CLEFT MAXILLARY HYPOPLASIA: A REVIEW OF TREATMENT OPTIONS

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Abstract
Maxillary hypoplasia is a frequently observed secondary deformity in individuals born with cleft lip and palate. The impairment of maxillary growth coupled with extensive post-operative scarring as a result of multiple previous surgeries brings about maxillary deficiency in all planes viz vertical, horizontal and transverse. The resulting class III skeletal malocclusion together with maxillary hypoplasia produces structural imbalance, functional insufficiency, and aesthetic disharmony. Two types of options are available to manage this deformity viz non-surgical and surgical. The non-surgical options include Orthodontics alone or orthopedic protraction using face mask/reverse pull headgear. The surgical options include conventional orthognathic surgery at Lefort I level or distraction osteogenesis either complete or anterior maxillary distraction. This article highlights all the options available for managing this complex deformity. An effort has also been made to highlight the merits and demerits of each procedure as well as its adverse effects on speech present if any.

Keywords: Cleft maxillary hypoplasia, face mask, orthognathic surgery, distraction osteogenesis, anterior maxillary distraction

Introduction
Maxillary hypoplasia is the most commonly encountered secondary deformity as a result of cleft lip and palate with a reported incidence of about 15-50%¹. The reasons understood for this hypoplasia are twofold, firstly that the cleft maxilla’s intrinsic potential of growth is less due to the developmental deficiency. Secondly, the iatrogenic factor created due to the surgical repair of cleft lip and palate causes scarring which inhibits the normal growth of the maxilla². The resulting class III skeletal malocclusion together with maxillary hypoplasia produces structural imbalance, functional insufficiency, and aesthetic disharmony. Two types of options are available to manage this deformity viz non-surgical and surgical. The non-surgical options include Orthodontics alone or orthopedic protraction using face mask/reverse pull headgear. The surgical options include conventional orthognathic surgery at
Lefort I level or distraction osteogenesis (DO) either complete or anterior maxillary distraction (AMD).

**Non-Surgical Options**

The non-surgical options currently available are the use of Orthodontics alone\(^3\)\(^4\) or orthopedic protraction using face mask\(^5\)\(^6\). Orthodontic treatment consists of extraction of teeth, the proclamation of maxillary incisors and retraction of mandibular incisors in patients with a mild underbite. However, orthodontic treatment only camouflages the malocclusion and can result in a deficient amount of supporting bone for the dentition\(^3\)\(^4\).

Use of face mask was first reported by Molina et al\(^7\) and was largely inspired by Delaire reverse face mask originally conceived to orthopedically advance the maxilla in growing children presenting severe maxillary retrusion and class III malocclusion. Face mask therapy has been used to stimulate sutural growth at circum-maxillary suture sites in patients with mild to moderate maxillary hypoplasia\(^8\). A downward and forward movement of the maxilla, an increase in overjet and backward rotation of mandible with increased anterior facial height have all been documented with face mask therapy\(^5\)\(^9\)\(^10\).

The transmission of orthopedic force from protraction face mask to the maxilla is brought about with the use of intra oral devices like a labiobuccal arch, quad helix and bonded rapid maxillary expansion. The Rapid Maxillary Expansion is primarily used to disrupt the circum-maxillary sutural system increasing the effects of the orthopedic face mask and initiating downward and forward movement of maxillary complex\(^9\)\(^14\).

For orthopedic correction of a short maxilla in the cleft lip and palate patient, face mask forces range from 300 to 700 grams of force and the time period of treatment can be as early as 6 years of age and as late as 12 years. The treatment modality offers favorable outcomes in the early mixed dentition period (aged 7-9) than in late mixed dentition\(^5\). This treatment can correct class III malocclusion with a negative overjet 4-5 mm and advances the maxilla an average distance of 2.1 mm (range 1-4mm)\(^4\)\(^5\).

The use of protraction face mask is not without disadvantages. Usage of maxillary dentition as anchorage brings about adverse effects like lab version of maxillary incisors, extrusion of maxillary molars, counter-clockwise rotation of palatal plane, and eventual clockwise rotation of the mandible. It was recommended that it would be more advantageous to perform maxillary protraction with a mini plate placed in the infra zygomatic crest area in severe class III patients who need more advancement in the middle part of the zygomaticomaxillary complex, and maxillary protraction with a mini plate in the lateral nasal wall area in patients who need more advancement in the paranasal area and the lower part of zygomaticomaxillary complex. Several studies showed that the maximum stresses were seen at frontonasal, front maxillary, zygomaticomaxillary, zygomaticotemporal, and pterygomaxillary sutures. Some of the investigators claimed, that the infra zygomatic area could transfer the orthopedic force more effectively to the sutures than the lateral nasal wall and cause a slight tendency for counter clockwise rotation of the Naso maxillary complex. In contrast, some of them suggest that the lateral nasal wall might be favorable to minimize the counter-clockwise rotation of the maxilla and the lateral nasal wall of the maxilla might be a proper site for mini-plate placement because it is anterior to the center of resistance of the Naso maxillary complex, allowing the force vector to be near the centre of resistance. Therefore, changing the force application point to a more forward position and the force vector to a more downward direction might be recommended to minimize the unwanted
counter-clockwise rotation tendency of the Naso maxillary complex\textsuperscript{15-23}. Secondly, the compliance of the patient is very low due to the long treatment duration as well as the extra oral appliance design\textsuperscript{5}. Another drawback to early correction during growth is a recurrence of underbite with mandibular growth thereby allowing the occlusion to revert back to class III malocclusion during adolescence.

**Surgical Options**

It has been clearly documented in the literature that in nearly 25-47 \% of patients with cleft lip and palate, orthodontics alone might not be able to address the issue of maxillary hypoplasia and that these patients might require surgical intervention to achieve desirable aesthetics and functional results\textsuperscript{24-26}. The most traditional of all surgical treatment modalities is orthognathic surgery in the form of Lefort I maxillary advancement. Maxillary surgeries render themselves to correction of three-dimensional deformities unlike mandibular surgeries\textsuperscript{27} and are hence performed for cleft lip and palate patients who suffer from deformities in not only the sagittal plane but also the transverse and vertical planes. While traditional orthognathic surgery has the advantage of being a one stage procedure, it is not an easy task and is riddled with the problem of relapse. Relapse noted in few studies ranges from 22 \% to 40 \% in a horizontal plane and from 19 \% to 70 \% in vertical plane\textsuperscript{28-30}. Causes of high relapse rates are scarring from previous surgical repair, soft and hard tissue deficiency, aberrant dentition and larger advancements\textsuperscript{29}. Another major disadvantage is that maxillary advancement may predispose cleft lip and palate patients to velopharyngeal function deterioration as soft tissue moves forward especially when there is evidence of borderline insufficiency before surgery\textsuperscript{31, 32}. A study by Chua et al reported that advancement as small as 4 mm can result in deterioration of velopharyngeal function\textsuperscript{33}. Moreover, orthognathic surgery is usually reserved for patients in whom growth completion has occurred using at 16-18 years of age. Although somewhat variable in literature, the maximum recommended advancement in cleft lip and palate patients is around 6 mm\textsuperscript{34-36}. In patients presenting with severe maxillary hypoplasia (>10 mm), combining maxillary advancement with mandibular setback has been used to reduce the relapse and make the procedure more stable. But as cleft lip and palate patients have been shown to have normal to slightly small mandibles, this strategy may lead to facial disharmony depending on the starting mandibular position\textsuperscript{37-39}. The limitations observed with conventional orthognathic surgery were effectively dealt with by applying the principles of DO in the management of cleft maxillary hypoplasia. DO allows for the slow regeneration of bone accompanied by an expansion of the surrounding soft tissue envelope which makes larger movements possible with better long term stability thus lessening the risk of relapse. Horizontal relapse following DO has been found to be in the range of 5.5\% to 23 \% in horizontal plane\textsuperscript{40-43} which mostly occurred in the first 6 months after consolidation. DO allows for high osteotomies and a lack of rigid fixation techniques which avoids injury to tooth follicles and allows for advancement in growing patient\textsuperscript{37, 40, 44-45}. The majority of patients treated with maxillary DO report in the literature have been aged 6 to 15 years\textsuperscript{46}. Distraction osteogenesis provides a treatment modality for growing patients with severe maxillary hypoplasia. Both external and internal distraction devices have been considered for distraction osteogenesis. The Rigid External Distraction (RED) system in the mid face was first reported by Rachmiel et al\textsuperscript{47} which was then modified by Polley and Figueroa\textsuperscript{34} for maxillary advancement with predictable functional and aesthetic results. The principle advantages of RED...
are that it allows adjustment of the orientation of the distractor's vector during the entire duration of distraction period and that it is easy to install and remove. Despite the advantages and predictable results with RED, internal distraction devices have made tremendous progress, the principal reason being that these devices decrease the physical and psychological stresses caused by the bulkiness and management of the RED. Nevertheless, internal distraction devices have disadvantages. An inability to alter the vector during the distraction phase, difficulty in placing right and left distractors parallel to each other, discomfort related to stretch of buccal tissues by distractor rods, the limitation of distractor length and the need for a second operator to remove the devices are some of the disadvantages cited in the literature.

Whatever be the method chosen for DO, it should be kept in mind that it also causes or worsens velopharyngeal incompetence in cleft patients and there has been found to be no statistically significant difference between velopharyngeal incompetence seen after Lefort I maxillary advancement and maxillary DO. Hence, when considering DO for treating cleft maxillary hypoplasia, internal DO is a successful alternative in patients that require an advancement under 10 mm whereas RED is more suited for larger advancements. The distraction of the anterior maxillary segment is a novel surgical technique that brings about improvement in facial balance and aesthetics and provides stable occlusion without any detrimental effect on speech and velopharyngeal function. Block and Brister first reported on the clinical application of AMD using an intraoral tooth borne distractor in dogs in 1994 followed by Dolanmaz et al in humans in 2003. In contrast to conventional orthognathic surgery and DO of the entire maxilla, only the anterior maxillary segment is advanced by distraction, keeping the posterior aspect of the maxillary segment in contact with the posterior pharyngeal wall.

AMD has the distinct advantage of not worsening or affecting velopharyngeal function as the muscles of velopharynx are not affected by anterior maxillary advancement and the velopharyngeal closure remains intact. In a study by Richardson et al a 64% decrease in Velopharyngeal incompetence after anterior maxillary distraction as well as posterior movement of the posterior maxilla as evidenced by the posterior movement of the molars was seen bringing about substantial improvement in speech post distraction. Also, it does not have deleterious effects on growth of facial skeleton if done at early age as opposed to effects on growth of Lefort I advancement is performed. AMD can be carried out using either RED or internal distractors or tooth borne palatal distractors. Tooth borne distractors compared to RED or internal distractors offer the advantages of less conspicuousness, non-surgical fixing of the appliance in the mouth and a procedure that is financial, socially and psychologically acceptable to the patient. However, similar to internal distractors, the vector cannot be altered during the distraction phase. Furthermore, maintenance of oral hygiene can be difficult in the presence of the distractor.

Advancement up to 13 mm (average 9.42 mm) has been reported in a study by Richardson et al in 147 cleft lip and palate patients with cleft maxillary hypoplasia. He noted a relapse in 7 patients (4.76%) over a period of the 1-4 year follow up period.

**Conclusion**

In conclusion, numerous treatment options exist for managing cleft maxillary hypoplasia. An effort has been made to explain each treatment modality as comprehensively as possible. Each technique has its inherent merits and demerits. It is imperative that the technique chosen should be stable and be able to tackle the problem of cleft...
maxillary hypoplasia effectively without causing a relapse or without causing or worsening pre-existing velopharyngeal function and speech.

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