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Cleft Lip and Palate Primary Repair

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Preface

Congenital cleft lip and palate is the most common cranio-maxillo-facial birth defect. It can markedly affect the morphology and almost all the functions in the facial area except vision. Religion, superstition, invention and charlatanism characterized the early history of cleft lip. Spartans and Romans killed cleft lip children, whom they considered harbour evil spirits, while the Greeks simply ignored them. As saner senses prevailed, Fabricius ab Aquapendente (1537-1619) was the first to suggest an embryological basis for clefts. The knowledge of cleft lip and its surgical correction expanded significantly between the Renaissance and the 19th century. During this period, Pierre Franco's Petit Traité and Traité des Hernies described the cleft condition as "lievré fendu de nativité" (cleft lip present from birth). The first ever documented cleft lip surgery is from China, in 390 BC, performed on an 18 year old patient named Wey Young-Chi, who wanted to be a soldier. Albucasis of Arabia and his fellow surgeons used cautery instead of the scalpel, and Yperman in 1854 recommended dissecting the cleft margins with a scalpel before suturing them with a triangular needle dipped in wax. The repair was reinforced by passing a long needle through the two sides of the lip and fixing the shaft of the needle with a figure-of-eight thread over the lip. Germanicus Mirault was the originator of the triangular flap, which was later modified by C.W. Tennison in 1952 and Peter Randall in 1959. In the late 1950s, Ralph Millard presented his "cut as you go" technique. Similarly, the protruding premaxilla of a bilateral cleft lip has also been dealt with in several ways throughout the ages—from being totally discarded, to being pushed back by a wedge resection of the vomer, to finally being left to the orthodontists.

Though Tennison and Millard, who established the foundations of modern cleft lip repair, significantly improved the previous technique of cheiloplasty and achieved more stable, long-term outcomes, there remained a remarkable amount of secondary deformities after the application of their techniques. Furthermore, it is especially difficult for junior surgeons to expertly practice their methods and obtain consistently favorable results. In order to resolve these problems, other researchers have carried out much methodological work characterizing cleft lip

deformities in the last twenty years. They partially improved previous methods, especially the rotation advancement method, and created several distinct methods to repair cleft lip. These improvements have made the operations easier to perform and the postoperative results more consistent. These technical improvements in surgery prompted a number of questions, however. What is the soul of cheiloplasty design? In other words, how to harmonize the incision design and the surgical purpose? How can the labial peak on the cleft side be precisely rotated downward, no matter who the surgeon is? What is the value of the horizontal incision at the alar base on the cleft side and how to apply it? What causes the deficiency of columella height and how to correct it? How to design and rebuild the labial tubercle? Cleft lip repair has developed from a purely morphological rebuilding to a procedure which restores the labial form based on functional reconstruction, and aims to lead to normal growth of the upper lip – hence, the academic debate as to whether the surgical method or the surgeon's experience is the more important.

We believe that clinicians can further understand the methods and the underlying principles of different researchers through studying their theories in depth. They can then compare and choose to apply the optimal theories in their daily practice to improve final outcomes.

Primary cleft lip repair is the most important part in the whole therapeutic sequence. Its influence is the widest and lasts the longest. Therefore, it is always meaningful to emphasize the importance of primary lip repair and pursue related research to improve its outcome. This is also the reason why this book mainly focuses on primary repair.

With regard to surgery of the palate, this was virtually nonexistent in the 16th and 17th centuries in Europe. Cleft palate was first repaired in 1556 by Franco. Though the understanding of the velar function and cleft repair was still quite primitive, the first doctor who really succeeded in repairing cleft palate was the French dentist, Le Monnier, who operated on a child with a palatal cleft "from the velum to the incisor teeth", as described in 1766 by Robert. The preliminary procedures to repair soft palate cleft were based on the continuous endeavors of Von Graefe and Roux. Dieffenbach and von Langenbeck further established the method of repairing complete cleft palate, which is also most commonly used by plastic surgeons today. Sir William Fergusson first described the function of palatal muscles in 1844 in a presentation to the Royal Medical and Chirurgical Society of London. Philip Gustav Passavant of Frankfurt made significant contributions to the understanding of palatal function in speech. In one of his monographs on "closure of the palate in speech", written in 1863, he stressed the movement of the soft palate and noted a "forward swelling (of the posterior pharyngeal wall) at the level of the base of the uvula". This later became known as Passavant's ridge. Passavant was also one of the first to attempt to surgically improve velopharyngeal dysfunction. At the same time, some other phenomena

correlated with cleft palate repair were discovered. In the early 1800s, it was noted that a cleft palate was rather a separation of parts of normal size and potential than an underdevelopment or absence of normal parts. External pressure on the cheeks followed by palatal closure was described by Montin in Paris in 1836. When the borders of the cleft were approximated, they were "denuded with a red iron which was dipped in boiling water", and it was noted that "this pressure may result in fracturing of the maxillary bones".

In order to improve the postoperative velopharyngeal function, the pushback procedure invented by Harold Gilles and Kelsey Fry was once regarded as a logical method in theory and widely applied. The V-Y palatal lengthening invented by Ganzer was also widely used.

A most important improvement in palatoplasty is intravelar veloplasty, which originated from the discoveries of the autopsy by Otto Kriens on an autopsy patient with cleft palate. He made the point that the dislocated levator palatini muscles along the fissure should be restored to their normal horizontal position in palatoplasty. It also accords with the principle of "normal to normal", expressed by Millard. Based on this opinion, Leonard Furlow created the double opposing Z-plasty for cleft palate closure.

Modern palatoplasty is no longer the simple matter of closing the cleft only. Account should be taken of how to repair the cleft with less out releasing incisions and on how to recover maximum velar function through the levator reconstruction. It involves several aspects, such as the patient's age, surgical principles and ideas, and the arrangement of sequential therapy.

In a word, the concepts and techniques of modern cleft surgery are almost unrecognizable compared with those of earlier times. The global efforts of clinicians have taken the treatment of cleft lip and palate on to a completely new level. In order to promote continued patient management and development of technique and procedure, a number of experts in the field of craniofacial cleft have contributed their theories and techniques in this book.

The satisfaction for the clinician in treating cleft lip and palate lies in the potential to achieve a successful outcome, although in reality it can be extremely difficult to regain normal morphology and function compared with the ideal results one can imagine and hope for. Therefore, many experts attribute different outcomes to the competence or experience of different surgeons rather than different surgical techniques. The innovation of cleft surgery is increasingly focused on scientific theory rather than on formal surgical technique. We are aware that mere technical advances can bring improvements of a minor nature, while theoretical developments can bring about revolutionary changes. Furthermore, some earlier surgical procedures which originally seemed logical and understandable nevertheless came under scrutiny: naturally, only those of proven worth can be widely accepted and advocated, and this applies even to those that previously might have been regarded as incomprehensible by the majority of

investigators.

Cleft lip and palate is a globally significant birth defect and people can compare the effectiveness of treatment in different countries or areas. The sheer frequency of this condition and the significance of successful treatment for the patient challenge plastic surgeons to develop optimal and effective treatment plans. We all know that primary repair is the most important procedure for patients with cleft lip and palate, which emphasizes not only improved appearance and simplified follow-up treatments, but also the beneficial effects on normal mental development. Therefore, one should never over-estimate the importance of primary repair.

The long history over 150 years of cleft repair has proved and is still proving that the treatment of clefts is not easy. Therefore, I have invited some of the leading world experts who have developed new ideas in primary cleft repair to collaborate in this book, to introduce their theories, techniques and experience in primary cleft repair. The purpose of this book is to help the reader to rapidly acquire experience from worldwide medical institutions, to help them modify their treatment strategy where appropriate and finally improve overall treatment. Much to my delight, I received generous support from a significant number of specialists in this field and I would like to gratefully acknowledge their contributions and thank them.

In order to realize the ideas mentioned above, we have tried to explain primary cleft treatment from theory to technique. I hope this combined approach will enable the reader not only to understand the result but also the reasoning behind the practice.

Bing Shi
Chengdu, China
Mar. 2013

Contents

Part I

Pre-Surgical Treatment of Cleft Lip and Palate

1

The Role of Nasoalveolar Molding in the Presurgical Management of Infants Born with Cleft Lip and Palate

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1.1 Introduction

While the conventional nomenclature of cleft deformity would suggest that only the "lip and palate" are affected, recognition and management of the potentially substantial nasal deformity is necessary in order to achieve an esthetic and functional treatment outcome. In most cleft treatment protocols, the nasal deformity is addressed surgically and requires a number of secondary surgical revisions as the affected individual grows to maturity.

Although surgical techniques to repair cleft lip and palate have improved over the years, arguably the most significant advance in surgical cleft rehabilitation has been the presurgical management of the cleft alveolar and nasal deformity with nasoalveolar molding (NAM) and the subsequent primary nasal reconstruction.

This chapter will describe the clinical indications, clinical management, and how to avoid or manage the potential complications of NAM.

1.2 Treatment Objectives and Clinical Indications of NAM

NAM is the only presurgical infant orthopedic technique that directly addresses

both the cleft nasal and alveolar deformity [1]. The rationale for performing NAM is to reduce the severity of both the hard and soft tissue deformities, thereby facilitating optimal results from the primary surgical correction. Enhanced esthetic outcome of the primary repair should lead to improved early social integration of children with clefts and a reduction in the need for repeated surgical revisions.

NAM uses an intraoral molding plate to gradually approximate the displaced alveolar segments, which results in approximation of the cleft lip segments as well as the medial and lateral aspects of the nose. Once the underlying alveolar segments are approximated, the overlying lip and nasal soft tissues attain a degree of laxity that permits reshaping and alignment with the introduction of the nasal stent. In addition, owing to the plasticity of the alar cartilage during infancy, the achieved changes can become permanent [2].

The nasal stent reshapes and projects the nasal tip while correcting the corresponding deformity of the lower lateral alar cartilage. As the immature nasal cartilage is maintained in the correct shape and position, these positive changes become permanent. In cases with a bilateral cleft deformity where the columella is notably deficient, nonsurgical columella elongation can be performed with the nasal stent, providing the surgeon with adequate columella tissue to achieve the primary surgical repair of the nose.

While not every infant that is born with a cleft lip and/or palate is a candidate for NAM, specific clinical findings may be used to determine which infants are good candidates for this treatment. Nearly every infant with complete unilateral cleft lip and palate (UCLP) and the associated nasal deformity is a candidate for NAM. In complete UCLP, the nasal floor is absent on the cleft side and the greater alveolar segment is often found to be protruding anteriorly, lacking its normal dental arch curvature. The cleft side lower lateral alar cartilage is subsequently prolapsed with deviation of the nasal tip and columella towards the non-cleft side. Asymmetry is the hallmark of complete UCLP as evidenced by comparing the nasal apertures, nasal apices, and columellar heights between the cleft and non-cleft sides (Fig. 1.1a). NAM has demonstrated the ability to improve nasal symmetry in complete UCLP up to the age of 9 years [3, 4] (Figs. 1.1b – 1.1c).

Infants born with complete bilateral cleft lip and palate (BCLP) often present with the premaxillary segment positioned outside the oral cavity, a wide nasal tip and alar base, and a severely deficient columella. The prolabium appears to extend directly from the tip of the nose (Fig. 1.2a). NAM has been shown to obviate or reduce the need for surgical construction of the columella and reduce the number of surgical revisions in complete BCLP cases up to the age of 8 years [5]. Further, it has been demonstrated that NAM can lead to a normalized columella length, nasal tip protrusion, and alar base width at the age of 12.5 years following only the primary nasal reconstruction [6] (Figs. 1.2b – 1.2c).

While the cleft lip and palate deformity has variable expression, the majority of the patients with them could benefit from NAM. For patients with unilateral clefts, the lower lateral alar cartilage deformity, reduced nasal tip projection, and a wide lip and/or alveolar cleft are often improved with NAM. In bilateral cleft