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Long-Term Impacts of Cleft Lip and Palate: Challenges and Management

Omar S. Fattel-Servin[1], Valentina Prieto-Vargas[2], Lino E. Ramirez-Sosa[1], Jorge Flores-Orduña[3], Gabriela A. Seltrao-Migon[2] and Rogelio Martinez-Wagner[1]

[1]Department of Plastic and Reconstructive Surgery, Dr. Manuel Gea González General Hospital, México City, Mexico
[2]Plastic and Reconstructive Facial Surgery, Houston, USA
[3]Tecnologico de Monterrey, School of Medicine and Health Sciences. Monterrey, N.L. Mexico

Corresponding author: Omar S Fattel-Servin, Department of Plastic and Reconstructive Surgery, Dr. Manuel Gea González General Hospital, México City, Mexico

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Abstract

Background: From an early age, patients with cleft lip and palate undergo numerous procedures and reinterventions, facing a variety of sequelae and complications that impact both their present and future, significantly affecting their lifestyle and that of their families. Analyzing the sequelae of cleft lip and palate requires prior knowledge of the anatomy of the nose, lip, and palate, the types of deformities specific to each structure, and the procedures indicated for each type.

Methods: A bibliographic review of articles published between 2010 and 2023 was conducted with the primary objective of describing these various sequelae, addressing their specific anatomical aspects, and proposing both invasive and non-invasive strategies for their management from a reconstructive, functional, and aesthetic perspective.

Results: The bibliographic review reveals different sequelae in crucial areas such as nutrition and feeding, surgical aspects, orthognathic issues, speech and language, otorhinolaryngological concerns, and psychosocial aspects. Each of these areas shows complications that could be prevented with early interventions and multidisciplinary therapeutic approaches that primarily address the current quality of life of the patient, taking into account the future repercussions of each type of procedure and how to prevent or treat them.

Conclusions: Effective management of cleft lip and palate requires a comprehensive, multidisciplinary approach. From surgical interventions addressing functional and aesthetic concerns to ongoing speech therapy and psychological support, each aspect plays a crucial role in enhancing outcomes and quality of life for patients. Early intervention and continuous evaluation are key to mitigating long-term complications and maximizing the overall success of treatment.

Keywords

Sequelae; Lip; Nose; Palate; Cheiloplasty; Palatoplasty; Lipoinjection; Plastic surgery.

Introduction

Cleft lip and palate constitute one of the greatest challenges for the plastic surgeon due to their complex and multidisciplinary management. From an early age, patients face numerous difficulties during feeding, growth, and development. This is compounded by the large number of procedures and reinterventions they undergo from the moment of diagnosis, leading to a wide variety of complications and sequelae that directly impact their quality of life and that of their families. The different sequelae to be analyzed in this review article are grouped as follows:

- Nutrition and feeding
- Surgical
- Orthognathic
- Speech and language
- Otorhinolaryngological
- Psychosocial

Thus, the importance of a multidisciplinary team for proper management is considered crucial.

Sequelae in nutrition and growth

Difficulty in proper feeding is closely related to the size and severity of the cleft, affecting the ability to create an adequate vacuum during suction. According to [1] with proper feeding technique and positioning from an early stage, this difficulty can improve over time. The same article reports that in patients with cleft palate, breastfeeding has a protective effect against otitis media. Early incorporation of a feeding support program has been shown to reduce the growth delay rate in infants with cleft palate from 49% to 26%.

Surgical sequelae

Lip

At this level, sequelae mainly derive from facial growth disturbances, poor wound healing, deficiencies in surgical technique, or poor adherence of family members to postoperative care during primary repair. It is important to perform a thorough analysis of nasal structures (dorsum, tip, ala, base, and columella) as well as the lip, evaluating the symmetry and position of the philtrum, Cupid's bow, vermilion, and mucosae.

Primary surgery is crucial; if the technique is inadequate, it can result in aesthetic and functional stigmata or sequelae. [2] classify lip sequelae into superficial and muscular. When both components are compromised, a complete revision is required to improve the scar and realign the muscle.

Surgically, five groups can be listed:

1. Complete lip revision including muscular realignment.
2. Local flaps: Z-plasty, V-Y or Y-V plasty, transposition flaps, Kapetansky flap.
3. Regional flap: tongue, Abbe flap, cross-lip vermilion flap, island flap from the lower lip.
4. Autografts, dermal substitutes, alloplastic materials.
5. Combination of procedures.

Superficial sequelae

Previous studies by [3] show that this type of sequelae can present as hypertrophic, wide, depressed, contracted, or pigmented scars; and describe the use of fractional CO2 laser or pulsed light laser as minimally invasive measures for their treatment. Similarly, surgical procedures such as lipoinjection with micro and nano grafts, resection and direct closure, oval, diamond, Z, or W plasty can be employed, the latter also useful for small defects in vermilion alignment. According to previous studies by [4] excess mucosa can be corrected with Y-V or transverse wedge resection.

Muscular sequelae

The orbicularis muscle provides mobility and volume to the lip. According to [5] poor repairs are associated with various distortions including philtral deformities and whistle deformity. Misalignment causes bulging on both sides of the scar as well as notable deformity, requiring revision through a full-thickness incision. The orbicularis muscle is isolated from the overlying skin/vermilion and the underlying mucosa, and abnormal attachments to the maxilla are released if necessary, allowing for repair and proper transverse anatomical repositioning. Surgical techniques vary depending on whether the primary repair was unilateral or bilateral.

Vermilion

Whistle deformity can be defined as a deficiency of tissue in the central tubercle, resulting from factors such as the inability to restore muscular continuity at the vermilion level, mucosal and vermilion continuity, muscle dehiscence, or severe postoperative scars. According to [6] treatments depending on severity, ranging from superficial corrections to more complex measures like the Abbe flap. The Abbe flap involves rotating a pedicled flap from the lower lip, dependent on the labial artery, to cover a central defect in the upper lip. After 2 to 3 weeks, the pedicle is divided, and the labial artery is ligated. During its design and transfer to the recipient site, it is crucial to align the muscle fibers before external closure in the vermilion, thus offering a full-thickness reconstruction of the upper lip’s central aesthetic unit, replacing deficient and scarred tissue and restoring functional oral sphincter capacity.

Philtrum

Primary cheiloplasty involves the end-to-end approximation of the orbicularis oris muscle, often resulting in a flat lip appearance. When treating this sequela, appropriate management generally involves adding volume to the philtral crest as indicated by [7]. This can be achieved by resecting the scar contributing to the low prominence of the crest associated with muscle eversion sutures at the philtral crest level. A less invasive strategy consists of adding volume directly to the philtral crest using lipografts or hyaluronic acid.

Nose

Nasal deformity in cleft lip and palate represents a significant challenge, affecting all nasal components (skin, cartilage, mucosae, and skeletal framework). To achieve a satisfactory result, it is crucial first to understand the anatomical characteristics of the nasal deformity, as described by [8] in unilateral and bilateral clefts.

Unilateral Cleft: This type shows the dorsum deviated towards the healthy side, the nasal tip identified by a shorter medial cru on the cleft side, a longer lateral crus on the cleft side, and a flattened and laterally displaced alar dome on the cleft side. The columella is shorter on the cleft side, with its base displaced towards the healthy side. The nasal fossa appears horizontalized on the cleft side, with the alar base displaced laterally, posteriorly, and inferiorly, and the caudal portion of the septum deflected towards the healthy side and its cephalic portion deviated towards the cleft side.

Bilateral Cleft: This type shows the dorsum central and wide, both medial crura short, lateral crura elongated, and domes without definition or projection. The columella is short and has a wide base. The nasal fossa is bilaterally horizontalized with a wide alar base, displaced laterally, posteriorly, and inferiorly, and the septum is central with deviation towards the more affected side.

Depending on the timing of the intervention, repairs can be classified as described by [9] into primary, intermediate, and definitive repairs.

Primary rhinoplasty: This is any correction performed at the time of primary cheiloplasty. In unilateral cases, it aims to reposition the lower lateral cartilages and achieve columellar symmetry. In bilateral cases, it seeks to elongate the columella and narrow the alar bases.

Intermediate rhinoplasty: These procedures are performed before completing nasal growth to correct the deformity and minimize psychological pressure. This is preferable between 8 and 12 years, after completing orthodontic alignment and/or alveolar bone grafting, providing a stable skeletal base for correcting severe nasal deformities. Intermediate rhinoplasty is more conservative than definitive rhinoplasty techniques.

Definitive Rhinoplasty: This should be the final step in the process, also called secondary rhinoplasty, performed after completing maxillary and nasal growth. It is important to note that nasal cartilages have a memory and tend to revert to their original state. Therefore, a complete dissection, the use of strong grafts, and well-planned skin coverage are essential. Definitive rhinoplasty is the most difficult procedure in cleft lip and palate due to significant secondary distortion of cartilages. These distortions may necessitate using cartilage grafts from other body regions such as the ear or ribs.

Definitive rhinoplasty is typically performed after orthognathic surgery because maxillary advancement improves many of the characteristics of nasal support. Facial skeletal growth is generally complete between 14 and 16 years for girls and between 16 and 18 years for boys.

Rhinoplasty Approach

While closed technique can be used, open rhinoplasty is preferred according to [10] as it allows maximum visualization of structures and better exposure for graft placement and suturing. Proper manipulation of the bony pyramid and septum, as well as cartilage graft placement for nasal support and reinforcement, is crucial. Measures to achieve these objectives include:

1. Procedure to Improve Airflow: Reduction of the inferior turbinate, with resection/repositioning of the septum.
2. Correction of the Bony Vault: Osteotomies in the nasal bone.
3. Manipulation of the Lower Third: Centralization of the septum, septal extension grafts, V-Y chondromucosal advancement, vestibular web corrections, repositioning of the alar base and nasal fossae.

Cartilage grafts are useful for maintaining, reconstructing, shaping, and reinforcing the structural framework. These grafts include:

- **Dorsal grafts:** To improve the position and height of the dorsum or as spreaders to increase the angle of the internal valve.
- **Caudal septal extender:** For structural support.
- **Tip grafts:** To reinforce the medial and middle crura, essential for providing additional support to the alar cartilage and better positioning the nasal tip against postoperative scar tissue forces.
- **Crural Grafts:** To shape the lateral crura and support the vestibule. They are inserted beneath the lateral crura to reinforce the ala and correct its collapse, thus opening the external valve.

Reconstruction of the main tip support mechanisms to improve tip projection and correct lower lateral cartilage deformity generally requires rib cartilage, either autologous or cadaveric. Auricular and septal cartilage often lack sufficient strength for adequate correction and resistance to relapse. Septal extension grafts and/or lateral crural extension grafts are commonly made from rib cartilage due to septal cartilage deficiency.

Rhinoplasty Complications

According to [11], the most common long-term complication is residual asymmetry and poor tip position due to soft tissue retraction. Septal deviation can also recur or remain as a residual issue caused by the initial deformity's severity. Differences in the size of the nasal fossae and residual deformity apparent in the basal view are common. Clear communication with patients about these potential complications is crucial to avoid unrealistic expectations.

Palate

Oro-nasal fistulas and velopharyngeal insufficiency are the most common sequelae requiring secondary surgeries in cleft palate patients.

Fistulas

According to [12], the incidence of postoperative palatal fistula varies from 0.5% to 58%, with a recurrence rate of 33% to 37%. Small fistulas, especially at the junction of the hard and soft palate (Pittsburgh Type III), followed by those at the premaxillary-maxillary junction (Type V), are more common due to high tension areas during primary repair.

No universal classification for palatal fistulas exists, but the Pittsburgh system (2007) is one of the most widely used today. This system provides an anatomical description of the oro-nasal communication, suggesting a standardized scheme that includes seven types based on location:

- Small: 1 to 2 mm
- Medium: 3 to 5 mm
- Large: > 5 mm

Functional: Symptomatic, nasal escape, speech alterations, regurgitation, velopharyngeal insufficiency.

Non-functional: Asymptomatic, clinically insignificant.

Fistula treatment

[13] highlight that early postoperative fistulas can be managed conservatively and may close without intervention. Asymptomatic fistulas should also be treated conservatively as most small fistulas close spontaneously. Functional fistulas, regardless of size, can impair speech and progress to velopharyngeal insufficiency, making treatment essential.

Management depends on the location and size of the fistula. Isolated soft palate fistulas can often be repaired directly, frequently along with intravelar veloplasty or Furlow palatoplasty if velopharyngeal dysfunction is present. Larger hard palate fistulas may require more than direct excision and closure due to a lack of pliable local tissue. Well-vascularized tissue from other donor sites, such as tongue flaps or pedicled flaps like the facial artery myomucosal flap (FAMM) and even the Bichat bag flap, are preferred for covering large palatal fistulas.

[14], propose the addition of acellular dermal matrix to enhance nasal closure, especially in patients with local or regional tissue deficiency. For complex cases with large fistulas or failed previous procedures, free flaps, particularly the radial forearm flap, are considered.

Velopharyngeal Insufficiency (VPI)

Velopharyngeal function depends on the muscular structures of the soft palate, lateral pharyngeal wall, and posterior pharyngeal wall, which act together as a muscular valve creating a seal between the velum and the posterior pharyngeal wall during swallowing and speech. Failure of this mechanism is known as VPI, often due to structural anomalies resulting in inadequate closure of the velopharyngeal sphincter. This condition is common in patients with cleft palate and submucous cleft palate, with persistence in 20-50% post-repair due to inadequate palatal length or poor muscle repair.

Hypernasality and compensatory sounds, where patients substitute normal sounds for abnormal ones for better speech intelligibility, are characteristic of VPI. Diagnosis involves speech evaluation by an expert in

phoniatrics and language therapy, including a perceptual speech evaluation to assess articulation errors, compensatory articulations, abnormal resonance, nasal emission, and dysphonia.

VPI treatment

Treatment options for VPI include speech therapy, prosthetics, and surgery. The choice of treatment should be individualized based on patient history and clinical findings. The three most common surgical procedures are posterior pharyngeal flap, double-opposing Z-plasty Furlow palatoplasty, and dynamic sphincter pharyngoplasty. Understanding the patient's velopharyngeal closure pattern and the size of the velopharyngeal space is essential for selecting the appropriate procedure. The different management approaches described by Glade & Deal are based on closure patterns.

Closure Patterns

Three main velopharyngeal closure patterns, defined by the primary contribution of each structure to the closure, can be evaluated by nasal endoscopy or multi view video fluoroscopy:

- **Coronal Pattern:** Typical closure with the velum contacting the posterior pharyngeal wall in an anteroposterior manner with minimal or no lateral pharyngeal wall contribution (55% prevalence).
- **Sagittal Pattern:** Lateral pharyngeal walls move toward the midline.
- **Circular Pattern:** Combination of coronal and sagittal patterns with velar posterior movement and medial movement of lateral pharyngeal walls.

Closure Pattern	Procedure
Sagittal	Gap < 9 mm: Furlow / > 9 mm: Posterior pharyngeal flap
Coronal	Dynamic sphincter pharyngoplasty
Circular	Gap < 9 mm: Furlow / > 9 mm: Posterior pharyngeal flap

Table 1: Define the primary contribution of each structure.

Posterior pharyngeal flap

This involves creating a tissue bridge from the soft palate to the pharynx using a superiorly pedicled posterior pharyngeal myomucosal flap. The donor site is closed or left to heal by secondary intention if wide. Adequate lateral pharyngeal wall movement (sagittal or circular pattern) is required for proper function. Preoperative tonsillectomy and adenoidectomy are suggested to avoid airway obstruction, reported in up to 65% of cases.

Furlow palatoplasty

Not indicated for gaps > 9 mm, Furlow palatoplasty is suitable for poor lateral wall closure but good velar mobility. Posterior-based myomucosal flap transposition reorients the levator muscles from a sagittal to a horizontal position, reconstructing the palatal elevator sling.

Dynamic sphincter pharyngoplasty

A dynamic technique using superiorly based myomucosal flaps from the posterior pillar of the soft palate (palatopharyngeus muscle). Vertical incisions anterior to the posterior tonsillar pillars expose the palatopharyngeus muscles. Vertical posterior incisions create flaps about 1 cm wide, which are crossed, overlapped at the midline, and inserted into a transverse incision in the soft palate. Reported effectiveness is 80-85%.

Posterior pharyngeal lipoinjection

For small defects with adequate velar dynamics, volume augmentation of the posterior pharyngeal wall using alloplasts or cartilage grafts has been described. However, lipoinjections are currently most used due to their adequate integration and easy acquisition.

Lipoinjection aims to reduce the velopharyngeal gap, generally achieved with 20 cc of volume, facilitating velar approximation to the posterior pharyngeal wall. The procedure is performed submucosally in the midline and paramedian areas, avoiding excessive injection to prevent airway obstruction.

Non-Surgical Management of VPI

The non-surgical management of velopharyngeal insufficiency (VPI) includes support from a speech therapist, which must be individualized and closely monitored to assess language progression. Additionally, prosthetics are included to occlude areas with tissue deficits in patients who are not candidates for surgery or who have had previous failed procedures.

Orthognathic sequelae

Patients with cleft lip and palate (CLP) require multiple interventions that can potentially inhibit maxillary growth through the disruption of facial growth centers and postoperative scarring of soft tissues. This frequently leads to transverse maxillary hypoplasia. Additionally, the alveolar segments may collapse unilaterally or bilaterally, causing narrowing or alteration of the maxillary arch. Since the mandible continues to grow, the dentofacial relationship is further altered, usually resulting in a Class III malocclusion that accentuates midfacial retrusion.

According to [15], the suggested surgical treatment is maxillary advancement via Le Fort I osteotomy, which is necessary in 48% of unilateral clefts and 65% of bilateral clefts. Alternatively, [16]. suggest maxillary distraction osteogenesis, which can be initiated at younger ages compared to Le Fort I osteotomy, without the need to wait for skeletal maturity. Severe maxillary hypoplasia present at early ages can be corrected with distraction, taking into account the normal growth of the mandible.

Speech sequelae

Due to the dysfunction of the palatal muscles involved in phonation, speech sequelae are observed. Abnormal nasal resonance and articulation difficulties are common characteristics in the majority of patients with cleft lip and palate. Recent findings by [17] highlight the importance of speech therapy to improve speech and avoid compensatory articulations, which are independent of surgical repair and require early stimulation and re-education to counteract them.

Otorhinolaryngological sequelae

Numerous studies by Goudy et al. have demonstrated the relationship between cleft palate and anomalies in Eustachian tube function, leading to recurrent otitis media in up to 95% of patients. This is related to chronic obstruction and decreased angulation of the Eustachian tube, which favors its occurrence. The main complication is hearing loss, with an estimated 20-30% of cleft palate patients presenting some degree of hearing impairment.

Despite some controversies, it is considered that palatoplasty reduces the risk of chronic serous otitis media and therefore hearing loss. However, serous otitis persists in most cleft patients even years after palatoplasty. In persistent or recurrent cases, myringotomy with tube placement remains the cornerstone of treatment for this issue, necessitating joint management with otorhinolaryngologists.

Psychosocial sequelae

Alterations in social relationships affect the rehabilitation and overall development of patients with CLP. Psychological issues such as low self-esteem and social integration problems have been reported. Data from [18] reveal the prevalence of depression, which is present in 2% of patients during childhood and reaches 15-20% during adolescence and adulthood. Another common disorder among these patients is anxiety, present in up to 50% during adolescence.

Therefore, it is crucial to provide adequate psychological support to patients from an early age to promote psychosocial development and maximize the positive outcomes of surgery and rehabilitation. The adaptation of a child with CLP depends partly on the acceptance and support environment provided by family and close relations; thus, it is necessary to educate and raise awareness among the family and people around the patient.

Conclusion

Effective management of cleft lip and palate (CLP) requires a comprehensive, multidisciplinary approach. This involves collaboration among surgeons, speech therapists, otolaryngologists, orthodontists, psychologists, and other healthcare professionals to address the wide range of challenges faced by these patients. The intricate interplay of surgical interventions, speech therapy, and psychosocial support is essential to enhance functional outcomes and improve quality of life. Early intervention and continuous support throughout the patient's developmental stages are critical for achieving optimal results. By addressing the anatomical, functional, and emotional aspects of CLP, a holistic treatment plan can ensure better rehabilitation and integration for these patients, ultimately leading to successful long-term outcomes.

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