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Technical Note

# Surgical management of lower lip pits in Van der Woude syndrome

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ABSTRACT

Van der Woude syndrome (VDWS) is characterized by the presence of lower lip pits which may be of concern to patients due to aesthetic considerations. By presenting three clinical cases, we provide an overview of the surgical techniques currently available to treat labial pits. Fusiform excision with dissection of the entire pit is still the most commonly used procedure and it generally yields good functional and aesthetic outcomes. The split-lip advancement technique and the inverted T-lip reduction nonetheless represent good surgical alternatives. Proper management of the lower pits that occur with VDWS requires thorough knowledge of the available surgical procedures.



## 1. Introduction

The association between labial pits and cleft lip was first reported by Demarquay in 1845 [1]. Van der Woude described the syndrome (VDWS) and its mode of transmission in 1954 [2]. With an overall incidence of 1/60,000 individuals, VDWS represents the most common form of syndromic cleft lip/palate (CL/P).

Management of the cleft in VDWS does not differ from the surgical planning that is usually used. In regard to the labial pits, some teams favour treatment concomitant with surgical closing of the cleft [3] while others prefer a delayed surgery when treating children who are aged 10–12 months or older [4]. Regardless of the surgical planning and the technique employed, treatment of the lower lip often requires more than one procedure [5,6], and the afflicted children and their parents need to be made aware of this.

The main indication for surgery is cosmetic, although it can sometimes also be required as a result of chronic secretion or infection [7]. Facial surgeons generally consider the lip reconstruction after the surgical excision to be a significant challenge [5,8]. It consists of a complete excision of the fistula with its salivary environment, while preserving the oral sphincter [9]. The surgical treatment has to respect mucous unity of the lip, as well as the anatomical integrity of the orbicularis oris muscle and the mucocutaneous junction of the lower lip.

There are very few reports in the literature regarding the surgical management of the lower lip pits. By presenting three clinical cases, the aim of this work was to provide an up-to-date description of the most relevant techniques for lip reconstruction in VDWS.

## 2. Case report number 1

A twenty-one year old man presented with two paramedian and symmetric fistulas of the lower lip (Fig. 1). The surgical treatment consisted of a single stage fusiform excision of the pits and complete resection of the tracts.

## 3. Case report number 2

A young girl presented with VDWS associated with a complete bilateral CLP and symmetrical bilateral pits on the mucous side of the lower lip (Fig. 2). Primary surgery of the cleft was performed at 6 months of age. Surgical management of the pits was carried out in two stages, first by fusiform resection at 9 years of age, and secondly by mucous excision at 17 years of age, with good outcomes at the end of the follow-up period.

## 4. Case report number 3

This was a patient who exhibited VDWS with a left unilateral CLP and asymmetrical bilateral labial pits of the lower lip (Fig. 3). The left fistula was located on the mucocutaneous junction of the lip. The labial and palate cleft was operated on at 6 months of age. The surgical correction of the pits had to be undertaken in three stages, with a first excision of the sinuses associated with Z-plasties of the mucocutaneous line at 5 years of age. A second procedure consisting of an inferior labial mucous excision was

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**Fig. 1.** Van der Woude syndrome with lower labial pits in a 21-year-old man. Frontal (a) and lower (c) views of the lips. Postoperative clinical results after fusiform resection in frontal (b) and lower (d) views. Iconography: Prof. Mercier.

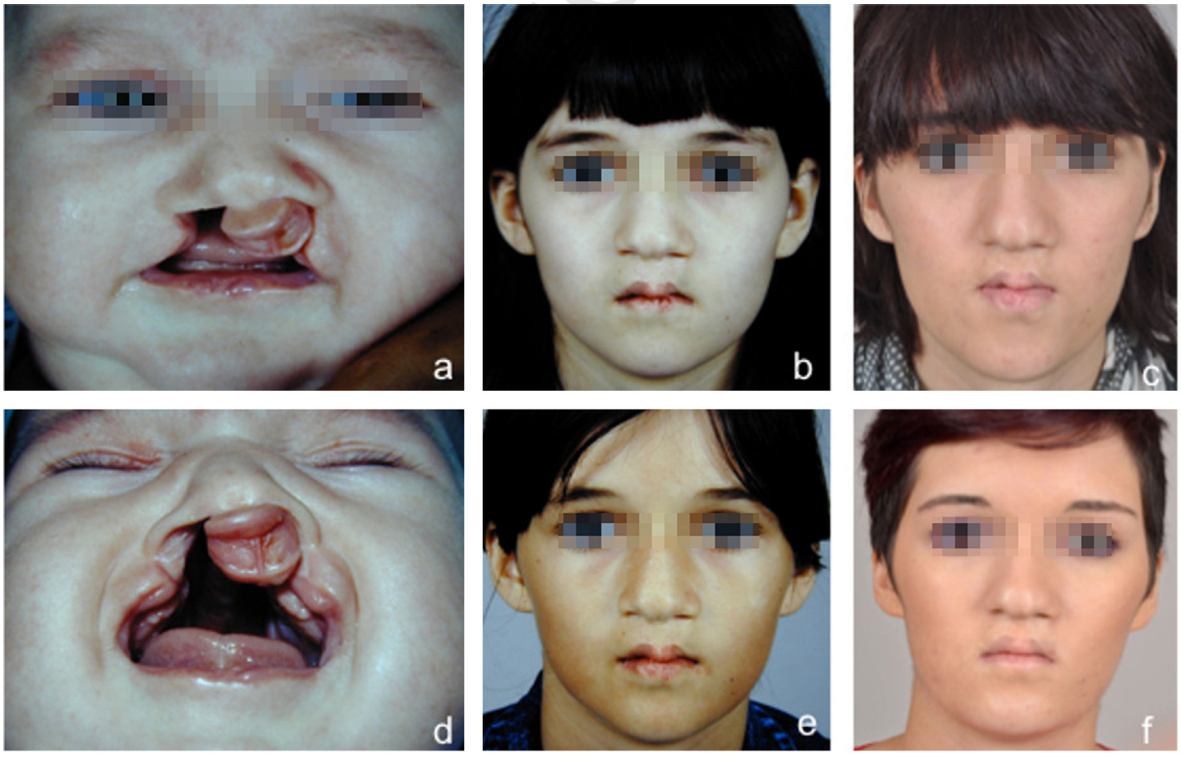


**Fig. 2.** A case of Van der Woude syndrome associated with a complete bilateral CLP and fistulas of the lower lip in a young girl. Pictures at the time of birth (a and d), before (b) and after (e) surgical excision at 9 years of age, before (c) and after (f) mucous resection at 17 years of age. Iconography: Prof. Mercier.

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