



Introduction

Cleft lip and cleft palate together are the most common congenital facial defects that manifest in about one in every 800 live births, and the majorities are males. Combined lip and palatal defects represent 86% of cases while isolated bilateral clefts of the lip are distinctly uncommon (*Kirschner and LaRossa, 2003*).

In bilateral cleft "like a tent without insertion of its front pole, the nasal tip never rises, leaving the angle of its alar cartilages flattened and the feet of its medial crus splayed" (*Seibert, 2000*). The surgical management of bilateral cleft nasal deformity remains a functional and aesthetic dilemma for patients, their families, and reconstructive surgeons. It appears that no one procedure has achieved preeminence within the plastic surgical community (*Bardach and Salyer, 1991*).

Although techniques continue to evolve over the decades, the basic principles of cleft surgery remain the same. The main principles are to attain muscular continuity, to achieve an appropriate philtral size and shape, to position the cartilages in a more optimal position, and to and symmetry for optimal appearance and function (*Annette and Craig, 2006*). Ideally, bilateral cleft lip nasal repair should include the following features: (1) a single stage operation at primary lip repair; (2) a nose that is aesthetically pleasing, falling within the normal



anthropometric range and normal physiologic function; (3) technically reasonable with a low complication rate and little need for secondary corrective surgery or highly specialized appliances; (4) minimal scarring that is hidden as much as possible; (5) no interference with growth; and (6) be a cost effective procedure that equates the burdens and demands on the patient and caregivers with the final results (**Seibert, 2000**).

History teaches that operative correction of complex malformations usually evolves from multistage to single-stage procedures (**McComb, 1990**). This is possible because of gradual formulation of surgical principles as guides to improved techniques. This development is exemplified in the history of repair of cleft nasal deformity (**Mulliken, 2004**).

The traditional stratagem for correction of the bilateral complete cleft lip is staged operative repair. The primary focus, however, has always been on closure of the lip, with nasal deformities postponed for "secondary" repair, commonly until childhood or early adolescence. These delayed or secondary surgical efforts are usually termed "columellar lengthening" procedures. All the techniques for secondary columellar lengthening leave their own particular stigmata and problems. In most instances, new scars are introduced across the columella-labial junction, resulting in a circumferential scar around the prolabium that causes it to bulge (**Mulliken, 2004**).



It is essential to emphasize that the bilateral cleft lip nasal deformity is the result of both primary dysmorphogenesis (hypoplasia and deformation) and secondary consequences of well-intentioned surgical repair (**John et al, 2001**). In short, surgical repair of the bilateral cleft lip without the nose can cause the nasal deformity to look worse. Mulliken repairs the bilateral cleft lip, positions the alar cartilages, and closes the alveolar clefts (gingivoperiosteoplasty) in a single stage, usually at 3 to 5 months of age, after completion of premaxillary orthopedics (**Mulliken, 1997**). Although at this age the alar cartilages are small and thin, with careful dissection they can be secured into their proper position overlying the upper lateral cartilages. The alar domes (genua) and medial crura are opposed, and if necessary, the medial footplates are fastened to the caudal septum. The alar bases must be transposed medially so that the interalar width is normal for an infant. The preliminary results with such synchronous repair are very encouraging. It is technically possible to repair synchronously a bilateral cleft lip and the nasal malformation, usually in conjunction with premaxillary gingivoperiosteoplasty (**Mulliken, 2004**).

There is no evidence that careful surgical manipulation of the lower lateral cartilages causes distortion or stunts the growth of these structures (**Anthony et al, 2004**). The future requires learning from the past. The child is branded by the broad nasal tip, slumped nostrils, and flared alae nasi. The



columella is either diminutive or excessively long and abnormally shaped, depending on the type of “lengthening” procedure. Too often these stigmata cannot be erased by revisions. The appearance of a repaired bilateral complete cleft lip and nasal deformity should equal (*Afify and Handesty, 2000*).

Children born with bilateral cleft lip and palate deserve to be cared for by superspecialists. Less than 10% of cleft lips are bilateral, and thus, even high-volume surgeons are privileged to see only a few affected babies per year. In a professional career, a surgeon will follow up only a small number of these children to adulthood. Therefore, it is the obligation of every surgeon who undertakes the care of these children to learn from the past and to analyze results periodically, including comparison with other centers. No infant born with this deformity should grow up exhibiting the consequences of conceptual and technical misadventures (*Mulliken, 2004*).

The standard Mulliken single-stage lip repair technique involves preoperative orthopedics with a Latham device followed by repair of the lip and nasal deformity. Primary nasal correction is completed via rim incisions. Interdomal stitches are placed between the lower lateral cartilages and the interalar distance is narrowed. Excess skin created by this maneuver at the soft tissue triangle is excised in a horizontal crescentic fashion. Postoperative care of the nose includes 19-gauge



catheters acting as nasal stents for 48 hours (*Annette and Craig, 2006*).

Previous attempts concerning primary repair of bilateral cleft nasal deformity aren't enough. Accordingly we are trying through this study to assess whether primary repair of bilateral cleft nasal deformity is of value or not, in the absence of presurgical orthopedics of the premaxilla.





Aim of the Work

The aim of this thesis is to evaluate and compare outcome of nasal morphology in patients who underwent primary bilateral cleft nasal deformity repair with those who underwent isolated bilateral cleft lip repair.



Historical Background

The first cleft lip repair to be described in the world literature was that of the unknown Chinese surgeon who successfully closed the cleft lip of a poor farm boy in the fourth century AD.

For more than a millennium thereafter, cleft lip surgery did not advance much beyond simple cauterization or paring of the cleft edges and reapproximation of the segments (*Kirschner and La Rossa, 2000*).

Von Graefe in 1825 was one of the first surgeons to propose the use of curved incisions to allow lip lengthening (*Noordhoff, 1984*).

His work would later provide the basis for the straight-line closure techniques of **Thompson** (1912) and **Rose** (1991). In 1844, Mirault described the use of a local flap from the lateral lip element to gain length for cleft lip closure. Modifications of Mirault's technique would later become the foundation for the methods described by **Blair** (1930) and **Brown** (1945). These methods, among the first to produce reliable reproducible results remained popular in many parts of the world into the 1950s. Each of these techniques however failed to reconstruct the Cupid's bow and the philtrum

adequately on the cleft side, thereby producing unnatural results (*Kirschner and LaRossa, 2000*).

In the 1920s **Le Mesurier** developed a technique of constructing an artificial Cupid's by turning down a rectangular flap, a technique that remained unpublished until 1948. The triangular flap technique of unilateral cleft lip repair, introduced by **Tennison** in 1952 was among the first to preserve the normal Cupid's bow. Later in 1959, **Randall** described the geometry of the technique, thereby providing a reproducible pattern for lip repair. The **Tennison-Randall** repair was widely used and produced predictable results. The technique has the disadvantages however, of taking the main flap from the already deficient lateral element, of discarding too much tissue, of introducing unnatural zigzag scar into the lip, and of providing little correction of the nasal deformity (*Kirschner and LaRossa, 2000*).

Several surgeons have advocated the addition of a small triangular or rectangular flap from the lateral element into the lower aspect of the repair to provide added length and to preserve the contour of the vermilion-cutaneous roll (*LaRossa, 1998*).

Others have used a triangular lateral vermilion flap to augment the deficient vermilion of the medial element.





Early attempts to repair bilateral clefts relied on complete excision of the premaxillary segment, a technique that uniformly produced unnatural horizontally deficient lips (**Noordhoff, 1984**).

Later techniques preserved the prolabial skin in the upper central lip, adding flaps from the lateral elements to the lower aspect to the prolabium, producing lips that were too long. Recognition of the ability of the vertically deficient prolabium to stretch and to grow when secured to the lateral lip elements was critical to the development of modern techniques of bilateral cleft lip repair (**Kirschner and LaRossa, 2000**).

Evolution of single stage repair:

In 1991, **Trott** and **Mohan** began working in rural Malaysia where single-stage nasolabial repair was a necessity. They opened the nasal tip by lifting the prolabium and columella, dissected the anterior surface of the middle crura, and corrected the displaced alar cartilages, secured the alar cartilages in proper position and redraped them.

The Japanese introduced presurgical moulding of the bilateral cleft nasal deformity. **Cutting** and co-workers extended this scheme to preoperative stretching of the columella by an acrylic, double outrigger and prolabial band, attached to a palatal moulding plate, and secured to the cheeks



with tape. They also use an open-tip approach. However they elevate the prolabial-columellar flap at a deeper plane (membranous septum) so that placement of interdomal sutures is visualized from the underside of the cartilages (***Singh and Nandini, 2009***).



Embryology

At the end of the third week of gestation, the ectoderm in the region of the neural plate begins to fold to produce the neural tube. At the same time, specialized neural crest cells derived from the neuroectoderm appear as paired columns dorsolateral to the neural tube. Despite their ectodermal origin, these neural crest cells make a major contribution to the mesenchyme of the head and neck, the so-called ectomesenchyme. Migration and proliferation of these cells is critical to the formation and the fusion of the facial prominences that will later form the structures of the lip & nose (*Sperber, 1989*).

Early in the fourth week of gestation, five facial primordia develop around the stomodaeum of the embryo: a median frontonasal prominence formed by mesenchyme ventral to the forebrain and paired maxillary and mandibular prominences derived from the first branchial arch mesenchyme (figure 1).

By the end of the fourth week, paired nasal placodes develop on the lower part of the frontonasal prominence. Proliferation of mesenchymal tissue at the periphery of these ectodermal thickenings produces the medial and lateral nasal prominences (figure 2) (*Jones, 1998*).



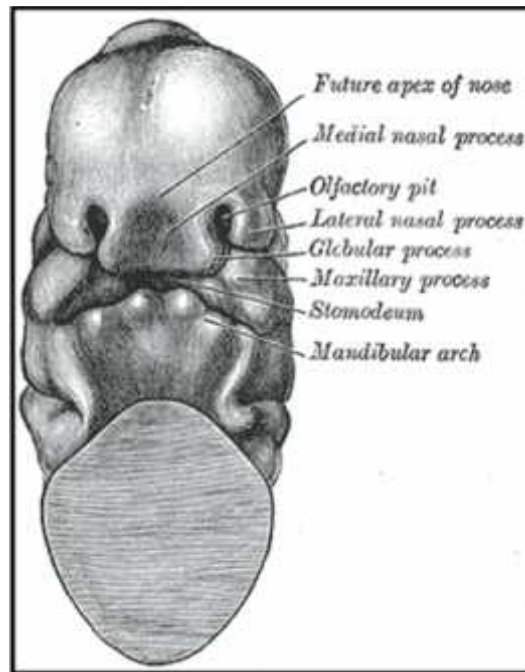


Figure (1): Head end of human embryo of about thirty to thirty-one days (*Susan et al, 2008*).

By the end of fifth week, the maxillary prominences have enlarged and merged with the lateral nasal prominences, establishing continuity of the lip. Failure of one or both of the maxillary prominences to merge with the fused medial nasal prominences results in a unilateral or bilateral cleft lip, respectively (figure 5) (*Moore, 1982*).

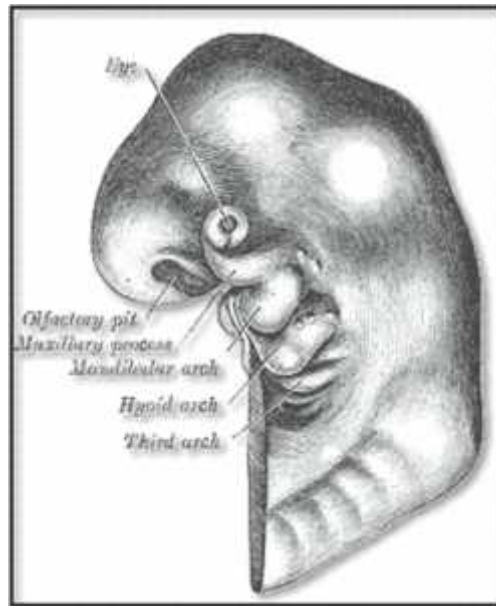


Figure (2): By the up growth of the surrounding parts, the olfactory areas are converted into pits, the olfactory pits, which indent the fronto-nasal process and divide it into a medial and two lateral nasal processes (*Susan et al, 2008*).

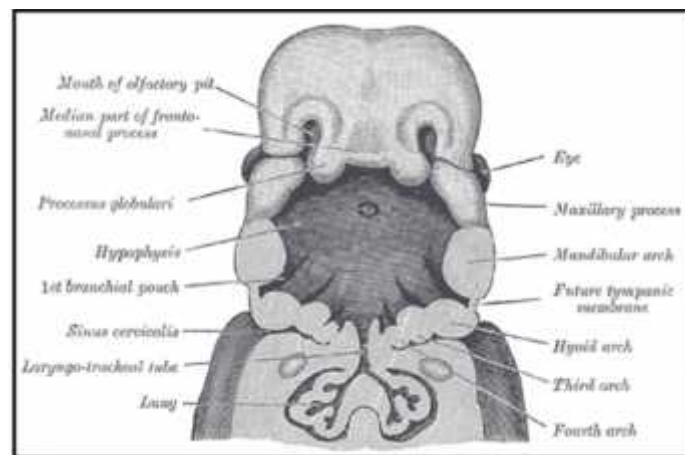


Figure (3): Under surface of the head of a human embryo about twenty-nine days old (*Susan et al, 2008*).

A midline flange (the nasal septum) grows down from the base of the forebrain capsule. Also, from each maxillary process of flange known as the palatal process grows medially across the dorsum of the tongue. The two palatal processes and nasal septum meet and unite from before backwards to form the nasal capsule, so the nasal cavities are separated from each other and they are separated also from the mouth. In mesoderm, chondrification of nasal capsule occurs (figure 4) (*Jones, 1993*).

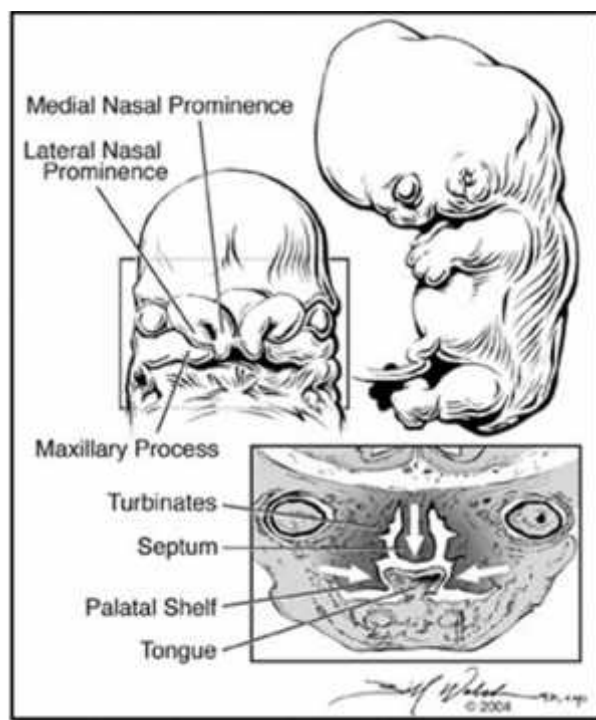


Figure (4): A seven week gestational age embryo. Fusion of the medial nasal prominence with the maxillary process forms the upper maxilla and the philtrum of the upper lip. Cross-section in bottom right shows the formation of turbinates and the separation of the nasal cavities by fusion of the nasal septum with the palatal shelves (*Walsh and Kern, 2004*).

By the sixth week, ossification of the cartilaginous nasal capsule begins in the upper part of the nose, but in the lower part of the septum and in the hard palate, there is deposition of membranous bone on each surface of the cartilage.

The nerve supply of all these structures is derived from the Trigeminal nerve. The frontonasal process is supplied by ophthalmic division, the maxillary process by the maxillary division and the lower jaw by the mandibular division (**McMinn, 1994**).

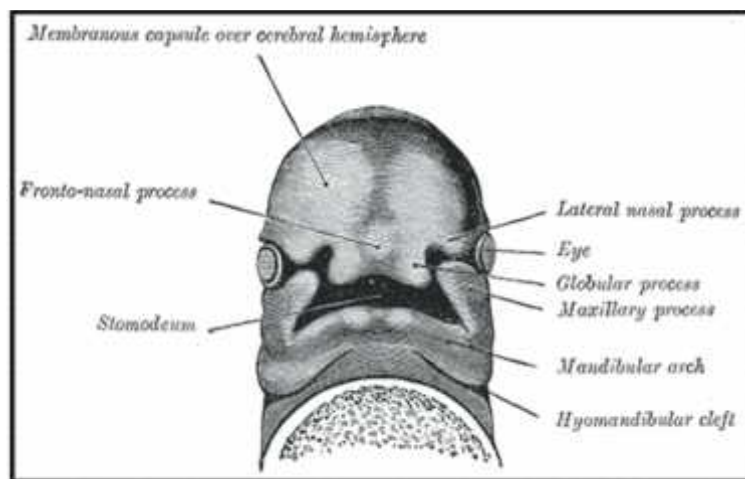


Figure (5): The head and neck of a human embryo thirty-two days old, seen from the ventral surface (**Susan et al, 2008**).

There have been numerous theories as to the cause of the pre-maxillary protrusion in bilateral clefts (**Chandra et al, 1995**).

