

Appendix A: Detailed Surgical Procedure

1. Palate Repair with Bipedicle Flaps and Continuous Circular Closure

The first incision to produce a bipedicle flap on the cleft side started medially of the alveolar crest with an oblique angulation to the bone in order to protect the deciduous tooth buds (**Fig A.1., ①**). The incision stopped anteriorly at the starting point of the alveolar arch curvature, and posteriorly lateral to the hamulus. The mucoperiosteal flap was then lifted bluntly towards the cleft edge. The second incision (**Fig A.1., ②**; **Fig. A.2a., ...**) started on the vestibular side at the bottom of the alveolar ridge to give access for blunt elevation of the nasal mucosa. The elevation extended around the ridge of the palatal shelf to reveal the oral–nasal mucosa junction line, and the third incision was made along this line (**Fig A.1., ③**). The second and third incisions met around the free end of the alveolar ridge.

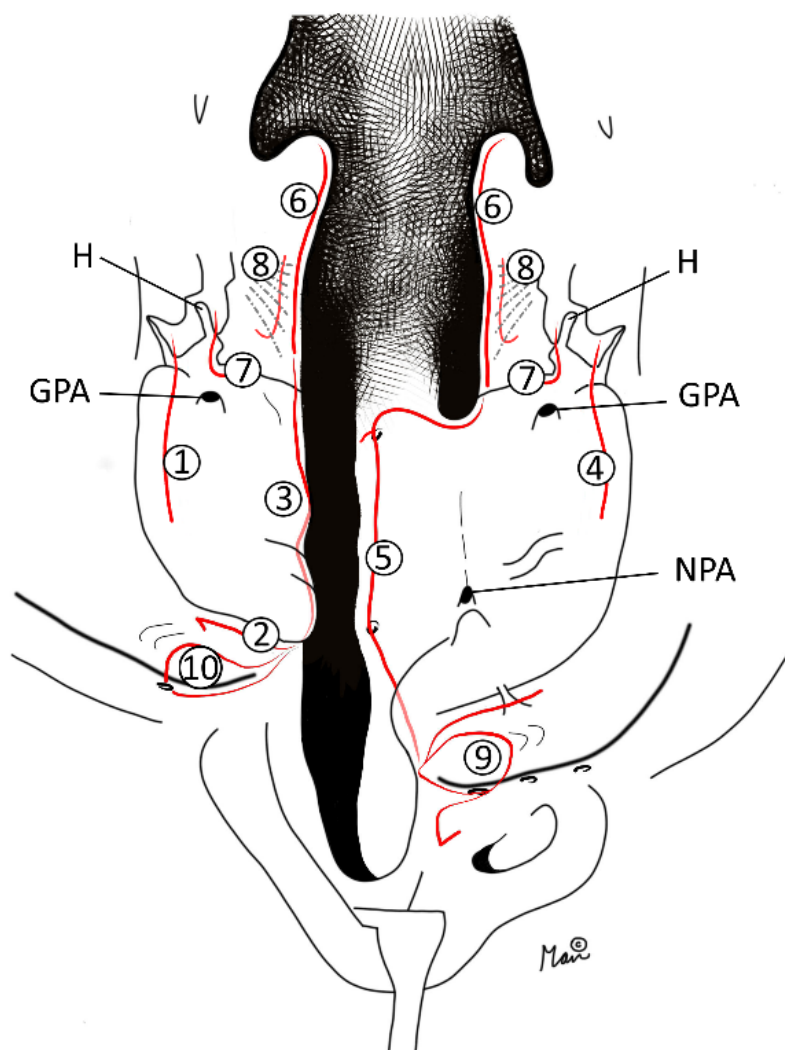


Fig A.1. Sketch of the surgical incisions. The incisions are marked as red lines and labelled in sequential order using circled numbers. H, hamulus; GPA, greater palatine artery; NPA, nasopalatine artery).

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Journal of Cranio-Maxillofacial Surgery, 2021, <https://doi.org/10.1016/j.jcms.2021.07.002>.

The lateral incision made to create a bipedicle flap on the healthy side also stopped at the beginning of the alveolar ridge curvature and lateral at the hamulus (**Fig A.1.**, ④). The mucoperiosteal flap was elevated over the palatal shelf and curved vomer to its posterior end (**Fig A.2b.**, -->). In relation to the mobilized mucosa layers on the cleft side, it was now possible to estimate the position of the anterior (**Fig A.2c.**, →) and posterior (**Fig A.2c.**, -->) vomer incisions to achieve a balanced split of the mucosa to achieve tension-free closure of the palatal vault and nasal floor (compare (Veau, 1931)(Observation 162)). The posterior curve could also form a posterior triangular extension if additional oral tissue was needed at the junction between the hard and soft palates. Anteriorly the vomer incision (**Fig A.1.**, ⑤) passed medially to the incisive foramen and preserved the nasopalatine neurovascular bundle of the nasopalatine artery (**Fig A.1.**; **Fig A.2c.**, ■). The incision then followed slightly cranially the alveolus–vomer furrow – which is also called the innominate sulcus ((Malek, 2001)(p178ff), (Veau, 1931) (p35ff, p448)) – around the alveolar process to the vestibular side. The exact course was selected according to the need to allocate sufficient tissue on either side of the cut for closing the nasal and oral layers. Posterior-to-anterior 4-0 nasal-layer sutures were placed in the hard palate. Using a 5/8-circle round-body 10-mm needle was helpful in cases with a narrow cleft (Serafit, 4/0, FR 10, Ref 6O15132T, Serag-Wiessner, Naila, Germany).

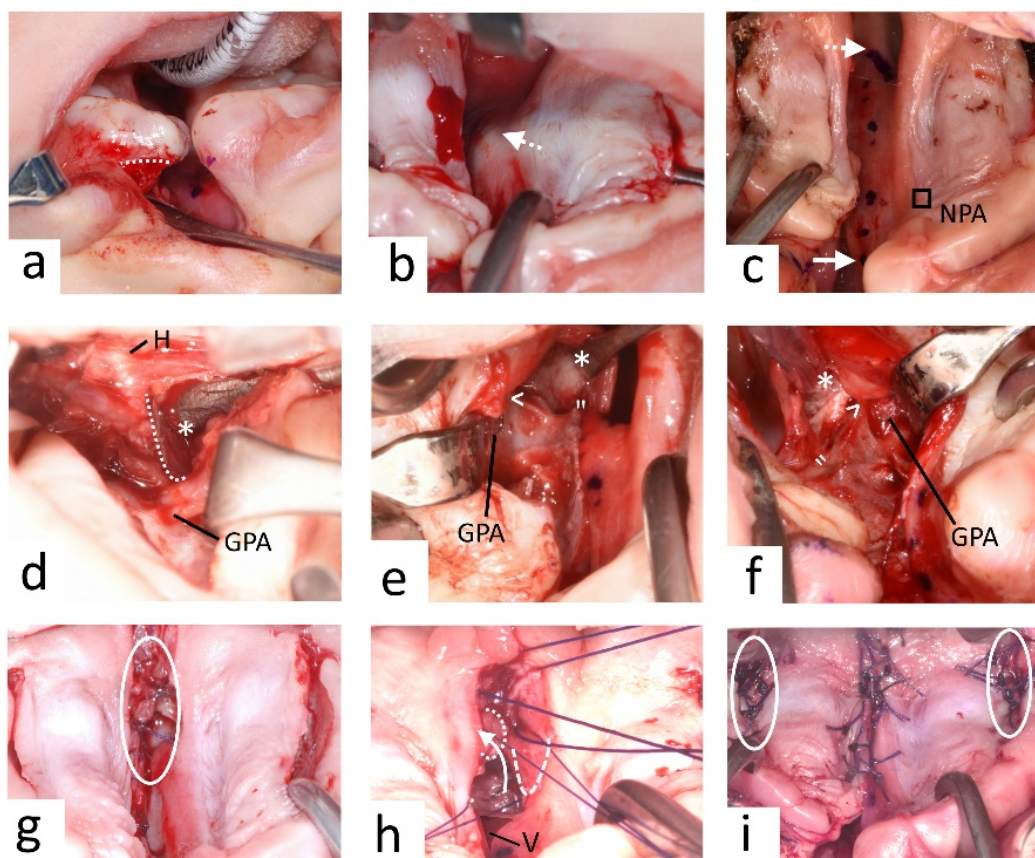


Fig A.2. Surgical steps applied to the hard and soft palates for repair in one single surgical intervention and continuous circular closure . (a) The vestibular approach (···) allowed access for elevating the nasal mucosa over the cleft-side palatal shelf. (b) The mucoperiosteum elevation extended in the medial direction (···>, tip of periosteal elevator) over the vomer and preserved the nasopalatine artery (NPA). (c) In relation to the mobilized mucosa layers on the cleft side, it was now possible to estimate the vomer incision outline from posterior (···>) to anterior (→) to achieve a balanced split of the mucosa to the closure of the oral and nasal layers (■, nasopalatine neurovascular bundle).

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(d) Medial pterygoid detachment with an incision (...) medial to the hamulus (H) to the posterior edge of the palate (*, medial pterygoid plate; GPA, greater palatine artery; ", posterior nasal spine). (e, f) Subperiosteal medialization of the soft palate on both sides (<, fibrous bands behind the GPA). (g) Gapless sutures of the nasal mucosa (0) in the hard and soft palates allowed for primary healing on the nasal side. (h) The anterior portions of the palatopharyngeus muscle and levator veli palatini muscle (---) were dissected off the nasal layer and reoriented (→) into a transverse direction (...). Excess of triangular vomerine mucosa (V)- (i) Gapless sutures over the hard palate allowed for primary healing (0) on the oral side.

The incision made along the soft-palate edges to the uvula exposed the full thickness of the muscle layer but without further dissecting it (**Fig A.1., ⑥**). The approach for periosteal detachment from the medial pterygoid plate was through the medial and lateral incisions of the bipedicle flaps. The periosteal incision ran medially to the hamulus (**Fig A.1., H; Fig A.2d., H**) to the posterior edge of the palate (**Fig A.1., ⑦; Fig A.2d., ...**) and transected the cranial portion of the tensor aponeurosis (Veau, 1931)(p50–61)(Kriens, 1990; Losee et al., 2008) while maintaining the portion at the tip of the hamulus. The blade was stopped by the posterior hard-palate shelf. Access from a lateral direction was preferred when making this incision. After making the incisions, the dissection was preferentially performed through the medial incision of the bipedicle flaps. Subperiosteally elevating along the medial aspect of the medial pterygoid plate detached the bundle of the levator and palatopharyngeus muscles (medial) from the tensor and constrictor aponeurosis (lateral).

The subperiosteal mobilization was directed vertically along the medial pterygoid plate (**Fig A.2d,e,f, ***) to the cranial base, and retrogradely and medially along the nasal side of the palatal shelf around the posterior nasal spine (**Fig A.2e,f, "**) (cf (Veau, 1931)(p170)). Transecting the fibrous bands behind the greater palatine artery (GPA; **Fig A.2d,e,f**) assured complete mobility of the bipedicle flap. A direct view and the surgical access to these fibrous bands were easier through the medial incisions (**Fig A.1., ③** and **⑤; Fig 2e,f, >**) than through the lateral incisions (**Fig A.1., ①** and **④; Fig A.2d.**). The lateral circumference of the choanae was now free on both sides. Subperiosteal elevation avoided damage to the hamular branch of the ascending palatine artery (Huang et al., 1998; Wilhelm, n.d.) that runs between the hamulus and muscle layer.

The soft-palate nasal mucosa suture was started gapless to the nasal-layer suture of the hard palate (**Fig A.2g, 0**) and ran posterior to the uvula. The anterior portions of the palatopharyngeus muscle and levator veli palatini muscle intertwined while running towards the posterior edge of the bony palate (**Fig A.2h, ---**) (Kriens, 1990; Veau, 1931). This muscle area was infiltrated with 0.9% saline and adrenaline (10 µg/mL), and then the dissection commenced 5 minutes later to allow vasoconstriction to occur. This muscle portion (“faisceau musculaire de la fente”, (Veau, 1931) (p50ff)) was then sharply lifted from the nasal mucosa from the anteromedial direction to the posterolateral direction on both sides (**Fig A.1, ⑧; Fig A.2h, curved →**). No or minimal dissection from the oral mucosa was performed. A noticeable release was perceived when the muscle was laterally freed from the rudiment of the velar aponeurosis (Kriens, 1990). The intertwined muscle portion of the palatopharyngeus and levator muscles then swung backward to a transverse orientation in the middle third of the soft palate (**Fig A.2h,...**). The muscle was sutured by using forceps to pull the muscle and assess the sufficiency of its transverse orientation and freedom to

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move. The muscle suture joined identical sites of muscle traction to form a clear anterior and posterior tonsillar pillar.

All muscle stitches (polydioxane suture (PDS) 4-0, 1/2-circle round-body 10-mm needle; Novosyn® 3-0, 5/8-circle round-body 26-mm needle) were first placed and then sequentially knotted. The oral mucosa of the soft and hard palates was then sutured, alternately using single knots and oblique mattress sutures. If an excess of triangular vomerine mucosa (**Fig A.2h**, V) formed at the junction between the hard and soft palates, it was trimmed off. Finally, the lateral surgical access incisions (**Fig A.2i**, 0) were gapless sutured and allowed for complete primary healing on the oral side.

1.2. Combined Alveolar, Lip and Nose Repair

After removing the mouth gag, the two-layer closure in the alveolar cleft area was completed. The nasal-floor suture (**Fig A.3a**, -----) was completed anteriorly to reach the nasal sill. Temporarily compressing the inferior concha (**Fig A.3a**, b; *) with a freer periosteal elevator provided space for suturing. The oral layer between the alveolar ridges was closed using sutures between the vomerine mucosa and the palatal edge on the cleft side (**Fig A.3a**, —) and extended to the buccal side of the alveolar cleft (**Fig A.3a**, Δ). This avoided the use of the lip mucosa for wound closure between the alveolar ridges. The release for ensuring symmetric points of cupid's bow came from a back cut in the columella–labial angle (**Fig A.1**, ⑨; **Fig A.3b**, 0), with an inferior back cut added if necessary (**Fig A.3b**, —). The decision about the necessity of an inferior triangle was postponed until the end of skin closure.

Primary rhinoplasty comprised four steps to construct the four boundaries that form the nasal channel in three dimensions (**Fig A.3c**). The first boundary comprised the medial wall, which was aligned by caudal release of septal cartilage from the bone and mucosa on the healthy side (**Fig A.3c**, +) and refixation to the septal mucosa on the cleft side (not yet performed in the photograph). The second boundary was the roof formed by the depressed nasal dome, which was corrected by separation of both medial alar crura (**Fig A.3c**, > and <), elevation of the cleft-side nasal dome and refixation between the alar crura in the lifted position. The third boundary was the lateral wall consisting of the lateral alar crura, which was moved into a stable convexity and anterior position. Therefore, the lateral alar crura cartilage was bluntly mobilized from the overlying skin and from the upper lateral cartilage as well as subperiosteally lifted to allow for tension-free realignment. Furthermore, the incorrectly inserted nasalis musculature (levator labii alaeque nasi, transverse nasalis and alar nasalis) (**Fig A.3c**, C) was completely released from the inner surface of the ala (**Fig A.3c**, ----) in order to avoid any appearance of tissue webbing. The mobilized nasalis muscle bulge was reoriented and fixed to the opposite alar base (**Fig A.3c**, ⊙), which defined the symmetry of the alar base. The reoriented muscle supported and stabilized the sutured nasal floor (the fourth boundary) and sill. Buried compressing stitches reduced the dead space and defined the tip and ala facial groove and formed a normal vestibular nasal valve. The skin at the nasal sill and columella was sutured.

The procedure for achieving lip closure started with suturing of the oral vestibule. The releasing cut in the oral vestibule mucosa on the healthy side stopped at the frenulum of the upper lip (**Fig A.3c**, d; ->). On the cleft side the cut was also made only just long enough (**Fig A.3d**, →) to close the vestibule mucosa with equal length and mobility on both sides of the cleft, and to construct a deep upper sulcus

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(Fig A.3d., O). Making a temporary stay stitch between the two parts of the lip at the junction between the inner and outer lip mucosae (Fig A.3d., \diamond) facilitated checking that the vestibule mucosa was the same length on both sides and the construction of a deep and supple lip vestibule. The stitches were made strictly between the mucosa layers and did not involve the periosteum to maximize the lip mobility. Muscle stitches were placed from the cranial direction to the caudal direction (PDS 4-0), and the last muscle stitch was very superficial under the mucosa at the junction between the inner and outer lip mucosae (Fig A.3d., \diamond). The aim at the end of surgery was for the two cupid points (Fig A.3e., o) to lie in a position horizontal and parallel (Fig A.3e., ----) to the alar base points (Fig A.3e., Δ). In some cases achieving this required an inferior back cut (Fig A.3b., —) to be cut before the final closure of the lip skin and mucosa. Nasal shape definition was achieved by nostril stenting by a silicon sheet (0.5 mm Silicone Sheeting Custom-Cut™, Invotec®, Jacksonville, United States) and transmural fixation to eliminate dead space. Milk or porridge was consumed immediately after surgery. Arm restraints and feeding tubes were not used, and no blood transfusions were needed. Nostril retainers (Koken CO. LTD, Tokyo, Japan) placed later than 1 week post-operatively were used for 4 months, but some patients or parents refused their use.

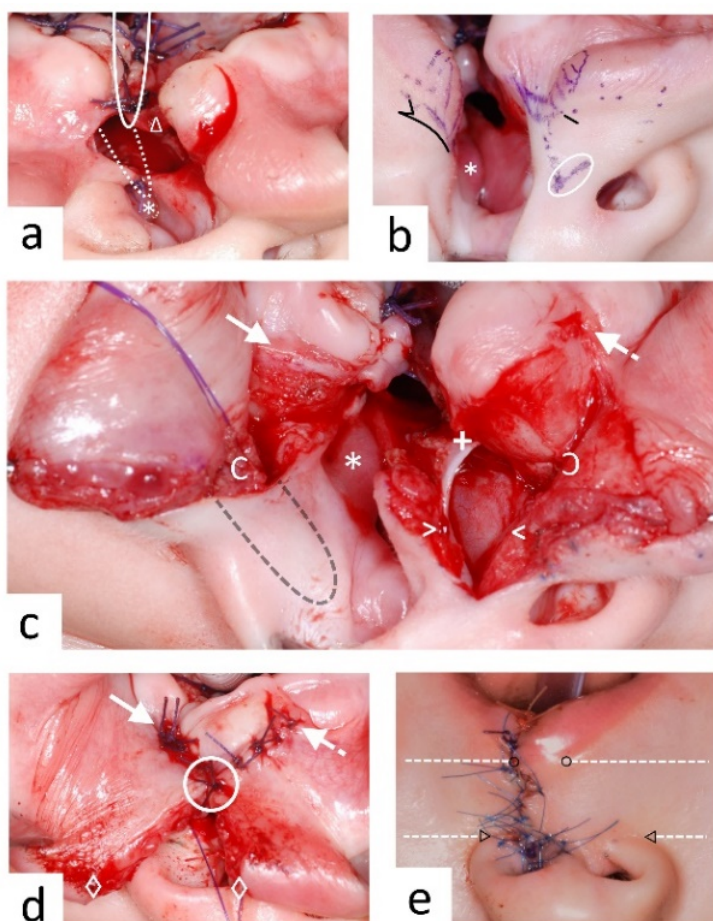


Fig A.3. Surgical steps for combined nose and lip repair. (a) Anterior nasal-floor suture (*, inferior nasal concha). (b) Incision outline on the lip. (c) Primary rhinoplasty (+, septal cartilage; > and <, medial alar crura; C and O, lateral crural cartilage). (d) Closure of the oral vestibule (\diamond , junction between the inner and outer lip mucosae). (e) Closed lip (o, cupid points; Δ , ala base points).

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