



NOMA

Surgical guidelines

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This survey represents not a scientific or general analysis of all surgical procedures, suitable for NOMA surgery, but rather the personal preferred procedures in typical, often seen NOMA-defects. Cases with short- and long-term results demonstrate the difficulties of achieving acceptable results. Nearly all cases and surgical procedures have been performed in cooperation and in hospitals belonging to the “German NOMA-Hilfsaktion” foundation.

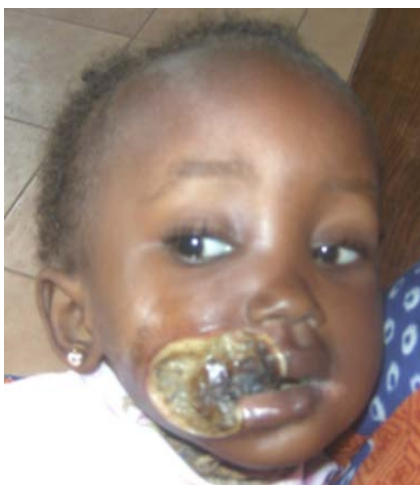
Prof. Dr. Anton Schwabegger, Prof. Dr. Milomir Nincovic, Dr. Harald Kubiena and Dr. Ousmane Issa have been involved in surgery or pictures taking of some of the presented cases.

1.-Introduction

Noma is a type of tissue gangrene in the face that destroys mucous membranes of the mouth, soft cheek tissue and bone structures of the maxilla and mandible. This disease mostly begins like an untypical periodontal infection in severely malnourished children between the age of 2 and 5 years. The gums and lining from the cheek become inflamed and develop necrotizing ulcers). Then the infection spreads to the skin (Pat.1.1) and bone of the cheek and may cause extensive tissue loss in the face (Pat:1,2), general sepsis and death. The disease is caused most often by malnutrition and illness reducing the general health condition of children, such as measles, scarlet fever or tuberculosis. NOMA occurs mainly in the Sub-Sahara region and in other underdeveloped countries.

Healing of gangrene with loss of mucosa and underlaying layers in the cheek is followed by scarring and constriction of the surrounding soft tissue and often of tissue between the remaining maxilla and mandible resulting in trismus.

Trismus , lockjaw or mandibular ankyloses leading to restricted mouth opening , is a frequent consequence of a healed NOMA infection



. Pat:1,1



Pat:1,2

During the healing period the extensive soft tissue loss of the cheek is reduced to a rather small defect of the upper lip on the affected side. This happens by constriction of the surrounding cheek tissue. At the same time the volume of the affected cheek reduces (Pat.1,3)



Pat:1,3

(Pat:1,3) Five years after the onset of the NOMA infection

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1.1 - Surgical correction of a facial NOMA defect requires the following preoperative considerations:

Regarding the patient:

1. The post infection healing period needs to be completed (around 1 year after the infectious tissue damage).

1.2- Classification of tissue defects in Noma

NOITULP classification

(This method was described by Cariou (1986) , developed at the NOMA Children Hospital Sokoto, Nigeria and is now often used)

Rearranging the initials of the affected anatomical units gives the classification title: NOITULP

Affected area:

N - nose

O - outer lining of the cheek

I - inner lining of the cheek

T – trismus

U – upper lip

L – lower lip

P – particularities

Qualification of tissue loss

0 – no loss

1 – up to a quarter lost

2 – from a quarter to half lost

3 – from half to three quarter lost

4 – more than three quarter lost

2. - Cases and surgical Procedures

2.1- NOITULP – T, 0 (Trismus, no loss)



Pat:2,1

(Pat:2,1) This 11 year old patient suffers with trismus and is unable to move his jaw. Constricted scars of the cheek tissue below the skin limits his jaw movements. Trismus is a frequent complication of healed Noma infection and is caused by soft tissue and muscle contraction.

Free flap surgery is recommended to replace the missing oral mucosa and soft tissue.

Since significant complications can occur if this surgery is performed in places like Africa, where conditions are usually less than optimal, we started to treat cases of trismus in Niamey, Niger by distracting the soft tissue and muscles constriction between the jaws with a bone distractor (Image: 1,2) with the aim

of achieving a mouth opening of 3 cm.

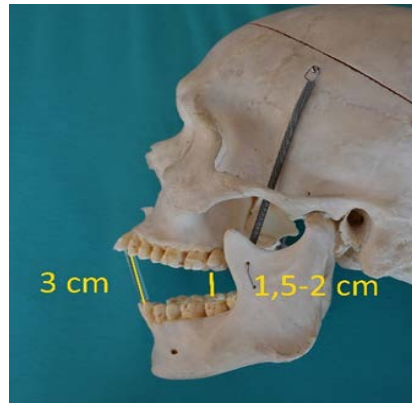


Image 1.1:

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Cadaver skull measurements (Image 1.1) demonstrate that an interincisal distance of 3 cm means that the distance between the molars, where the soft tissue constriction is usually located, is only between 1.5 and 2 cm.

This is the amount of scar distraction needed to achieve an acceptable mouth opening of 3 cm.



Image:1,2

A special bone distractor (image:1,2) is positioned in the zygoma bone, 1 cm caudal of the lateral eye canthus and in the mandibular bone, lateral to the mental foramen (Image 1.3).

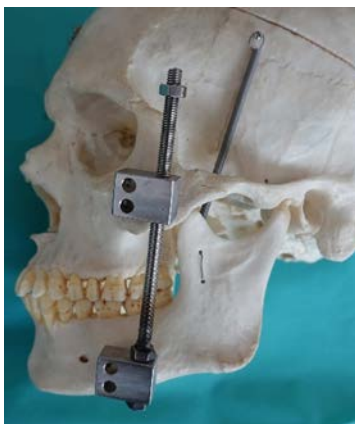


Image:1,3

Until a satisfactory mouth opening of 3cm interincisal distance (image:1,1) is accomplished the distraction of the soft tissue and muscle constriction is done continuously for 1mm per day. Minor complications such as infection in the pin holes were easily managed.

A study analyses of the results of distraction therapy was published in 2020.

(Distraction Therapy to Correct Trismus Following Noma.

Holle J, Kubiena H, Issa OH. J Craniofac Surg. 2020 Mar/Apr;31(2):488-491.)s

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After distraction therapy a medical grade silicon block with the widest dimension of 3 cm should to be used as a spacer between the incisors at night.

(image 1.4)



Image 1.4.:

The results of distraction therapy can be seen in an eight year old patient with trismus following NOMA-Infection before treatment (Pat.2.1) , with the distractor in place 3 weeks after beginning of distraction (Pat.2.2), and 6 month after treatment (Pat.2.3).

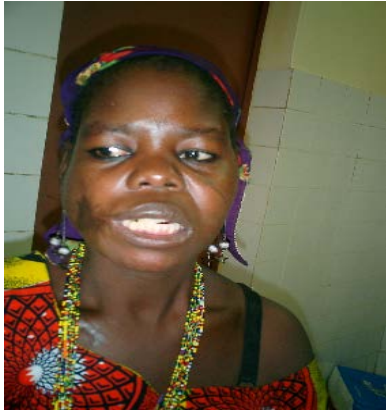


Pat: 2,1



Pat. 2.2

Another patient with trismus before (Pat. 3.1-2) and two years after distraction



Pat:3,1



Pat.3.2

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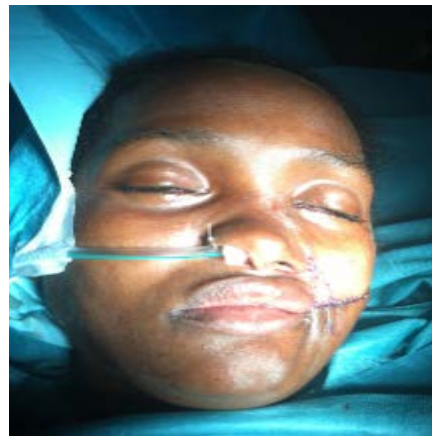
2.2 - NOITULP – O+I,1

(cheek, outer and inner lining, up to a quarter lost)

A primary closure of small defects in the cheek is possible if the symmetry of the face does not change (Pat.4.1). All layers of the face, the mucosa, the cheek muscles and the skin, have to be closed separately



Pat. 4,1



Pat.4,2

Pat. 4.2 : Result at the end of primary surgical closure

Bigger defects need tissue from the surrounding areas to be transposed.



Pat:5,1

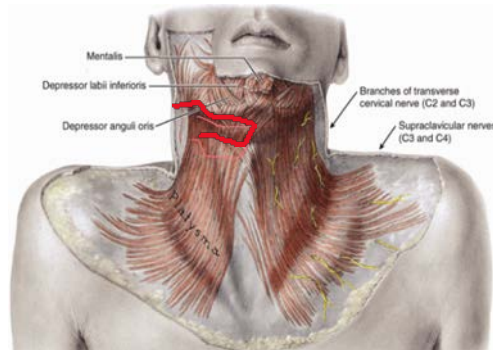


Image 2.1

A rather small defect in the lower part of the cheek can be treated with transposition of a Dermis-platysma-flap from the neck. Image 2.1 : schematic drawing of the dermis-platysma-flap

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The Dermis-platysma-flap can be used to replace missing tissue of the cheek and lip in the area below the level of the oral commissure (Pat.5.1-2, Pat.61-2).



Pat:5,1



Pat:5,2



Pat.6.1



Pat.6.2

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2.3 - NOITULP - O+I, 2

(cheek, outer + inner lining, from a quarter to half lost)



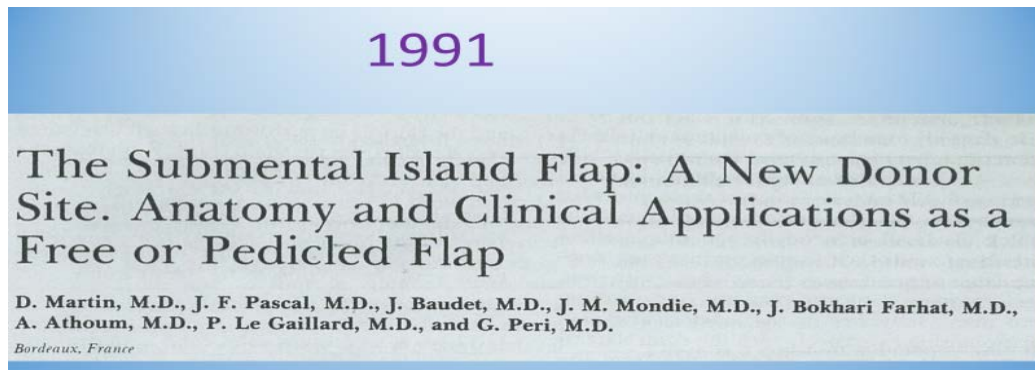
Pat. 7.1



Pat:7,2

Bigger tissue defects in the cheek cranial of the oral commissure (Pat. 7,1-7,2) need a free flap or an extended submental muscle-skin-flap to be transposed in order to close the mucosa and cheek tissue.

The Submental-Island-Flap as described by D. Martin et alii. 1991 may be used in this case.



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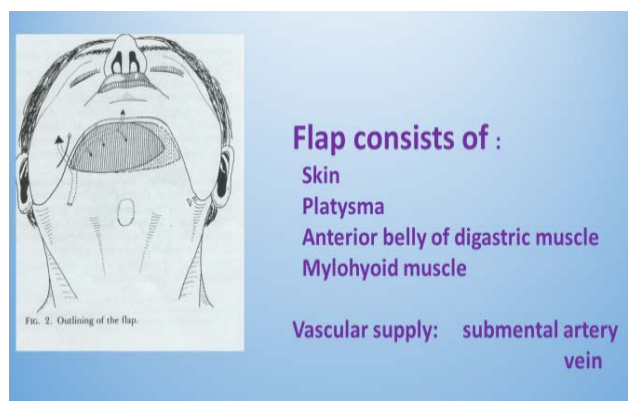


Image 3.1

The vascular supply are branches of the facial artery and vein, the submental vessels. The submental artery is a constant vascular branch that arises from the facial artery. It courses forward and medially between the submandibular gland and the mylohyoid muscle, travels either deep (70%) or superficial (30%) to the anterior belly of the digastric muscle, and terminates behind the mandibular symphysis. Along its course, cutaneous perforators pierce the platysma and anterior belly of the digastric muscles to constitute the subdermal plexus forming extensive anastomoses with the contralateral artery. The submental

vein drains into the facial vein, communicating with both the internal and external jugular veins.” (image: 3,2)

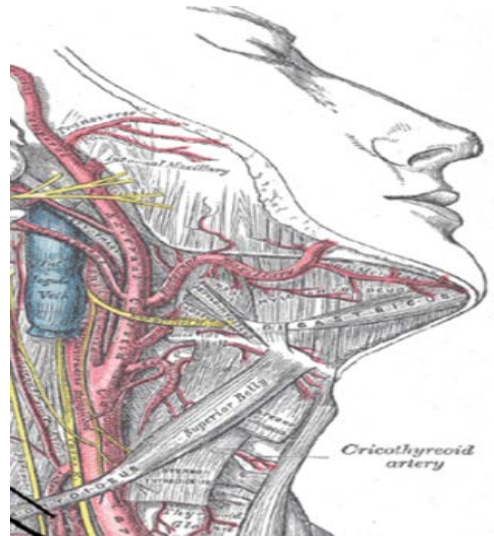
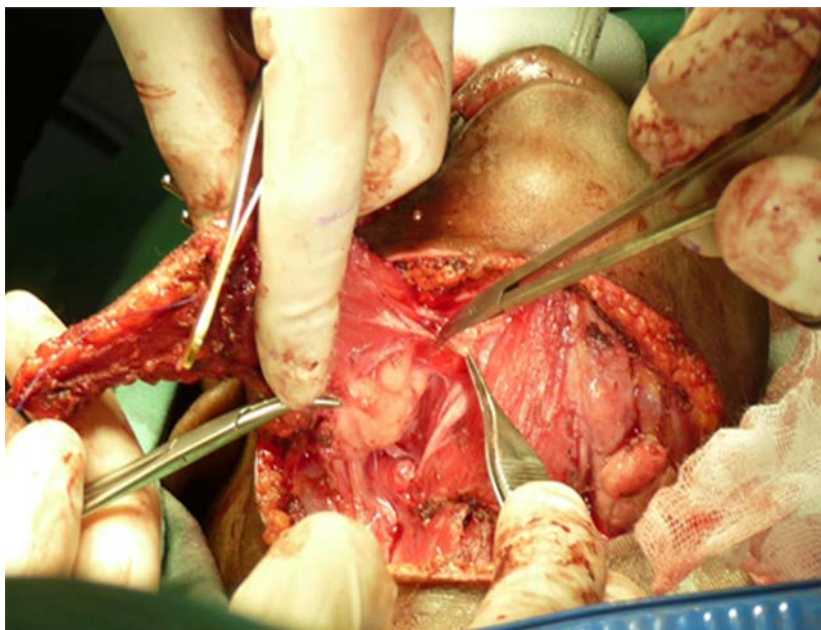


image: 3.2

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Surgery :



Pat.7,3

The Submental-Island-flap including the mylohyoid muscle and the anterior belly of the digastric muscle is carefully dissected. Supplying submental vessels are identified underneath the submandibular gland on the right side (Pat.7.3).



Pat.7.4

The flap is transposed to close the huge cheek defect. The distal part of the flap is folded and used to replace the missing oral mucosa lining (Pat.7.1-7,2).



Pat: 7,5

(Pat. 7.5) Three weeks after Submental-Island-flap transposition with distractor in place

To release the contracted scar tissue and contracted chewing muscle we fixed one distractor on each side between the zygoma and the mandible and opened by the mouth slowly, by distracting one millimeter per day (Pat 7.5).

Another example of the use of the Submental-Island-flap to close the tissue loss on the right cheek of a girl (Pat 8.1). 8 years of age.

Pat:8,2 demonstrates the same child two years later.



Pat:8,1



Pat: 8,2



Pat:8,3



Pat:8,4

Pat. 8.:3-4 The situation in the submental area before and after the flap transposition.

Another example of the useful application of the Submental-Island-flap
(Pat 9.1-2):



Pat 9.1



Pat. 9.2

(Pat:9,1-2) the folded flap has been used to replace the missing mucosa and skin in cheek area..

Concluding: The Submental-Island-flap is a very useful and safe tool (if performed properly) to reconstruct extensive soft tissue loss in the face.

It can easily reach the lower eyelid level and can be used for enoral lining simultaneously. The donor defect is neglectable even in children.



Pat:10,1



Pat:10,2

In this patient (Pat. 10.1-3) part of the upper lip is missing and the remaining rest is retracted. The left ala nasi is cranially transposed.



Pat 10.3

(Pat:10,3) demonstrates the early postoperative situation. Direct adaption of the retracted upper lip parts and transposition of the retracted left nasal ala to improve nasal symmetry have been performed

2.5 - NOITULP- U,1-2

(Upper lip, up to a quarter loss)

Naso-labial-flap transposition

A patient with missing columella, nasal floor and part of the upper lip.

(Pat 11.1-6).



Pat. 11.1



Pat 11.2

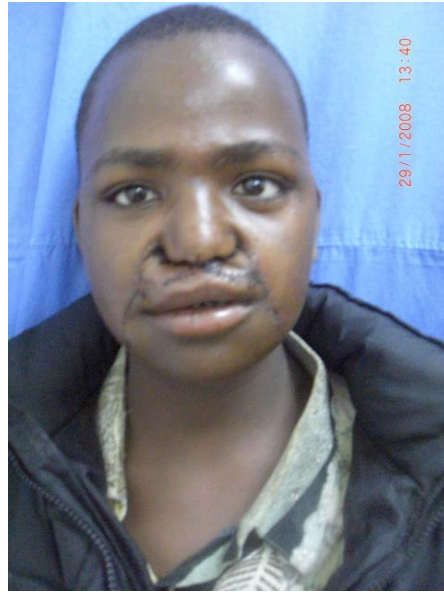
Construction of the upper lip and the nasal floor with transposition of two nasolabial flaps (Pat 11.3). The columella has not been reconstructed



Pat:11, result 10 days after surgery



Pat 11.4



Pat 11,5

(Pat. 11,4-5) the result several month later.

(Pat:12) shows a 14 years old girl with a similar philtrum tissue defect of the upper lip. like (Pat:11)



Pat:12,1



Pat 12.2



Pat. 12.3

Situation one month (Pat. 12.2) and one year (Pat. 12.3) after reconstruction of the missing philtrum tissue with two nasolabial flaps.

Lower lip fan flap transposition for partial upper lip defects

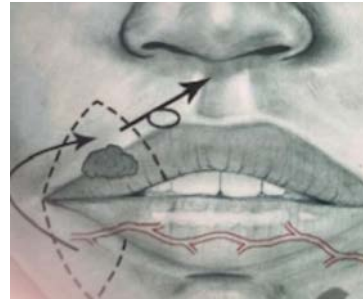
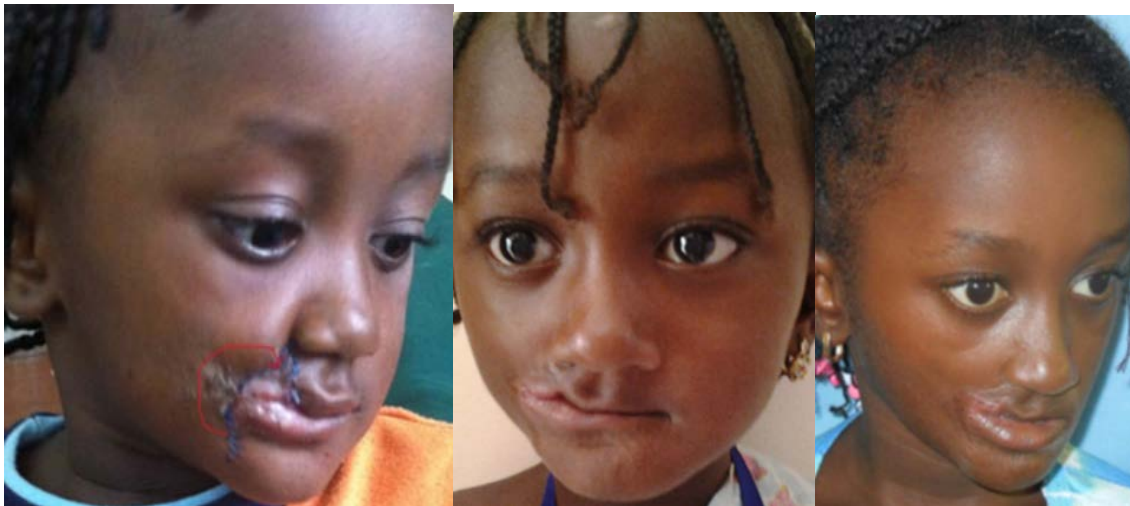


image:4.1

Image 4.1: schematic drawing of the
lower lip fan-flap

Patient (Psat:1,4) with a rather small defect of the upper lip . The right cheek volume is evidently reduced.

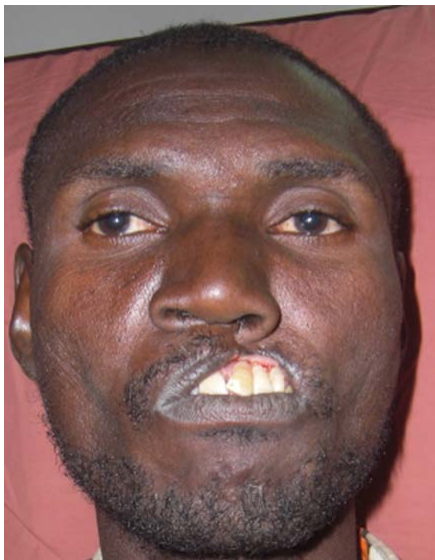
Reconstruction of the upper lip defect with a lower lip fan flap.



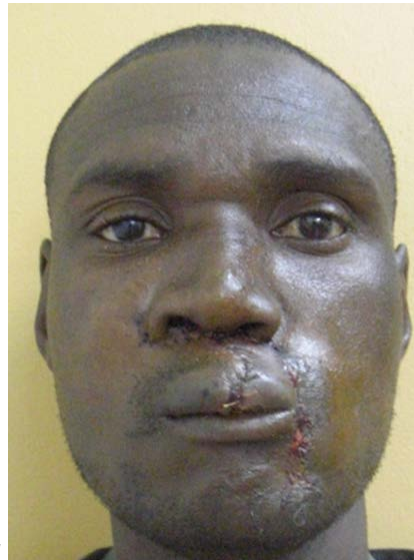
Pat:1.4-6 ,1 week after surgery, 7 years later, 9 years later

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(Pat:14,1-2) One pre and postoperative example for upper lip defect and correction with a lower lip fan-flap.



Pat. 14.1



Pat. 14.2

2,7 – NOITULP – U, 1-2

(central defect of the upper lip)

Abbe-Estlander flap transposition

An Abbe-Estlander flap is used to correct a median defect of the upper lip. The lower lip pedicle can be transected weeks later.

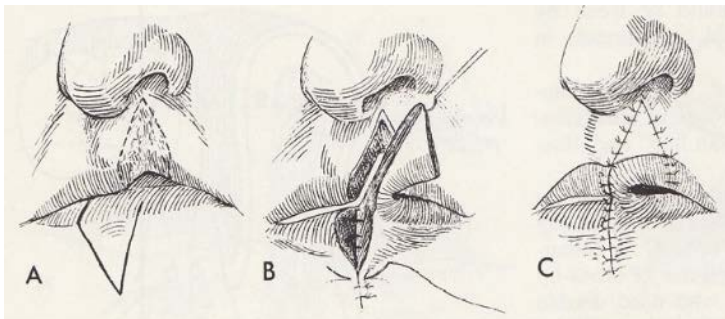


Image 5.1 : Abbe-Estlander flap



Pat. 15.1-2

Flap-pedicle transection 3 weeks later.

2,8 – NOITULP – U,4, N. 2-3

(part of the nose and the hole upper lip is missing)

Total upper lip reconstruction



Pat. 16.1



Pat 16.2

A NOMA -patient with missing frontal alveolar ridge and total upper lip (Pat. 16.1-2). First step of reconstruction is the lip-reconstruction with a free flap. A submental pedicled flap may be possible as well, but other regional flap transpositions are not suitable in young NOMA patients.

2.7 – NOITULP - L, 4 the lower lip is missing)

Total lower lip reconstruction

To perform a total lower lip reconstruction, a free flap is recommended. In some case a submental flap may be acceptable

A female NOMA patient, 15 years of age with loss of the whole lower lip (Pat: 17,1).



Pat:17,1

(Pat 17.1-5). In this case a radial forearm flap has been transferred. A submental flap would also have been possible but seemed less suitable in this case.



Pat:17,2



PPat:17, 3



Pat:17,4



Pat:17,5

(Pat:17, 4-5) Result two years later

3 – NOITULP –Cheek, O+I,3, U+L,1-2
(Cheek, outer +inner lining, from half
to three quarter lost + upper and lower
lip, one -two quarter lost)



Pat.18,1



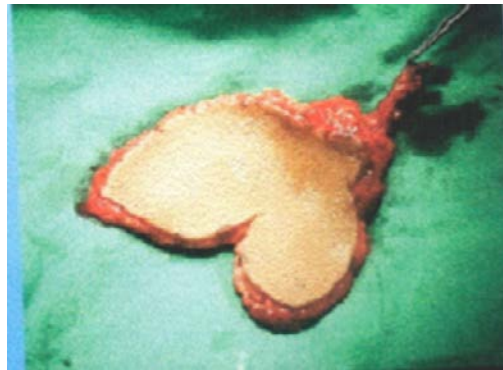
Pat:18,2

(Pat. 18.1-2).NOMA patient.13 years of age with an extensive defect of the left cheek, upper and lower lip.

Reconstruction of the missing oral mucosa and the cheek skin has been performed with a free combined scapula-parascapula flap. A big submental flap may have been possible as well.



Pat:18,3



Pat:18,4

The flap in position two days after surgery (Pat. 18.5). The oral commissure and the lower eyelid require further refinements.



Pat. 18.5

Another NOMA-patient before and after free flap reconstruction of the missing cheek tissue and lip. (Pat. 19. 1-2).



Pat:19,1

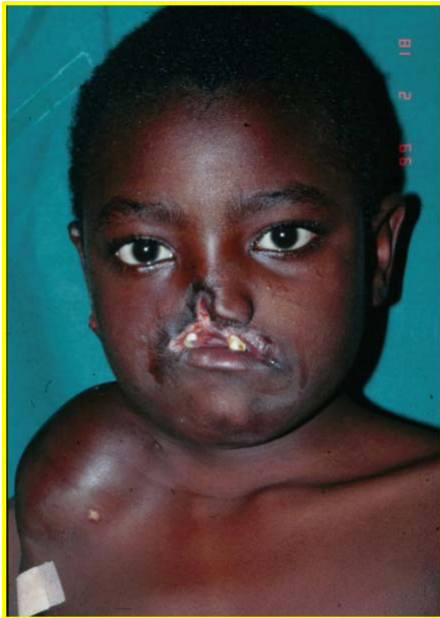


Pat:19,2

3,1 – NOITULP- N. 1

(Partial defect of the nose in children)

A 9 year old female patient (Pat.20,1) with loss off the left nasal ala has been reconstructed with a prefabricated supraclavicular flap. The flap, pedicled on the temporal vessels has been used to reconstruct the upper lip and the right nasal ala. (Surgery performed in Austria). Another option in such a case would a frontal flap be to do the nostril and an Abbe-Eslander-Flap for reconstruction of the lip.



Pat:20,1



Pat:20,2

(Pat:20,2) Result one year later.

The reconstruction of partial nose defects in children (Pat:20,2) should be done with limited dissection of the nose tissue. Don't resurface the whole dorsum as it is recommended for grown-ups. Restricted growth may lead to retraction problems of the whole nose in the years after reconstruction. This may end up with results, worse than before. (Pat: 21:1-4)

Example:



Pat. 21.1



Pat:21,2

(Pat. 21.1-2). A girl of 6 years with a right nasal ala defect before and after reconstruction with a frontal flap to replace the whole nasal skin together with the missing part



Pat: 21,3



Pat:21,4

(Pat 21.3-4). The result 5 years later demonstrates the low growing capacity and the consequential retraction of the reconstructed nose.

3,2 – NOITULP- N,1 +Cheek,I-O2+ U,2 (Combined cheek, lip and nose defects)

A female patient, 10 years of age with extensive tissue loss of the nose, the hard palate the upper lip and left cheek (Pat. 22.1-1).



Pat. 22.1



Pat:22.2

In the first stage a free flap was performed for reconstruction of the cheek and left upper lip (Pat. 22.2)-



Pat. 22.3

In a second step a frontal flap and left lower lip fan-flap were transposed to improve symmetry of the nose and correct the missing part of the lip (Pat.22.3)



Pat: 22,4 Result after several month



Pat. 22.5 Result one year later. Some revisions to improve the result have been performed.

3,3– NOITULP- N,4 + U,4
(Total Nose reconstruction and
simultaneous upper lip
reconstruction)
(performed in Austria)

Total Nasal Reconstruction

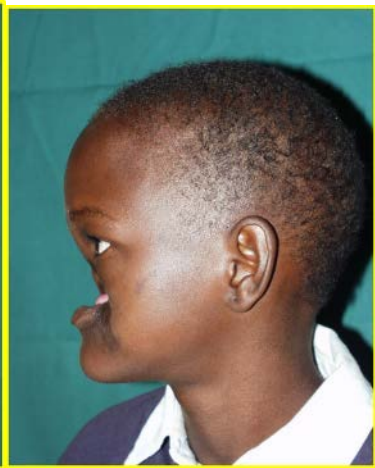
1. Lining:
 - a. Transposition flap
 - b. Free flap
2. Framework:
 - a. Rip-bone
 - b. Rip-cartilage
3. Forehead-Flap:
 - a. Transposition
 - b. Separating the proximal cover flap (3 weeks)
 - c. Division of the pedicle (6 weeks)



Pat:23.1



Pat:23.2



Pat: 23.3

In patient 23,1-3, a girl, 8 years of age, the whole nose, the hard palate and upper lip are missing.

A free flap transfer to reconstruct the nose-lining and rip bone and rip-cartilage were used for the nasal structure. (image:5.1-2, image: 5,3-4)

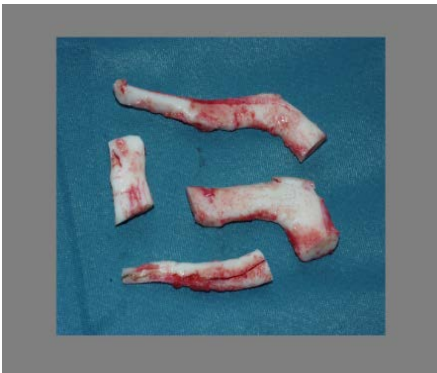


image: 5,1



image:5,2

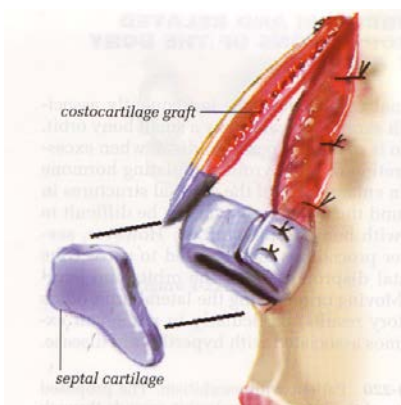


image:5,3

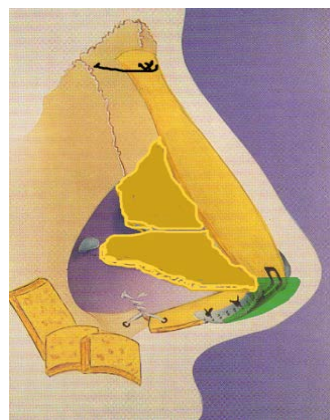


image:5,4

A frontal flap was used to build up the nasal skin cover and a free radial flap for the upper lip. (Pat: 23,4 – 5)



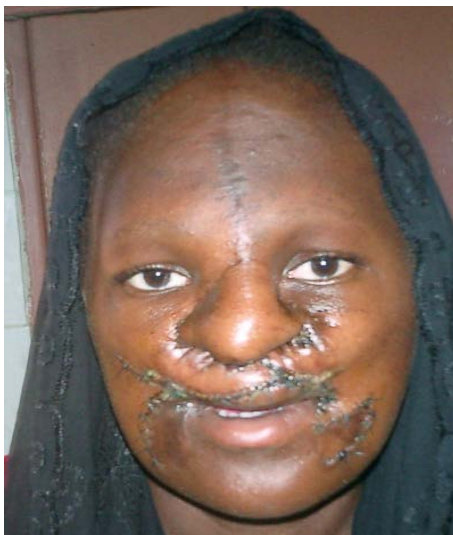
Pat: 23,4



Pat:23,5

Pat: 23.4-5 demonstrates the results one year after surgery, (performed in Austria.)

The result of Pat 23 (Pat: 23, 6-7) 10 years later.



Pat.23,6



Pat:23,7

Some revision surgery of the retracted tissue and scars have been performed in Afrika.

3,4 – NOITULP- N,4+U,4+ cheek-
I,2+O,2-3

(Nose and upper lip are missing
and half of the left cheek
together with the lower left eyelid)

A girl, 6 years of age (Pat:24,1-2) with a huge defect of
the whole face.

(The reconstruction has been performed in Austria.)

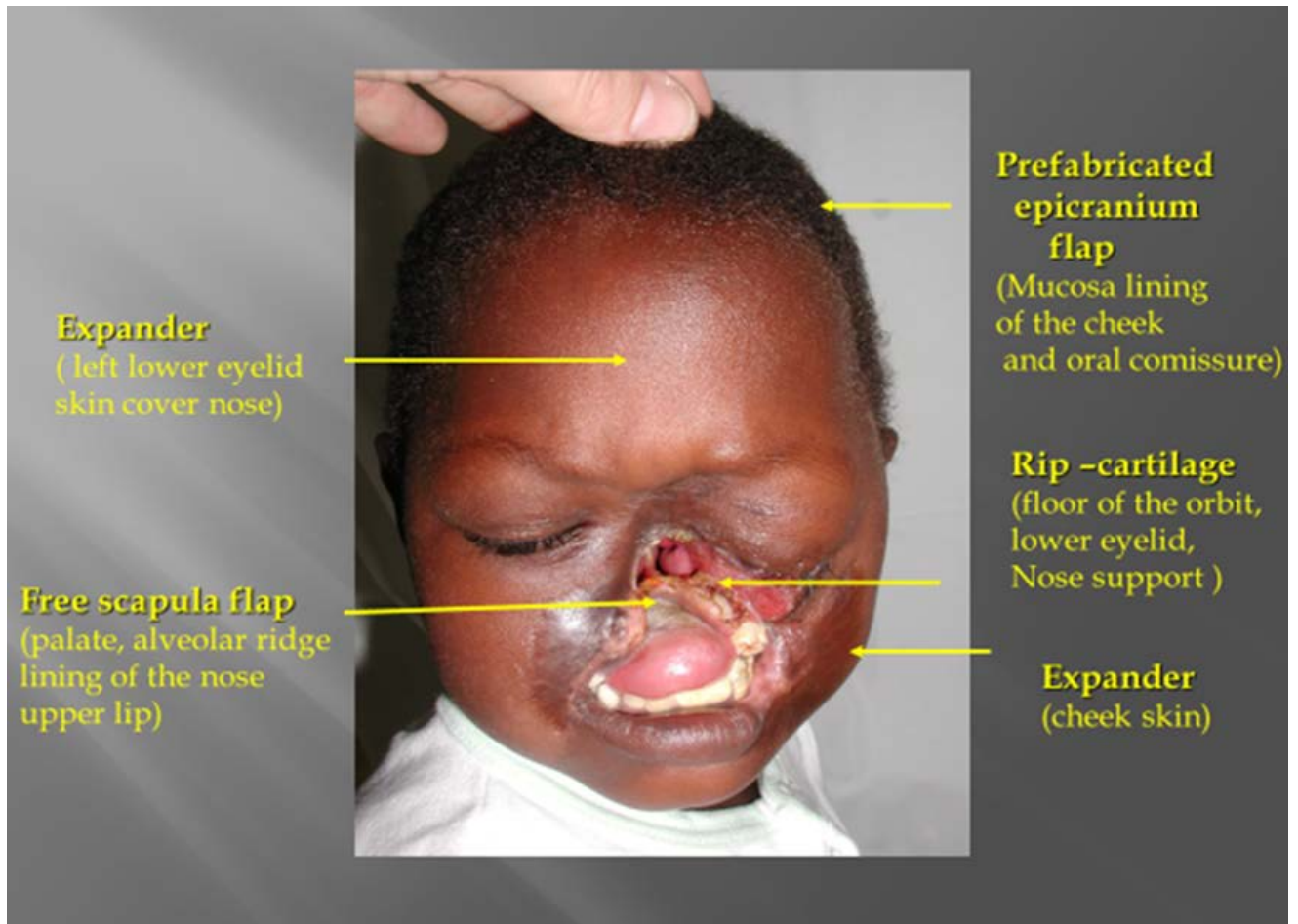


Pat:24.1



Pat:24,2

Scheme of reconstruction (Imag.7.1)



Imag.7.1



Pat:24,3

Pat: 24.3 shows the situation with a free radialis flap used for lining of the nose and an expander in place to prepare the frontal skin for nasal skin cover.



Pat:24.4

(Pat:24.4) The frontal flap is transposed and another free flap is used to build up the upper lip.

10 years later



Pat:24,5

The patient (24,5) 10 years later, became a nurse in an Austrian institution

