CLEFT LIP AND PALATE: A CASE SERIES ANALYSIS

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Orofacial clefts (OFC) are common birth defects of complex aetiology resulting in disruptions of normal facial structure. They represent one of the most usual birth defects and occur in 1 per 500 to 2,500 births depending on ancestry, geographic residential location, maternal age and prenatal exposures, and socioeconomic status. Aim of this retrospective study is to assess the clinical outcome in a series of patients affected cleft lip and palate and discuss the pertinent literature. In the period between January 2001 and December 2010, 56 patients underwent to cleft lip and/or palate correction at the Pediatric Surgery Unit, S. Anna Hospital, Ferrara, Italy. Patients included 25 females and 23 males. There were 7 cleft lip, 24 cleft palate and 17 cleft lip and palate. All patients were surgically corrected under general anesthesia. Millard and Skoog techniques were used for cleft lip anomalies, whereas Langebeck, Vidmayer-Perko and Furlow techniques were used for cleft palate defects. In our series a multidisciplinary approach was used and several surgical techniques were performed. Functional and aesthetic results were satisfactory in most cases. The need of a specific dedicated team is mandatory for treating this group of patients.

Orofacial clefts (OFC) are common birth defects of complex etiology resulting in disruptions of normal facial structure (1). They represent one of the most usual birth defects and occur in 1 per 500 to 2,500 births depending on ancestry, geographic residential location, maternal age and prenatal exposures, and socioeconomic status (2, 3).

The common forms of OFC involve disruption of tissue planes above the lip extending into the nares and/or the palate (hard and/or soft). Thus, OFC can occur in three main types: cleft lip only (CL), cleft lip with palate (CLP), and cleft palate only (CP). More than 60% of cases with OFC have CL or CLP (4). More males than females were affected, and more males had complete clefts. Furthermore, unilateral clefts were most common on the left hand side (5).

Because the defects arise early in embryological development, have a complex and multifactorial etiology with both several genetic and environmental factors contributions interacting to shift the complex process of morphogenesis of the primary and secondary palates toward a threshold of abnormality at which clefting can occur (6).

Approximately 70% of cases with cleft lip with/without cleft palate (CL/P) occur in isolation i.e. in isolated entities with no other major birth defects, developmental disabilities, apparent cognitive and structural abnormalities, commonly termed "isolated, non-syndromic OFC" (7, 8). However, they can still occur as part of a broad range of chromosomal, Mendelian, or teratogenic syndromes (1) including other anomalies such as cardiovascular (24–51%), musculoskeletal, facial dysmorphia or genitourinary system disturbance (9). There are over 200 syndromes with CL and /or CP as features. However, children with associated anomalies are more likely to have combined CLP or CP, rather than CL alone. They are often of lower birth weight (10).

Closure of the cleft in the lip and palate requires a surgical operation. The general programme for OFC

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treatment is characterized by a close "multidisciplinary teamwork" and a consistent continuity in methods and follow-up. There are a variety of surgical techniques and timings. Any surgical protocol has to satisfy 1)cosmetic restoration at an appropriate time; 2)functional restoration of the lip and particularly the palate to provide a normal eating and drinking and the development of normal speech; 3)optimum facial growth and development (11).

Generally, the surgical treatment consists of primary lip repair, with simultaneous vomer flap closure through a cleft alveolus, a primary posterior palatal repair and a scheduled later bone grafting of the alveolar defect. In addition, various secondary corrections of nose, lip, sulcus, palate and pharynx are done when indications arise. Moreover, palatal surgery can improve Eustachian tube function. There is a well-recognized association between cleft palate and middle ear disease that is related to failure of the ventilatory function of the Eustachian tube. Eustachian tube dysfunction causes otitis media with effusion (OME), commonly known as "glue ear". However, the right management of OME involves also the use of ventilation tubes inserted through the tympanic membrane under general anesthetic (12).

Several treatments including not only surgery but also speech therapy, dental care and psychological support are available for OFC. However, care of the patient within an indispensable multidisciplinary team requires the development of a protocol of care or care pathway to enable the team members to function most effectively and to maximise the benefits of this system to the patient.

Aim of this retrospective study is to assess the clinical outcome in a series of patients affected cleft lip and palate and discuss the pertinent literature.

MATERIALS, METHODS AND RESULTS

In the period between January 2001 and December 2010, 48 patients underwent to cleft lip and/or palate correction at the Pediatric Surgery Unit, S. Anna Hospital, Ferrara, Italy.

Patients

Patients included 25 females and 23 males. There were 7 cleft lip, 24 cleft palate and 17 cleft lip and palate (Fig. 1).

Treatment

All patients were surgically corrected under general anesthesia (Fig. 2). Millard and Skoog techniques were used for cleft lip anomalies, whereas Langebeck, Vidmayer-Perko and Furlow techniques were used for cleft palate defects. Clefts palate alone were corrected by means of Langebeck (9 cases), Widmayer-Perko (8 cases) and Furlow techniques (7 cases). Clefts lip were closed by using Skoog (2 cases) and Millard (5 cases) techniques. Clefts lip and palate were surgically treated by means of a combination of the above mentioned techniques. Clefts palate were corrected by means of Langebeck (8 cases), Widmayer-Perko (5 cases) and Furlow techniques (1 case). Clefts lip were closed by using Skoog (5 cases), Millard (5 cases), Straigth line (5 cases) techniques. Two clefts lip were corrected in other surgical unit.

DISCUSSION

Orofacial clefts (OFC) are etiologically heterogeneous defects and this has critical implications for understanding the biology of facial development, how environmental risks interact with genetic factors and how we can incorporate known etiologic variables to improve clinical care (1).

Although the OFC mode of inheritance has been investigated for many years, the results obtained are controversial. The discrepancy is probably a result of both the sample and the models used. OFC investigations are limited by the small pedigrees usually available, the reduced number of affected individuals in the pedigree (possibly a result of low penetrance), and the genetic heterogeneity exhibited by this malformation (13). Furthermore, environmental factors seem to play a role in the onset of the malformation. Maternal smoking has been associated repeatedly with increased risk of OFC (14) and the meta-analysis by Little et al.(15) strongly supports an overall odds ratio (OR) for having OFC of ~1.3 among offspring of mothers who smoke. Moreover, increased risks from exposure to maternal smoking during the peri-conceptual period raises the possibility that genes in certain metabolic pathways may play a role in the development of OFC (16). Exposure to maternal alcohol consumption has also been suggested as a risk factor, however, studies also suggest that 'binge' drinking patterns (high doses of alcohol in short periods of time) increase risk, and this is supported by associations with variation in the ADH1C alcohol dehydrogenase gene (17).

Besides nutrients and toxins other environmental exposures have been assessed for possible roles in clefting. These exposures include hyperthermia, stress, maternal obesity, occupational exposures, ionizing radiation, cortisone or phenitoin and infection (18). Food fortification programs using folic acid have shown detectable decreases in the rates of clefting in some (19) but not all studies (20).

The outline of care begins with antenatal diagnosis and continues to adulthood. The patient may be under active surgical treatment by several techniques. An outline of the treatment pathway adopted by the Pediatric Surgery Unit at S. Anna Hospital, Ferrara, provided *Millard* and *Skoog* techniques for cleft lip anomalies, *Langebeck*, *Vidmayer-Perko* and *Furlow* techniques for cleft palate defects.

The "Millard's lip repair" is based on rotation-advancement technique. One of the main advantages of

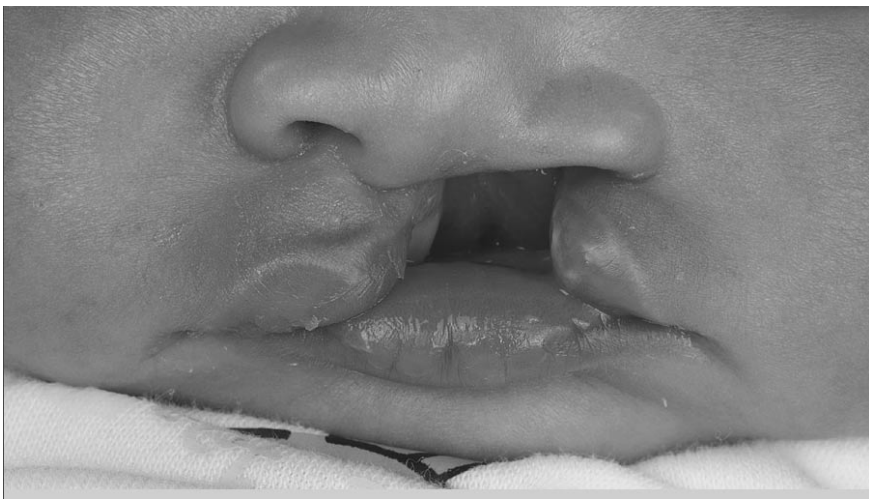

Fig. 1. *Cleft lip and palate*

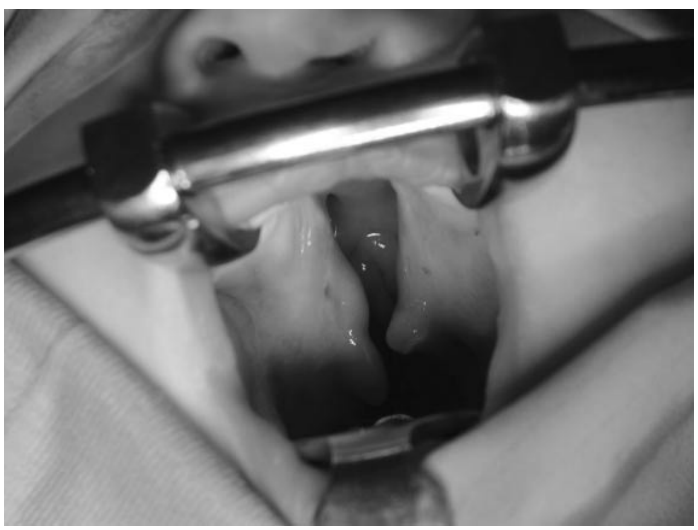

Fig. 2. *Surgical treatment*

this surgical procedure is good access to the nose by way of incision around the ala on the cleft side. Medial and lateral elements are cut to maintain the flaps vermilion. The incision is made on the non-cleft side and the segment is rotated inferiorly to establish the proper position of the Cupid's bow. A back-cut, combined with Z- plastic, is used to rotate the medial portion of the lip. It's important to release the orbicularis muscle from its abnormal attachment. After incision the underlying muscle will expand to its normal dimensions, then it can be sutured by a separate layer.

The "Skoog technique" is based on the creation of the triangular flaps. It's important to make precise measurements in which the triangular flap is placed in the lower portion of the lip. Its dimensions are calculated to fit the gap of the non-cleft side to establish symmetric height on both side of the lip. It's performed a small triangular flap at the base of the ala on the cleft side designed to fit into defect at the base of the columella. Two-triangular flaps are designed, one in the lower and one in the upper portion of the lip. The small upper flap is used to help the reconstruction of the nostril floor. At the free border a full-thickness triangular flap is made to bring the cupid's bow at the normal level. In this way the columella is straightened and the alar base is repositioned to create a symmetric aspect to the controlateral side.

It is important to say that the correction of bilateral cleft lip is not a double correction of a unilateral cleft. Orthopedic devices are often used preoperatively to try to approach the edges of the cleft and expand the maxillary lateral segments. This is a simultaneous and bilateral reconstruction, which uses mucosal flaps taken from the sides of the cleft, turned caudally and advanced towards the middle line, to create a deep buccal groove and to avoid that the center of the lip is firmed with the premaxilla. The mucosa on the medial side of the cleft is turned down. Vermilion is left attached to the white line of the prolabio. The side flaps are united under the central mucosa to create the central tubercle. The closure is then performed in layers, the nasal floor is closed by bilateral vomerine flaps, turned sideways and united under the periosteum of the palate.

In the "Von Langebeck" technique the operative principle is to mobilize the tissues lateral to the cleft, moving along the midline and suturing. In detail, the purpose of an early intervention with this technique approach is to detach the tissue proceeding with an incision at the level of both sides of the cleft in the anterior-posterior direction to reach the base of the uvula to the edge of the rear pillar. Once created the engravings must mobilize the tissues prior use of special periosteal able to penetrate between the bone and the mucous. However, this process meets the palatine artery, which should be isolated to avoid the heavy bleeding caused by his interruption. If the dissection is performed to a sufficient extension and depth, the two side edges of the palate can be conducted to meet in the midline without any difficulty or tension.

The "Widmeyer-Perko" technique involves two stages of intervention, on the soft palate and hard. The incision of the mucosa, starting from the posterior third of the hard palate, get two separate flaps with particular attention to maintaining adequate blood supply. This manoeuvre allows for two flaps or "wings" to strip the muscular structures and to create the conditions to reconstruct the nasal floor through a proper dissection of mucosal surfaces, the approaching on the midline of the parties, and pack a longitudinal suture. Then, should be detached the anterior insertions of the elevator muscles of the palate to the edge of the hard palate and then rejoined the two free portions together in order to rebuilding a new structure of the transverse muscle to make the performance criteria of the palate. With this background it is now able to reconstruct the nasal planum and the mouth with the operations of detachment and approach of the flaps in the midline. The second half of the technique involves the reconstruction of the oral and nasal plan mobilizing and detaching a single flap which is sutured with the portion of the mucosa of the opposite side. The technique, in fact, provides a single manoeuvre mobilization and transposition of the flap from the intact side to the affected by cleft and the subsequent creation of a suture with the only trick to reduce to minimum tensions.

In the "Furlow" technique the sequence of manoeuvres to be performed provides for the incision of the oral mucosa and to peel off portions of the muscle-aponeurotic plane from nose to get, from both sides of the cleft, two flaps that are able to be combined for the reconstruction. The manoeuvres of detachment allow to free the nasal mucosa, and appropriate incisions allow to create two triangular flaps. Next step is the combination of flaps which leads to the creation of a Z-plastic, where it is important to recreate the muscle-aponeurotic conditions, both on the nose and oral side. A simple translation of the flaps on the plane puts them in a position to approach creating a margin of suture to Z, whose function is not only to join the two edges, but also to allow the plane to stretch to fill completely the affected cleft.

In our series, functional and aesthetic results were satisfactory in most cases by using these specific techniques to cleft lip and/or palate correction. However, the need of a specific dedicated multidisciplinary teem is mandatory and indispensable for the care and the treatment of this group of patients.

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