

## Systematic Review Craniofacial Anomalies

# Treatment of lower lip pits in Van der Woude syndrome: a systematic review

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**Abstract.** The presence of lower lip pits in individuals with Van der Woude syndrome (VWS) may cause discomfort due to saliva secretion. Furthermore, one of the main complaints in relation to lip pits is poor aesthetics, which often affects quality of life. The aim of this systematic review was to identify the best technique for the surgical removal of lower lip pits in terms of aesthetic and functional characteristics. A search of the PubMed, Embase, Web of Science, Science Direct, and Scopus databases was performed on December 27, 2016, which retrieved 88 records without duplicates. Among these papers, three ultimately met all eligibility criteria. The three studies included a total of 61 individuals, with follow-up ranging from 6 months to 10 years and sample collection from 10 to 24 years. The findings demonstrated that the outcome of surgical removal of lower lip pits was better with the use of vertical wedge excision, inverted-T lip reduction, Mutaf–Goldstein technique and modified simple excision than with simple excision. Simple excision may result in postoperative complications, such as mucocele and pit recurrence.

**Key words:** cleft lip; cleft palate; congenital abnormalities; reconstructive surgical procedures; Van der Woude syndrome; review; systematic.

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Van der Woude syndrome (VWS) is a developmental disorder with autosomal dominant inheritance, characterized by cleft lip and palate and congenital lower lip pits (OMIM 119300; Online Mendelian Inheritance in Man), with high penetrance ranging from 61%<sup>1</sup>, 75%<sup>2</sup>, 80%<sup>3</sup>, and 96.7%<sup>4</sup>, to close to 100% when considering individuals with lip pits and submucous cleft palate<sup>5,6</sup>. The prevalence is within the range of 1.65/100,000<sup>1</sup> to 1/100,000–1/75,000 individuals<sup>3</sup>. VWS is

the most common syndrome among individuals with cleft lip and/or palate, affecting 0.5% to 2% of all such individuals<sup>1,2,7–9</sup>. Mutations in interferon regulatory factor 6 (IRF6) have been identified as the cause of VWS, interrupting orofacial development<sup>9,10</sup>. Hypodontia is also observed in individuals with VWS (15.5–86%), especially affecting the maxillary incisors, canines, and premolars, as well as other dental anomalies that may cause malocclusions<sup>4,6,11–13</sup>.

Congenital lip pits were described by Demarquay in 1845, who reported pits ranging from small cavities in the lip vermilion border up to large structures in the midline, predominantly affecting the lower lip. The association with cleft lip and palate was reported in 1954 by Anne Van der Woude<sup>2</sup>. Histologically, the pathway of congenital lower lip pits is lined with a stratified squamous epithelium in continuity with the lip epithelium<sup>14</sup>. They are associated with minor salivary

glands in the lower lip mucosa, with a predominance of mucous acini and some seromucous units<sup>15</sup>.

The most common presentation is two pits in the lower lip vermillion, equidistant from the midline; however, they may be asymmetric or single, and centrally or laterally positioned in relation to the midline. The types of pits may vary in each family member with the syndrome<sup>3</sup>. Also, mild expressivity may involve unilateral or bilateral conical elevations without secretion, called microforms<sup>1</sup>, associated with submucous cleft palate and bifid uvula<sup>11</sup>. The depth may vary between 1 mm and 25 mm, and the location may range in anteroposterior direction from the lip vermillion border to the lip mucosa. With regard to the frequency of pits, 47–50% occur bilaterally, 18–27% occur in other mixed types, and 35% occur as microforms<sup>1,12</sup>.

These pits are usually asymptomatic, but they may continuously or intermittently drain small quantities of saliva<sup>14</sup>. There are cases of involuntary secretion that seem to be stimulated by the same mechanism as salivation, especially by mastication<sup>16,17</sup>. Involuntary secretion may also result from digital pressure, or be related to crying in children<sup>18</sup>, and may also increase in the wintertime<sup>17</sup>.

The surgical removal of lower lip pits may be indicated in the presence of emotional and social problems resulting from an unfavourable lip appearance. This may occur in the case that the anomaly represents a physical problem in terms of chronic and uncontrollable inflammation due to secondary infections<sup>16</sup>, mucous secretions, other aesthetic issues<sup>18</sup>, or severe lip hypotonia affecting the marginal and peripheral orbicularis muscles of the medial portion of the lower lip, leading to protrusion. There may also be associated intermaxillary problems, such as cross-bite<sup>19,20</sup>, and suction and feeding problems in children with conical lip protrusion<sup>21</sup>.

The possible treatments for this condition are surgical excision, surgical diathermy, and electrocautery<sup>11,18,22</sup>. Results with the latter two techniques have been unsatisfactory<sup>23</sup>. Furthermore, during surgical excision, it is important to remove the entire pathway of the pits to prevent recurrence and later formation of mucocele<sup>24,25</sup>.

Information regarding the best surgical technique for the excision of congenital lower lip pits in individuals with VWS is scarce, as are analyses of aesthetic and functional outcomes. Such information might be useful for dental professionals,

to identify the characteristics of the syndrome in the first years of life and allow referral to specialized services for the rehabilitation of these individuals, thereby enhancing their quality of life.

This systematic review was performed to identify the best surgical technique for the treatment of lower lip pits in individuals with VWS, as well as the functional and aesthetic outcomes.

## Methods

This systematic review was conducted in accordance with the PRISMA guidelines (Preferred Reporting Items for Systematic Reviews and Meta-Analyses)<sup>26</sup>.

All studies met the criteria established by the PICO strategy: participants (P) were individuals with VWS requiring the removal of lower lip pits; the intervention (I) was surgical techniques other than the control (C) simple excision surgical technique; the outcomes (O) were functional and aesthetic outcomes related to the removal of lower lip pits.

## Records and protocol

The protocol of this systematic review was registered in the PROSPERO database (CRD42017056047) and is available at <http://www.crd.york.ac.uk/PROSPERO/>. PROSPERO is an international database of systematic reviews in the field of health and social care, which was developed and is managed by the National Institute for Health Research at the University of York, UK.

## Identification of studies

A thorough literature review was conducted to identify studies on the removal of lower lip pits in individuals with VWS. The identification of studies was based on a search strategy for each of the following electronic databases: PubMed, Embase, Web of Science, Science Direct, and Scopus.

A meticulous search strategy was performed, structured with Boolean operators (AND/OR) and designed to identify all relevant studies on VWS published up to December 27, 2016. The following descriptors were used: Van der Woude syndrome, lip pits, cleft lip, and cleft palate. Boolean operators were used, namely 'OR' to include similar or synonymous terms, and 'AND' to relate the aforementioned terms (Van der Woude syndrome AND lip pits AND cleft lip OR cleft palate). The strategy was the same for all databases except for Science

Direct, in which the 'Journal' filter was applied. The following search strategy was used: ((("Van der Woude syndrome"[All Fields])) AND ((Lower[All Fields] AND ("lip"[MeSH Terms] AND pits[All Fields]) OR "Lip pits"[All Fields] OR (Lower[All Fields] AND ("lip"[MeSH Terms] OR "lip"[All Fields]) AND fistulae[All Fields])) OR ((("congenital"[Subheading] OR "congenital"[All Fields])) AND lower[All Fields] AND ("lip"[MeSH Terms] AND ("congenital abnormalities"[MeSH Terms] OR "malformation"[All Fields])) AND ((("cleft lip"[MeSH Terms] OR "harelip"[All Fields]) OR ("cleft palate"[MeSH Terms] OR ("cleft"[All Fields] AND "palate"[All Fields]) OR "cleft palate"[All Fields])). The grey literature was also searched to include any additional paper that might eventually meet the eligibility criteria.

The records were collected and saved in the reference management software End-Note Web (<http://www.myendnoteweb.com>) and checked for duplicate references.

All collected studies were analyzed independently by two examiners (MPM and ATP), to avoid biases and ensure that they met the eligibility criteria for the review ( $\kappa = 0.96$ ). The identification of studies for inclusion was divided into two stages: in the first stage, the title and abstract of each paper were read; in the second stage, the full-text articles were read. Discordance during the identification of studies at each stage was resolved by discussion with a third examiner (IFRB). The detailed search strategy is presented in Figure 1.

## Selection criteria

This study included clinical studies on the removal of lower lip pits in individuals with VWS, as well as studies comparing simple excision with other techniques for the surgical removal of lower lip pits.

Review papers, clinical case reports, and case series were excluded, as well as epidemiological studies addressing the phenotypic variability of VWS, studies on the histological characteristics of lower lip pits, genetic studies, analyses of genetic penetrance or genetic maps, studies on associations between VWS and tooth abnormalities or on dental management, papers about the differential diagnosis with other syndromes, and studies that did not include two groups, i.e. a group undergoing simple excision as the control group, and a group undergoing other surgical techniques as the experimental group.

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