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The management of children born with cleft lip and palate

NM King, N Samman, LLY So, LK Cheung, TL Whitehill, H Tideman

Although cleft lip and palate is a single anomaly, its consequences affect several systems and functions of the child as well as the social and psychological problems that impact on the child and parents. Therefore, the services of a team of specialists are required to care for a child with cleft lip and palate. Empathic counselling and help with feeding ensures that the infant can cope with the primary surgery to the lip and palate. If speech problems occur, a nasendoscopy can be performed to determine the nature of the speech abnormality and to assess the appropriateness of additional palatal surgery. Nasendoscopy may also be required later because osteotomy surgery can compromise speech. Alignment of the teeth may be necessary before bone grafting of the residual alveolar cleft, and is always needed prior to and after orthognathic surgery. The development and regular practice of a range of clinical skills is essential if the team of specialists are to plan and deliver the appropriate high quality care needed by children and adolescents with cleft lip and palate.

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Introduction

In the past, many children born with cleft lip and palate (CLP) received inadequate care as a result of diagnostic errors, failure to recognise the full spectrum of problems associated with CLP, and the use of inappropriate or ill-timed procedures. Although CLP is a single anomaly, its possible consequences affect several systems and functions that include facial growth, dentition, speech, hearing, and genetic aspects because of the complex mode of

inheritance. The inevitable social and psychological impact on the child, their parents, and the family, must also be paid attention. To avoid these problems, specialists throughout the world have elected to gather together in multi-disciplinary teams to manage children with CLP. Consequently, the team is exposed to a sufficient number of patients each year to maintain clinical expertise, and is readily available to provide care for new cases.

The appropriate specialists within a team provide a rapid initial evaluation, often within hours of birth. Subsequent regular contact with family members through frequent review clinics ensures that social and psychological problems are identified and resolved early. Treatment plans are formulated and implemented in collaboration with fellow specialists. Regular follow up appointments enable the accumulation of data on the outcome of clinical procedures. This information, the psychological well-being of the patient, the effects of treatment on growth, post-operative function and appearance can be used to help future patients under the care of the team.

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It is generally accepted that CLP is more prevalent in Asians.¹ During the period 1977 to 1979, it was calcu-

lated (based on figures from three major general hospitals in Kowloon) that in Hong Kong, cleft lip and cleft palate occurred in 1.23 and 1.35 per 1000 live births, respectively.² At the time, these figures were comparable to those obtained in Beijing, China, (1.25 and 1.38 per 1000 live births, respectively) for cleft lip and cleft palate.³ However, local factors also influence the incidence of CLP because the rate of occurrence is much lower in different parts of China, such as Chengdu and Wuhan.⁴

In 1992, there were 70 949 births in Hong Kong. Figures on the prevalence of CLP in Hong Kong gathered then indicate a prevalence of 1.2 per 1000 births. From these data it is estimated that there are approximately 80 new cases each year.

At the Prince Philip Dental Hospital, Hong Kong, the precursor of the Prince Philip Dental Hospital/Hong Kong University Cleft Lip and Palate Centre (PPDH/HKU CLP Centre) was opened in 1981. Since that time the composition of the team has evolved to include specialists in many different fields of CLP management. These fields of expertise include counselling, assistance with feeding, surgical closure of the lip and palate, speech and

hearing assessment and therapy, orthodontics, periodontics, restorative, prosthetic, and preventive dentistry for the child and adult patient, maxillofacial bone osteotomies, and aesthetic lip and nose surgery. This paper describes the role of each specialist and gives the timing for their respective contribution in the long term management of a child born with CLP.

Once a child is born with CLP, the services of a team of specialists are needed to care for it until adolescence (Table). During that time, a number of primary and secondary surgical procedures need to be performed; each being preceded by a period of pre-surgical preparation of the child. Careful planning by the team members is essential to ensure that any proposed procedure is appropriate and that the timing of the procedure is in keeping with the development of the child.

The neonatal period

In the early stages after birth, the parents need to be counselled about the immediate and long term implications of the cleft defect and the necessary surgery. Although nasogastric tube feeding can be

Table. The procedures that may need to be performed for a patient with cleft lip and palate and the optimum time for the procedure

Age of patient	Operative procedure
0 - 3 days	Counselling Feeding plate fitting
4 - 6 months	Surgical closure of lip
1 ¹ / ₂ - 2 years	Surgical closure of palate
1 - 3 years	Indirect speech therapy - by parents under guidance of speech therapist
3 - 6 years	Speech therapy Nasendoscopy ± pharyngoplasty
9 - 11 years	Alveolar bone grafting ± pre-grafting orthodontic therapy
14 years	Comprehensive orthodontic therapy
18 years	Orthognathic surgery Lip/nose revision surgery

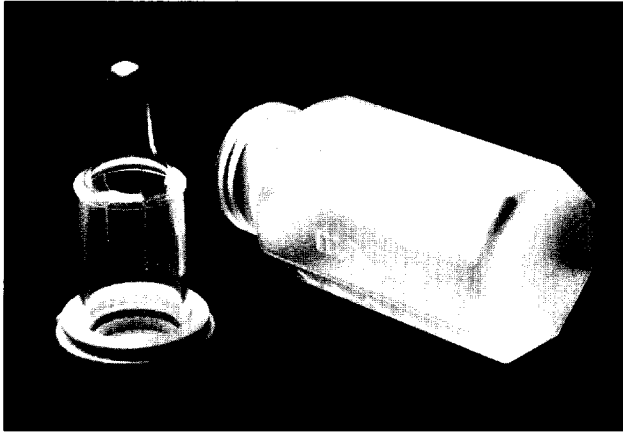


Fig 1. A purpose-designed feeding bottle with a cross-cut nipple

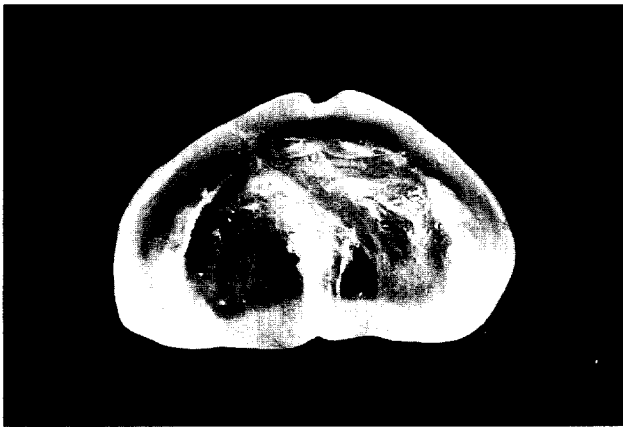


Fig 2. A feeding plate that helps to improve the speed and quality of feeding

used initially, it is best reserved for those neonates who have other central disorders. Breast feeding is desirable and should be encouraged,⁵ but it may not be successful, and the expressing of milk by the mother may be the most appropriate approach.

Bottle feeding can usually be achieved by most neonates, but this is often with some difficulty because of their inability to generate sufficient suction. The nipple of the feeding bottle should be positioned on the non-cleft side and moved about, or even squeezed by the parent. The cutting of a cross in the nipple of the feeding bottle increases the volume of milk that flows, but only when the nipple is compressed: hence, the neonate does not need to create suction in order to feed. Commercially available products can be used to achieve these objectives, however, they are expensive and not always readily available in Hong Kong (Fig 1). Custom-made acrylic feeding plates can be used to enhance

the quality and speed of feeding.⁶ With bilateral CLP, the plates can be modified to help prevent collapse of the buccal segments of the palate; if delayed closure of the hard palate is planned, these allow the development of normal speech and swallowing patterns. The feeding plates need to be made by a specialist and fitted as soon as possible after birth to ensure their acceptance by the neonate (Fig 2).

Careful counselling by an empathic specialist is needed by the parents at this early stage. It must be remembered, however, that parents have not had the benefit of a medical education and are unable to understand anatomical and medical terminology. It will also probably be the first time that they have seen an unrepaired cleft of the lip or palate. Consequently, counselling must be performed in a sympathetic but authoritative manner.⁷ Parents often appreciate and benefit from the assistance available from members of a parent support group, such as the Hong Kong Parent's Association of Cleft Lip and Palate Children.⁸ Once the child is feeding well, indicated by a progressive weight gain and the degree of healthiness, the child will be able to cope with the primary surgery to the lip and subsequent surgery to the palate.

Many surgical techniques have been described for the primary closure of CLP. Unfortunately, there is still controversy regarding the precise timing of surgery and which is the most reliable technique that is consistent with the restoration of adequate appearance and function. Two important considerations with all of the techniques are the effects of the timing of surgery on the ensuing growth of the face and development of speech.

Lip repair is less controversial than is palate repair. Lip repair aims to restore the continuity of the orbicularis oris muscle of the lip, and with it, the appearance and function of the upper lip (Fig 3). Recently introduced techniques that include the additional re-attachment of the muscles at the base of the nose have improved both the aesthetic result as well as the growth potential of the midface. Such techniques have been used at the PPDH/HKU CLP Centre since 1989, and the results are encouraging.

Palate repair aims to reconstruct the abnormally inserted musculature of the soft palate to normalise movements of the soft palate and permit the development of normal speech. Early repair aids speech development, but is likely to disturb the subsequent growth of the midface, whereas delayed repair minimises the growth disturbance but has a negative influence on speech development. A compromise is needed and this may be



Fig 3. Neonate with a complete unilateral cleft lip and palate. 3a. Before. 3b. After surgical closure.

either to repair the palate late, but before speech patterns develop at one year six months to two years of age or to repair the soft palate only at the time of lip repair, leaving the hard palate open for later closure and using an acrylic plate in the meantime. Despite these alternatives, there are still important disadvantages to the latter approach and our centre has adopted the first option.

In addition, many infants with CLP suffer one or more congenital abnormalities that often affect the heart or lungs. These infants, quite apart from the separate treatment which may be required for the other anomalies, may be at additional risk from operation. Such patients require specialist paediatric assistance in the post-operative period and may benefit from intensive care overnight after surgery. Hence, adequate paediatric support is essential in many instances.

Childhood

During childhood, care needs to be exercised to monitor hearing and middle ear function to ensure that speech and language are developing appropriately. When necessary, detailed speech and language

assessments and interventions should be provided. Speech problems that occur after the age of three years need to be carefully assessed to determine the contributing factors. The possible causes of speech problems in this group include developmental delay, hearing problems, oronasal fistulae, dental malocclusion, and velopharyngeal incompetence (VPI).^{9,10}

If a cluster of speech symptoms indicates VPI as the cause, direct examination of the velopharyngeal sphincter should be performed using nasendoscopy. If a structural deficiency in the velopharyngeal sphincter is identified, then pharyngeal flap surgery is needed. These flaps are designed to connect the posterior or lateral pharyngeal walls with the soft palate, which reduces nasal air flow during speech. The assessment and surgical correction for VPI should ideally be performed before the age of six years. This is generally followed by a period of speech therapy based on several established principles. Although surgery (velopharyngeal closure and the closure of palatal fistulae) can be expected to improve structural obstacles which

hinder the development of normal speech, post-operative speech therapy may be needed to correct poor speech habits that develop because of the structural deformity. The optimal time for surgery is before the age of six years so that the most benefit can be gained from speech therapy. Some children have speech problems that are resistant to traditional methods of speech therapy and they can benefit from recently developed techniques that use visual feedback.

Regular dental care, both preventive and therapeutic, is essential during childhood to minimise morbidity due to the loss of teeth. This includes oral hygiene instruction and prophylactic measures to prevent the development of dental caries and periodontal disease. In addition, restorative treatment to repair carious or malformed teeth is often needed.

Orthodontics may be applied in selected cases at this early stage. It is sometimes necessary to align the teeth in the arch before bone grafting of the residual alveolar cleft prior to the eruption of the per-

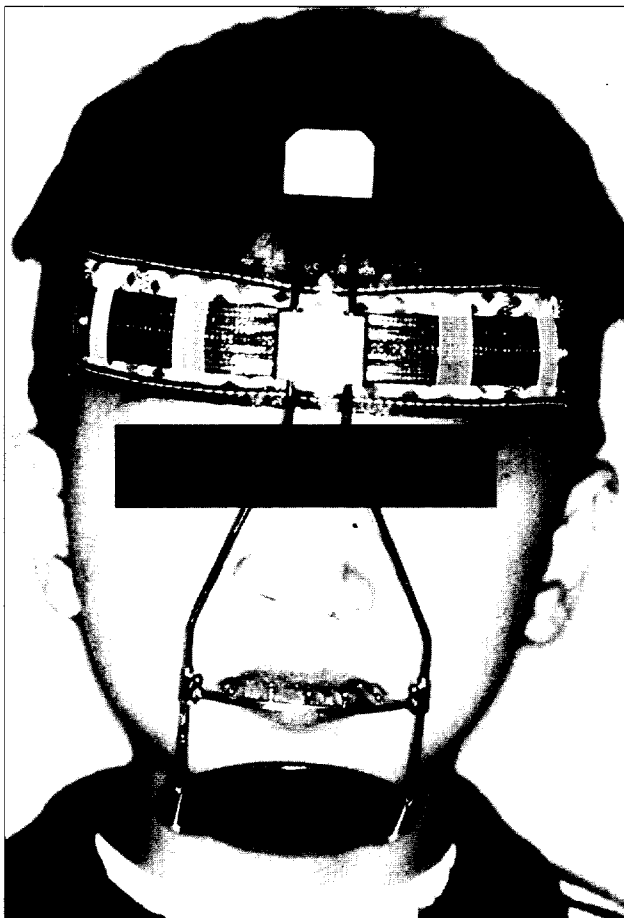


Fig 4. Early orthodontic intervention using reverse headgear, a type of dentofacial orthopaedic device



Fig 5. A radiograph showing a residual cleft of the alveolus. 5a. Before. 5b. After alveolar bone grafting.

manent canine or lateral incisor teeth. Thus, there may be a relatively short period of pre-grafting orthodontic treatment with simple fixed appliances to obtain optimal arch shape, improve access for the surgical procedure (bone grafting) and to help with stability during the initial stage of bone remodelling. Regular and careful observation and guidance of the erupting permanent teeth then follows.



Fig 6. A patient with a repaired unilateral cleft lip and palate. 6a. About to undergo orthodontic treatment to align the teeth into a normal arch form following alveolar bone grafting. 6b. The end result of orthodontic therapy.

Definitive orthodontic treatment is usually necessary once active skeletal growth has ceased. However, children with grossly abnormal jaw development may benefit from earlier intervention using dentofacial orthopaedic appliances (Fig 4).¹¹ Pre-surgical and post-surgical orthodontics are also essential when orthognathic surgery is performed if the best result is to be achieved. The orthodontic treatment of CLP patients requires expertise and experience which is often not available, even from some specialist orthodontists.

Alveolar bone grafting should be timed to precede the eruption of the canine tooth on the cleft side, which usually occurs at approximately 11 years of age. The aim is to provide a bony bridge across the cleft so that the canine tooth can erupt into this new bone, and to allow the tooth to have a healthy supporting periodontium (Fig 5). The dental arch is then aligned by orthodontic therapy (Fig 6). Bone for the grafting procedure may be obtained from a number of donor sites. The main site is the ilium, but the mandibular symphysis, tibia, rib, and the

calvarium can be used. Clinical experience has shown that the timing of the operation, pre-operative gingival health, and careful tissue and graft handling during the surgical procedure are important in determining the success of the operation—more so than the anatomical source of the bone graft.^{12,13}

Late adolescence and early adulthood

Bone grafting and the alignment of the grafted dental arch may not be the end of skeletal problems. If the maxilla has a growth disturbance, it will still require treatment after growth has stopped. A Le Fort I osteotomy with modification may be performed to correct the occlusion and facial appearance.¹⁴ More severe skeletal deformities require a staged procedure involving a Le Fort II and subsequently, a Le Fort I osteotomy and frequently, an additional mandibular procedure. When a patient is referred late, and has not had the benefit of bone grafting at the optimal time, the osteotomies may be combined with the alveolar bone grafting as a single-stage procedure. Experience with many such cases confirms that the results are stable and reliable.^{13,15} If necessary, an aesthetic rhinoplasty and cheiloplasty should be performed as a final surgical stage. Occasionally, however, functional lip and nose revision is combined with alveolar bone grafting at an earlier stage to reduce the impact of the aesthetic deformity on the growing child and to take advantage of the remaining growth potential in the midface.

For those patients who are referred late, or after sub-optimal results have been obtained, surgery is needed to correct what can be severe deformities (Fig 7). These are deformities that could, in many instances, have been avoided by detailed treatment planning, thoughtful liaison, and coordinated team work by the specialists responsible for the management of the affected child.

The above procedures are only some of the many that are needed by a patient with CLP. The acquisition of the clinical skills required to treat affected children, and the need to regularly practice them, are essential if the care that is to be delivered to these patients is to be of sufficient quality. It must be remembered that, at the time of birth, the parents want to know about their child's problems. The parents, however, are confused and disturbed—it is most important that they be attended by a team of specialists who are supportive and able to provide accurate information in a sympathetic manner and in terms that they can understand.

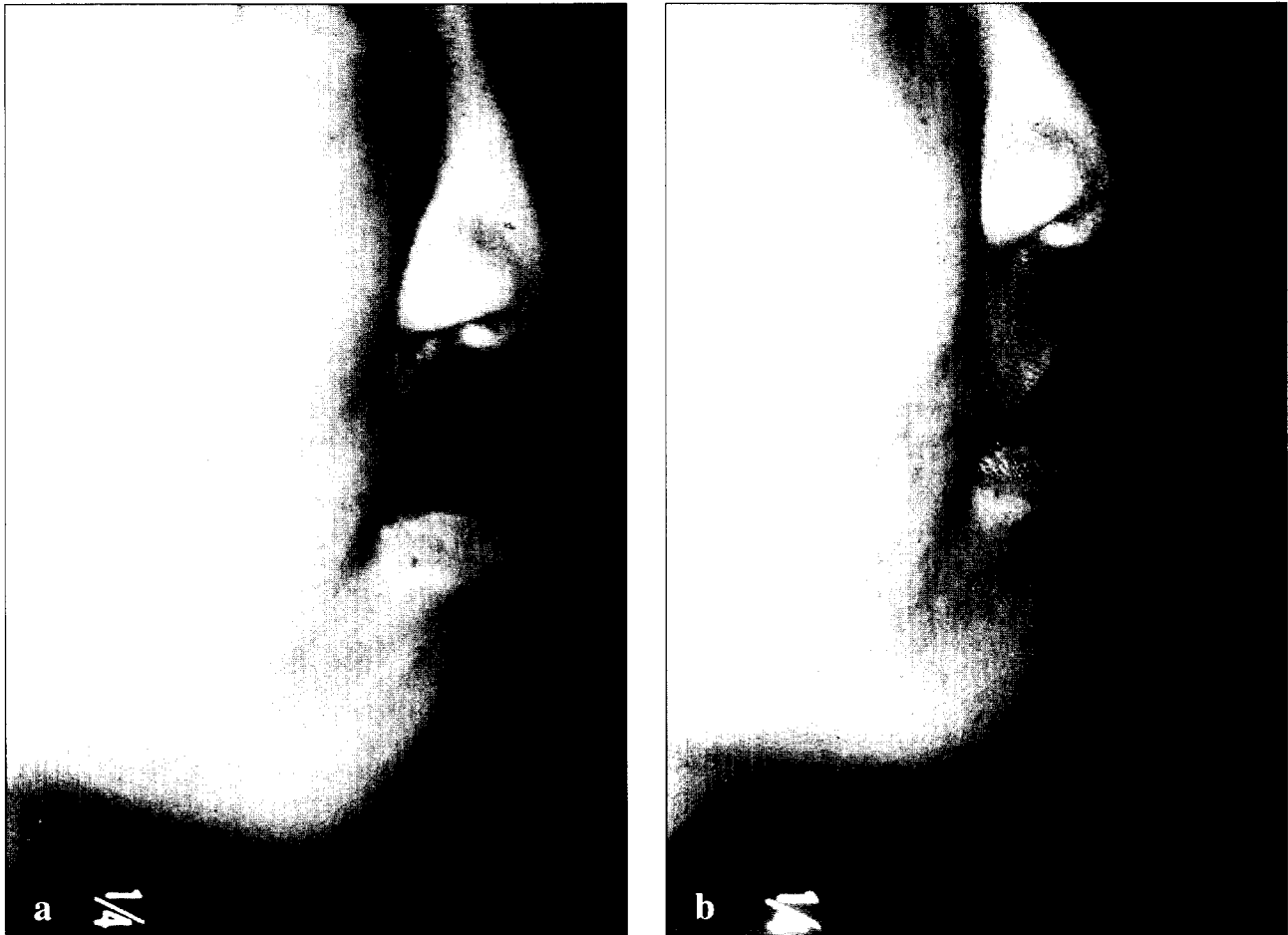


Fig 7. An adult patient referred because of a severe skeletal deformity due to sub-optimal treatment in childhood. 7a. Before. 7b. After orthognathic surgery.

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