VAN DER WOUDE SYNDROME: REPORT OF A CASE

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SUMMARY
A rare case of Van Der Woude Syndrome, which is characterized by pits in the lower lip and bilateral cleft of the lip and cleft palate is presented. A multidisciplinary approach to treatment produced an aesthetically pleasing and functional outcome.

INTRODUCTION
Congenital lip pits are developmental defects that occur on the paramedial portion of the vermilion border of the lower lip. They may be unilateral or bilateral and may occur as an isolated condition or in association with cleft lip and or cleft palate. When the labial pits occur in association with cleft lip and/or palate the condition is referred to as Van der Woude Syndrome[1].

Although the lip pits are inherited as an autosomal dominant trait, their pathogenesis is not well understood[2]. They are thought to develop from notching of the lip at an early stage of the labial development with fixation of the tissue at the base of the notch or from failure of a complete union of the embryonic lateral sulci of the lip, which persist and ultimately develop into the typical pits[3].

The surface opening of the lip pit may present as a circular or transverse slit or be located at the apex of nipple-like elevations and measure up to three millimetres in diameter. The depth of the pits ranges from one to fifteen millimetres[4]. Minor salivary gland orifices open into the pits hence the salivary exudate. Although the cleft lip and palate are the major esthetic problems for these patients, exudation of mucous from the lip pits onto the lower labial skin are a source of embarrassment to the patient.

We present a rare case of Van Der Woude syndrome with variable expressivity in a Ghanaian adolescent.

CASE REPORT
A sixteen-year-old female was referred from the Department of Oral and Maxillofacial Surgery of the University of Ghana Dental School to the Department of Orthodontics, for correction of her anterior cross bite. She was shy, withdrawn and continually held a handkerchief over her mouth. She had a history of a cleft lip and palate repair as a toddler.

Extraoral examination revealed mid face retrusion (Fig 1) and a repaired bilateral cleft lip of the upper lip. Her lower lip had bilateral lip pits at the base of two nipple-like elevations (Fig. 2). These lip pits were about three millimeters in diameter, ten millimeters deep and exuded mucous secretions continuously (Fig. 3).

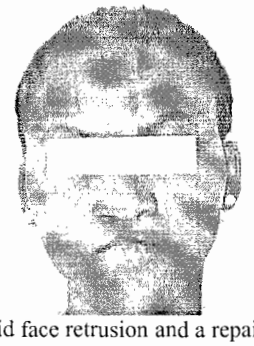

Figure 1 Mid face retrusion and a repaired bilateral cleft lip of the upper lip

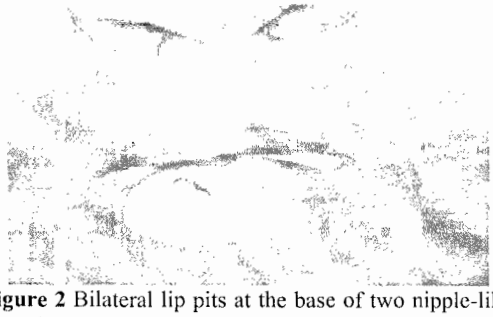

Figure 2 Bilateral lip pits at the base of two nipple-like elevations

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Figure 3 Lip pits of about three millimeters in diameter, ten millimeters deep and exuded mucous secretions continuously

To stop herself from drooling saliva down her chin, she had to bite on her lower lip and hold a handkerchief up against it hence her speech was barely audible.

Intraoral examination revealed a repaired maxillary cleft palate. Both maxillary lateral incisors and an upper right second bicuspid were congenitally missing. There was marked hypoplasia and labial abrasion facets on the maxillary left central incisor. The mandibular teeth however were all present and well aligned. Analysis of her occlusion revealed an anterior cross bite extending from the maxillary right first bicuspid to the maxillary left cuspid (Fig 4).

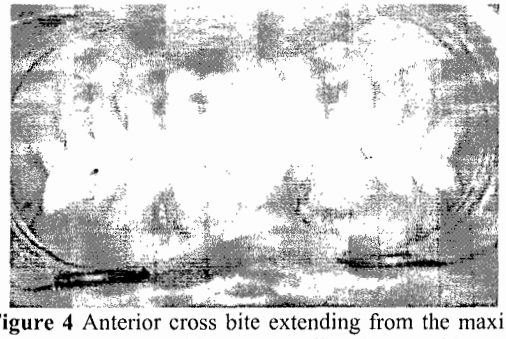

Figure 4 Anterior cross bite extending from the maxillary right first bicuspid to the maxillary left cuspid

The lip pits and the associated salivary gland tissues were excised under local anaesthesia (2% lignocaine hydrochloride with 1:50 000 epinephrine). Post-operatively, she was put on prophylactic antibiotics (500mg of amoxycillin capsules, three times a day for a week) and analgesics (200mg of ibuprofen three times a day for three days). Sutures were removed after one week. Healing of the surgical site was satisfactory with no mucous exudation being observed. She was again referred for orthodontic correction of the anterior cross bite (Fig. 5).

This was achieved with a removable maxillary appliance worn continuously over a six week period. At the completion of the treatment, she had a more normal closure pattern and decreased maxillary retrusion. Labial veneering of the maxillary central incisors was performed with composite restorative material. Spacing due to the congenitally missing maxillary left lateral incisor and maxillary right second bicuspid was closed with a removable partial denture pending fixed prosthodontics.

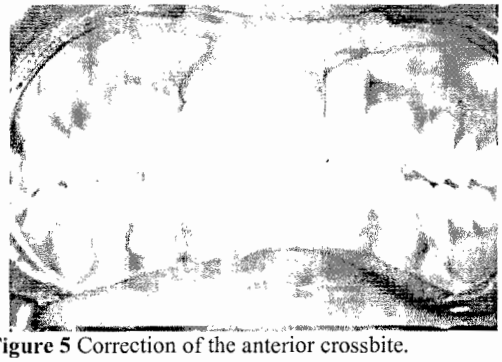

Figure 5 Correction of the anterior crossbite.

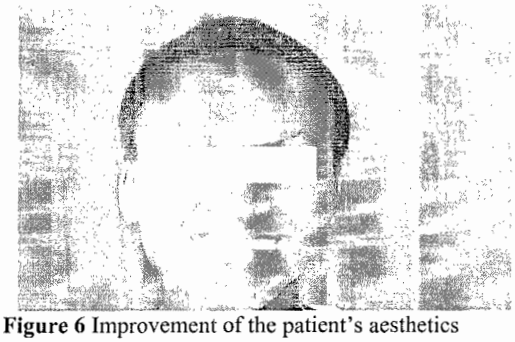

Figure 6 Improvement of the patient's aesthetics

DISCUSSION

The main clinical features of Van der Woude syndrome is well documented in the literature[1-7]. In addition to the classical clinical presentation, maxillary and mandibular adhesions, "peg-shaped" or missing maxillary lateral incisors may be observed. Ankyloglossia and cleft uvula were reported by Shawaf and Mani[8]. Two additional clinical features of enamel hypoplasia and multiple congenitally missing teeth observed in this case report indicate the variable clinical presentation of this syndrome.

The majority of the labial and commissural pits occur without mucus exudation[3]. However, a small percentage may be associated with cleft lip and palate and therefore not a thorough clinical examination is recommended in individuals with pits to

rule out a submucosal cleft palate which may not be obvious to the clinician.

Surgical excision of the labial and commissural pits is indicated if the aesthetics of the individual is appreciably affected and exudation of mucous secretions can not be controlled[9]. Surgical excision should include the total removal of the minor salivary glands that exude secretions at the base of the pits to prevent the formation of mucoceles or cysts.

In order to carry out the philosophy of 'total treatment", the collaboration of the departments of plastic surgery, oral and maxillofacial surgery, orthodontics and restorative dentistry were involved in the management of this case. This interdisciplinary collaboration resulted in a remarkable improvement of the patient's aesthetics (Fig 6). The quality of this patient's life improved considerably after treatment and earlier management of this case regarding the excision of labial pits, anterior crossbite correction and replacement of her missing teeth could have improved her self-esteem at a much earlier age. It is hoped that the multidisciplinary management of patients with oro-facial defects will be encouraged.

CONCLUSION

A case of Van Der Woude syndrome is presented. Additional clinical features noted were enamel hypoplasia and multiple congenitally missing teeth. Management of this case involved the collaboration of different specialists in dentistry. Earlier management of this case would have earned this patient years of improved self-esteem.

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