The Progress on 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 actually as noma. This name was given to this particular disease in the eighteenth century. It derives from the ancient Greek word νομ?, meaning the 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, for those who have seen a few patients who survived the initial stages, the diagnosis can be made almost immediately by the typical appearance of the residual lesions.

RECENT INVESTIGATIONS

Noma is a gangrenous stomatitis causing high mortality and devastating facial lesions with severe functional and esthetic consequences, affecting mostly children age 2 to 10.[1] 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 6-year, prospective, matched case–control study in a rural region of Niger.[2] The sample consisted in 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 patient with 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 *Peptosreptococcus* genus are associated with both diseases (noma and ANG). This study did not provide evidence for the existence of 1 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.

The sequels of noma usually encountered can be classified in 4 types:

Type I is a localized cheek defect, involving sometimes part of the lips (Fig. 1).
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 (Fig. 2).
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 (Fig. 3).
Type IV is a major cheek and maxillary defect that sometimes can spread to the orbit and the nose and destroy the whole hemiface (Fig. 4).

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 sequel 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, which requires a different treatment.

Once the healing process is achieved, reconstructive surgery is usually delayed for 1 to 3 or 4 years, depending of the type of lesion, the age of the child, and the medical facilities. If in some minor patients, it can be achieved by 1 operation (ie, 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 (Fig. 5). 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.[3–6]

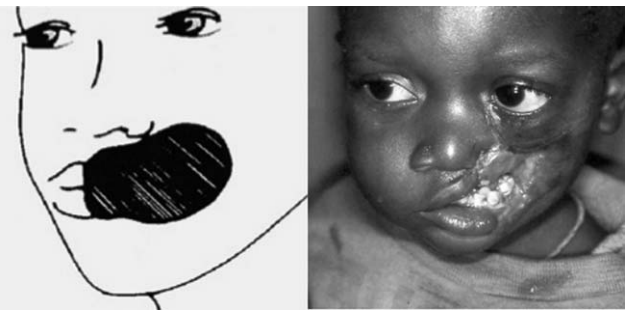

FIGURE 1. Type I defect.

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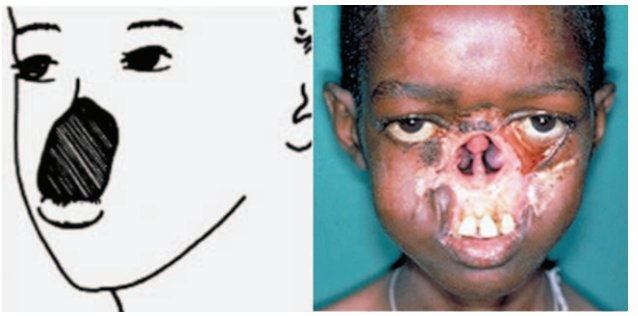

FIGURE 2. Type II defect.

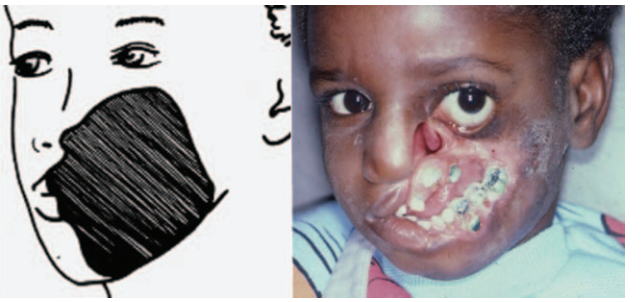

FIGURE 4. Type IV defect.

THE HISTORY

Although subjects (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 that shows that it was quite common in Europe up to the nineteenth 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 a research that Mengele could publish under his own name. Epstein accepted and proposed a research project on the treatment of noma. A "Noma ward" (*Nomaabteilung*) was established in the camp, where 45 to 70 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 other died and their heads sent for postmortem examination. The result of this "experiment" was 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.[7] During his three-and-a-half years stay in Ibadan (Nigeria), he treated more the 300 children and related his experience in a classical essay under the title *Cancrum oris*. He also gave a historical introduction and a description of the state of the art as regard the etiology and the treatment of this mutilating disease 50 years ago. In 2003, 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.[8]

Although the incidence of this impressive illness, devouring 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, 1 may find the following account corresponding to Type I sequel.

> Case 15: A cheek wound. "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 sequel:

> Τω παιδίω τω φαγεδαινωθέντι...
> "In the child suffering from phagedenic affection, the teeth of below and above, in front, have fallen down,

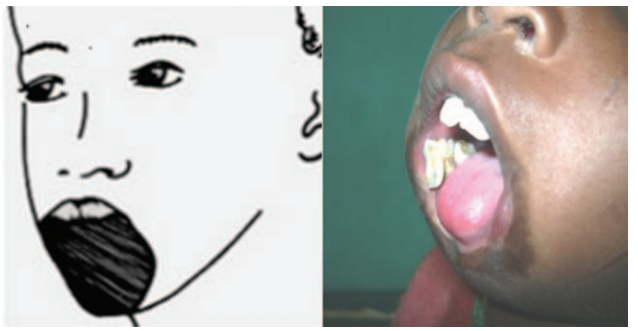

FIGURE 3. Type III defect.

FIGURE 5. Example of type II reconstruction of nose, cheeks, and upper lip with various skin and mucosal flaps and calvarial bone graft to the nose.

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 Age, 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 4 pages chapter of his book "Handboeck der Chirurgijen": "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 2 centuries, similar descriptions of the disease can be found in the English, French, Dutch, Swedish, German, and Italian medical literatures, accompanied sometimes with suggestive and dramatic illustrations.

FIRST OPERATIONS

While working in Switzerland as a protestant refugee, the famous surgeon Pierre Franco described in 1561 very precisely how he was able to cure a man suffering most certainly from noma sequel.[9] His account is a model of surgical report as it includes diagnosis, previous medical history, methods used for surgery (skin and mucosal flaps), follow up, and functional and cosmetic result.

"A certain Jacques Janot Savagny who lived near Neufchastel-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

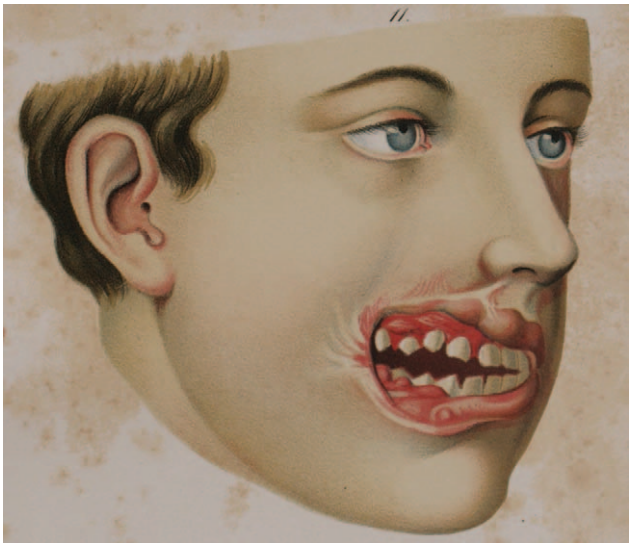

FIGURE 6. Von Brun's atlas: type I sequel.

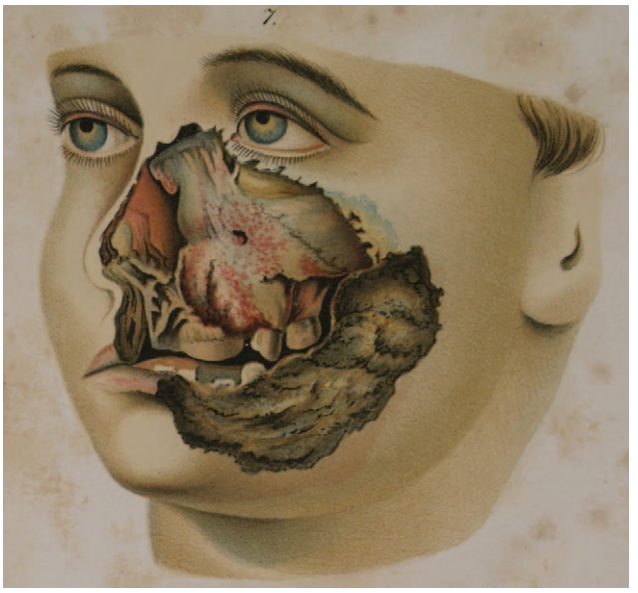

FIGURE 7. Von Brun's atlas: type IV sequel.

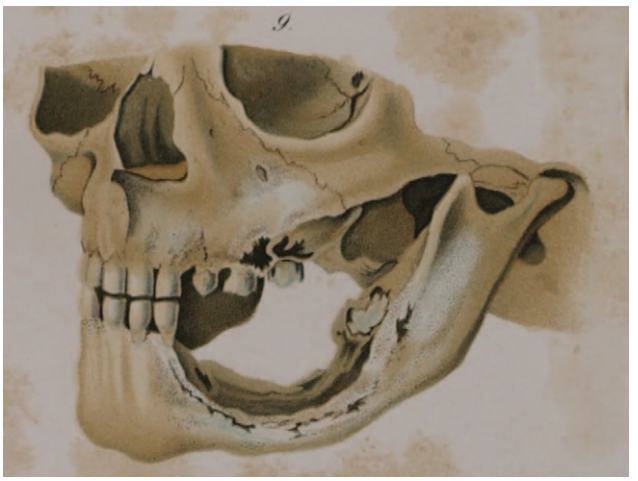

FIGURE 8. Von Brun's atlas: bone loss in noma.

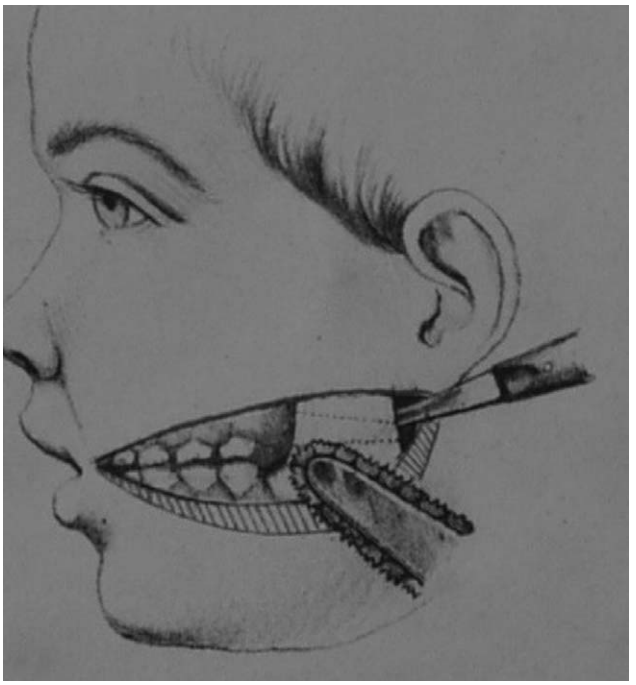

FIGURE 9. Von Brun's atlas: surgical cure of a cheek defect following noma.

'cauteres' nearby in a basin on a fire, ready to be used. 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 "cauteres" 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 seventeenth century to find other attempts to reconstruct the lips and the cheeks in noma patients. 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 (Figs. 6–9).

CONCLUSION

Although the disease was and is still infrequent compared with 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-maxillofacial field provide it.

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