Cleft Lip and Palate and Alveolar Bone Grafting in the United Kingdom: A Brief Overview for Non-specialists

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ABSTRACT

Cleft lip and palate is a common congenital defect that is associated with various functional, aesthetic and psychosocial problems. The objective of this article is to share a brief overview of cleft and lip palate classification, aetiology, complications and management, particularly aimed at non-specialists. The literature suggests that the use of iliac crest bone in secondary alveolar bone grafting (ABG) still seems to be the favoured technique for cleft lip and palate repair. In conclusion, we suggest that advances in bone morphogenic proteins may be the key to further advancement in ABG.

Keywords: Alveolar bone grafting, cleft lip, cleft palate

Labio Leporino Acompañado de Paladar Hendido en el Injerto Óseo Alveolar en el Reino Unido: Un Breve Panorama para no Especialistas

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RESUMEN

El labio leporino acompañado del paladar hendido es un defecto congénito común asociado con diversos problemas funcionales, estéticos y psicosociales. El objetivo de este artículo es compartir un breve panorama de la clasificación del labio y paladar hendidos, su etiología, complicaciones y manejo, dirigido especialmente a los no especialistas. La literatura sugiere que el uso de hueso de la cresta iliaca en el injerto óseo alveolar secundario (IOA) sigue siendo al parecer la técnica preferida para la reparación del labio y paladar hendidos. En conclusión, sugerimos que los avances en relación con las proteínas morfogénicas del hueso pueden ser la clave para avances ulteriores en el IOA.

Palabras claves: Injerto óseo alveolar, labio leporino, paladar hendido

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INTRODUCTION

Cleft lip and palate is the commonest congenital facial defect in the United Kingdom (UK), associated with around one in 700 live births. In understanding cleft lip and palate, it is important to appreciate both embryological development and anatomy of the facial hard and soft tissues. Cleft lip and palate form due to failure of the medial, lateral and nasal maxillary processes to fuse during development. This can occur unilaterally or bilaterally. Cleft lip occurs due to failure of fusion of the medial nasal and maxillary processes at five weeks gestation. Cleft palate forms as a result of a problem relating to the palatal shelves; partial or complete lack of fusion of the palatal shelves can occur. The secondary palate forms from the right and left palatal processes fusing, at eight to 12 weeks gestation (1).

Cleft lip and palate can affect the left, right, or can be bilateral. Anatomically, clefts extend from the lateral lip through the philtrum and, if the maxillary alveolus is involved, it is normally in the lateral incisor/canine region. Lateral incisor teeth can often be missing or peg shaped. Clefts extending to the incisive foramen, and no further, can be considered a cleft lip. Should the cleft continue beyond the incisive foramen, then a cleft lip and palate is present.

Classification

There is significant variability in cleft types, the two main categories being; cleft lip and cleft palate. Cleft lip may or may not be associated with a cleft palate and *vice versa*, cleft palates

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may or may not be associated with a cleft lip. Isolated cleft lips account for 25% of all cleft lip and palate cases. Twenty-five per cent are made-up of unilateral cleft lip and palate. Ten per cent are bilateral cleft lip and palate. The remaining 40% are isolated cleft palate (2). Cleft lips which extend to the nasal floor are considered complete cleft lips; those which do not are considered incomplete (Fig. 1).

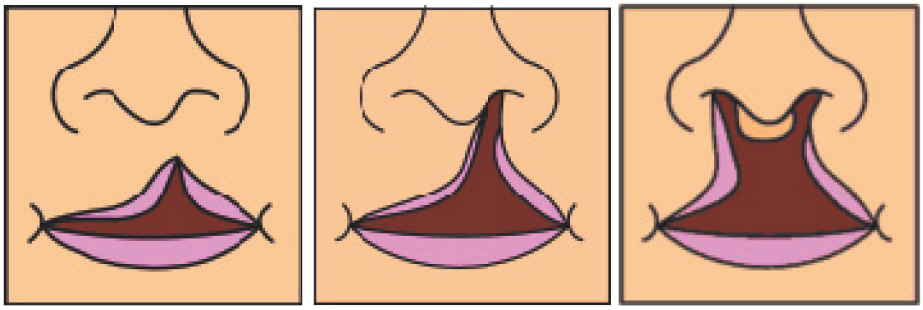

Fig. 1: Cleft lip classification.

Cleft palates which extend through the hard palate and entire soft palate are complete. Those that do not extend through the entire soft palate are considered partial. Complete cleft palates generally extend through the uvula (Fig. 2).

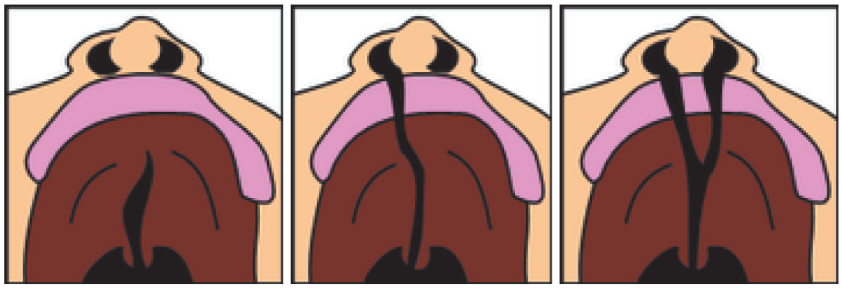

Fig. 2: Cleft lip and palate classification.

Submucous cleft palates can also occur; these generally present with a bifid uvula, a furrow along the soft palate and a notch at the posterior hard palate (3). Cleft lip can be diagnosed *in utero* with ultrasound at the 20-week antenatal screening; however, cleft palates are difficult to diagnose *in utero* (2).

Aetiology

There are both genetic and environmental aetiologies associated with cleft lip and palate. Environmental factors include anti-convulsant therapy including; phenytoin and phenobarbital, alcohol, cigarette smoking, hypoxia, folic acid deficiency and corticosteroids. Genetic factors are numerous, including many chromosomal abnormalities such as IRF-6, TGF-α, MSX-1 and many point mutations. Around 15% to 30% of individuals have clefts as part of a syndrome; these include Van der Woude syndrome, hemifacial microsomia, velocardiofacial syndrome, Stickler's syndrome, Loeys-Dietz syndrome and others (2).

Complications/difficulties

Cleft patients can suffer a variety of problems to a varying extent, depending on the severity and type of cleft. The associated problems can be split into functional, aesthetic and psychosocial. The aesthetic problem in relation to cleft lip, that is often immediately apparent, is the vertical soft tissue deficiency. Nasal involvement can often be seen *via* rotation of the septum, columella and nasal spine, away from the cleft. Alar base displacement is also commonly seen, along with displacement of the lateral nasal cartilages.

Alveolar clefting at the canine/lateral incisor region, if left untreated, it will affect the patient's dentition. This often manifests in a peg shaped or missing lateral incisor and the canine may fail to erupt due to lack of alveolar bone at the cleft. Isolated clefts of the palate present less of an aesthetic problem, as these tend to be less visible.

Functional problems can result early on in babies, with difficulty feeding in some cases, although most tend to manage fine. Babies with clefts associated with Pierre Robin sequence also have micrognathia and abnormal tongue size; these babies may have a higher risk of airway obstruction. Production of bilabial sounds can be a problem; these are the 'pa' and 'pi' sounds. Cleft palate can produce many of the functional problems related to clefts. Central to the problem is velopharyngeal incompetence (4). The majority of phonemes in the English language are produced *via* oral airflow, hence the velopharynx should be closed. In cleft palate patients this is not possible, which results in difficulties with speech. Soft palate dysfunction due to inadequate muscle function can also lead to problems with the Eustachian tubes. In a normal palate, the levator veli palatini and the tensor veli palatini act to open and close the Eustachian tubes. Eustachian tube dysfunction can lead to chronic otitis media, tympanic membrane perforation and atelectasia of the middle ear (5).

Cleft patients can potentially develop psychosocial problems; however, if clefts are repaired optimally and in a timely manner, then this can be avoided. The main problems can be considered in terms of self-concept, appearance and peer relationships. Issues for patients can arise in relation to visual deformity and speech impediments.

Management

It is clear that the problems relating to cleft lip and palates can be variable and diverse. Thus, the management of these patients requires a multidisciplinary team (MDT). Many cleft lip and palate units exist across the United Kingdom; the specialties/healthcare professionals who make-up the team include:

- Orthodontics
- Paediatric dental care
- Restorative dentistry
- Specialist cleft nursing
- Paediatrician
- Plastic surgeon
- Otolaryngologist
- Maxillofacial surgeon
- Psychology
- Speech and language therapy
- Geneticist
- Psychologist
- Social worker

The second part of this writing will concentrate on the surgical management of cleft lip and palate, specifically alveolar bone grafting (ABG). The main surgical specialties involved in cleft repair in the UK are Oral and Maxillofacial Surgery, and Plastic Surgery. The timing and protocols for the multiple stages of cleft repair is variable across the world. In the UK, lip repair is normally the first significant surgical procedure, which is carried out at three to five months. The favoured techniques are the Millard and Tennison techniques. As well as the lip, the nose and anterior palate may be repaired at this stage. In some cases, lip repair may be preceded by a tracheostomy in babies with airway compromise; as mentioned previously, these babies will often have an associated syndrome.

Lip repair is followed by palate repair, which is commonly done at six to nine months in the UK. The favoured technique in many UK units is the Sommerlad repair. Commonly used techniques in the United States of America (USA) are the Bardach double flap and Furlow double opposing Z-plasty techniques (2).

The next important stage is ABG. The timing of ABG is dependent on the development of the canine root and does not follow any specific time frame. There are many other surgical procedures, which may or may not be required. These include lip revision, palatal fistula closure, palatal lengthening, pharyngoplasty, pharyngeal flap, columellar lengthening, rhinoplasty, septoplasty and Le Fort 1 orthognathic procedures (2).

Alveolar bone grafting and the literature

Alveolar bone grafting is an important stage in cleft repair where there is a cleft involving the maxillary alveolus/premaxilla. It is thought that Axhausen first introduced the concept of modern ABG in 1952; prior to this, bone grafting was carried out, however, it is thought that Axhausen's ideas were the first to imply restoration of function at the cleft site (6).

The aims and benefits of successful ABG are numerous. Alveolar bone grafting helps stabilize the alveolar arch and restore arch integrity, it allows teeth to erupt into the optimal position, it permits future orthodontic alignment, the ABG can help close an oronasal fistula simultaneously, it allows future consideration of dental implants, it can optimize the maxilla for future orthognathic surgery, and it can also help create support for the alar base and lip (2, 6).

There is much debate with regards to ABG timing and donor sites. The preferred age for ABG in the UK is around nine years of age. Early bone grafting introduced by Axhausen was primary bone grafting; this is bone grafting at the time of lip repair, at three to six months of age. Later the concept of secondary bone grafting was introduced, which can be early (2–5 years) or later (6–13 years). Boyne and Sands seem to have been the first to identify and intentionally explore osetogenic cell survival in fresh autografts placed in cleft sites, in 1972. Later in 1986, Bergland *et al* (6) presented a significant paper outlining secondary bone grafting in 378 consecutive patients. This technique was initially introduced in Oslo in 1977. The age range of patients who underwent bone grafting was 8–18 years; they concluded that the optimal age for bone grafting was 9–11 years of age (6).

Prior to the introduction of secondary bone grafting, primary bone grafting was used. Primary bone grafting, prior to modification, was carried out at the time of lip repair with wide dissection and split rib grafting used (2). It became clear that this technique had the significant complication of inhibiting maxillary growth. This was the result of interference with maxillary sutural growth (6). Primary bone grafting is scarcely used today, but is still part of the protocol in some units in the USA, although in a modified form (2).

Early secondary bone grafting at two to five years of age has also been considered, but this is not widely done due to a lack of long-term maxillary growth outcome data. As mentioned above, the favoured technique is secondary bone grafting at 6–13 years of age. The timing of surgery is dependent

on canine development. Grafting is carried out when the canine root has one-half to two-thirds formed; the significant advantage being that the canine can erupt into the newly grafted area. This technique is well established with favourable long-term maxillary growth and dental development outcomes (2).

Late bone grafting in the permanent dentition has been considered and subsequently shown to have poor outcomes. This has been shown to result in compromise of the periodontium, higher fistula rates and a higher rate of tooth loss adjacent to the cleft (2). Bone grafting at the time of maxillary osteotomy is also contraindicated, for the obvious reason that managing a maxilla with multiple segments complicates surgery. Gingivoperioplasty has been another consideration; this is where gingival flaps are raised and sutured together. There is some evidence to suggest that this may decrease the need for an ABG later. However, recent studies have shown that secondary bone grafting is superior.

The other important factor in ABG is the donor site from which the graft should be harvested. Another consideration should be whether autogenous, alloplastic, xenogenic, or allogenic grafts should be used. Many sites for autogenous bone harvest have been considered over the years, all of which have had advantages and disadvantages. Bone that has been harvested for ABG includes calvarial bone, tibial bone, mandibular symphyseal bone, rib bone and iliac crest bone. The aims of ABG should be to achieve optimal uptake of the graft, good function, minimal complications and minimal disruption of maxillary growth (6).

Iliac crest bone grafting is considered to be the gold standard in ABG. The main benefits of iliac crest bone are that it is rich in osteogenic cells, large volume can be harvested, it rapidly transforms into alveolar bone and good long-term outcomes are reported (2, 6). The main reported disadvantages relate to morbidity at the donor site. These include postoperative pain, gait disturbance, long postoperative hospital stay, hip scar and neuropraxia of the lateral cutaneous nerve of the thigh. Bergland *et al* (6) concluded that fresh autologous cancellous iliac chips transformed into alveolar bone very rapidly and also that the bone responded normally to migration and orthodontic movement of teeth.

Calvarial bone grafting has largely come about in the last two decades. The main advantages are minimal scarring beneath the hairline, minimal postoperative pain and mesenchymal bone. However, it is thought that calvarial bone for ABG has poorer outcomes than iliac crest bone. The main disadvantages are cortical bone with little cancellous bone, only small volumes can be harvested and dural tears can result. Also, only one team can work when calvarial bone is being harvested, as opposed to two with iliac crest grafts (2, 6).

Tibial bone has the advantage of being easily harvested, minimal associated morbidity, good long-term outcomes and two teams can operate. However, tibial plateau fractures can occur, a scar will result and limited bone can be harvested. The long-term outcomes in tibial bone for ABG have not been reported (2, 6).

Mandibular symphyseal bone advantages include no visible scar, it is mesenchymal bone, minimal postoperative pain, minimal postoperative hospital stay and satisfactory reported long-term outcomes. The disadvantages are that only a single team can operate, there is risk of damage to dentition and the mental nerve and only limited bone can be harvested (7).

Rib grafting is now no longer used, due to the significant resorption and poor long-term outcomes.

The literature and long-term outcomes seem to support iliac crest bone grafting as the favoured bone harvest site in ABG. Autologous bone substitutes have the obvious advantage of no associated harvest site and hence no associated postoperative site morbidity. However, graft revascularization is slower, long-term results are variable, bone formation is not predictable and resorption is unpredictable. Also, bone substitute use is not advised in children. Bone morphogenic proteins are a significant advancement; however, a suitable carrier in humans has yet to be found (7).

Assessment of ABG success is based on success of fistula closure, canine eruption and assessment *via* radiographs or cone beam computed tomography (2). Commonly used scales include the Bergland, Kindelan and Chelses scales (8).

CONCLUSION

Alveolar bone grafting has advanced somewhat over the last half century or so, since initial discovery. However, the use of iliac crest bone in secondary alveolar bone grafting, which was recommended by Bergland *at al* in 1986, still seems to be the favoured technique. Perhaps advances in bone morphogenic proteins are where the key to further advancement in ABG lies.

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