

HISTORY

THE HISTORY OF NOMA DISEASE AND ITS SURGICAL TREATMENT



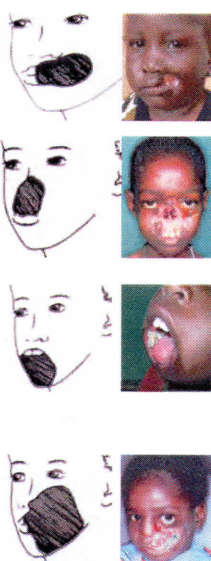
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It is not uncommon for plastic surgeons engaged in humanitarian aid in poor countries of Africa, South America or the Asia-Pacific region to encounter children affected with severe facial disfigurement due to a disease known as noma. This name was given to this particular disease in the 18th century and derives from the ancient Greek word νομή, meaning feeding from the devouring of pasture by herds and by extension a spreading lesion or a devouring ulcer.

Several other names like “water kanker” or “cancrum oris” have been abandoned, as they might suggest cancer-like lesions. Although little is known about the origin of the disease (see text box), for those who have seen a few cases who survived the initial stages, the diagnosis can be made almost immediately by the typical appearance of the residual lesions. Initially located in the gingiva, the sloughing of the necrotic tissues leaves a defect which always affects the mucosa, but may spread to the soft tissues and bones of the whole middle and lower third of the face.

Four types of sequelae are usually encountered:

- Type I is a localized cheek defect, sometimes involving part of the lips.
- Type II is a centro-facial defect, which affects mainly the nose and upper lip. It may involve the whole palate, nasal septum and nasal bones.
- Type III is a midline defect of the lower third of the face. It can affect only the lip or the chin or presents itself as a complete loss of the mandible.
- Type IV is a major cheek and maxillary defect that sometimes can spread to the orbit and the nose and destroy the whole hemiface.



In addition to the disfigurement, full thickness lips and cheek defects lead to constant drooling of saliva and mouth feeding is

always a difficult task. Another important feature of noma sequelae is the contraction of the tissues following the initial necrosis and the sloughing of the eschar. This retraction induces a forceful closure of the mouth, a lockjaw that should be differentiated from a TM joint ankylosis or trismus, that require different treatments.

Once the healing process is achieved, reconstructive surgery is usually delayed for one to four years depending of the type of lesion, the age of the child and the medical facilities. If in some minor cases it can be achieved by one operation (i.e. Estlander or submental flap), staged reconstruction is often indispensable. Complex defects represent the most difficult challenges in reconstructive plastic surgery of the face. They may necessitate the combined use of cranio-maxillo-facial procedures, free composite flaps, nasal reconstruction with forehead skin expansion, bone distraction and the whole panoply of local and distant flaps in multiple staged operations. Moreover, as this surgery is usually performed during childhood, a long-term follow-up is necessary leading sometimes to other corrections at a more advanced age.



The History

Although the people (mostly children) affected by noma are nowadays almost exclusively found in tropical or subtropical regions, the disease is not related to climate as demonstrated by the history showing that it was quite common in Europe up to the 19th Century. Even during the Second World War, a series of Gypsy children suffered from noma in the concentration camp of Auschwitz. Interested by this disease, which he thought was genetically or racially induced, the notorious doctor Josef Mengele approached a Jewish prisoner pediatrician, Professor Berthold Epstein, proposing that, in return for “an extension of his life,” he helps to carry out research that Mengele could publish under his own name. Epstein was granted a day to think the

matter over. He was indignant that Mengele “wanted to rob him of his soul.” But his colleagues convinced him that an attitude of this kind at a distance of three hundred meters from a crematorium was far from being realistic and that, under the pretext of “scientific research,” he could do a great deal of good for these inmates. Epstein then proposed a research project on the treatment of noma. Mengele agreed, and a “Noma ward” (*Nomaabteilung*) was established where forty-five to seventy children were kept and given a special nutritious diet, vitamins and sulfa drugs, as requested by Epstein. The children were photographed before, during and after treatment and SS doctors were brought to the ward to observe the work. A few of these children made good recoveries, the others died and their heads were sent for postmortem examination. The results of this “experiment” were never published. Having survived the war, Epstein later testified in the Soviet war crimes trials on genocide. He lived in Prague for the remainder of his life, serving as chair of the city Hospital pediatric clinic until his death in 1962. The excessively high number of children affected by noma in Auschwitz clearly shows the link of this disease to extreme malnutrition and debilitation.

If numerous papers are nowadays published on noma by plastic and maxillofacial surgeons, we are indebted to the British plastic surgeon Michael N. Tempest for having drawn the attention of our specialty to this disease in 1966. During his three and a half year stay in Ibadan, Nigeria, he treated more than 300 children and related his experience in a classical essay under the title *Cancrum oris*. He also gave a historical introduction and description of the state of the art as regards the etiology and treatment of this mutilating disease 50 years ago. In 2003, K.W. Marck, a Dutch plastic surgeon, published a book entitled *Noma the face of poverty* with an extensive chapter on the history of this disease, but failed to find the first descriptions of this pathology in antiquity.

Although the incidence of this impressive illness that devours beauty and life is relatively rare, it has been described a number of times throughout the ages. Already in the Egyptian Smith papyrus (1650 BC), translated by Breasted, one may find the following account corresponding to Type I sequelae.

Case 15: A cheek wound

Title: Practice for a perforation in his cheek.

Examination and prognosis: If you treat a man for a perforation in his cheek and you find a swelling on his cheek risen, black, and gone off, then you say about him: «One who has a perforation in his cheek: an ailment I will handle».

Treatment: You have to bandage him with alum and treat him afterward with oil and honey every day until he gets well.

In Hippocrates (400 BC), we find a typical description of a Type II sequela:

Τὼ παιδίῳ τῷ φαγεδαίνωθέντι...

“In the child suffering from phagedenic affection, the teeth of below and above, in front, have fallen down, the bone being eroded. The exposure of the palatine bone induced the collapse of the nose in the middle; the fall of the front teeth above induced the flattening of the tip of the nose.”

During the Middle Ages, physicians like the Arab Avicenna (980-1037) or the Flemish Jehan Yperman (1260-1301) mention in their books this spreading ulcer starting from the gums. The first author to describe noma as a clinical entity was the Flemish surgeon Carolus Battus in 1590 in a four-page chapter of his book *Handboek der Chirurgien*:

“These ulcerations in the mouth of children, beginning as a little spark, can devour the lips, the jaws and even the tongue, leaving in the course of a month the children horribly disfigured if they stay alive.”

During the next two centuries, similar descriptions of the disease can be found in the English, French, Dutch, Swedish, German and Italian medical literature, accompanied sometimes with suggestive and dramatic illustrations.

First Operations

While working in Switzerland as a protestant refugee, the famous surgeon Pierre Franco (see ISAPS News: vol. 8/1, 2016) described in 1561 very precisely how he was able to cure a man suffering most certainly from noma sequelae. His account is a model surgical report as it includes diagnosis, previous medical history, methods used for surgery (skin and mucosal flaps), follow up, functional and cosmetic result.

“A certain Jacques Janot Savagny who lived near Neuchâtel-on-the-Lake in Switzerland was afflicted with a cantabris (ulcer) which penetrated his cheek, so much so that the largest part of the cheek had been destroyed. The ulcer was so large and round that a goose egg would fit through it. The two mandibles showed no flesh and there were no teeth in the ulcerated side of his face.

The patient had to wear a dressing and a leather strap. Even with this bandaging he still could not prevent some of his food and beverage falling through the hole in his cheek. As a result, he did not dare to socialize; especially since the saliva was always oozing through the wound. During the seven or eight-year period of his disease, he searched for someone who would be able to cure him. However, when I first saw him, he had not yet found anyone willing to touch it, and had been told that he was incurable, for flesh could not be generated, nor could the edges of the hole be brought together.

He asked if I could cure him. I told him I would heal him with the help of God. I placed the patient against the trunk of a tree and attached his thighs firmly to it. I advised that he be tied well. I had my *cauterres* nearby in a basin on a fire, ready to be used.

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I took a small razor and cut all around the edges of the ulcer. I split the skin near the eye, towards the ear and towards the inferior mandible as quickly as I could, while making sure not to cut too far (so as not to create further damage and cut transverse muscles). I then cut the flesh around the ear and eye as much as I could, yet still the edges could not be joined. Next I cut the flesh inside the cheek sidewise and lengthwise, lacerating it, paying attention not to reach the outside area because one should not cut the skin. Where there was bleeding or a vein cut-off, I cauterized with the *cauter* mentioned above. I pulled the edges together and managed to join them. I then immediately applied seven needles in the same manner as for cleft lips. After four or five days, three of them fell out and did not need to be replaced because the edges were pulling. I used dressings and little cushions with bandages around the wound to draw the flesh to the center so that the edges would not break and the remaining needles would not fall out. I also applied a cloth soaked in *oxicatron* and used my *restraintifs* in a way that they would not interfere with the needles.

Within fourteen days, I had cured him. Some said that his cheek would pull and he would not be able to open his mouth; in fact, the flesh was abundant enough that he even grew a beard, which made the wound hardly noticeable."

Since Franco's time, one has to wait until the end of the 17th century to find other attempts to reconstruct the lips and the cheeks in noma cases. The most comprehensive reports on various aspects of the disease and their surgical treatment can be found in Von Brun's *Chirurgische Atlas* (1857), with a series of very realistic illustrations.



Conclusion

Although the disease was and is still infrequent compared to epidemics like leprosy, tuberculosis or plague, noma has haunted generations of people by the terrible disfigurement it may produce. The fact that it has disappeared nowadays in the wealthy countries should not let us forget that thousands of children are still possible victims among the undernourished populations of the globe. In addition to their functional and cosmetic burdens, they are sometimes considered as a malediction for their family and deprived of schooling and social relationships. Plastic surgery has a lot to offer to these children if fully trained surgeons in the cranio-maxillo-facial field provide it.

Bibliography

Marck KW: *Noma, the face of poverty*. MIT-Verlag GmbH, 2003
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Noma is a gangrenous stomatitis causing high mortality and devastating facial lesions with severe functional and aesthetic consequences, affecting mostly children age 2 to 10. It starts from a gingival lesion that spreads to the underlying bone. The corresponding facial region develops edema and becomes necrotic, leading to the destruction of large parts of the soft and hard facial tissues. Findings from observational studies suggest several risk factors such as poverty, stunting and malnutrition, low birth weight, poor sanitation, endemic infections (measles, malaria, AIDS), high number of previous pregnancies in the mother, poor oral hygiene and proximity of livestock. Although these factors might play a role in a cascade of events leading to a catastrophic derailment, this disorder is essentially an opportunistic infection to poor health status. In 2013, the Gesnoma (Geneva Study Group on Noma) published the result of a six year, prospective, matched case control study in a rural region of Niger. The sample consisted of 82 children younger than 12 years presenting a typical initial noma infection, that is exposure of the maxillary bone, edema or initial facial necrosis. For each case of noma, 4 children of similar age from the same village were included in the study and had similar investigations. Structured interviews took place with the children's family members, demographic, clinical and nutritional data were collected, as well as viral and microbiological samples of gingiva and blood. Acute necrotizing gingivitis (ANG) appears to be a precursor of the lesion. Analysis of bacterial composition showed that the flora present in the lesion and in the healthy mouth harbor fundamentally the same bacterial communities, but differ in the prevalence of a limited number of species. *Prevotella intermedia* and members of the *Peptostreptococcus* genus are associated with both diseases (noma and ANG). This study did not provide evidence for the existence of one bacterial pathogen as a cause for the disease, but strongly suggests that an altered oral microbiota increases the risk of the disease, independently of socio-demographic and environmental factors.

Baratti-Mayer D, Gayet-Ageron A, Hugonnet S, François P, Pittet-Cuenod B, Huyghe A, Bornand JE, Gervais A, Montandon D, Schrenzel J, Mombelli A, Pittet D; Geneva Study Group on Noma (GESNOMA) : *Risk factors for noma disease: a 6-year, prospective, matched case-control study in Niger*. Lancet Glob Health. 2013 Aug;1(2): 87-96.

The history of noma: Illustrations

1. Type I defect
2. Type II defect
3. Type III defect
4. Type IV defect
5. Example of type II reconstruction of nose and upper lip with various skin and mucosal flaps and calvarial bone graft to the nose.
6. Von Brun's atlas: type I sequel
7. Von Brun's atlas: type IV sequel
8. Von Brun's atlas: bone loss in noma
9. Von Brun's atlas: surgical cure of a cheek fistula

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