Van der Woude syndrome: Management in the mixed dentition

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Abstract

This article presents the case of a patient with Van der Woude syndrome treated with orthodontic and orthopedic intervention in the mixed dentition stage. The patient had a bilateral cleft of the lip and alveolus accompanied by lip pits on the lower lip. Intra-orally, there was bilateral anterior and posterior cross-bite with a collapsed maxilla. The maxillary transverse deficiency was managed with orthopedic expansion and the second phase of treatment involved secondary alveolar bone grafting followed by retention with functional regulator-3. The mild maxillary retrognathia and deficient lip support was managed with dental compensation.

Keywords: Cleft lip, cleft palate, secondary bone grafting, Van der Woude syndrome

Introduction

Van der Woude syndrome is an autosomal dominant syndrome characterized by cleft lip or cleft palate, distinctive pits of the lower lips, or both. It is the most common syndrome associated with cleft lip or cleft palate. The degree to which individuals who carry the gene are affected widely varies, even within families. In general, Van der Woude syndrome affects about 1 in 100,000-200,000 individuals. About 1-2% of patients with cleft lip or cleft palate have Van der Woude syndrome. Hypodontia (absent teeth) also has been increasingly recognized as a frequently associated anomaly.[1]

We present a case of Van der Woude's syndrome, which was managed by both orthodontic and surgical means in the mixed dentition period.

Case Report

A female patient aged 6 years and 7 months, reported to our department with the chief complaint of malaligned upper and lower front teeth. The patient did not give any history of medical conditions. Patient's parents were extremely concerned about the patient's esthetics and speech defects. Lip repair was done at the age of 3 months. The extra-oral examination revealed a surgical scar on the upper lip, bilateral symmetrical lower lip pits, obtuse nasolabial angle and a concave profile due to recessive upper lip and everted lower lip [Figures 1a and 1b].

A bilateral cleft of the alveolus was observed intra-orally with an overhanging mobile premaxilla. An oronasal fistula was present in the cleft on the right side. Posterior cross-bite on both the sides confirmed a collapsed maxillary arch. Mild crowding with retroclination of the anteriors was noticed in the lower arch. The patient was in mixed dentition stage with permanent central incisors still in erupting stage [Figures 1c].

Orthpantomograph [Figure 1d] showed congenitally missing upper lateral incisors. Also, lower second premolar tooth buds were missing.

The treatment was carried out in two phases.

Phase I of the treatment consisted of expansion of the maxillary arch to correct the posterior cross-bite [Figure 2]. Expansion was continued for 6 months until the posterior cross-bite was overcorrected and the quad helix was retained in place. Total expansion achieved was approximately of about 5 mm. Maxillary central incisors and canines were bonded and 0.016 niti arch-wire was ligated to prevent an anterior cross-bite from developing [Figure 3]. A posterior bite block was given in the lower arch to raise the bite.

Phase II of the treatment involved secondary alveolar bone grafting to provide adequate bone support for the eruption of canines. At the cleft site, mucoperiosteal flap was elevated and nasal floor was exposed. A pouch was created for receiving the graft. For the bone graft, iliac crest was exposed and cancellous bone was harvested. The bone graft was then

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placed in the pouch and the flap edges were approximated and sutures placed. The oronasal fistula on the right side of the cleft was completely obliterated by the graft.

No tooth movement was initiated for a period of 6 months post-surgery. On reviewing the case after 6 months, it was decided that maxillary protraction would not be required as the profile had become sufficiently straight. However, to maintain the arch width and to facilitate unhindered growth of maxilla till the permanent canines erupted, functional regulator-3 (FR-3) was delivered [Figures 4a and 4b].

It was observed that the premaxilla had immobilized completely and the profile had improved esthetically to a considerable extent [Figures 4c and 4d]. The patient was followed-up regularly for a period of 6 months after which she had to discontinue the treatment at our institution as her parents had migrated.

Discussion

Treatment of cleft patients in mixed dentition requires an accurate judgment on the diagnosis and treatment plan priorities.

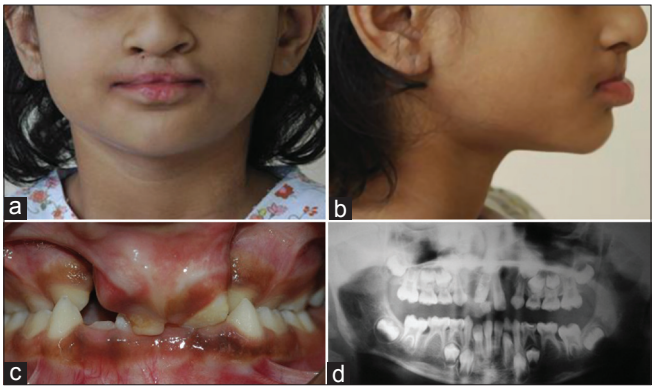

Figure 1: (a) Pre-treatment extraoral frontal view, (b) Pre-treatment extraoral profile view, (c) Pre-treatment intraoral frontal view, (d) Pre-treatment orthopantomograph

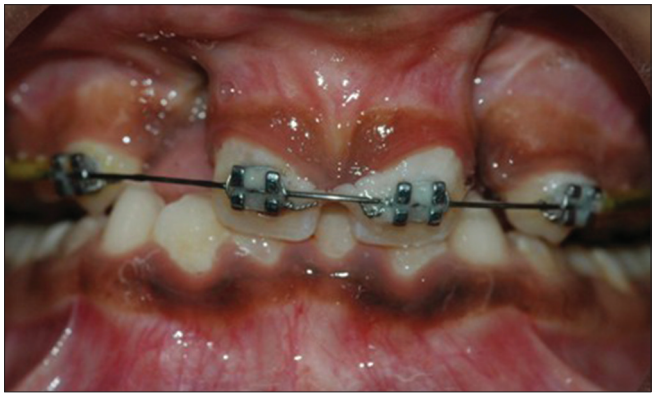

Figure 3: 0.016" Niti archwire ligated to prevent development of anterior crossbite

Teeth if made to erupt through the grafted bone further increase bone stock and respond well to orthodontic forces.

Need for secondary alveolar grafts:
Closing oronasal fistulae[2,3]
Stabilizing the maxillary segments[2,4]
Restoring the alveolar ridge[5]
Providing a bony support for teeth adjacent to the cleft[6]
Minimizing maxillary growth disturbance.

Bone grafting is typically performed between the ages of 9 and 11 years of age shortly before canine eruption.[6] In this case also the secondary alveolar grafting as done to achieve adequate bone level for canine eruption.

The anterior iliac crest is the most common donor site used today (gold standard). This site is preferred as adequate amount of bone can be mobilized, which has high particulate cancellous bone content.[5] Calvarium and mandibular bone[5] has been

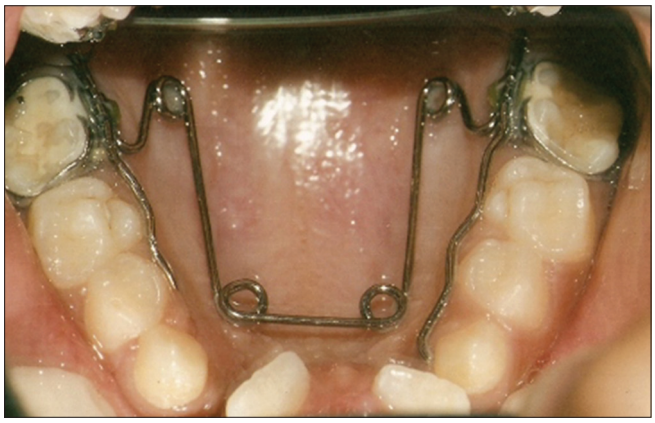

Figure 2: Phase I treatment with quad helix for maxillary expansion

Figure 4: (a) Status of the dentition 6 months post-surgery, (b) Functional regulator-3 appliance delivered to maintain the expansion (5 mm) and to promote unimpeded maxillary growth, (c) Extraoral frontal view 6 months post-functional regulator-3 appliance therapy, (d) Extraoral profile view post-functional regulator-3 appliance therapy

advocated, as being a superior donor however, there are inconsistent clinical results.

Radiographic follow-up demonstrated adaptation of the cancellous bone of the iliac crest to the host area, making it impossible to distinguish the mesial and distal limits of the cleft. The findings of present case agree with other studies in which teeth erupted through the grafted bone.[7,8] Cancellous bone graft is quickly incorporated and vascularized and most importantly, does not interfere in presence of the tooth contributes to the preservation of the grafted bone and to the differentiation of the periodontal support.[9,10]

The use of FR in cleft lip and palate patients has not been studied in detail. It has been said that Frankel appliances promote periosteal growth in either of the jaws depending on the type of appliance used. In this case, we decided to use FR-3 as a retention appliance to promote maxillary growth and to maintain the changes achieved till the permanent teeth especially, the canines erupt. This has given a better lip support, enhancing facial and dental esthetics and dental occlusion. Also the patient was quite comfortable and cooperative wearing the functional appliance.

One thing which really encouraged us was the increased positive psychologic impact on her personality and self-esteem.

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