Chapter 14

NOMA

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Noma, or cancrum oris, is an overwhelming invasion of micro-organisms of the oral orifice into the face leading to gangrene, sepsis and death. This is a necrotizing fasciitis-like wound. It may be localized to the mouth and oral cavity or it may extend up into the nose or laterally to the ears. It was first described by Carolus Barthus in 1595 in the Dutch textbook Handboek der Chirurgijen. The Greek term “noma” that means meadow/grazing/devouring, metaphorically “quickly spreading,” was introduced in 1680 by Cornelis van Voorde. It is often referred to as “the disease of the poor” and was common in Europe and the United States in the previous centuries but disappeared from the developing world a century ago when even the poorest could feed their children. It, however, reappeared under the atrocious conditions in the concentration camps of World War II and in the Netherlands with the severe food shortage at the time.

Epidemiology
The global incidence, according to the World Health Organization, is estimated at 140,000 people with active disease, mainly children. The mortality rate of noma is 90% and mostly occurs in children between the ages of 1-3 years. 3000 - 4000 children are likely to survive this infection yearly and live with the sequelae of the disease. It is estimated that presently there are 770,000 people who have survived noma and are living with the consequences of the disease. However the data is not completely reliable as “societies only counts its dead when it can feed its living” as it is indeed a disease of the poorest of countries of the world. The areas most affected are East and West Africa, parts of Asia, and Latin America.

Fig 1                                        Fig 2                                                Fig 3
Sequelae of noma in survivors (Courtesy Dr. Gary Parker, Mercy Ships)
Noma is a totally preventable disease; food and vaccination are the critical components in the prevention of the disease.

The Predisposing Factors

Malnutrition is the major predisposing factor. This combined with a concomitant infection particularly measles, chronic malaria, tuberculosis, HIV and typhus, and the third component, poor oral hygiene or stomatitis, set the scene for a devastating and, most often, fatal disease. Malnutrition and other concomitant infections such as measles weaken the immune system. This depressed immune status combined with the high bacterial load of the mouth has the potential to cause an acute necrotizing gingivitis with rapid gangrene of the facial tissue. This may be followed by sepsis and death. An additional predisposing factor appears to be environmental, as noma is seen more frequently in dry, arid, desert-like conditions.

The child’s family often reports that the child was bitten by a hyena but this was not witnessed by anyone. This is very unlikely since the child should have been eaten and killed.

Fig 4
Presented as a hyena bite in childhood, most likely noma
(From The Surgical Treatment of Noma)

Bacteriology

There are no specific organisms associated with noma. It is a polymicrobial infection caused by anaerobic organisms that produce enzymes as well as pro-inflammatory mediators that degrade the intracellular matrix—a mixed infection of oral and extraoral opportunistic pathogens. Spirochetes and species of Fusobacterium have long been suggested to play a role in the disease process. The evidence available suggests that it is caused by the normal flora in the mouth. Although Fusobacterium nucleatum has been implicated in this condition, numerous other anaerobes have been identified. Noma appears to be simply an opportunistic infection in tragic circumstances.

Clinical Presentation

The first signs of the disease are:

♦ swelling of cheek
♦ excessive salivation
♦ foetor ex ore/bad breath/halitosis
within days, dark areas and ulceration appear on the cheeks. Unlike most infections, noma is able to spread through anatomic barriers such as muscle.

extensive intraoral slough follows that typically breaks through skin a week after onset with full thickness gangrene that can extend to the ear, eye, sinuses, nose and to the contralateral side.

Ninety percent of children affected die from sepsis. The 10% of patients that survive are left with varying degrees of facial deformities and can have severe problems with eating, drinking, and speaking. As with leprosy and vesicovaginal fistula patients, victims are often rejected by their own communities. Of these patients that survive the acute infection, 50% will be left with ankylosis of the temporomandibular joint (TMJ) adding a further challenge to survival. Many will die of starvation as a result of the late complications of the infection since they are unable to eat due to the facial mutilation and ankylosis.
Medium and long-term consequences of noma
Healing complete but with severe contracture of the surrounding surviving tissues.
(From The Surgical Treatment of Noma)

Hypertrophy of the coronoid impinging on the zygomatic arch and bony fusion between the maxilla and mandible
(Figures 11 and 12 From The Surgical Treatment of Noma)
Treatment

♦ Prevention
Prevention begins with adequate nutrition, immunization and oral hygiene education. In an ideal world, feeding the hungry and malnourished is the overall answer. Vaccination campaigns against childhood infections and the use of mosquito nets must be intensified. Infectious diseases wreak havoc in poor communities. Noma is a socio-political disease and must be addressed from the governmental level down to the level of each individual family. Effective prevention includes all three interventions: feeding, vaccination and education on the importance of oral hygiene.

♦ Acute phase
Noma patients rarely present to a medical centre in the acute stage of infection due to their poor socioeconomic status. In addition, in most areas where noma is seen, there is a lack of adequate medical facilities and surgical expertise to advise on early management. Survival, however, can be significantly improved with prompt treatment and staged débridement, should the patient arrive early at a sufficiently equipped medical facility.

The following steps should be considered:

- The patient must be stabilized with appropriate hydration and nutritional support. Intravenous fluids must be carefully selected to correct dehydration and electrolyte imbalance as indicated by the patient's clinical condition. A most important aspect of the management protocol is dietary supplementation; the patient is given a high-protein diet. Multivitamin preparations should be given as well. A naso-gastric feeding tube may be necessary for adequate nutritional intake.

- Laboratory investigation: Once the patient is moved to a medical centre, the usual routine investigations, where available, include full-blood count with white blood cell differentials, serum electrolytes, urea and creatinine levels, stool microscopy and cultures, serum protein level and HIV screening. Hypoproteinaemia and anaemia (usually of nutritional origin) are common findings. The stool examination often reveals worms, especially hook worm.

- Address predisposing infections such as measles, typhoid, HIV, etc...

- Administer appropriate antibiotics: As with any necrotizing infection, the patient with noma must be treated for gram positive, gram negative and anaerobic organisms. This mixture of aerobic and anaerobic bacteria include Group A streptococcus, Clostridium perfringens, Fusobacterium, resistant Staphylococcus, and others. If available, high doses of intravenous antibiotics should be used. Drug therapy of choice in our setting is penicillin (ampicillin), gentamycin, and either metronidazole or chloramphenicol to cover anaerobic organisms. Newer drugs, as clindamycin and vancomycin can be used if available and not cost prohibitive. These would replace ampicillin and metronidazole.
• The patients are routinely treated for malaria and dewormed using appropriate antimalarial and anthelmintic drugs.

Fig 13
Patient with acute noma with full thickness necrosis of the cheek, approximately 1 week after onset.
(Courtesy Dr. Abi Boys)

• Effective local management of the ulcer demands regular/repeated débridement and frequent dressing changes. The wound should be kept moist as described in the chronic wound chapter. This may be with saline dressings, honey or even negative pressure therapy if the wound does not communicate with the mouth. Loose teeth and loose bony sequestra must be removed.

• Treatment of sequelae: chronic complications from acute noma include fibrosis and formation of sequestra. The fibrosis leads to ankylosis of the temporomandibular joint with limitation of mouth opening. If a sequestrum is not loose it is allowed to remain in hopes that an involucrum, new bone, will form around it. In the case below, the malar sequestrum is allowed to remain until one is ready to begin reconstruction. It is then removed and final débridement is carried out to prepare the wound for reconstruction. Most sequestrum as this will loosen and fall out in time.

Fig 14
The same patient in Fig 13 after repeated débridement of the wound. The sequestrum has not been removed yet.
(Courtesy Dr. Abi Boys)
**Principles of Surgical Treatment**

♦ **Timing**
Ideally, one should wait for 1-2 years before initiating any reconstructive surgery. Most patients, however, are not seen early and come in many years after the acute episode. The orofacial defect is usually very extensive and requires multiple and highly specialized surgical reconstructions. There is an advantage to delay the surgery so that the patient is mature enough to cooperate in the postoperative period. Delayed surgical reconstruction also ensures adequate tissue for reconstruction, and it also allows the defect to contract and reduce in size.

♦ **Anaesthesia**
With these orofacial defects there is scarring of soft tissues and frequently trismus. Some will have a bony block between the mandible and maxilla or hypertrophy of the coronoid (Fig 36). Such cases will require nasotracheal anaesthesia. A blind nasotracheal intubation can be attempted, but usually a fiberoptic bronchoscope is needed. The use of the latter requires trained anaesthesia personnel. The patient may require a tracheostomy and postoperative ICU care. Surgery under ketamine is not recommended. Without good postoperative tracheostomy care, a tracheostomy should not be done. See the “Regional Anaesthesia” chapter.

♦ **Débridement**
The edges of the defect must be sharply débrided back to viable and bleeding, well perfused tissue. If one plans to use a local turnover flap for mucosal reconstruction, the side of the proposed turnover flap is left intact. Only minimal freshening of the wounds on the face is necessary.

♦ **Temporomandibular Joint**
If there is trismus, the TMJ may need to be released unilaterally or bilaterally. This can be performed though a preauricular incision with care to protect the frontal branch of the facial nerve—CN VII. The reader is referred to plastic surgery or ENT books for this technique.

♦ **Mucosal lining**
In most cases the mucosa will need to be reconstructed. This is always necessary for nasal defects. A common method is to use local skin turned over for mucosal closure. If this good skin is available for turnover, it is ideal. If muscle is used it will epithelialize over. Skin or fasciocutaneous flaps can be skin grafted on the inner or mucosal side. The thin platysma muscle flap from the ipsilateral neck can be used for large defects (Fig 38).
Figure 15: Fully matured wound of patient in Fig 13. Note improvement of the nutritional status. Figure 16 and 17: Defect closed with temporalis muscle flap and skin graft. The muscle on the oral side gradually epithelialized over. (Courtesy Dr. Abi Boys)

Healed appearance but further contracture formation required scar revision and lower eyelid release (Courtesy Dr. Abi Boys)

♦ Closure

Any local flap may be used as seen with the rhomboid flap seen in Fig 7 and 8 and the cervical and Abbé Estlander flaps below. Regional flaps may be necessary such as the forehead, deltopectoral, pectoralis major myocutaneous, temporalis muscle, visor (Fig 32), or supraclavicular flap. Microvascular flaps give an outstanding reconstruction if one expertise in microvascular surgery.
The forehead flap based on the superficial temporal artery is an excellent flap for reconstructing the mid-face. The color match is perfect. Unfortunately, the donor defect deformity is significant. In cultures where head coverings or turbans are worn, the defect would not be conspicuous. The case below (Fig 28, 29) was a hemangioma in a young girl. She came from a thousand kilometres away. There was uncertainty about her return, and there was a high likelihood she would wear a turban in adulthood based on the culture in her home country. This shows how a forehead flap can provide excellent reconstruction and it has been used extensively in noma treatment.
Forehead gives an excellent color match for reconstruction of the midface.
Donor defect is skin grafted on periosteum with poor cosmetic result.
(This is not noma but shows the excellent color match with a forehead flap)

The **visor flap** can be used for large defects. This is a bilateral superficial temporal artery flap turned down as a visor to reconstruct upper and lower lips. Lip mucosa can be reconstructed with **cheek turnover flaps** which should be delayed twice in most cases. In such a case, the surgeon raises a portion of the flaps at the first stage. At the second stage, the full flap is raised. Finally, the flap is turned over to reconstruct the lip mucosa, and the forehead flap is used to reconstruct the skin of the upper and lower lips. In the case below, the forehead was expanded with hopes that the hair bearing skin would create the upper lip and hairless forehead skin would be for the lower lip.

Large defect with trismus: Turnover cheek flaps used to reconstruct mucosa. **Visor flap**, bilateral superficial temporal artery flaps, elevated after forehead expansion. In this case, the expander became infected and had to be removed, leaving only hair bearing tissue for the flap. Ideally, the expander would have expanded both hairless forehead skin for lower lip and hair bearing skin for the upper lip and mustache. The forehead flap would also cover the defect left by the cheek flaps which were turned over for mucosa.
Pre-operative ankylosis and flattening of cheek: coronoideectomy to release ankylosis and temporalis flap (blue arrow) and calvarial bone graft (green arrow) used for reconstruction.

This patient with right cheek noma and trismus required nasotracheal intubation with a fiberoptic scope, removal of the bone block (arrow above) between maxilla and mandible, and TMJ release through bilateral preauricular incisions. Mucosa was reconstructed with a platysma flap (donor defect green arrow and flap prior to insetting blue arrow) and the cheek reconstructed with a deltopectoral flap. The challenging fiberoptic nasotracheal intubation took 2 hours and the removal of bony block another 2 hours.

Same patient as in Fig 36 with flaps in place Fig 39 and postop in Fig 40 with 20 mm of mouth opening.

♦ Other defects--Nose
As mentioned earlier, nasal lining is critical with any nasal reconstruction. Bone grafts from the dorsum may be taken from the calvarium, rib or iliac crest. In the chapter on “Facial Trauma and Reconstruction,” there are guidelines for nasal reconstruction. Mucosal lining for the nose may be obtained by local
turnover flaps from the cheek (within the nasal labial fold), nasolabial flaps, or forehead flaps. In some cases, the forehead flaps used for external reconstruction may be skin grafted as the primary step and then turned down in the second stage for both mucosal and external reconstruction. Cartilaginous reconstruction for the alar rim and columella may be harvested from the ear or nasal septum. After the forehead flap is turned down for external reconstruction, a second stage in a week to ten days is done to defat the flap in order to give a better cosmetic result. This is done before the flap is divided. Please refer to the chapter above or to a chapter on nasal reconstruction in a plastic surgery or ENT text.

![Fig 41](image1.jpg) ![Fig 42](image2.jpg)

Noma with nasal loss. Nose reconstructed with a turnover midline forehead flap for mucosal lining, calvarial bone graft for nasal dorsum, right lateral forehead flap based on contralateral superficial temporal artery, Mustardé cheek flaps to reconstruct cheek, and lateral lip advancement. Maxilla was not reconstructed. Secondary division and inset of lateral forehead flap

**Summary**

Noma deformities are some of the most challenging plastic surgery problems in Africa. The disease process begins in childhood, but the patients present later in childhood or even as young adults. Most victims of noma die during the acute phase of the disease, not having access to adequate medical care. Those who do survive are left with these severe facial deformities. When the patient with chronic sequelae understands his condition and when the surgeon feels the patient and family will be compliant, then operative treatment may be considered. Ideally, reconstruction would be completed prior to school age and socialization. However, delayed presentation usually precludes this possibility. Microsurgical flaps, as the radial forearm flap, have been used by microsurgical teams to give an excellent reconstruction.

**Sources:**

The Surgical Treatment of Noma by Kurt Bos and Klaas Marck. (Kurt Bos and Klaas Marck, founder and President of Dutch Noma Foundation, have written this
publication aimed at surgeons and anaesthetists working with Noma as part of a surgical aid programme. The book offers a very in-depth description of the procedures used in the reconstructive surgery.)