

A New Concept to Relate Theories, Pathology and the Clinical Degree of Unilateral Cleft Lip Nasal Deformity

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ABSTRACT

The surgical management of cleft lip nasal deformity remains a functional and aesthetic dilemma for the patients, their families and reconstructive surgeons, and a clear knowledge of the formation of this deformity will be extremely helpful in achieving a final reproducible satisfactory surgical correction. The aim of this study is to prove the new concept which clarify the link between the theories, pathology and the clinical degree of Unilateral Cleft Lip Nasal Deformity. The study included 38 patients. Scoring system was reconstructed to analyze the deformity based on the clinical examination (preoperative photographs). The results showed that the scoring system proves the proportionate and disproportionate relation between the nasal deformity and the degree of cleft lip and palate. In conclusion, the nasal deformity is not always proportionate to the degree of cleft lip. The degree of Nasal deformity in patient with unilateral cleft lip is due to: The Balance between the Extrinsic and Intrinsic factors, meaning that, Patients with nasal deformities proportionate to the degree of clefting are those who are affected by the extrinsic factor more than the intrinsic factor. On the other hand patients with nasal deformities out of proportion to the degree of clefting are those who are affected by the intrinsic factor more than the extrinsic factor.

INTRODUCTION

Cleft lip nasal rhinoplasty is one of the most challenging procedures in plastic surgery domain, due to its complex three dimensional deformities involve the skin, cartilage, vestibular lining, skeletal platform, and a clear understanding of the pathological anatomy is required to achieve a satisfactory aesthetic and functional result [1].

Blair, 1925 [2] was one of the first surgeons to discuss the cleft lip nasal deformities in the scientific literature. With extreme accuracy and based only on his clinical experiences, Blair described many of the abnormalities that we still appreciate today. Blair also acknowledged the role of the deviated cartilaginous septum on the shape of the nose, writing that it can lead to divergence of the entire nose at birth.

Pathogenesis of unilateral cleft lip nasal deformity as described in previous innumerable great studies is due to either the intrinsic, extrinsic or both of these factors.

The intrinsic theory state that during embryogenesis, alterations occur in cells originating from the neural crest. The alterations are accompanied by disordered migration, differentiation, and proliferation resulting in mesenchymal impairment of the nasofrontal process and appearance of the cleft and nasal deformity. Extrinsic theory state that the forces exerted by the perioral muscle has disorganized dynamics caused by the cleft, acting on the nasal wing, contribute to the appearance of nasal deformity.

There is a considerable debate as regard the theories of cleft lip nasal deformities. Proponents of the intrinsic theory stated that the deficiency of the alar cartilage is due to alterations in the cells originating from the neural crest [3-6].

The extrinsic theory was supported by [7-20]. The combination of intrinsic and extrinsic theories contributing to the development of cleft lip nasal deformity was explained by Reynolds and Horton, 1965 [21], and others [22-29].

Typical cleft lip nasal deformity may be associated with complete cleft lip, alveolus, and palate or cleft lip and alveolus with the palate intact. Even incomplete cleft of the lip may lead to severe cleft nasal deformity Byrd et al., 2007 [30]. There is a missing link between the theories, pathology and the clinical degree of nasal deformity in relation to the degree of unilateral cleft lip. The aim of this study is to clarify the relation and clarify the link between theories, pathology and the clinical degree of unilateral cleft lip nasal deformity.

PATIENTS AND METHODS

This study was performed in the Department of Plastic Surgery at Ain Shams University (Cairo, Egypt) on patients who were operated upon for primary lip repair. Preoperative clinical examination and photographs were taken and a scoring system was reconstructed based on eleven points to analyze the deformity.

The study included thirty eight patients, aged between four months and six years. Twenty four males and fourteen females. Ten patients show non syndromatic isolated unilateral cleft lip (seven left and three right side), and twenty eight patients show non syndromatic unilateral cleft lip and palate (twenty two left and six right side). Twenty two patients had complete unilateral cleft lip and sixteen patients had incomplete cleft lip. All patients were not previously operated upon for nasal reconstruction.

All parents signed consent for the surgical procedure, medical photography and their children to be included in the study.

The scoring system based on the clinical examination included 11 points to analyze the lip nose deformity as follows:

- 1- The whole nasal framework, the septum and the anterior nasal spine are shifted toward the non cleft side.
- 2- The nasal tip is ill-defined, broad, and deviated toward the non cleft side.
- 3- Alar base in wide clefts often appears to be broadened and everted at the alar base. It is usually but not always lower, than the non cleft side. In complete clefts, the width of the nose is greater than normal, but each alar base is displayed equally from the mid line.
- 4- Alar rim is lower on the cleft side than the non cleft side, with loss of upward concavity of the alar rim, changed to drooping, downward convexity. There is a skin curtain without a cartilage which droops over the alar rim like a web.
- 5- Alar Dome moves downward, backward, and laterally.
- 6- Alar facial angle is flattened.
- 7- Alar Arch is flattened, and the ala joins the columella at an oblique angle.
- 8- The nostrils are asymmetric. On the cleft side it is horizontally oriented with increased cir-

cumference, and retropositioned because of the deficiency in the underlying maxillary bone.

- 9- The Columella: In the cleft side the columella is shortened in the vertical height. It lies more caudally, tilted and displayed away from the midline toward the non cleft side, and lies obliquely along the edge of the displayed septum.
- 10- Vestibular lining shows the "Plica Vesitbularis".
- 11- Maxilla and premaxilla: When the platform of the nose is clefted, their will be projection and outward rotation of the premaxilla and the retro position of the lateral maxillary segment.

RESULTS

I- According to the scoring system described above, patients with cleft lip nasal deformity proportionate to the degree of cleft lip scored as follows:

- Patient with complete cleft lip scored not less than 8 points out of 11 points.
- Patients with complete cleft lip and palate scored not less than 10 out of 11 points.
- Patients with incomplete cleft lip scored 1 up to 4 points out of 11 points.

II- According to the scoring system described above, patients with cleft lip nasal deformity not proportionate to the degree of cleft lip (incomplete cleft lip) score ranged from 7 up to 10 points out of 11 points.

Examples of patients with cleft lip nasal deformity proportionate to the degree of cleft lip are shown in Figs. (1,2) and Tables (1,2).

Table (1): The score system for patient No. (1).

	Present or Not
Deviated whole nasal framework	+ve
Tip of nose	+ve
Alar base	+ve
Alar rim	+ve
Alar dome	+ve
Alar facial angle	+ve
Alar arch	+ve
The nostrils	+ve
The collumela	+ve
Vestibular lining	+ve
Maxilla and premaxilla	+ve
Total number of points	11



Fig. (1): A- Preoperative anterior posterior. B- Preoperative basal view, patient number one (Right complete cleft lip and palate).



Fig. (2): A- Preoperative anterior posterior. B- Preoperative basal view, patient number two (Left incomplete cleft lip).



Fig. (3): A- Preoperative anterior posterior. B- Preoperative basal view, patient number three (Left incomplete cleft lip).



Fig. (4): A- Preoperative anterior posterior. B- Preoperative basal view, patient number four (Left incomplete cleft lip).

Table (2): The score system for patient No. (2).

	Present or Not
Deviated whole nasal framework	-ve
Tip of nose	-ve
Alar base	-ve
Alar rim	-ve
Alar dome	-ve
Alar facial angle	-ve
Alar arch	+ve
The nostrils	+ve
The collumela	+ve
Vestibular lining	+ve
Maxilla and premaxilla	-ve
Total number of points	4

Examples of patients with cleft lip nasal deformity proportionate to the degree of cleft lip are shown in Figs. (3,4) and Tables (3,4).

Table (3): The score system for patient No. (3).

	Present or Not
Deviated whole nasal framework	+ve
Tip of nose	+ve
Alar base	+ve
Alar rim	+ve
Alar dome	-ve
Alar facial angle	-ve
Alar arch	+ve
The nostrils	+ve
The collumela	+ve
Vestibular lining	+ve
Maxilla and premaxilla	-ve
Total number of points	8

Table (4): The score system for patient No. (4).

	Present or Not
Deviated whole nasal framework	+
Tip of nose	-ve
Alar base	+ve
Alar rim	+ve
Alar dome	+ve
Alar facial angle	-ve
Alar arch	+ve
The nostrils	+ve
The collumela	+ve
Vestibular lining	+ve
Maxilla and premaxilla	-ve
Total number of points	8

DISCUSSION

There is a missing link between the theories, pathology and the clinical picture of the nasal deformity in patients with unilateral clefts of the lip and the palate.

Stenstrom and Thilander, 1965 [10] according to their experience in treating patients with UCLND, they found that there is no question of any abnormal shape of the alar cartilage. Furthermore they mentioned that the lateral crus of both sides are similar, but if occasionally the lateral crus on the cleft side were smaller; this does of course tend to accentuate the deformity. So they concluded that there is no reason that the nasal deformity is a consequence of inherent abnormality of the alar cartilage, and they explained it by saying "The deformities can probably be regarded as secondary to (concealed) partial cleft lips".

Boo-Chai and Tange [12], reported five Asian adults with isolated nasal deformities. The alar cartilage was exposed through rim incision during the repair, they examined the alar cartilage as regard their size and shape, and there was no difference between the both sides. They concluded that there should be a clinical sub group of the nasal deformity with the clinical intact lip, and the nasal deformity was due to extrinsic rather than intrinsic defect in the alar cartilages.

Modolin et al., [21] examined specimens of the lateral portion of both the healthy and the cleft side of the alar cartilages. They concluded that the nasal deformity is due to the action of extrinsic forces, from the time of embryogenesis, on the nasal wing on the side of the cleft, and the muscles on the cleft side have a dyskinetic mechanism whose resulting disordered, fragmentary, or incomplete forces exercise a deforming effect on the nasal wing and consequently on the alar cartilage. They mentioned that a thorough study is needed, especially to assess the composition of the cartilage matrix, not only of the combinations of glycosaminoglycans, but also of collagen. They also noted that "The supposition of the presence of collagen with different degrees of consistency permits us to formulate a third hypothesis that links the intrinsic and extrinsic theories, i.e., that alar cartilage of a different consistency would be more susceptible to dyskinesia of the perioral muscles deformed by the presence of the cleft".

In our previous study, analysis and comparison of the cleft and non cleft side lower lateral cartilage was done through; Intraoperative anthropometric measurement using the digital sliding caliber

(length, width and thickness), One mm punch biopsy was taken for histological Staining (Periodic acid Schiff, Toluiden Blue and Masson's Trichrome stains), and immune histochemical staining (Fibroblast growth Factor 18, Collagen type I & II) and these stains were processed using the image analyzer. It was concluded that pathogenesis of the cleft lip nasal deformity is not only due to an extrinsic factor, which is presented in the misdirected pull of the muscle, but also due to an intrinsic deficiency of the alar cartilage, which makes it prone to the deformational forces by the malinserted perinasal and orbicularis oris muscle [28].

Summary and Conclusion:

Theories of the cleft lip nasal deformity is not only due to an extrinsic factor, which is presented in the misdirected pull of the muscle, but also due to an intrinsic deficiency of the alar cartilage.

According to the results of this study a new concept which link the theories, pathology and clinical presentation of the nasal deformity state that: What determines the degree of the nasal deformity is the balance between the Extrinsic and Intrinsic factors, meaning that:

- Patients with nasal deformities proportionate to the degree of clefting are those who are affected by the extrinsic factor more than the intrinsic factor.
- Patients with nasal deformities not proportionate to the degree of clefting are those who are affected by the intrinsic factor more than the extrinsic factor.

REFERENCES

- 1- Guyuron B.: Late Cleft Lip Nasal Deformity. Plastic and Reconstructive Surgery, 121 (4): 1-11, 2008.
- 2- Blair V.P.: Nasal deformities associated with congenital cleft of the lip. JAMA, 84: 185-187, 1925. Quated from Millard D.R.: Cleft Craft. The Evolution of its surgery. Vol I: The unilateral deformity. Millard D.R. (ed.), Little brown and company Boston 1976; Chapter 49-50-51: 639-689, 1976.
- 3- Tulenko F.J.: Cleft lip nasal deformity in the absence of cleft lip. Plast. Reconstr. Surg., 41 (1): 35-37, 1968.
- 4- Spira M., Hardy S.B. and Gerow F.J.: Correction of nasal deformities accompanying unilateral cleft lip. Cleft Palate J., 7: 112-123, 1970.
- 5- Avery J.K.: The evolution of its surgery, In R.D. Millard (Ed.), Cleft Craft, Boston: Little, Brown, 1976.
- 6- Millard D.R.: Cleft Craft. The Evolution of Its Surgery. Vol I: The unilateral deformity. Editor: D. Ralph Millard, Jr. Little brown and company Boston, Chapter 2: 19-40, 1976.

- 7- Huffman W. and Lierle D.: Studies in the pathological anatomy of the unilateral hare-lip. *Plast. Reconstr. Surg.*, 4 (3): 225-234, 1949.
- 8- Stenstrom S.J. and Oberg T.: The nasal deformity in unilateral cleft lip. *Plast. Reconstr. Surg.*, 28: 295-305, 1961.
- 9- Stenstrom S.J. and Thilander B.: Cleft lip nasal deformity in the absence of cleft lip. *Plast. Reconstr. Surg.*, 35 (2): 160-166, 1965.
- 10- Atherton J.: A descriptive anatomy of the face in human fetuses with unilateral cleft lip and palate. *Cleft Palate J.*, 4: 104 -114, 1967.
- 11- Boo-Chai and Tange: The isolated cleft lip nose. *Plast. Reconstr. Surg.*, 41 (1): 28-34, 1968.
- 12- Latham R.A.: The pathogenesis of the skeletal deformity associated with unilateral cleft lip and palate. *Cleft palate J.*, 6: 404-414, 1969.
- 13- Hogan V.M. and Converse J.M.: Secondary deformities of cleft lip and nose. In Grabb W.C., Rosenste in S., Bzoch K. (eds): *Cleft lip and palate*. Boston: Little, Brown, 1971. Coated from Goodman W. and Zoran M.: The unilateral cleft lip nose. *The Journal of Otolaryngology*, 11 (3): 198-203, 1982.
- 14- Fara M., 1977: The musculature of cleft lip and palate. In Converse J.G., McCarthy J.G. (eds): *Reconstructive Plastic Surgery*. Philadelphia: Saunders, 1977. Coated from Bardach J. & Cutting C.: *Anatomy of the unilateral and bilateral cleft lip and palate. Multidisciplinary management of cleft lip and palate*. Janusz Bardach, Hughlett L. Morris (eds). W.B. Saunders, 1990.
- 15- Heckler F.R., Oesterle L.G. and Jabaley M.E.: The minimal cleft lip revisited: Clinical and anatomical correlations. *Cleft Palate J.*, 16 (3): 240-247, 1979.
- 16- Green M.F.: The embryological development and functional importance in the repair of the nasal mucosa to reduce the deformity of the cleft lip nose. *Scand J. Reconst. Surg.* 21 (1): 1-5, 1987.
- 17- Sadove R., Ladaga L. and Magee W.P.: Cartilaginous histology of the cleft lip nose: Proving the extrinsic etiology. *Plast. Reconstr. Surg.*, 8 (5): 655-660, 1988.
- 18- Beyoung-Yun P., Dae-Hyun L. and Young-Hoo L.: Anatomy of the lateral crus of the alar cartilage in unilateral cleft lip nasal deformity. *Plast. Reconstr. Surg.*, 101 (4): 915-920, 1998.
- 19- Ai-Qun Li, Sun Y., Wang G., et al.: Anatomy of the Nasal Cartilages of the Unilateral Complete Cleft Lip Nose. *Plast. Reconstr. Surg.*, 109 (6): 1835-1838, 2002.
- 20- Modolin M., Baracat G. and Kamakura L., et al.: Histological comparison of the alar nasal cartilages in unilateral cleft lip. *Rev. Hosp. Clin. Fac. Med. S. Paulo.*, 57 (4): 143-146, 2002.
- 21- Reynolds J.R. and Horton C.E.: An alar lift procedure in cleft lip rhinoplasty. *Plast. Reconstr. Surg.*, 35: 377-384, 1965.
- 22- Berkeley W.T.: Correction of secondary cleft-lip nasal deformities. *Plast. Reconstr. Surg.*, 44 (3): 234-241, 1969.
- 23- Malek R.: Nasal deformities and their treatment in secondary repair of cleft lip patients. A clinical study of 175 cases. *Scand J. Plast. Reconstr. Surg.* 8 (1-2): 136-141, 1974.
- 24- Mc Comb H.: Treatment of the unilateral cleft lip nose. *Plastic Reconstructive Surgery*, 55 (5): 596-601, 1975 .
- 25- Sawheny: The nasal deformity in unilateral cleft lip. *Plast. Reconstr. Surg.*, 58 (1): 291- 295, 1976.
- 26- Millicovsky G., Ambrose LH. and Johnston M.C.: Developmental alteration associated with spontaneous cleft lip and palate in CL/FR mice. *J. Anatomy*, 164 (1): 29-44, 1982.
- 27- Rifley W. and Thaller S.: The residual cleft lip nasal deformity. What's new in plastic surgery?. *Clinics in Plastic Surgery*, 23 (1): 81-92, 1996.
- 28- Byrd H.S., El-Musa K.A. and Yazdani A.: Definitive repair of the unilateral cleft lip nasal deformity. *Plast. Reconstr. Surg.*, 120 (5): 1348-1356, 2007.
- 29- Saleh M.A., Elshahat A., Emara M., et al.: Objective Tools to Analyze the Lower Lateral Cartilage in Unilateral Cleft Lip Nasal Deformities, *The Journal of Craniofacial Surgery*, 22 (4): 1435-1439, 2011.