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Review Article

Functional and aesthetic correction of secondary unilateral cleft lip nasal deformities

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ABSTRACT

The treatment of patients with unilateral cleft lip has undergone significant development during the last decades. With better understanding of the anatomy of the unilateral cleft lip and nasal deformities, primary correction of the nasal deformity at the time of lip repair, critical evaluation of short and long-term results following various treatment protocols, and constant striving for perfection in both aesthetics and function, we have been able to design improved treatment strategies and more accurate surgical techniques so as to achieve overall superior and long-lasting results. In this review article, we present our protocols and experience for functional and aesthetic correction of secondary unilateral cleft lip nasal deformities and a retrospective review of 219 consecutive patients treated at our Craniofacial Centre for correction of secondary unilateral cleft lip nasal deformities. The protocols used in the treatment of 219 consecutive patients at our Craniofacial Centre for correction of secondary unilateral cleft lip nasal deformities were reviewed. In addition, analysis of the most recent 51 consecutive patients who underwent complete clinical and functional evaluation with rhinomanometry followed by correction of the cleft lip nasal deformity was performed. A variety of time-honoured techniques of rhinoplasty were applied in the correction of the residual deformities to achieve symmetry, aesthetic balance, and functional correction of the nose. Follow-up ranged from 5-11 years. Analysis of the data revealed that 39 patients (76.47%) had significant functional and aesthetic improvement; seven patients (13.07%) had significant aesthetic improvement but a modest functional improvement; and five patients (9.8%) required additional surgery to improve their appearance and had no functional improvement. Further analysis demonstrated that five out of seven patients in the second group had pharyngeal flaps in place that were primarily responsible for the airway obstruction. No attempt was made to revise the ports of these flaps because the speech was excellent. The surgical plan is based on the information gained from our extensive clinical evaluation and is tailored to the patient's specific functional and aesthetic needs.

KEY WORDS

Cleft rhinoplasty; Cleft nasal deformity; Secondary cleft deformities

INTRODUCTION

The treatment of patients with unilateral cleft lip has undergone significant improvement during the last decades. With better understanding of the anatomy of the unilateral cleft lip and nasal deformity, incorporation of rhinoplasty at the time of lip repair, critical evaluation of short and long-term results following various treatment protocols, and constant striving for perfection in lip and nasal aesthetics and function, we

have been able to design improved treatment strategies and more accurate surgical techniques to achieve overall superior and long-lasting results.

Preoperative management and repositioning of the maxillary segments with active and passive orthopaedic protocols and manipulations of the cleft side lower lateral cartilage and columella with cartilage moulding devices has contributed to improved final results. Cumulative experience and critical analysis of long-term outcomes have further assisted us in improving our techniques and subsequent outcomes of primary unilateral cleft lip and nasal deformities, achieving lips with excellent appearance and function while significantly reducing the number and extent of secondary procedures and revisions.

Most surgeons agree that despite these major improvements in cleft lip surgery, secondary rhinoplasty will be indicated in a significant number of patients for aesthetic and/or functional reasons.[1,2] Furthermore, a number of patients are still referred to major centres with suboptimal results following the initial surgery. Thus, secondary correction of residual nasal deformities associated with unilateral cleft lip remains an important procedure for the total habilitation of the patient with a facial cleft.

Historically, many surgical procedures and modifications have been described to correct the residual cleft nasal deformity. Several of these have resulted in very good objective and subjective aesthetic results. It is well known, however, that a significant number of patients with repaired unilateral cleft lip also suffer from varying degrees of airway obstruction that may have significant implications on the quality of life.[3] Surprisingly, little attention has been paid to the functional aspect of the deformity and the functional outcomes associated with these corrective procedures. In this review article, we present our protocols and our experience for functional and aesthetic correction of secondary unilateral cleft lip nasal deformities.

Treatment protocol

Timing

In an effort to streamline care and to address both the aesthetic and functional issues associated with the patient's residual unilateral cleft nasal deformity, 20 years ago, our centre established a comprehensive, multidisciplinary protocol for care[4] [Table 1]. It is important to stress from the onset, however, that one must remain flexible and that the protocol should be regarded only as a framework and that treatment plans should be individualized based on extensive analysis of a given patient's specific deformity and needs.

Timing of the definitive nasal deformity correction is critical and is determined by several factors. It is deferred until the following procedures have been completed: closure of the possibly coexisting oronasal/palatal fistulas, bone grafting of the alveolus and the hypoplastic maxilla, and orthodontic alignment of the maxillary dentition. If the patient is to undergo orthognathic surgery, final nasal reconstruction is deferred until after this as well. There are several reasons for this timing protocol:

To achieve a symmetric result it is critical that the alar bases start at a spatially symmetric level. Prior to nasal reconstruction the depressed base on the cleft side is raised to a favourable level. This might be accomplished through alignment of the maxillary segments, alveolar bone grafting, or osteotomies during orthognathic surgery. If the maxilla and dentition are in acceptable position but the alar base is still depressed, then additional onlay cortical bone grafting might be indicated in order to raise the alar base to a symmetric height before rhinoplasty.

When oronasal and/or palatal fistulas are present, saliva and food particles regurgitate into the nasal cavity, chronically irritating the nasal mucosa and creating tissue thickening which further exacerbates the airway obstruction. Closure of fistulas eliminates the irritation and swelling of the nasal lining, thus reducing one element of the airway obstruction and assisting the

Table 1: Timetable for definitive repair of residual nasal deformities in patients with unilateral clefts of the lip

Procedure	*Age**
First-stage orthodontic care with arch alignment	7-9 years
Closure of oronasal/palatal fistulas and bone graft of the alveolus and hypoplastic maxilla	7-9 years
Second-stage orthodontic treatment	9-13 years
Definitive correction of residual cleft nasal deformity	14-16 years**

*Ideal age for each intervention/surgery, **If orthognathic surgery is indicated this takes place following dentofacial skeletal maturity; thus correction of residual cleft nasal deformity would be further delayed

surgeon in better evaluating and managing the airway obstruction.

Preoperative Evaluation and Planning

Ideally, for patients that have been at our Centre from early in their treatment, we plan the definitive procedure after most of the nasal growth has been completed, typically around 14 to 16 years of age. This might be later if the patient is to undergo orthognathic surgery. Each patient undergoes extensive examination to evaluate all aesthetic and functional elements that comprise the deformity. These elements are recorded and taken into consideration in formulating the surgical plan [Table 2].

The external nasal deformity is evaluated for asymmetries and deviations in the sagittal, horizontal, and coronal planes. More specifically, the following are assessed: degree of deviation of the nasal pyramid; nasal width; position of the dorsum; asymmetry; depression and/or deviation of the tip; tip projection; asymmetry of the nasal base; size, shape and inclination of the nostrils; condition of the columella and nasal cartilages; and presence of external scarring.

Intranasal rhinoscopic evaluation with nasal speculum and adequate lighting is performed in every patient. All intranasal abnormalities, including condition of the mucosa, presence of scars, septal deviation, previous resection of septal cartilage, septal perforations, condition and position of the upper and lower lateral cartilages, lining deficiencies, vestibular webs or synechiae, turbinate hypertrophy and clinical obstruction are also identified and recorded. Most patients with unilateral cleft lip nose deformity present with a variable degree of septal deviation. Typically, the caudal portion of the septum in patients with a unilateral cleft lip deviates away from the cleft side. This deviation can primarily be C-shaped or S-shaped. On several occasions the septum is dislocated outside the maxillary groove. In addition to deviation of the septum, one can encounter deviations of the perpendicular plate of the ethmoid and the vomer as well as bony spurs from the floor of the nose and the crest of the maxilla. Several patients will present with "iatrogenic" deformities due to previous surgical interventions, scarring, or even septal perforation that further accentuate nasal resistance and decrease airflow. Thus, as with the rest of the procedure, the surgeon should proceed according to the findings and individualize the septal work accordingly.

Intraoral examination is performed to identify the presence of residual oronasal/ palatal fistulas that could further contribute to the airway obstruction. Furthermore, several patients with clefts might have a pharyngeal flap or a pharyngoplasty in place that can also contribute to the airway obstruction.

Special attention is given to the subjective and objective evaluation of the airway. Prior to evaluation, patients and families are asked to complete a questionnaire adopted from symptoms listed by Thurston *et al*. to give an initial impression of the patient's airway issues.[4] If the nose is

Table 2: Preoperative evaluation of patients with residual unilateral cleft lip and nasal deformity

- Evaluation of external deformity, including:
  - asymmetric deviation (sagittal, horizontal, coronal planes)
  - projection and symmetry of the nasal tip
  - shape and size of nostrils
  - condition of the columella
  - skin and cartilage deficiencies
  - scars
- Functional evaluation, including:
  - condition of the nasal lining
  - septal deviation and other pathology
  - turbinate hypertrophy
  - other factors obstructing airway
  - scarring
  - tissue deficiency
- Oral examination:
  - presence of fistulas (oronasal, nasolabial)
  - presence of pharyngeal flaps/pharyngoplasties
- Questionnaire for nasal/airway obstruction, subjective evaluation of patients by family
- Nasal endoscopy for patients with significant airway obstruction and for pharyngeal flaps/pharyngoplasties
- Imaging with CT scan for patients with history of multiple infections and possible paranasal sinus pathology
- Objective evaluation of airway obstruction with component rhinomanometry

congested from allergies or a cold, a decongestive spray is used to yield a more accurate idea of the airway status. In most cases, airway obstruction is due to septal deviation and inferior turbinate hypertrophy. Other causes of obstruction include intranasal scarring, previous excision of lining and cartilage, and nostril collapse.

If other intranasal pathology, such as bony spurs and distant scarring are present, we use the endoscope to determine as accurately as possible the level of obstruction. Nasopharyngoscopy is useful in fully evaluating the contribution of previous flaps or pharyngoplasties to airway obstruction and determining the possible need for port size revisions.

Radiography and other imaging techniques (e.g. computed tomography) are employed when patients present with significant airway obstructive symptoms or symptoms and signs of sinus disease.

Component rhinomanometry is routinely used for the evaluation of the airway patency and resistance. Here we are able to obtain valuable objective information about the status of the airway and the location of the obstruction (anterior, posterior, left, right). This modality assists us in better understanding the status of the airway preoperatively and comparison with postoperative data provides us with objective information about the functional outcome of our procedures.

Surgical Techniques

The surgical plan is formulated based on the information gained from extensive clinical evaluation. This plan is designed to address and correct all external and internal aspects of the deformity and those factors contributing to the airway obstruction. For relatively minor deformities, a closed approach is employed.

For cases requiring significant reconstruction, the open approach is favoured. When significant inferior turbinate hypertrophy is present, we begin with turbinate reduction to enable better visualization of the nasal cavity. We then proceed with the open rhinoplasty. Detailed steps of our surgical approach have been described previously.[5,6]

In brief, a V-shaped columellar incision is used; the nose is skeletonized; and all cartilaginous and bony deformities are visualized. The entire septal cartilage is first exposed by lateral reflection of the medial crurae of the lower nasal cartilages. As soon as we visualize the caudal portion of the septum, we dissect the mucoperichondrium on either side of the septal cartilage to fully expose the septum, the perpendicular plate of the ethmoid bone, the crest of the maxilla, the vomer, and the anterior nasal spine. The septal anatomy and configuration are completely visualized and managed [Figure 1]. If only the caudal portion of the septum deviates from the midline, then we free this portion from the underlying maxilla and nasal spine. One can weaken the spring of the cartilage with scoring or with a crusher as needed. The septum is secured in the midline with sutures to the nasal spine. When the deviation is more severe, we remove the appropriate parts of the septum, leaving adequate cartilage for support. In extreme situations, the septum is completely mobilized from all its connections, removed en bloc, sculptured to a flat contour, and repositioned. Cartilage grafts and strut can also be used, if needed, to maintain the contour and provide additional support. Time-honoured techniques of rhinoplasty and liberal use of cartilage grafts are used to achieve the best possible results. Symmetry of the domes is achieved either with cartilage repositioning or with augmentation using cartilage grafts. Additional tip projection, when needed, is achieved with columellar struts while collapse of the middle vault and internal valve dysfunction are corrected with spreader grafts. Osteotomies are necessary when skeletal deformities and deviations are present and composite grafts from the ear are used to correct significant lining deficiencies. The incision is then closed in a V-Y fashion to provide additional columellar length. Further treatment of nostril asymmetry may require direct excisions. Finally, if the cleft side nostril is significantly smaller than that of the non-cleft side, it may be corrected with a composite graft

Figure 1: Intraoperative view of exposure of the septum in a patient with unilateral cleft lip and palate demonstrating significant septal deviation after reflection of the medial crura of the lower alar cartilage

from the conchal bowl of the ear.

DISCUSSION

For many years, surgeons were focusing primarily on the repair of the cleft lip and deferred correction of the coexisting nasal deformity. This was done for fear that early surgical intervention and manipulation of the nasal structures might result in growth disturbances of the nose. This rationale, however, was challenged and proven not valid by several surgeons who published their long-term results demonstrating that early nasal surgery does not carry long-term deleterious effects on nasal and facial growth.[7-9] Thus, the philosophy of surgeons has dramatically changed over the past years and correction of nasal deformity is currently considered to be an intricate part of the primary cleft lip repair procedure. Nostril size and symmetry, columellar length, and tip position and projection are currently addressed during the initial surgical procedure. This approach has resulted in superior outcomes, with less residual nasal deformities and improved symmetry and facial aesthetics.[10] Several surgeons have demonstrated that early intervention and correction of the nasal deformity at the time of lip repair, with or without preoperative orthopaedic alignment of the maxilla, produces overall long-lasting aesthetic improvement without detrimental effect on facial and nasal growth.[11]

Preparation with preoperative orthopaedics and the use of the nasoalveolar moulding device (NAM), while controversial among surgeons, appears to be an important addition to our armamentarium. Since its initial description by Grayson *et al.*, the technique has gained popularity and has been incorporated in the treatment protocols of many craniofacial teams. Advantages of the technique include an improved and tension-free lip repair as well as improved and stable changes in the nasal shape, with less scar tissue and better lip and nasal form.[12] Disadvantages include the need for multiple weekly preoperative visits for device adjustment and the attendant cost-related issues. Liou *et al*. demonstrated that nasal asymmetry significantly improved with the nasoalveolar moulding and this was further corrected to symmetry following primary cheiloplasty. Nasal symmetry significantly relapsed in the first postoperative year but remained stable thereafter.[13] Our clinical observations from the use of NAM are similar. We feel that with the use of this modality, we have witnessed significantly less residual deformity and required less effort in the final correction of the deformity. Further prospective studies with comparison of groups of patients undergoing management with and without the use of the nasoalveolar moulding will be necessary to answer all remaining questions on this issue.

Despite the use of these advanced techniques and modalities and the overall changes in treatment philosophy for patients presenting with unilateral cleft lip and simultaneous correction of the nasal deformity, some patients will require additional care for definitive correction of the residual unilateral cleft nasal deformity.[14] The severity of the residual nasal deformity varies, depending on the magnitude of the initial deformity and the degree, if any, of nasal correction and maxillary repositioning performed during the initial lip repair or during subsequent revisions. We rarely any more encounter patients with the “classic” residual cleft nasal deformity but rather patients who present with a variety of residual deformities and also iatrogenic deformities from previous interventions. As such, we would like to again stress that secondary procedures for patients with residual unilateral cleft lip nasal deformities should be individualized and based on the specific patient’s individual needs.

A large number of surgical techniques and modifications have been described for the definitive correction of the residual nasal deformity.[15] These techniques are used in combination with other time-honoured techniques of rhinoplasty to correct residual deformities, achieving symmetry and aesthetic balance of the nose within the rest of the face. Beautiful and aesthetically pleasing long-term results have been reported. Some authors have even attempted to objectively evaluate and validate their results through computer analysis of photographs and panels. Thus, the success of surgical techniques can be further validated and technical flaws identified and corrected, as indicated.[11,16,17]

Unfortunately, less attention has been paid in the literature to the correction of coexisting airway obstruction with definitive rhinoplasty. Several authors have recognized the need for simultaneous septoplasty or turbinate reduction at the time of correction of the nasal deformity, but very little attention has been given to the functional outcome of the reconstructive procedures and, hence, very little such information has been reported.[18] Yet, it is well known that patients with facial clefts have a variable

Figure 2A: 16-year-old man with residual unilateral cleft lip nasal deformity and airway obstruction

Figure 2B: Base view demonstrating significant septal deviation causing airway obstruction

Figure 3A: Two years postoperative- anteroposterior view

Figure 3B: Two years postoperative- base view

Figure 3C: Two years postoperative- lateral view

Figure 3D: Two years postoperative- lateral view

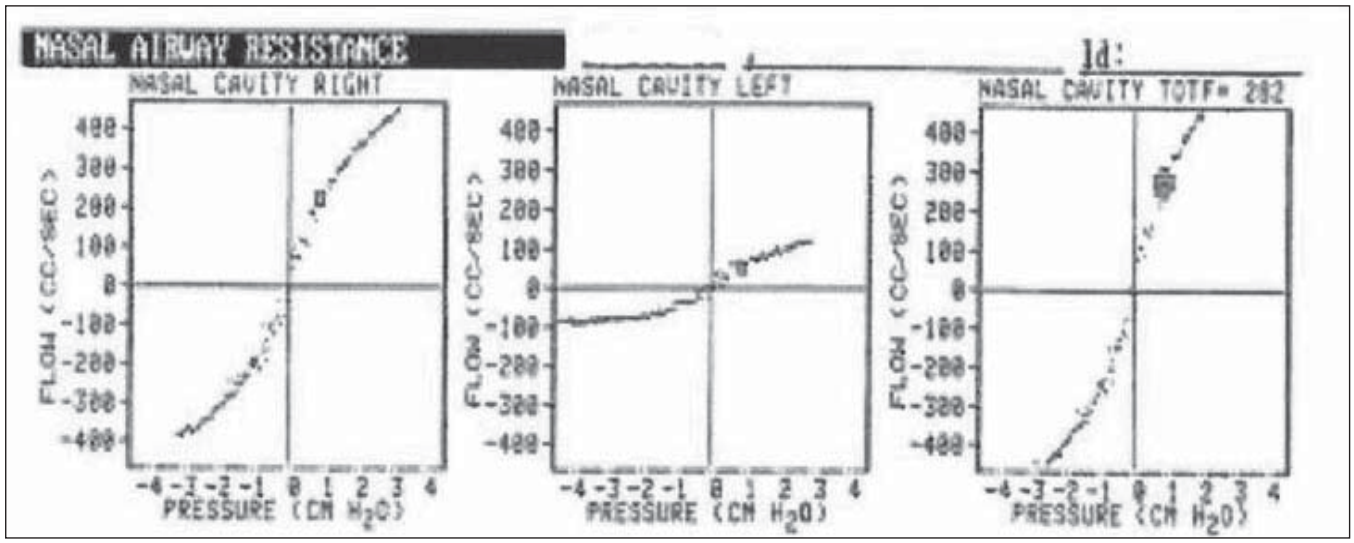


Figure 3E: Preoperative pressure flow plot demonstrating significant airway obstruction on the left side

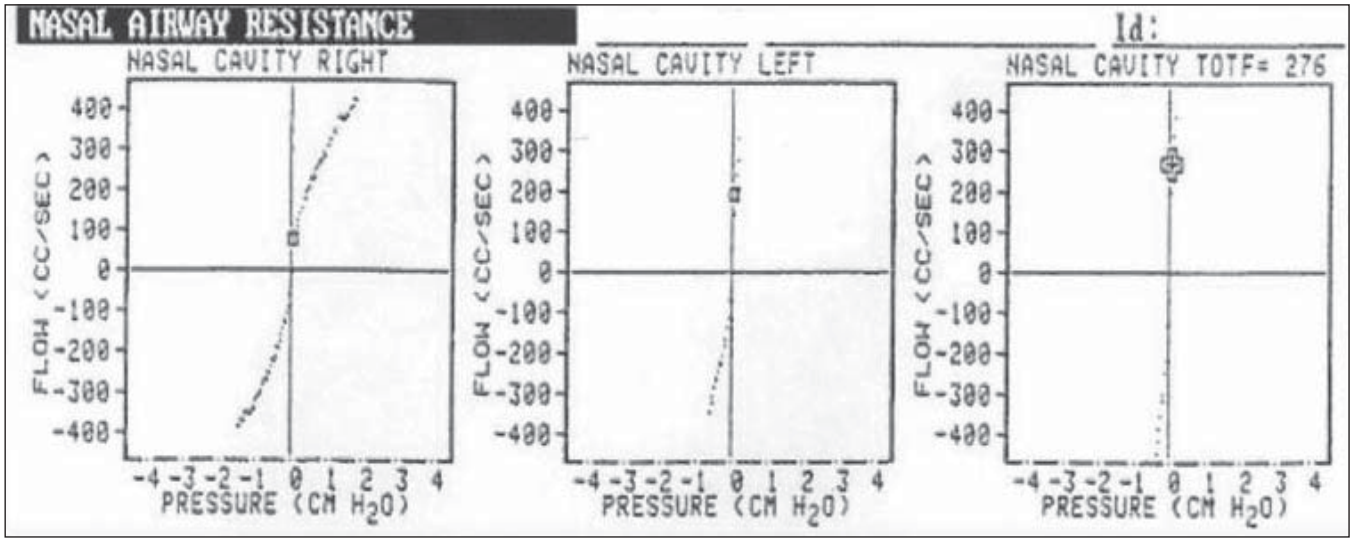


Figure 3F: Postoperative pressure flow plot demonstrating postoperative improvement of left side and total airway resistance

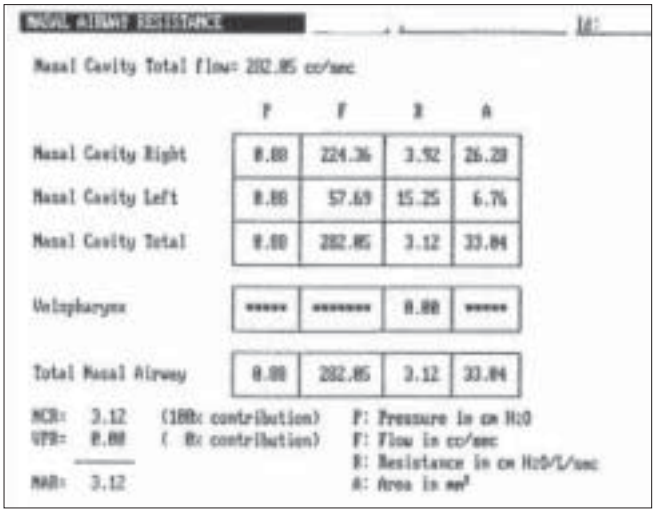


Figure 3G: Numerical representation of preoperative airway area, flow, and resistance demonstrating high resistance and decreased area on the left

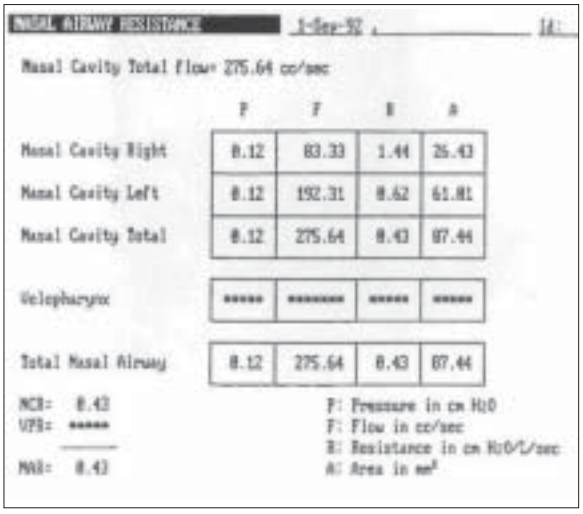


Figure 3H: Numerical representation of postoperative airway area, flow, and resistance demonstrating significant improvement in flow and area on the left, increase in area of total airway, and reduction in resistance on the left

Figure 4A: 18-year-old woman with residual unilateral cleft lip nasal deformity and airway obstruction

Figure 4B: Postoperative view- anteroposterior, after septoplasty, bilateral inferior turbinate reduction, and spreader grafts

Figure 4C: Postoperative views- lateral, after septoplasty, bilateral inferior turbinate reduction, and spreader grafts

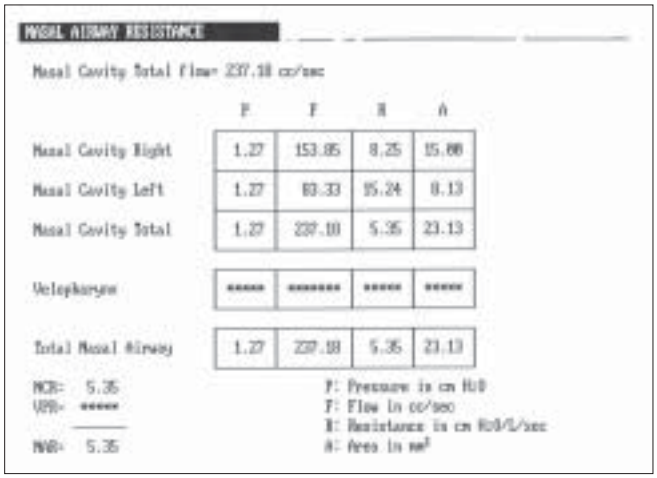


Figure 4D: Numerical representation of preoperative airway area, flow, and resistance demonstrating increased resistance and decreased area on the left

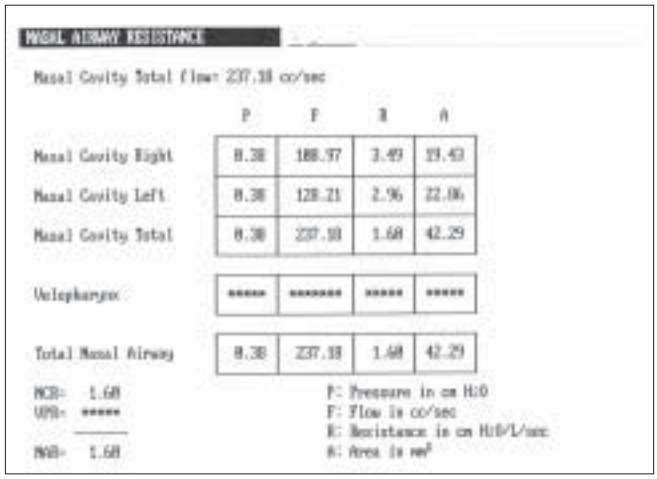


Figure 4E: Numerical representation of postoperative airway area, flow, and resistance demonstrating modest functional improvement in left airway resistance, total airway resistance, and left area

Figure 5A: 21-year-old woman presenting with residual cleft nasal deformity after multiple previous surgeries at another institution

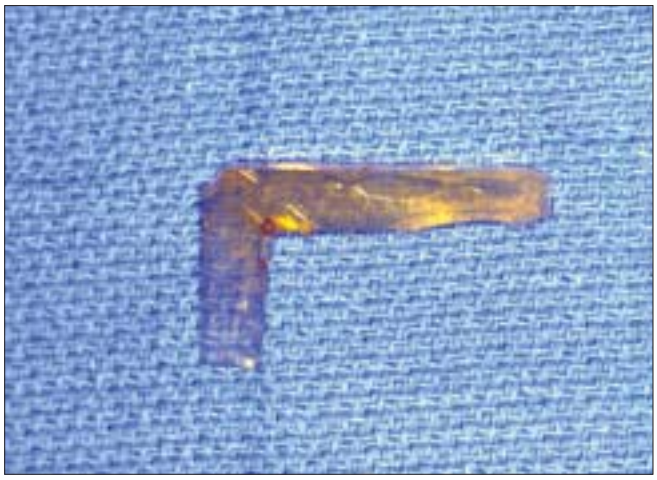
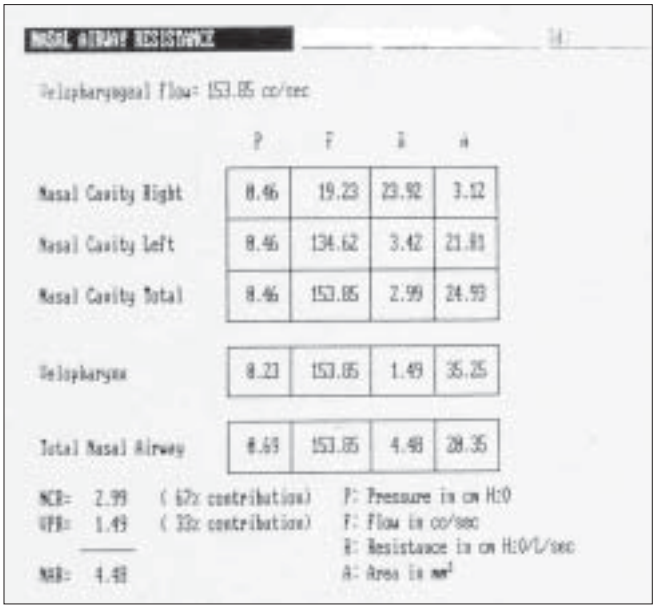


Figure 5B: Silicone strut that was eroding through the nose

Figure 5C: Postoperative view- anteroposterior

Figure 5D: Postoperative views- lateral



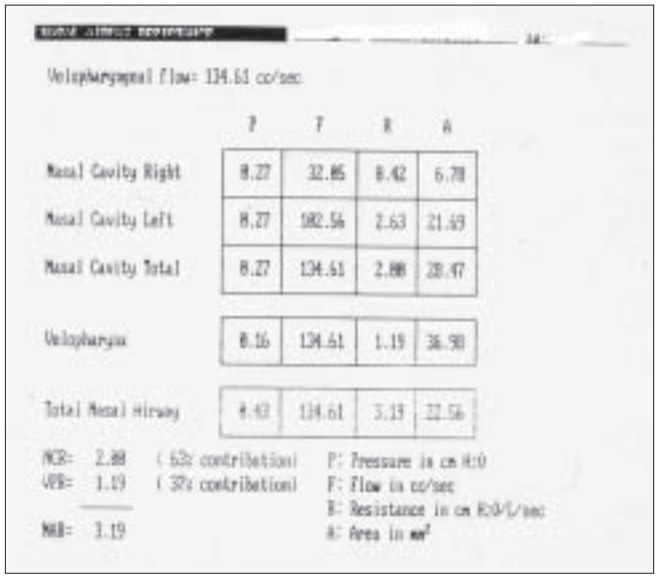
Velopharyngeal flow= 153.85 cc/sec

	P	F	R	A
Nasal Cavity Right	0.46	19.23	23.92	3.12
Nasal Cavity Left	0.46	134.62	3.42	21.81
Nasal Cavity Total	0.46	153.85	2.99	24.93
Velopharynx	0.23	153.85	1.49	35.25
Total Nasal Airway	0.69	153.85	4.48	28.35

NCR= 2.99 (67% contribution)
VPR= 1.49 (33% contribution)
NAR= 4.48

P: Pressure in cm H2O
F: Flow in cc/sec
R: Resistance in cm H2O/L/sec
A: Area in cm^2

Figure 5E: Numerical representation of preoperative airway area, flow, and resistance



Velopharyngeal flow= 134.61 cc/sec

	P	F	R	A
Nasal Cavity Right	0.27	32.85	8.42	6.78
Nasal Cavity Left	0.27	102.56	2.63	21.69
Nasal Cavity Total	0.27	134.61	2.00	28.47
Velopharynx	0.16	134.61	1.19	36.50
Total Nasal Airway	0.43	134.61	3.13	22.56

NCR= 2.00 (63% contribution)
VPR= 1.19 (37% contribution)
NAR= 3.19

P: Pressure in cm H2O
F: Flow in cc/sec
R: Resistance in cm H2O/L/sec
A: Area in cm^2

Figure 5F: Numerical representation of postoperative airway area, flow, and resistance demonstrating no functional improvement in total airway resistance or area

degree of airway obstruction due to reduced size of the airway, caused primarily from septal deviation, inferior turbinate hypertrophy and deficiency of the maxillary growth.[19] In addition, patients with clefts might have undergone pharyngeal flaps or pharyngoplasties, which further alter the nasopharyngeal space, increasing nasal resistance through decreasing the size of the airway. This obstruction can cause many symptoms including, but not limited to, nasal stuffiness, postnasal secretion, nasal bleeding, infections, loss of olfactory acuity, sinus conditions, headaches, pain in the distribution of the ophthalmic branch of the trigeminal nerve, snoring, and

sleep apnoea. Nasal obstruction can also cause systemic effects from increased pulmonary resistance with reduction of the forced expiratory volume and alteration of the flow volume loop.[3] Sleep apnoea can also result from airway obstruction.[20] This is a serious condition that can lead to enuresis, daily hypersomnolence, and, eventually, cor pulmonale.

Nasal airway obstruction in patients with clefts might also affect the growth of the maxillofacial skeleton and result in various deformities, including Class II malocclusion, open bite, a retrognathic mandible, and proclined upper incisors.[21]

We believe that management of the airway should be given equal importance during the corrective nasal surgery because of the significance of airway obstruction to the patient's wellbeing. Our multidisciplinary protocol of care was designed in an effort to achieve not only aesthetic but functional habilitation. In order to achieve that, we had to be able to gather appropriate subjective and objective information about the status of the nasal airway of a given patient, evaluate all contributing factors, and treat them accordingly. In addition to the extensive clinical evaluation and subjective findings revealed during the patient's interview, we rely on physiologic evaluation with rhinomanometry and pressure flow technique to measure airway resistance and cross-sectional breathing in an objective way.[22] For comprehensive assessment of the nasal respiration we use the component modification approach described by Smith *et al*.[23] This technique partitions the airway into its nasal cavities and velopharyngeal components, providing resistances and cross-sectional areas for the left and right nasal cavities, total nasal cavity, velopharynx, and total nasal airway(total nasal cavity + velopharynx). Computer plots are then generated and demonstrate air flow and resistance for each side along with the contribution of the total nasal cavity and velopharynx to the total nasal airway resistance. Postoperative testing and re-evaluation provides us with subjective and, above all, objective information about the outcome of our intervention. We find this feedback very important not only because we are able to objectively validate our results, but because we are also able to critically evaluate our techniques and adjust or improve as needed.

We continue to evaluate our long-term functional and aesthetic results. We have treated to date 219 patients. The most recent analysis of 51 consecutive patients who underwent complete clinical and functional evaluation with rhinomanometry at a follow-up ranging from 5 to 11 years revealed the following:

- Thirty-nine patients (76.47%) had significant functional and aesthetic improvement [Figures 2 and 3]
- Seven patients (13.07%) had significant aesthetic improvement but a modest functional improvement [Figure 4] and
- Five patients (9.8%) required additional surgery to improve their appearance; they had no functional improvement [Figure 5].

Further analysis of our results demonstrated that five out of seven patients in the second group had pharyngeal flaps in place that were primarily responsible for the airway obstruction. No attempt was made to revise the ports of these flaps because the speech of all these patients was judged as excellent and any revision might have compromised speech. Each patient in the last group had undergone several procedures prior to the final reconstruction, which resulted in significant intranasal scarring and both mucosal and cartilage deficiency. These findings remain in line with our previous analysis six years ago.[3]

Our current project employs the addition of three-dimensional photography in our pre- and postoperative evaluation. Thus, we will be able to present objective aesthetic data in addition to functional data.

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