Treatment of Bilateral Cleft Lip and Palate: Protocol for Surgical Treatment

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Bilateral cleft lip and palate (BCLP), although representing around 20% of cleft cases (Trindade and Silva Filho 2007), is one of the greatest challenges faced by craniofacial surgeons, well summarized by Dr. James Barrett Brown as "Bilateral cleft lip is twice as difficult to repair as unilateral, and the results are only half as good" (Brown et al. 1947). Bilateral clefts tend to represent the more severe cases of cleft lip and palate, for which reason an in-depth analysis is warranted, with special attention paid to treatment choice (Brown et al. 1947; Semb 1991). The major surgical challenges of treating BLCP stay from the technical difficulty of achieving symmetry of the lips, muscular continuity, lengthening of the columella, nasal projection, and proper positioning of the premaxilla. Of these challenges, nasal asymmetry, malpositioned or projected premaxilla, and prolabium underdevelopment, which are associated with a lack of muscular continuity, are some of the most difficult to overcome (Mulliken 1985; Millard 1977; Spina et al. 1978).

Of course the final result needs much more surgical skill to achieve the final goal of social reintegration of these patients. The dental occlusion and the speech have double attention for obtaining good functional results.

There are several main points of contention and discussion with regard to bilateral cleft repair: first, the time and type of lip repair, and whether to perform a staged or non-staged repair; secondly, the use of preoperative orthopedic devices, whether active or passive; thirdly, premaxilla repositioning; and finally the ideal time for primary rhinoplasty (Bishara and Olin 1972; Bittermann et al. 2016; Liou et al. 2007; Mulliken 2000).

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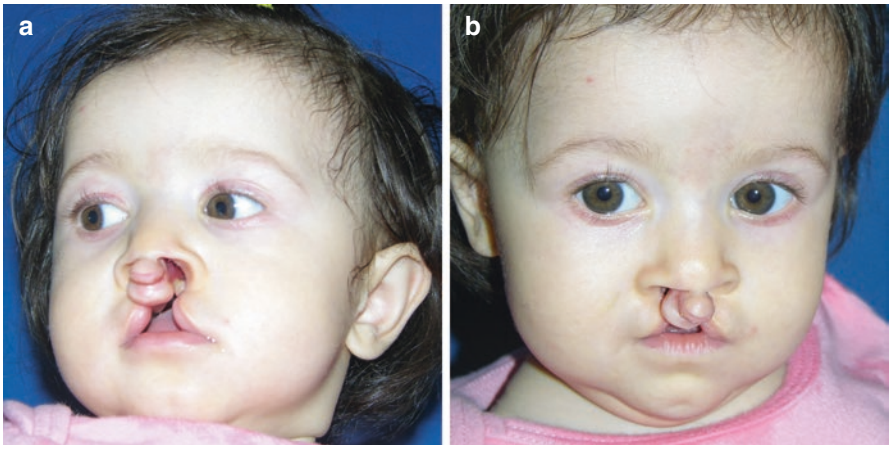

Fig. 8.1 (**a**, **b**) Protruding premaxilla in a complete bilateral cleft. Frontal and lateral view showing the severe projection of the central part of the lip

Bilateral cleft classification is initially determined by whether the cleft is complete or incomplete as defined by the presence or absence of a cutaneous band that maintains the continuity of the inferior aspect of the nares (Spina et al. 1972; Victor 1931). In the complete bilateral the position of the premaxilla is important, which is either projected or not. Nonprotruding premaxilla has the lip repair, nasal elongation, and muscular repositioning as surgical steps; added to all this the complete has the protruding premaxilla limiting the lip repair and primary rhinoplasty.

Bilateral alveolar clefts result in a premaxilla solely fixed to the vomer bone and freely mobile (Bittermann et al. 2016). The malpositioned premaxilla is, without doubt, the anatomic element that causes the greatest technical difficulty. These bilateral clefts can also be differentiated by the grade of development of the prolabium, without muscle and hypoplastic vermillion dry and wet (Spina 1966) (Figs. 8.1 and 8.2).

The protocol of the craniofacial service at the Hospital das Clínicas, University of São Paulo Medical School, is based on two anatomic aspects of the patient: the position of the premaxilla and the dry vermillion and prolabium development. When there is no projection of the premaxilla, primary queiloplasty is done in one stage. The technique for repair is determined by the second criterion, with a well-developed dry vermillion and prolabium allowing for the principles of Spina's technique, and if either is poorly developed, the principles of Noordhoff's technique modified in the department by the senior author (Spina et al. 1978; Spina 1966; Noordhoff 1986) (Figs. 8.3–8.5).

Principles of Spina's technique are used to avoid disruption of the white line in a well-developed dry vermillion. The technique involves using the lateral soft tissue of the lip to reconstruct the median tubercle whilst maintaining the mucocutaneous

Fig. 8.2 (**a**, **b**) Nonprotruding premaxilla with hypoplastic prolabium in two different patients

vermillion border. Principles of our modified technique allow for proper reconstruction of an underdeveloped white line of the dry vermillion. The entire reconstruction of the medial tubercle and medial dry vermillion is performed using the lateral soft tissue of the lip and the mucocutaneous vermillion border is recreated with the same lateral tissue. The muscular belt is done, and the orbicularis oris is sutured in the midline with nonresorbable suture, two guide stitches close to columella and at the upper transition of vermillion (Fig. 8.6–8.10).

In the case of a projected premaxilla, primary queiloplasty must be undertaken in a staged manner. The first repair, which is typically performed between 3 and 6 months of age, is simply a joining of the cutaneous lip borders to help guide the premaxilla to its proper position during facial growth. This first stage is either unilateral or bilateral, depending on the size of the cleft and what is possible, with larger clefts often not allowing for bilateral approximation of the cutaneous lip borders. If this is the case, the widest cleft is closed first and the second side closed 12 weeks later however, if the pre maxilla is not projected, a single stage queiloplasty is performed (less than 7 mm projected). (Spina 1966). Technique for the staged definitive repair, which typically is performed after palatal closure at 12 months of age, is determined once again by the development of the dry vermillion and the prolabium. Spina's repair is performed when the patient has a well-developed dry vermillion and prolabium and the modified bilateral local technique repair is performed when this is lacking. The original description of Spina technique, the final lip repair was done at the age of 5 years old.

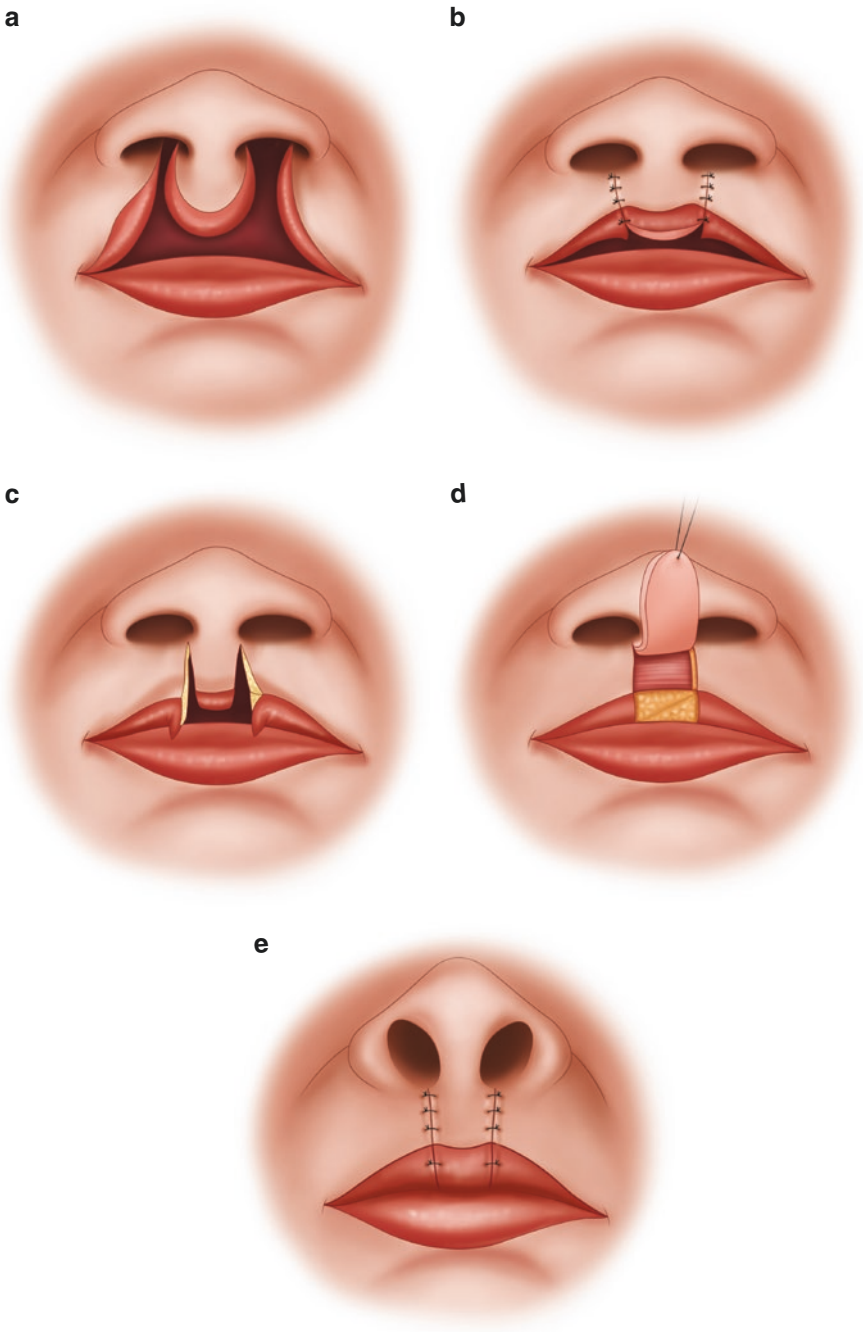

Fig. 8.3 Spina's technique drawings

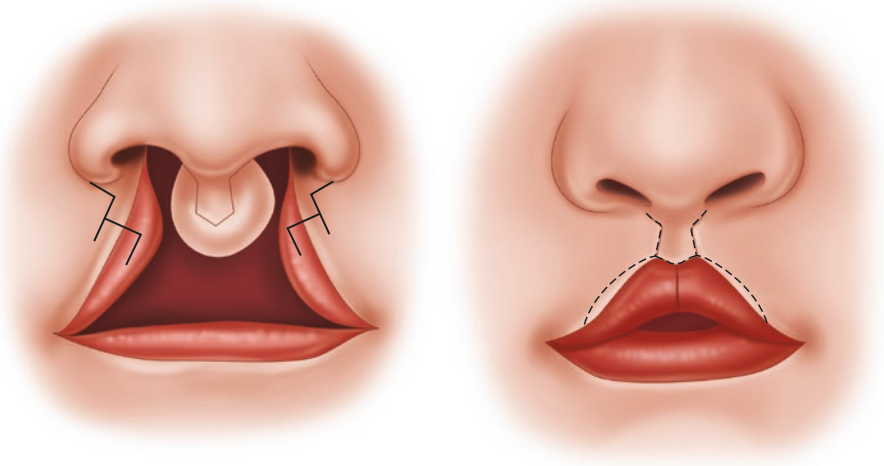

Fig. 8.4 Principles of Noordhoff's technique with some personal modifications

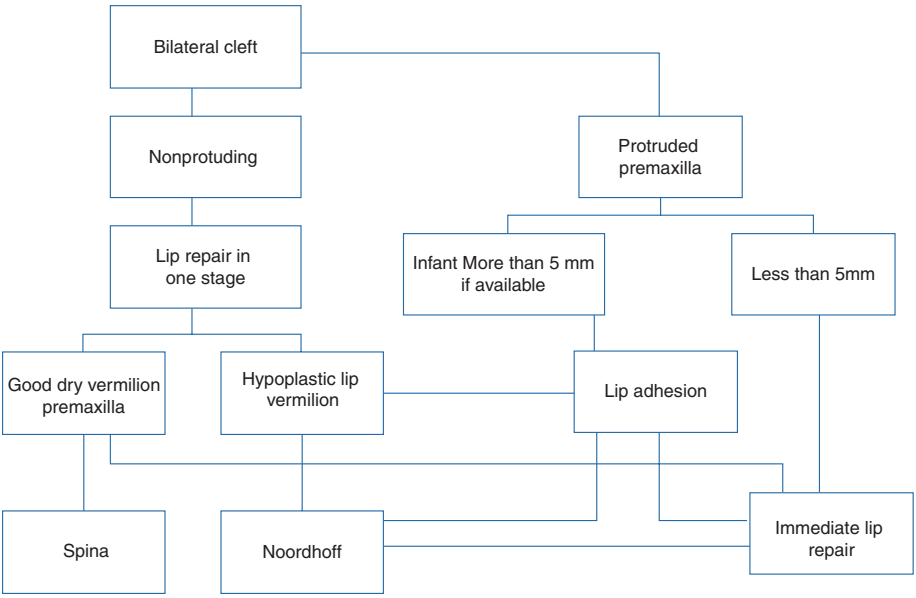

Fig. 8.5 Algorithm of bilateral cleft treatment

Fig. 8.6 Nonprotruding premaxilla with good dry vermillion Spina's one stage

Fig. 8.7 Two-staged premaxilla protruding premaxilla

Fig. 8.8 (**a**–**e**) (**a**) Spina three stages second-side adhesion, (**b**) premaxilla after lip adhesion, (**c**) Spina definite repair, (**d**) final position of premaxilla after adhesion, (**e**) demarcation of Spina definitive repair

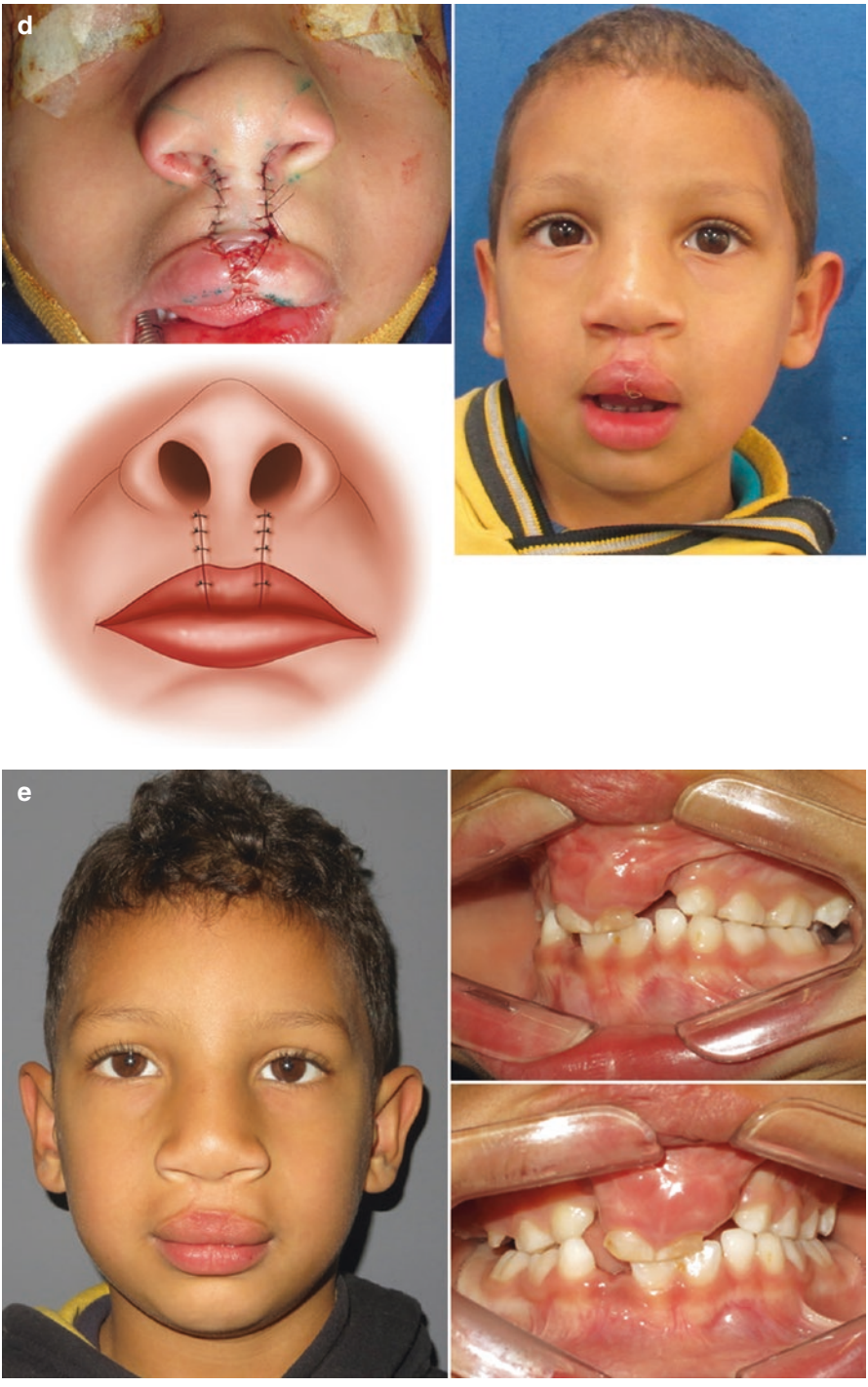

Fig. 8.8 (continued)

Fig. 8.9 Protruding and asymmetrical pre maxilla hypoplastic vermillion (less than 7 mm) Noordhoff's modification

Fig. 8.10 Noordhoff's modification long-term follow-up. (**a**, **b**) Noordhoff's modified one stage with nasal repair pre maxilla non projected

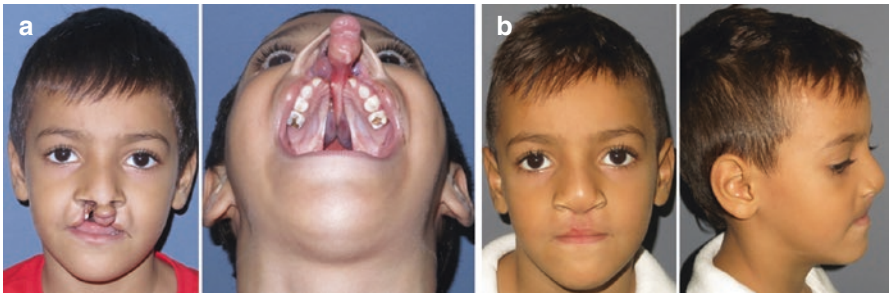

Fig. 8.11 Projected premaxilla nonoperated bilateral cleft patient with repositioning of premaxilla at 6 years old. (**a**, **b**) A projected premaxilla after lip repair; (**b**) useless external traction after surgical procedure and orthodontic

At the time of primary queiloplasty, if there is a projected premaxilla, it is not possible to perform an adequate primary rhinoplasty. The moment of the primary queiloplasty is, nevertheless, the best moment to reposition the nasal cartilage and elongate the columella (Mulliken 1985, 2000, 2001). Primary rhinoplasty is important for achieving final esthetic results in BCLP patients. Historically, nasal alveolar molds (NAM) were used with the desired outcome of approximating tissues preoperatively (Grayson et al. 1999). Many studies have shown no benefit to the use of preoperative devices compared to controls, besides the fact that in many places this practice is not possible (Semb 1991; Spina et al. 1978; Liou et al. 2007; Mackay 2016). In cases of delayed primary rhinoplasty due to a severe projected premaxilla, current practice is to perform a primary rhinoplasty after repositioning of the premaxilla.

There is much discussion surrounding the ideal age for repositioning of projected premaxilla, which is present in the vast majority (70%) of BCLP patients at our institution (Alonso 2016). Our protocol indicates surgical correction for the pre maxilla between 8 and 10 years of age, at the same moment as performing an alveolar bone graft to fix bony discontinuity of the maxilla. If surgical repositioning is performed prematurely, it often results in significant impingement on facial growth (Bishara and Olin 1972; Bittermann et al. 2016; Padwa et al. 1999). Between the moment of primary queiloplasty and surgical repositioning of the premaxilla, orthopedic devices have often been used to provide nonsurgical repositioning of the premaxilla. The presurgical devices are not used in our protocol; only external compression with tapes and elastics bands can be used. Concerns regarding the use of active orthopedic devices that exert traction on a projected premaxilla include possible restriction of the natural progression of facial growth, and data from reference centers show an increased incidence of orthognathic surgery in BCLP patients who previously used orthopedic devices (Good et al. 2007).

A caveat to the timing of performing surgical correction of excess projection of the premaxilla is delayed presentation of a bilateral cleft patient. If the patient has not yet undergone primary queiloplasty by 6–8 years of age, the temporal indication

for surgical repositioning of the premaxilla is as soon as possible with the lip repair. The vomer-premaxillary suture is closed by 6–8 years of age; therefore premaxilla repositioning after this age will not impair facial growth. The primary queiloplasty and premaxilla repositioning will take place at the same time (Bishara and Olin 1972; Padwa et al. 1999). In these cases, there is significant difficulty with respect to primary rhinoplasty, due to extensive local vascularization (Fig. 8.11).

With a large variety of clinical presentations and an array of repair techniques available, even though many improvement was seen still there are several points of contention that often arise with regard to care for BLCP patients. These points of contention are whether queiloplasty should be performed in a staged or non-staged manner, the use of preoperative orthopedic devices, the ideal age for surgical repositioning of a projected premaxilla, and the use of primary rhinoplasty. Evidences are scare as to the best manner in which to answer these questions, and as bilateral cleft patients represent a small percentage of all patients with cleft lip and palate, there has been to date a lack of randomized controlled trials evaluating treatment options. Despite this dearth of evidence, just like care for unilateral cleft, care for bilateral cleft is essential (Bittermann et al. 2016).

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