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Comprehensive review and management of cleft lip and palate

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Abstract
Cleft lip and palate (CLP) are among the most common congenital craniofacial anomalies, with a complex etiology involving genetic, epigenetic, and environmental factors. These anomalies result from a failure of normal facial fusion during embryogenesis and can significantly affect feeding, speech, hearing, dental development, facial growth, and psychosocial well-being. This detailed review integrates current evidence to provide an in-depth analysis of the embryology, etiology, epidemiology, clinical features, multidisciplinary management, and long-term outcomes associated with CLP. Despite advances in surgical techniques and team-based care, lifelong challenges persist, underscoring the need for comprehensive follow-up and ongoing research to optimize outcomes and address disparities in care.

Keywords: Cleft lip and palate, craniofacial anomalies, multidisciplinary management

Introduction

Cleft lip and palate (CLP) are among the most prevalent congenital malformations, affecting approximately 1 in 500 to 1,000 live births globally [1]. These anomalies involve a failure of fusion between the medial nasal, lateral nasal, and maxillary processes during early embryonic development [9, 11]. CLP can present as isolated cleft lip (CL), isolated cleft palate (CP), or combined cleft lip and palate (CLP), with wide variation in severity and functional implications [2, 6]. The pathogenesis of CLP is multifactorial, involving both genetic and environmental factors. Over 300 syndromic forms of CLP have been described, highlighting the role of genetic predisposition [6]. Environmental influences such as maternal smoking, alcohol consumption, folic acid deficiency, and certain medications (e.g., anticonvulsants) further modulate the risk [3, 4, 10]. Understanding these complex interactions is crucial for effective prevention and management.

CLP has lifelong implications, including feeding difficulties, speech and hearing problems, dental anomalies, facial growth disturbances, and psychosocial challenges [3,5,7]. Multidisciplinary care, incorporating surgical, dental, speech, audiological, and psychological expertise, is essential for optimizing outcomes and enhancing the quality of life of affected individuals [2, 5, 6]

Epidemiology

The incidence appears high among Asians (0.82 – 4.04 per live births) intermediate in Caucasian (0.9 – 2.69 per 1000 live birth) and low in Africans (0.18 – 1.67 per 1000 live births). Chinese showed 1.76 per 1000 live birth, while Japanese reported 0.85 to 2.68 per 1000 live birth of orofacial clefting.

Overall incidence of cleft lip and palate is approximately 1 in 600 to 800 live births (1.42 in 1000) and isolated cleft palate occurs approximately in 1 in 2000 live births. Thus, the typical distribution of cleft types are

- Cleft lip alone – 15%
- Cleft lip and palate – 45%
- Isolated cleft palate – 40%

CLCP occurs more frequent and more severe in boys than in girls. Unilateral clefts are more common than bilateral clefts with a ratio of 4:1, and for unilateral clefts, about 70% occur on the left side of the face. Cleft palate is seen more frequently in females than in males. CLP is frequently associated with other developmental abnormalities and majority of cases are presented as part of a syndrome. Syndromic clefts account for about 50% of the total cases in some reports with about 300 syndromes described. Although the percentage of cases directly linked to genetic factors is estimated to be about 40%, all clefts appear to show a familial tendency. Recent epidemiological studies [1, 7] have also highlighted disparities in access to care, with higher unmet needs and worse outcomes in low-resource settings [1]. This underscores the importance of early detection and comprehensive care.

Embryology and Pathogenesis

Facial development occurs between the 4th and 12th weeks of gestation and involves complex interactions between the neural crest-derived mesenchyme and ectodermal tissue [9, 10]. The critical fusion events include:

- **4th–7th weeks:** Fusion of the medial nasal and maxillary processes forms the upper lip and primary palate.
- **7th–12th weeks:** Fusion of the palatal shelves forms the secondary palate.

Disruption of these processes due to genetic mutations or environmental factors can result in clefting [4, 9]. Key genes implicated include IRF6 (interferon regulatory factor 6), MSX1 (muscle segment homeobox 1), and PVRL1

(poliovirus receptor-related 1) [4, 10]. IRF6 mutations, for instance, are linked to Van der Woude syndrome, the most common syndromic form of CLP (10). Environmental teratogens such as maternal smoking and alcohol can interfere with cellular signaling pathways during palatogenesis [3, 4, 10].

Recent advances in molecular genetics have also highlighted the role of epigenetic regulation in CLP pathogenesis [4]. For example, folic acid supplementation can modulate methylation pathways critical for craniofacial development, offering potential preventive strategies [4, 10].

Classification

Precise classification is essential for diagnosis, treatment planning, and outcome assessment. Several classification systems have been developed over time, each aiming to provide a comprehensive and standardized framework for clinical and research applications.

1. Cleft Lip (CL)

Cleft lip occurs when there is an interruption in the continuity of the upper lip. It is typically classified based on:

- **Extent**
- **Incomplete cleft lip:** Partial cleft that does not reach the nostril base.
- **Complete cleft lip:** Cleft extends through the entire vertical height of the lip into the nostril.
- **Laterality**
- **Unilateral cleft lip:** Occurs on one side of the lip.
- **Bilateral cleft lip:** Involves both sides of the lip.

2. Cleft Palate (CP)

Cleft palate refers to a defect in the roof of the mouth, which may involve the hard palate, soft palate, or both.

- **Extent**
- **Incomplete cleft palate:** Typically limited to the soft palate or part of the hard palate.
- **Complete cleft palate:** Involves both the hard and soft palate, extending to the incisive foramen.
- **Submucous cleft palate:** A special form where the cleft is covered by intact mucosa, often presenting with a bifid uvula, a notch in the posterior hard palate, and a translucent zone (zona pellucida).

3. Cleft Lip and Palate (CLP)

This involves a combination of cleft lip and cleft palate. It is further classified as

- **Unilateral cleft lip and palate:** The cleft affects one side of the lip, alveolus, and palate.
- **Bilateral cleft lip and palate:** Involves both sides of the lip, alveolus, and palate.

4. Veau's Classification (for Cleft Palate)

Veau's classification is widely used for describing the severity of palatal involvement:

- **Group I:** Soft palate only.
- **Group II:** Hard and soft palate.
- **Group III:** Unilateral cleft lip and palate, involving the soft palate, hard palate, alveolus, and lip on one side.
- **Group IV:** Bilateral cleft lip and palate, involving the soft palate, hard palate, alveolus, and lip on both sides.

5. Kernahan's Stripped Y Classification

Kernahan introduced the stripped Y diagram as a symbolic and practical representation of cleft anatomy. It divides the primary and secondary palates into numbered segments, providing a clear visual tool for recording the exact anatomical sites involved. (fig 1).

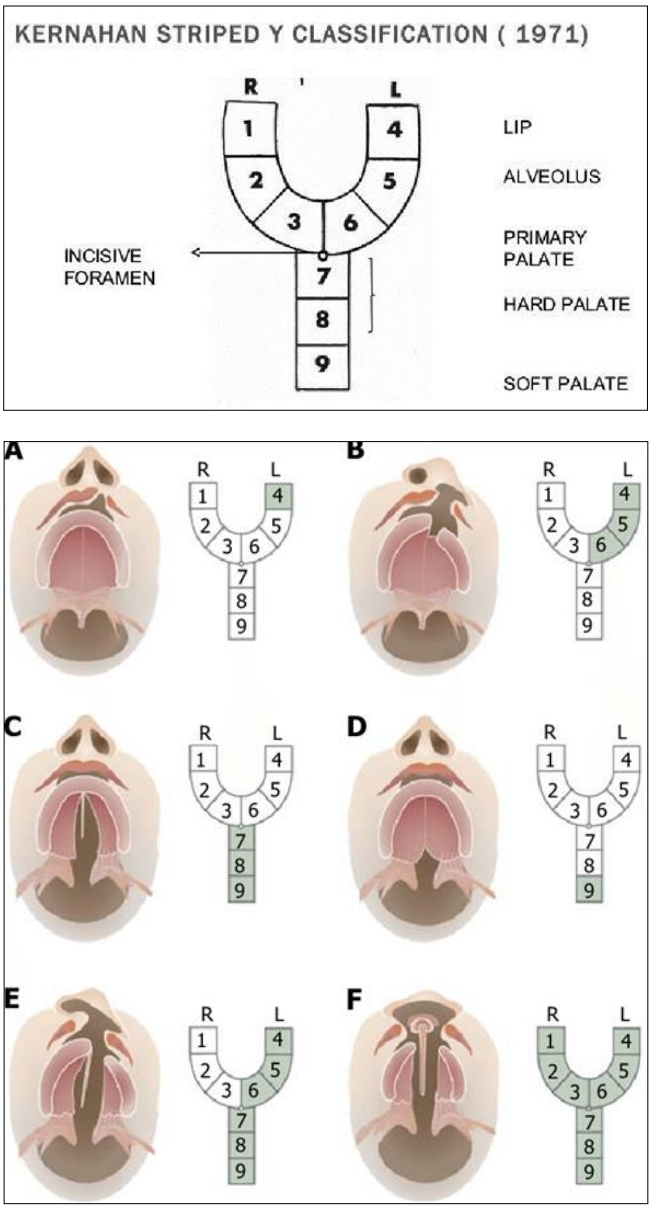


Fig 1: Kernahan's Stripped Y Classification

Etiology and Risk Factors

1. Genetic Contributions

Genetic factors play a substantial role in the pathogenesis of CLCP. Syndromic forms, accounting for approximately 30% of cases, are associated with well-characterized genetic syndromes, such as Van der Woude syndrome (linked to IRF6 mutations), Stickler syndrome (COL2A1 mutations), and Pierre Robin sequence [6]. These syndromes present with additional systemic anomalies, such as limb defects or ocular abnormalities, necessitating broader clinical management.

Non-syndromic CLCP, which accounts for the majority of cases, arises from complex polygenic interactions. Genome-wide association studies (GWAS) have identified several susceptibility loci, including IRF6, PVRL1, MSX1, and MAFB genes [1]. These genes are implicated in key processes such as craniofacial morphogenesis and cell adhesion. However, their effects are modulated by

environmental and epigenetic influences, highlighting the multifactorial nature of non-syndromic CLCP.

2. Environmental and Maternal Factors

A range of environmental factors can disrupt normal embryonic development, particularly during the critical window of facial formation (weeks 4–12 of gestation). Maternal smoking remains one of the most significant environmental risk factors, with meta-analyses indicating a 30–50% increased risk for CLP [1]. Similarly, alcohol consumption during pregnancy has teratogenic effects, impairing neural crest cell migration and disrupting palatal shelf fusion.

Nutritional deficiencies, especially folic acid, have been consistently linked to increased risk of neural tube defects and orofacial clefts [3]. Maternal folic acid supplementation reduces the risk, underscoring the importance of public health initiatives in prenatal nutrition.

Other maternal health issues, such as diabetes mellitus and obesity, further increase the risk of CLCP. Hyperglycemia and metabolic dysregulation can impact fetal development, while certain medications (e.g., anticonvulsants like phenytoin, isotretinoin, corticosteroids) have recognized teratogenic effects.

3. Socioeconomic and Ethnic Disparities

Socioeconomic status (SES) influences both the prevalence and outcomes of CL/P. Limited access to prenatal care, poor nutrition, and inadequate healthcare resources in low-SES populations can exacerbate risk and hinder timely intervention [1].

Ethnic variations in prevalence highlight both genetic predispositions and environmental exposures. Asians have the highest prevalence (1 in 500), followed by Caucasians (1 in 1000), and Africans (1 in 2500) [3]. These disparities reflect complex interactions of genetic diversity and differences in prenatal care and maternal health. (fig 2)

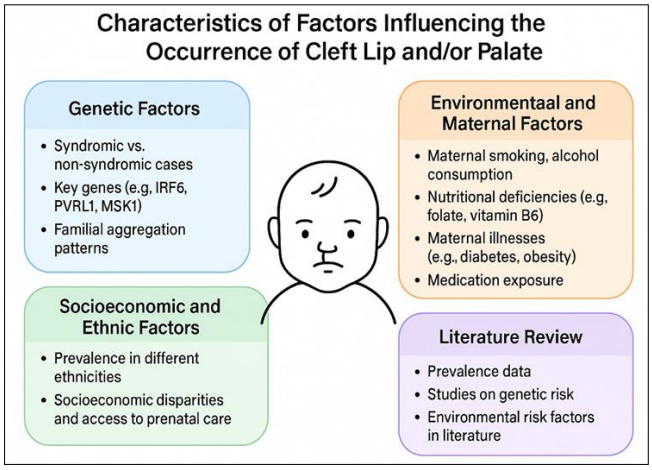


Fig 2: Etiology and Risk factors

Clinical Presentation and Functional Implications

The severity of CLP varies widely, with clinical presentations ranging from a microform notch in the lip to a complete bilateral cleft involving the lip, alveolus, and palate [6].

- **Feeding and Nutrition**

Infants with CLP often experience difficulties with breastfeeding due to poor lip seal and nasal regurgitation. Specialized feeding devices such as the Haberman feeder or cleft feeding bottles help optimize nutrition [5].

- **Speech and Hearing**

Velopharyngeal insufficiency (VPI) is a common consequence of cleft palate, leading to hypernasal speech and articulation errors [17]. Middle ear dysfunction and recurrent otitis media with effusion are prevalent due to impaired eustachian tube function.

- **Dental and Skeletal Anomalies**

Dental anomalies such as hypodontia, supernumerary teeth, enamel hypoplasia, and malocclusion are common. Alveolar clefts can affect tooth eruption and arch stability [8]. Maxillary hypoplasia may develop due to surgical scarring and growth disturbances [17].

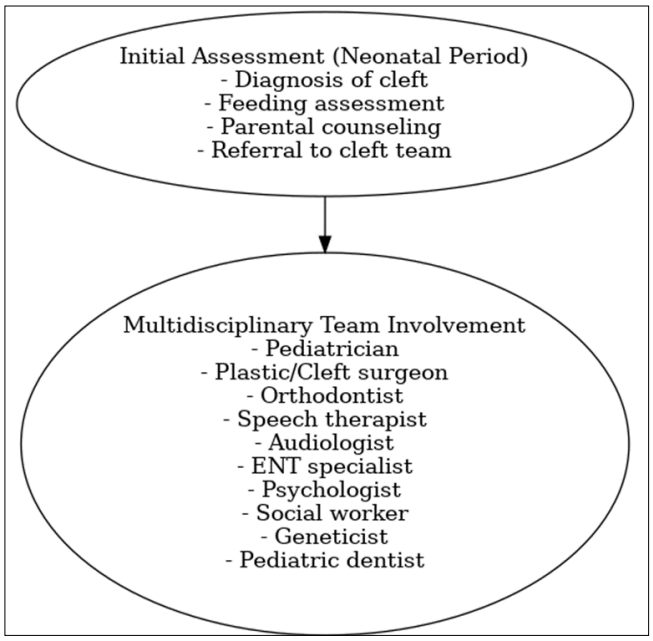
- **Psychosocial Impact**

Children with CLP often face social stigma, bullying, and low self-esteem [20]. Early psychosocial support and family-centered care are essential to mitigate these challenges [13].

Management of Cleft Lip and Palate

The management of cleft lip and palate (CLP) is comprehensive, complex, and lifelong, requiring a coordinated multidisciplinary team. The main goals of management are to restore normal function (feeding, speech, hearing), promote favorable facial growth, achieve optimal esthetics, and ensure psychosocial well-being.

The Multidisciplinary Cleft Team



Timeline of Interventions and Management Phases

a. Neonatal Phase (Birth to 3 months)

- **Feeding Support:** Infants with cleft palate cannot generate suction; specialized cleft feeding bottles (e.g., Haberman feeder) are used [5].
- **Airway Assessment:** Especially in Pierre Robin sequence, where glossoptosis and micrognathia can cause airway obstruction.
- **Parental Counseling:** Emotional support and education are essential to reduce anxiety and prepare families for the treatment journey [12].
- **Genetic Evaluation:** To rule out syndromic forms of CLP [10].

- **Pre-Surgical Orthopedics:** Nasoalveolar Molding (NAM) may be started to approximate cleft segments and improve nasal shape [8].

b. Infant Phase (3–18 months)

Surgical Phase I: Cleft Lip Repair (3–6 months)

- **Goal**: Restore lip continuity, muscular function (orbicularis oris), and nasal symmetry.
- **Techniques**

Millard Rotation-Advancement Flap
Tennison-Randall Technique
Fisher's Anatomical Subunit Technique [19].

Surgical Phase II: Cleft Palate Repair (9–18 months)

- **Goal:** Separate oral and nasal cavities, enable speech development, and minimize VPI risk.
- **Common Procedures**

von Langenbeck Palatoplasty
Furlow's Double Opposing Z-Plasty (preferred for lengthening soft palate)
Two-Flap Palatoplasty [19].

C. Toddler and Preschool Phase (18 months–5 years)

Speech and Language Intervention

- **Speech Therapy** begins as soon as speech emerges (~18 months).
- **Common Problems**: Hypernasality, nasal emission, articulation errors.
- **Velopharyngeal Insufficiency (VPI)** is assessed using
- Nasopharyngoscopy
- Videofluoroscopy

Audiological and ENT Care

- Middle ear effusion is common due to Eustachian tube dysfunction.
- **Tympanostomy tubes** (grommets) may be placed during palate repair or later to prevent hearing loss [17].

D. Mixed Dentition Phase (6–12 years)

Orthodontic and Dental Management

- **Interceptive Orthodontics**
- Maxillary expansion if crossbite is present.
- Alignment of incisors.

- **Dental Monitoring**
- Cleft site may have hypodontia or supernumerary teeth.
- Regular fluoride application and preventive care

Alveolar Bone Grafting (8–11 years)

- **Goal:** Stabilize maxillary arch, support eruption of permanent canine/incisor.
- **Donor Sites:** Iliac crest or mandibular symphysis.
- **Timing:** Ideally before canine eruption

Bone grafting also helps close oro-nasal fistulae and prepares for future prosthodontics or implants. [8].

E. Adolescent Phase (12–18 years)

Comprehensive Orthodontics

- Fixed appliance therapy for arch coordination and alignment.
- Orthopedic correction for skeletal discrepancies, often combined with orthognathic planning

Orthognathic Surgery (After Growth Completion)

- Many CLP patients have maxillary hypoplasia due to intrinsic deficiency or surgical scarring.
- Maxillary advancement via Le Fort I osteotomy is commonly performed

Rhinoplasty and Scar Revision

- Performed for esthetic refinement of nasal shape and lip scars.
- Typically done post-adolescence or along with orthognathic surgery [17].

F. Adulthood and Long-Term Follow-up

Prosthodontic Rehabilitation

- Dental implants or removable prostheses may be needed if teeth are congenitally missing or poorly aligned [8].

Speech and Psychological Support

- Ongoing support for speech clarity and social integration.
- Psychosocial assessments to manage self-esteem, anxiety, or depression [13, 20].

Reconstructive Revisions

- Some adults require late-stage corrections of fistulae, nasal deformities, or malocclusion.

Table 1: Cleft Management Timeline

Age	Intervention	Purpose
Birth–3 months	Feeding support, NAM, counseling	Nutrition, parental bonding, segment alignment
3–6 months	Lip repair	Esthetics, function
9–18 months	Palate repair, grommets	Speech, reduce VPI, prevent ear infections
18 mo–5 yrs	Speech therapy, ENT follow-up	Develop intelligible speech, hearing monitoring
6–12 years	Orthodontics, alveolar bone grafting	Tooth alignment, arch stability
12–18 years	Orthognathic planning, rhinoplasty	Skeletal correction, nasal symmetry
Adulthood	Prosthodontics, revision surgeries	Final esthetics, function, psychosocial care

Orthodontic Management in Cleft Lip and Palate Patients

Orthodontic treatment is a crucial component of the multidisciplinary approach in managing patients with CLP. The objectives evolve with age, dentition stage, and skeletal growth, and must be carefully timed to coordinate with surgical and restorative interventions [9].

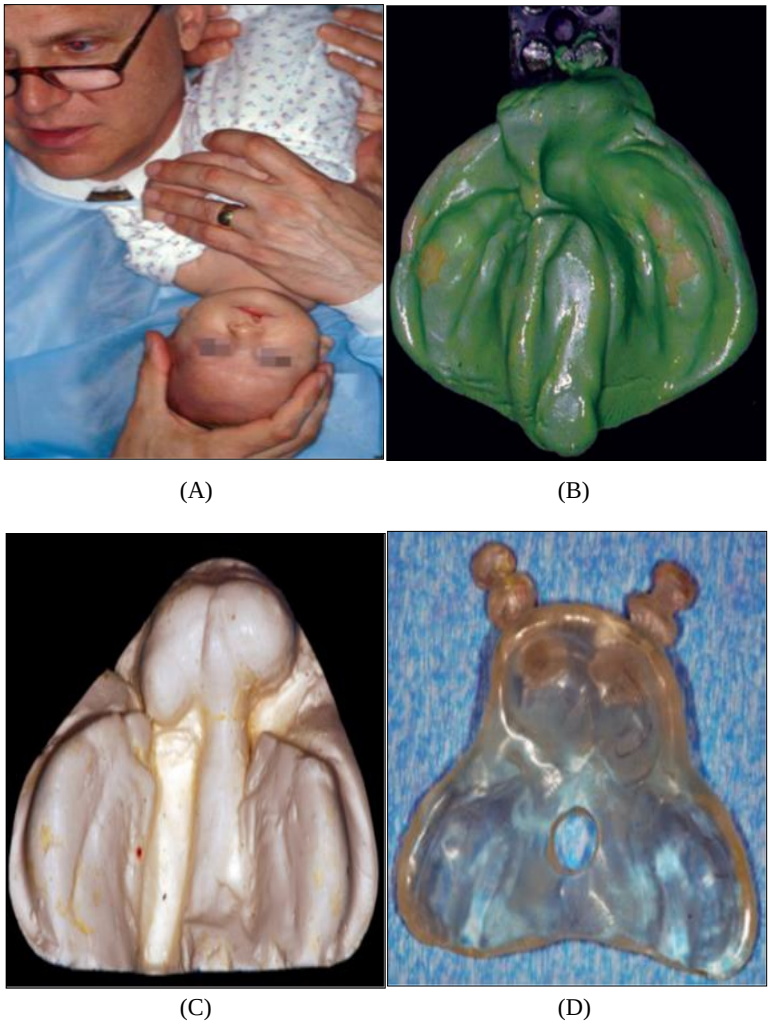
Phases of Orthodontic Management

1. Infant Orthodontics and Presurgical Orthopedics (Birth–6 months)

- **Nasoalveolar Molding (NAM)**

Introduced in infancy, NAM helps mold and align the alveolar segments of the maxilla before lip surgery.

NAM consists of a customized acrylic plate and nasal stents to reduce the alveolar gap, align segments, and improve nasal tip projection.
Benefits include better nasal symmetry and improved outcomes for primary lip repair [15]. (fig 3 & 4)



(A) (B)

(C) (D)

Fig 3: (A) Infant held in an inverted position during the impression process to prevent the tongue from falling back and to allow fl uids to drain out. (B) Impression of a unilateral cleft patient using a custom tray and heavy-body silicone impression material. (C) Plaster stone working model of a bilateral cleft patient for appliance fabrication. (D) Bilateral nasoalveolar moulding plate with retention buttons fabricated using self-cure acrylic resin

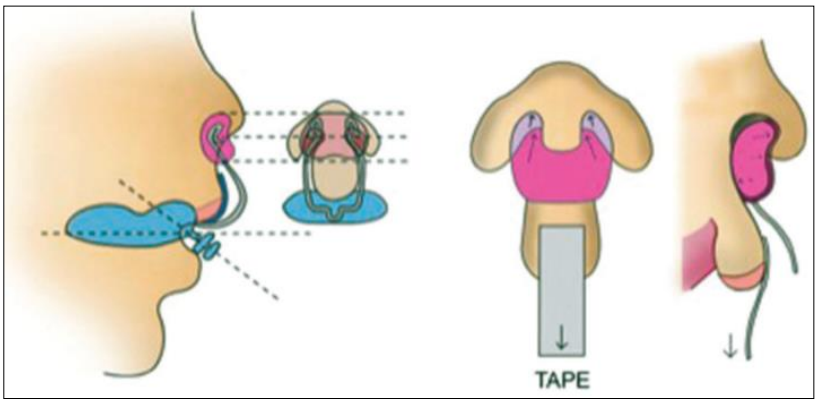


Fig 4: Figure showing the design of the nasal stent and the position of the nasal stent in the nostril

2. Early Mixed Dentition (6–9 years)

- **Goals**

Correct anterior and posterior crossbites.
Maintain arch form and width.
Facilitate eruption of permanent teeth.

- **Appliances**

Removable plates with expansion screws can be used in mild crossbites.
Fixed Rapid Palatal Expander (RPE) or quad-helix expanders are indicated in more severe transverse discrepancies.

- **Considerations**

Expansion is carefully timed before alveolar bone grafting to reduce the cleft gap and improve graft success [17].

3. Alveolar Bone Grafting Phase (8–11 years)

- **Timing**
- Alveolar bone grafting (typically secondary bone grafting) is performed when the permanent canine root is one-half to two-thirds formed [9].

- **Orthodontic Preparation**

Expansion of the maxillary segments prior to bone grafting helps reduce the alveolar gap and improve the bony support for erupting teeth [17].

Orthodontic appliances (fixed or removable) are used to align incisors and create space for canines.

- **Post-Grafting Orthodontics**

Once graft healing is confirmed (typically 4–6 months), orthodontics resumes to guide tooth eruption and alignment through the cleft site [9].

4. Comprehensive Orthodontic Treatment (Permanent Dentition, ~12–15 years)

- **Goals**

Establish a functional and esthetic occlusion.

Align teeth and coordinate arches in preparation for orthognathic surgery, if indicated.

- **Fixed Appliance Therapy**

Full fixed appliances (0.022" slot typically) are used for comprehensive alignment and leveling.

Archwires: Progression from light NiTi (0.014"–0.018") for initial alignment to rectangular stainless-steel wires for torque and finishing.

- **Anchorage Considerations**

Skeletal anchorage systems (e.g., mini-implants, TADs) can be helpful in managing asymmetries or retracting teeth across the cleft [17].

5. Orthognathic Surgery Coordination (Late Adolescence, 16–18 years)

- **Maxillary Hypoplasia Management**

Many patients with CLP develop maxillary retrusion due to intrinsic growth deficiencies or scarring from surgeries [7].

Le Fort I osteotomy is typically planned once facial growth is complete.

- **Orthodontic Preparation for Surgery**

Decompensation of dental inclinations (e.g., retroclined upper incisors) is crucial for accurate surgical planning.

Coordination of dental midlines and arch widths ensures optimal surgical positioning.

6. Post-Surgical and Retention Phase

- **Finishing and Detailing**

After orthognathic surgery, final detailing and settling of occlusion with finishing wires (e.g., 0.019x0.025 SS) is performed.

- **Retention**

Fixed retainers (bonded in the lower anterior segment) and removable retainers (Hawley or clear thermoplastic) are prescribed.

Retention protocols are adapted to the unique relapse risks in CLP, especially considering scar tissue and skeletal discrepancies.

Cleft management is a dynamic, multidisciplinary process that extends from infancy through adulthood. It requires staged interventions tailored to growth, development, and individual needs. With early, integrated care, most patients with CLP can achieve excellent functional and esthetic outcomes, although long-term psychosocial and dental follow-up remain essential. Modern treatment protocols emphasize family-centered care, patient-reported outcomes, and continuous quality improvement in both surgical and nonsurgical interventions.

Challenges in Orthodontic Management

1. Tooth Agenesis and Malformations

- Hypodontia of lateral incisors adjacent to the cleft site is common.
- Treatment plans may involve space closure or prosthetic replacement (implant or bridge) [6].

2. Asymmetries and Midline Deviations

- Asymmetrical growth of maxillary segments can cause midline discrepancies.
- Careful midline correction through asymmetric mechanics is often required [8].

3. Scarring and Soft Tissue Limitations

- Surgical scars may reduce soft tissue elasticity, complicating expansion or protraction of teeth.
- Forces are applied gradually to avoid dehiscence or gingival recession.

4. Psychosocial Considerations

- Adolescents with CLP may have heightened concerns about dental esthetics and facial appearance.
- Incorporating patient-centered goals is crucial for treatment acceptance [12].

Conclusion

Cleft lip and palate are complex congenital anomalies requiring lifelong multidisciplinary management. Advances in surgical and rehabilitative care have improved functional and esthetic outcomes, but residual challenges remain, particularly in speech, facial growth, and psychosocial adaptation. Ongoing research and patient-centered approaches are critical to optimizing outcomes and addressing disparities in care.

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