

Management of Lip Pits in Van der Woude Syndrome: A Clinical Classification With Difficulty Index



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Purpose: Numerous case reports have been published on lip pits in Van der Woude syndrome explaining the morphology and genetics in detail; however, thus far, no article has focused on the classification of lip pits as an aid in surgical management. Although the procedure for lip pits in Van der Woude syndrome appears straightforward, even in the best of hands, the excision can be very challenging with no guarantee of esthetically desirable results. Therefore, we have devised a classification based on a difficulty index in the management of lower lip pits to assist in predicting the treatment outcome before surgery, as well as to offer the choice of a particular technique in a specific situation.

Materials and Methods: We reviewed 19 cases of Van der Woude syndrome having lower lip pits that were operated on at our unit from May 2005 to June 2015 with a minimum follow-up of at least 6 months. The data analyzed included the patient's age and gender, location of the lip pits with regard to their proximity to the white skin roll, number of lip pits, presurgical depth of the lip pits, and discharge of mucous secretion from the pits, as well as timing of lip pit excision. Four techniques of excision were performed via routine excision, modified routine excision, vertical wedge excision, and inverted-T lip reduction. The data were tabulated and analyzed. On the basis of our experience in managing lip pits, a clinically relevant classification with a difficulty index was then proposed.

Results: Among the 12 patients having preoperative involvement of the white skin roll, 8 had distortion of the white skin roll when operated on by either routine excision (n = 2), modified routine excision (n = 3), or inverted-T lip reduction (n = 3). The remaining 4 patients had no distortion of the white skin roll after surgery when the vertical wedge excision technique was performed. The 7 patients who had no distortion of the white skin roll preoperatively presented with esthetic results when operated on by either the routine excision, modified routine excision, or inverted-T lip reduction technique. In 2 patients with a presurgical pit depth greater than 6 mm, mucocele formation was observed after surgery. Using the data obtained, we proposed a classification based on 2 parameters: involvement of the white skin roll and presurgical depth. A difficulty index also was proposed using these same variables.

Conclusions: Classification and evaluation of the difficulty of lip pit excision are essential in planning the surgical treatment to give improved esthetic results. This proposed classification and difficulty index will provide the operating surgeon with a standardized scheme to evaluate the difficulty of the excision as well as to predict the overall outcome of the procedure before surgery.

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Van der Woude syndrome is the most common form of syndromic orofacial clefting, accounting for approximately 2% of all the cleft cases; the cardinal features are lip pits with or without cleft lip or palate.¹⁻³ The occurrence of lip pits on the vermilion border of the lower lip represents the most consistent feature of the syndrome, seen in nearly 90% of cases. Classically, 2 paramedian pits, bilaterally symmetrical and on either side of the midline of the lower lip, are the most common presentation.⁴⁻⁷ They usually communicate with the underlying minor salivary glands by way of canals formed by these pits that penetrate the orbicularis oris muscle at a variable distance of 1 to 25 mm.⁵⁻⁷ Although they are mostly asymptomatic, intermittent to continuous drainage of watery or salivary secretions may be expelled from these pits on occasion, contributing to a source of embarrassment to the patient and an unappealing cosmetic problem.

There are an abundance of case reports documented in the literature wherein the location and morphology of lower lip pits are described precisely.⁵⁻¹⁰ However, no article has focused on a classification aiding in the management of lower lip pits. We, in the first study of its kind, have highlighted a classification based on a difficulty index in the management of lower lip pits that helps in predicting not only the treatment outcome, but also the choice of a particular technique for a specific situation.

Materials and Methods

We analyzed the records of 19 cases of Van der Woude syndrome presenting with lip pits that were operated on at our unit from May 2005 to June 2015 with a minimum follow-up of 6 months. The study was ethically approved by the hospital's ethical committee and followed the guidelines of the Declaration of Helsinki. The data analyzed included the patient's age and gender, location of the lip pits with regard to their proximity to the white skin roll, number of lip pits, presurgical depth of the lip pits, and discharge of mucous secretion from the pits, as well as timing of lip pit excision.

As part of the hospital's protocol, lip pit excision using any of the four techniques—routine excision, modified routine excision, vertical wedge excision, or inverted-T lip reduction—was carried out simultaneously with another surgical procedure including cheiloplasty, palatoplasty, or secondary alveolar bone grafting (SABG) depending on the age at presentation. After excision of lip pits by any of the 4 techniques, the patients were followed for at least 6 months and the results were analyzed. On the basis of our experience in managing these lip pits, a

clinically relevant classification with a difficulty index was then proposed.

Results

The data regarding the study population are presented in [Table 1](#). The study population included 11 male and 8 female patients with ages ranging from 3 months to 9 years. With regard to the location of the lip pits, the white skin roll was involved by the lip pits in 12 cases, whereas in the remaining 7 cases, the lip pits were localized to the vermilion border of the lower lip with no involvement of the white skin roll ([Figs 1, 2](#)). Eighteen patients presented with 2 paramedian pits that were either bilaterally symmetrical (n = 14) or bilaterally asymmetrical (n = 4). A single case presented with a midline lip pit. The depth of the lip pits was measured using a graded lacrimal probe in all cases and ranged from 3.6 to 17.3 mm. We further observed that expression of mucous secretion from the pits on palpation was noted when the depth exceeded 6 mm (n = 11).

With regard to the timing of lip pit excision, 6 patients underwent excision at the time of cheiloplasty, 6 underwent excision at the time of palatoplasty, and 7 underwent excision at the time of SABG. Among the 19 patients requiring lip pit excision, 4 were operated on by the routine excision technique, 6 by modified routine excision, 4 by vertical wedge excision, and 5 by the inverted-T lip reduction technique ([Fig 3](#)).

When we analyzed the results at 6 months, among the 12 patients who presented with involvement of the white skin roll, 8 had distortion of the white skin roll after surgery ([Fig 4](#)). These patients were operated on by either the routine excision (n = 2), modified routine excision (n = 3), or inverted-T lip reduction (n = 3) technique. The remaining 4 patients presented with favorable esthetic outcomes with maintenance of the regularity of the white skin roll after surgery. These patients were operated on by the vertical wedge excision technique. However, all 4 patients presented with visible linear scars in relation to the skin of the lower lip (bilateral in 3 instances and midline in 1 instance). At long-term follow-up, these scars were barely perceptible ([Fig 5](#)). In the remaining 7 patients, who had presented with no involvement of the white skin roll preoperatively, favorable results were achieved by either the routine excision, modified routine excision, or inverted-T lip reduction technique. In 2 instances, mucocele formation was noted in those patients who underwent lip pit excision by the routine excision technique. In both cases, the presurgical depth of the pits was more than 6 mm, and both patients had expression of mucous secretion from the lips on palpation preoperatively.

Table 1. DATA OF STUDY POPULATION

Patient No.	Age/Gender	Involvement of White Skin Roll Preoperatively	Presentation of Lip Pits	Presurgical Depth, mm	Discharge of Mucous Secretion From Pits	Timing of Lip Pit Excision	Difficulty Index Score	Technique of Excision	Maintenance of Regularity of White Roll After Surgery
1	3 mo/M	Yes	Bilaterally symmetrical	6.1	Yes	With cheiloplasty	4	Routine excision	No
2	3 mo/F	Yes	Bilaterally asymmetrical	3.6	No	With cheiloplasty	3	Routine excision	No
3	5 mo/M	Yes	Bilaterally symmetrical	4.7	No	With cheiloplasty	3	Inverted-T lip reduction	No
4	8 yr/M	No	Bilaterally asymmetrical	9.9	Yes	With SABG	3	Routine excision	Yes
5	2 yr/M	No	Bilaterally symmetrical	5.7	No	With palatoplasty	2	Modified routine excision	Yes
6	3.5 yr/F	Yes	Bilaterally symmetrical	5.8	No	With palatoplasty	3	Inverted-T lip reduction	No
7	10 mo/M	Yes	Bilaterally symmetrical	12.9	Yes	With palatoplasty	4	Vertical wedge resection	Yes
8	8 yr/M	Yes	Single median	15.1	Yes	With SABG	4	Vertical wedge excision	Yes
9	9 yr/F	No	Bilaterally symmetrical	5.2	No	With SABG	2	Modified routine excision	Yes
10	7 yr/M	Yes	Bilaterally symmetrical	15.9	Yes	With SABG	4	Vertical wedge excision	Yes
11	4 mo/M	No	Bilaterally asymmetrical	11.6	Yes	With cheiloplasty	3	Modified routine excision	Yes
12	5 mo/M	No	Bilaterally symmetrical	4.9	No	With cheiloplasty	2	Inverted-T lip reduction	Yes
13	7 yr/M	Yes	Bilaterally symmetrical	11.9	Yes	With SABG	4	Modified routine excision	No
14	8 mo/F	No	Bilaterally asymmetrical	4.4	No	With cheiloplasty	2	Routine excision	Yes
15	7.5 yr/F	Yes	Bilaterally symmetrical	17.3	Yes	With SABG	4	Vertical wedge excision	Yes
16	4 yr/F	No	Bilaterally symmetrical	9.2	Yes	With palatoplasty	3	Inverted-T lip reduction	Yes
17	1 yr/F	Yes	Bilaterally symmetrical	5.1	No	With palatoplasty	3	Modified routine excision	No

Table 1. Cont'd

Patient No.	Age/Gender	Involvement of White Skin Roll		Presentation of Lip Pits	Presurgical Depth, mm	Discharge of Mucous Secretion From Pits		Timing of Lip Pit Excision	Difficulty Index Score	Technique of Excision	Maintenance of Regularity of White Roll After Surgery
		Preoperatively	Postoperatively			Yes	No				
18	6.5 yr/F	Yes	Yes	Bilaterally symmetrical	14.3	Yes	No	With SABG	4	Inverted-T lip reduction	No
19	12 mo/M	Yes	Yes	Bilaterally symmetrical	10.1	Yes	No	With palatoplasty	4	Modified routine excision	No

Abbreviations: F, female; M, male; SABG, secondary alveolar bone grafting.

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PROPOSED CLASSIFICATION

A clinically relevant classification has been proposed based on 2 parameters, that is, involvement of the white skin roll and presurgical depth of the lip pit:

1. Involvement of white skin roll
 - A. Uninvolved
 - B. Involved
2. Presurgical depth of lip pit or pits
 - A. Shallow (≤ 6 mm)
 - B. Deep (> 6 mm)

DIFFICULTY INDEX

A difficulty index has been proposed based on the same 2 parameters:

1. Involvement of white skin roll
 - A. Uninvolved—assigned a score of 1
 - B. Involved—assigned a score of 2
2. Presurgical depth of lip pit or pits
 - A. Shallow (≤ 6 mm)—assigned a score of 1
 - B. Deep (> 6 mm)—assigned a score of 2

The difficulty index was scored as follows: score of 2, easy; score of 3, moderately difficult; and score of 4, difficult.

Discussion

A definite protocol for lip pit excision has not been mentioned in the literature so far. Only a single study, by Krauel et al,¹¹ has mentioned the excision of lip pits that was carried out at the time of cleft lip repair in their study population. At our hospital, lip pit excision was carried out simultaneously with either cleft lip repair or cleft palate repair or with SABG as part of the hospital's protocol. In developing parts of the world, most patients with clefts are from low socioeconomic backgrounds and therefore cannot afford the cost of the medical care. Such patients benefit from undergoing two surgical procedures at one time. Furthermore, performing the operation simultaneously with another procedure avoids the need for an additional procedure at a later date with the patient under general anesthesia, with its attendant complications.

Elimination of the cosmetic deformity is the most common indication for surgical correction of lower lip pits, but the presence of infection or chronic inflammation also may warrant intervention.^{12,13} Although the lip pits in Van der Woude syndrome give a perception of easy excision, this is not the case because it is very difficult to achieve esthetically pleasing results in all cases. We used 4 techniques of lip pit excision: routine excision, modified routine



FIGURE 1. A-D, Photographs showing lip pits involving white skin roll preoperatively.

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excision, vertical wedge excision, and inverted-T lip reduction. The techniques of routine excision, vertical wedge excision, and inverted-T lip reduction have been discussed extensively in the literature, with authors showing favorable results using all these techniques.¹³ In modified routine excision, the exterior markings of the tracts were first marked similar to routine excision, but were connected in the middle (Fig 6A). The lateral edges of the incision stopped just beyond the lateral aspect of the tracts in a wedge-shaped form. Anteriorly, the incision was kept parallel to the white skin roll in such a way that a constant width of the vermilion tissue remained anteriorly. The incision was then made along the markings and deepened into the labial mucosa. The sinus tracts were probed during excision with a mosquito forceps to ensure complete removal of the sinus tracts (Fig 6B). The incised fibers of the orbicularis muscle were sutured for correct restoration of the orbicularis ring followed by closure of the incision with either Vicryl Rapide or Vicryl sutures (Ethicon, Somerville, NJ) (Fig 6C). We modified the routine excision technique because of issues related to access when the

depth of the pit was found to be increasingly greater, making it difficult to excise the pit in toto. Furthermore, because of improved access, no residual tract tissue was left behind, thereby avoiding mucocele formation as seen with the routine excision technique.

As of now, there exists no clinical article that has discussed the classification of lip pits or the difficulty levels of the repair. Our report introduces a new classification and difficulty index for management of lip pits in Van der Woude syndrome. It also emphasizes the use of the difficulty index to preoperatively judge the outcome of the procedure. Furthermore, the choice of one particular technique of excision over another also is presented.

A difficulty index score of 2 includes those pits that do not involve the white skin roll and those that are shallow (depth ≤ 6 mm). In our view, such cases are the easiest to deal with and present a cosmetic surgical outcome in nearly all patients, as observed in this study, regardless of the technique used for excision. We believe that an amateur cleft surgeon learning the art of lip pit excision also can achieve good results



FIGURE 2. A-D, Photographs showing lip pits not involving white skin roll preoperatively.

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when the difficulty index is 2. Techniques such as modified routine excision and inverted-T lip reduction offer the best results because the scar remains hidden at the red line of the vermillion. Whatever the technique of lip pit excision used, it is of utmost importance to bring about complete elimination of the fistulous tracts to prevent pit recurrence or development of mucous cysts at a later date.

A difficulty index score of 3 includes either those pits that do not involve the white skin roll but have depths greater than 6 mm (deep) or those that involve the white skin roll and are shallow. In the first instance, a cosmetic outcome is possible provided that complete excision of the pits is carried out. With increasing depth, the fistulas show a high propensity for bifurcation, in which case using the routine excision technique might leave behind residual tract tissue, leading to mucocele formation at a later date, as observed in our study in 2 cases. In our opinion,

such cases should be dealt with by either the modified routine excision, vertical wedge excision, or inverted-T lip reduction technique because the surgical access offered by these techniques allows for complete excision of the fistulous tracts. In the second instance, although the excision of the lip pits is relatively easy because of the lesser depth, it is difficult to maintain the regularity of the white skin roll because of the preoperative involvement of the white skin roll. We observed distortion of the white skin roll in all cases after surgery performed by either the routine excision, modified routine excision, or inverted-T lip reduction technique. Maintenance of the regularity of the white skin roll is possible with the vertical wedge excision technique; however, linear scarring of the lower lip skin is a disadvantage of the procedure. Moreover, in patients with less lower lip tissue preoperatively, this technique can bring about shortening of the lower lip. Linear scarring is not an issue because scars were

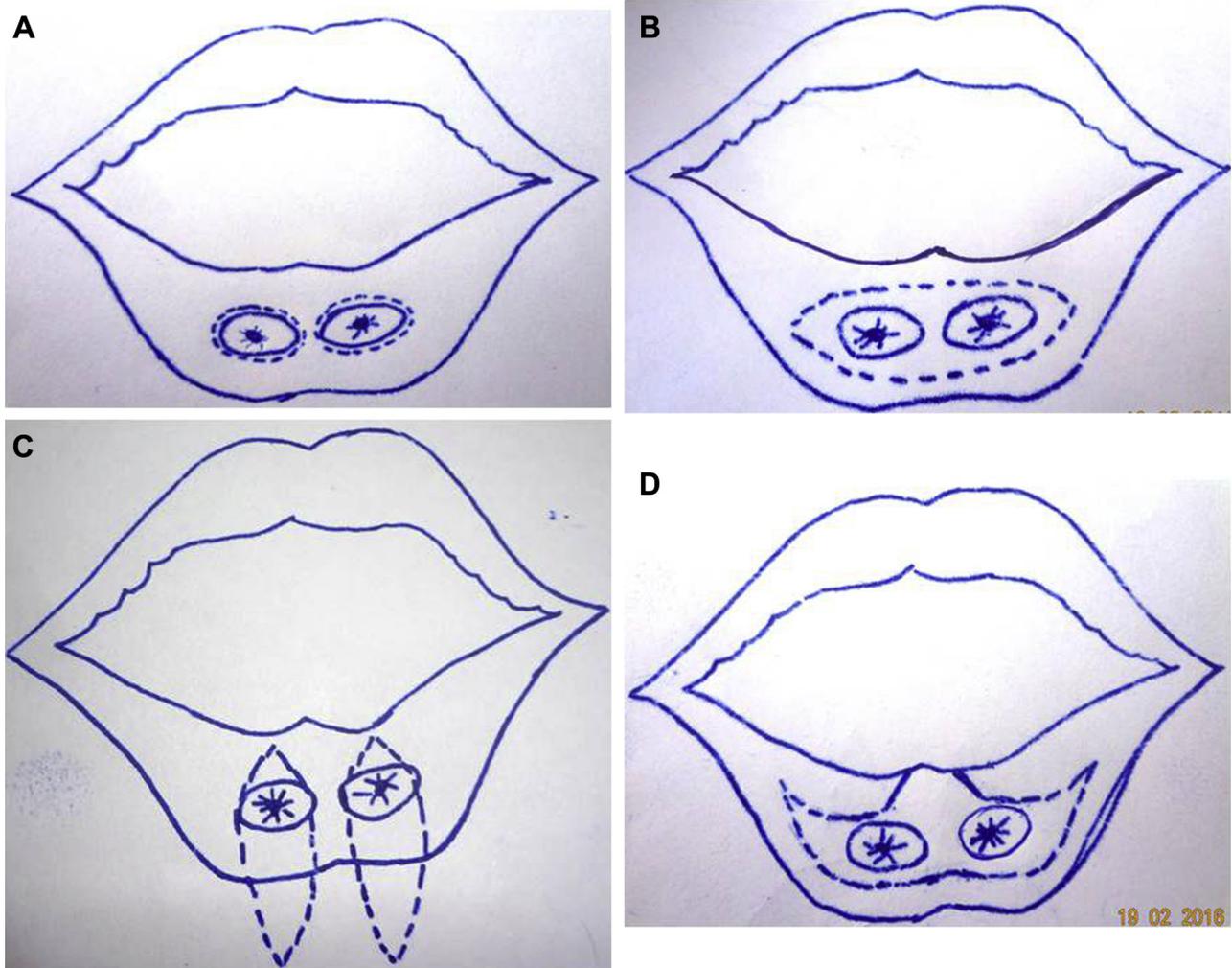


FIGURE 3. Diagrammatic representation of lip pit excision techniques. A, Routine excision. B, Modified routine excision. C, Vertical wedge excision. D, Inverted-T lip reduction.

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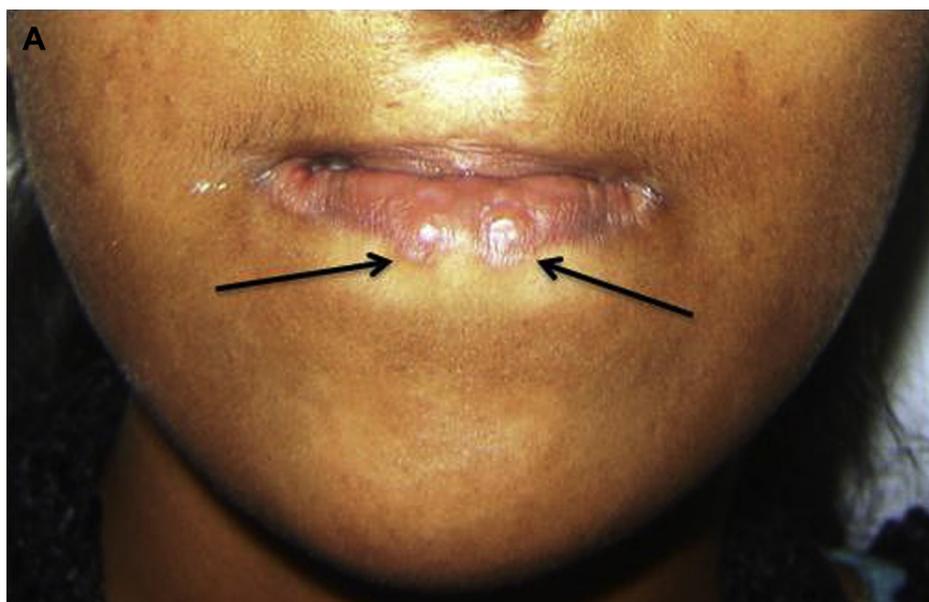


FIGURE 4. Distortion of white skin roll in 2 cases having preoperative involvement of white skin roll (arrows): patient operated on by routine excision technique (A) and patient operated on by modified routine excision technique (**Fig 4 continued on next page.**)

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FIGURE 4 (cont'd). (B).

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observed to become barely visible with increasing follow-up periods. The decision either to perform a vertical wedge excision that maintains the regularity of the white skin roll while risking a scar on the lip or to avoid a scar on the lip by choosing any other form of excision rests with the surgeon.

A difficulty index score of 4 includes those pits that involve the white skin roll and are deep (depth >6 mm). In our opinion, these cases pose considerable difficulty in achieving esthetically pleasing results. Distortion of the white skin roll is inevitable when techniques such as modified routine excision or

inverted-T lip reduction are used. We have observed favorable results using the vertical wedge excision technique in all 4 patients in whom this technique was performed.

Although lip pit excision appears to be a relatively straightforward procedure to perform, it is very difficult to achieve esthetically pleasing results in all cases. The proposed classification and difficulty index provide the operating surgeon with a standardized scheme to evaluate the difficulty of the excision as well as to predict the overall outcome of the procedure before surgery.



FIGURE 5. A case showing a lip pit involving the white skin roll treated by the vertical wedge excision technique. A, Intraoperative photograph. B, Postoperative photograph at 1 year showing barely perceptible scar and maintenance of regularity of white skin roll.

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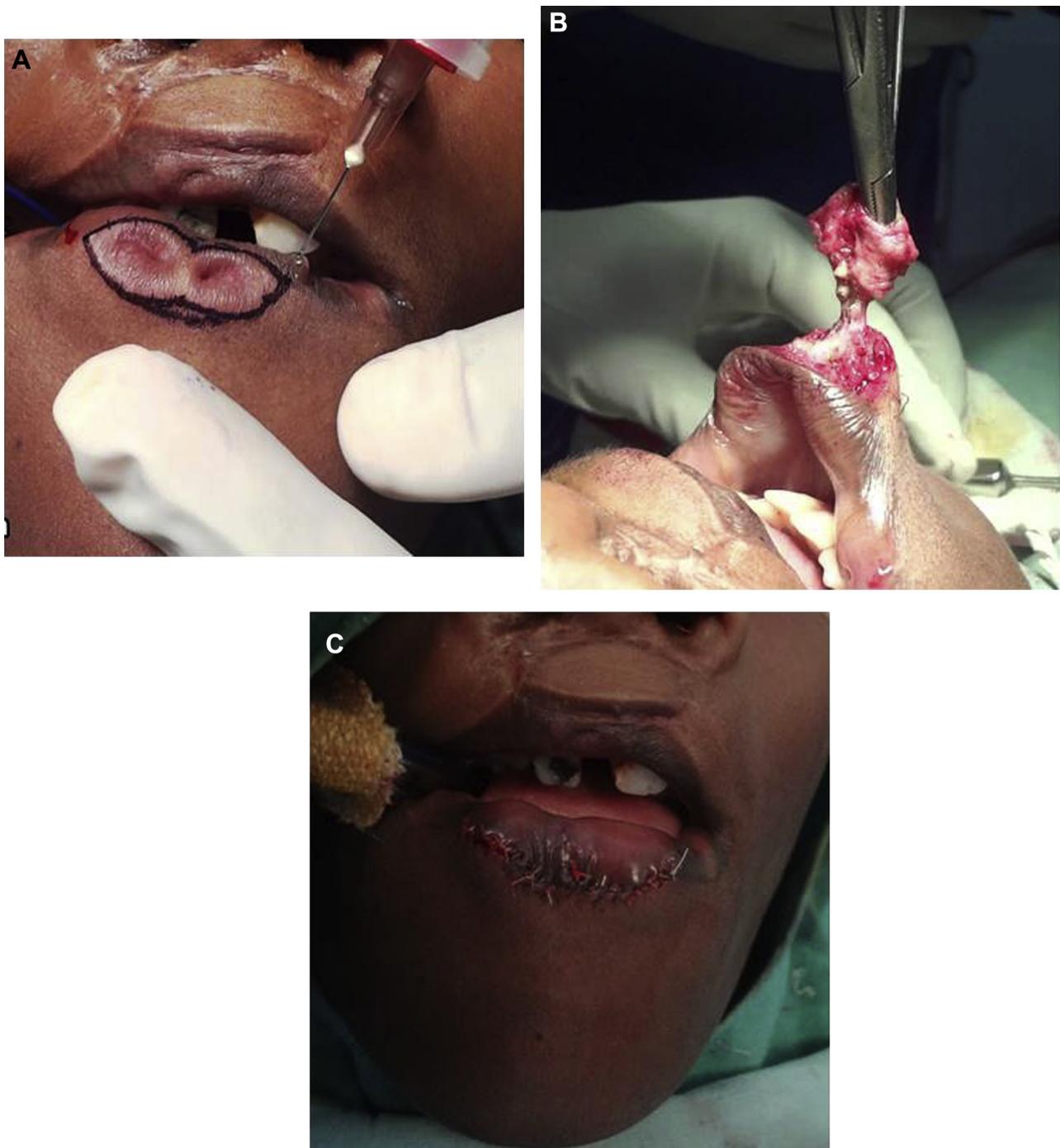


FIGURE 6. Technique of modified routine excision. A, Marking of lip pits. B, Dissection and checking any rupture and accidental leaving behind of the residual tract tissue with a mosquito forceps. C, Closure.

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