a nasoalveolar moulding in bilateral clefts is the same as in unilateral cleft lip and palate (Figure 10A and B). The molding of bilateral clefts involved three stages:

- Reposition of palatal segments and rotation of premaxilla.
- Reposition of ala cartilage.
- Lengthen the columella.<sup>[8]</sup>



**A. Bilateral cleft. B. NAM device for bilateral clefts.**  
**Figure 10: (A and B). Bilateral clefts and NAM in situ. (Burmer et al., 2011).**

**Orthodontic appliance**

To correct the contracted segment and cross bite, several appliances such as quad helix (Figure 11) and auxiliary appliances are used. This treatment starts after eruption of maxillary incisors (7 year) and first molar tooth.<sup>[8]</sup>



**Figure 11: Palatal view after placement of maxillary expansion appliance (Buemer et al., 2011).**

In addition, to treat malocclusion orthodontic treatment always is carried out after eruption of all permanent teeth and this involves the use of fixed appliance brackets are cemented on the teeth in order to move the teeth into correct position.<sup>[8]</sup>

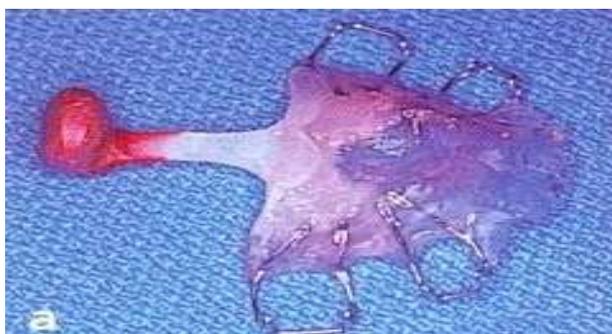
**Obturator devices**

Obturator prostheses are devices used to close the gap between oral and nasal cavities.<sup>[10]</sup> Two types of cleft lip and palate patients require the fabrication of obturator devices:

- Patients with hypernasal speech following breaking down of pharyngeal flap.
- Patients with secondary palate.<sup>[8]</sup>

**1. Clefts with secondary palate**

The process of fabrication of obturator prosthesis for patients with soft palate includes recording master impression; fabrication of acrylic base retained by adam clasps. Artificial teeth are set in case of edentulous and partially dentate and try in appointment is undertaken. After processing the first part of prosthesis, the second part of the prosthesis which covers the soft palate defect is attached to the first prosthesis using modeling compound (Figure12a and b).<sup>[8]</sup>



**Figure 12a: Provisional obturator prosthesis.**



**Figure 12b: Obturator prosthesis in situ.**  
**Figure12 (a and b). Provisional obturator prosthesis and obturator prosthesis in situ (Beumer et al., 2011).**

This prosthesis can be fabricated from of a base which made from metal (cobalt chromium) with extension of a metal loop into center of the soft palate defect.

## 2. Patients with hypernasal speech following pharyngeal flap procedure

Following pharyngeal flap procedure, patients may suffer from hypernasal speech as pharyngeal flap has broken

down. The gap between oral and nasal cavities results of passing air through the nasal cavity. this case can be treated with fabrication of obturator prosthesis (Figure 13).<sup>[8]</sup>



a. Pharyngeal flap broken down



b. Obturator prosthesis



c. Obturator prosthesis in situ

Figure 13(a - c). Soft palate defect with fabrication of obturator. (Beumer *et al.*, 2011).

### Removable partial denture

Any missing teeth in patients with repaired cleft palate can be replaced with removable partial denture. The process of fabrication of a denture for cleft palate patients is similar as in non-clefts patients. It involves recording master impression, followed by fabrication of a cobalt chromium design or, acrylic base, arrangement of artificial teeth and then processing with heat cure acrylic resin.<sup>[8]</sup>

### Fixed partial denture

Many cleft patient prefer fixed partial appliance than a removable partial denture for replacement of missing teeth. If the patient had undertaken a bone graft, implants can be placed to replace the missing teeth in the defect side. On the other hand, if the patient has not had a bone graft, preparation of two abutment teeth at least can be used for fabrication of fixed partial denture.<sup>[8]</sup>

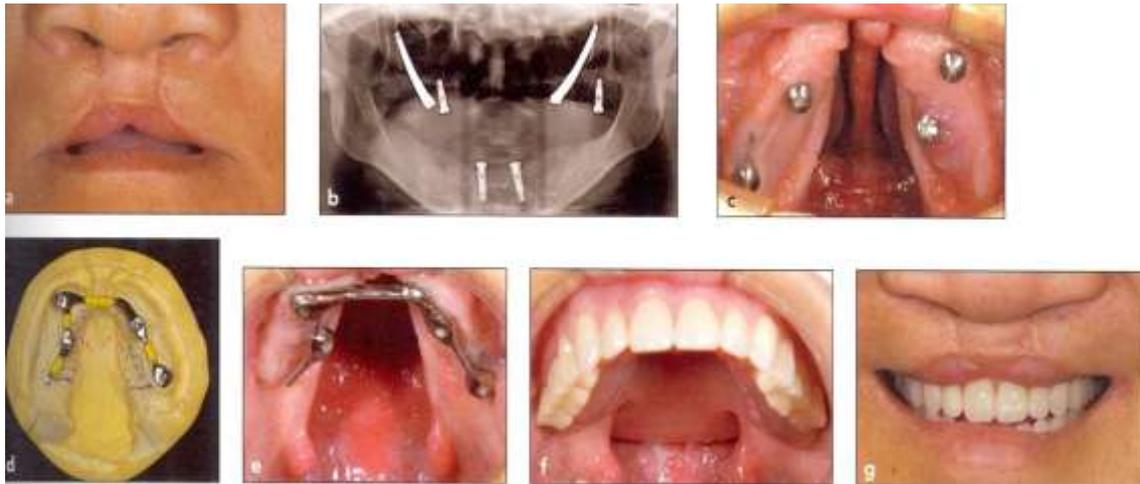
### Complete denture

The process of fabrication of a denture for edentulous patients with cleft palate is similar as in non clefts patients. It involves recording a primary impression, fabrication of customized tray followed by recording a master impression poured in dental stone. Fabrication of bite rim, recording of a face bow and mounting of a maxillary cast on the articulator are then completed.

Artificial teeth are arranged in the lab and try in appointment in undertaken for checking the setting of the teeth. Then complete denture is processed with heat cure acrylic resin.<sup>[8]</sup>

### Osseointegrated implants

Implants can be used for retention of obturator prosthesis in patient with cleft lip and palate both in dentate and edentulous area. Implants are placed either in the remaining maxilla in a good bone quality or in the zygoma (Figure 14a - g).<sup>[8]</sup>



**Figure 14:** (a) Edentulous patient with repaired cleft. (b and c) Four implants placed in the maxilla (d) Bar positioned on the cast (e) tissue bar placed orally (f) prosthesis with obturator extension in place. (g) Final result. (Beumer *et al.*, 2011).

### SUMMARY

Cleft lip and palate are the most common facial deformity and clefting may involve lip only, lip and palate and palate only. Environmental (such as smoking, alcohol, poor nutrition) and genetic factors (such as familial factors and chromosomes) are the main reasons of clefting in infants. Treatment of clefting involves a number of specialists who decide the best treatment plan depending on the site of defect and age of the infant.

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