

NOMA: A neglected disease!

By GESNOMA, Winds of Hope,
Sentinelles, and Médecins
sans Frontières

A boy having undergone rehabilitative surgery during the surgical camp, organised in collaboration between HUG (Hôpitaux Universitaires Genevois) and Sentinelles, in Ouagadougou, Burkina Faso, 2013.



Milène Zizzo, 2013



NOMA (cancrum oris and fusospirochetal gangrene or Necrotising Ulcerative Stomatitis), disfigures children rapidly, if they survive. It is one of the most devastating and disfiguring human diseases worldwide and was designated a health priority by the WHO in 1994 (1). NOMA is still a neglected disease, and there is not much known about its causes, prevention and optimal treatment. Fortunately, with simple interventions, NOMA can be addressed and contained and people with NOMA can be cared for. Thus, this disease deserves more attention from healthcare workers, nutritionists, researchers and policy-makers. In this article, we explain NOMA, explore the relationship between NOMA and nutrition and suggest how health workers in nutrition programmes can be involved in its identification and management. Two posters are included with the print edition of *Field Exchange*, one for an outpatient and one for an inpatient consulting room setting, in both English and French.

What is NOMA?

NOMA is a gangrene in the orofacial area. The course of the disease is very aggressive and fast. It starts as a gingivitis that develops into a gingival ulcer and/or necrotising gingivitis, spreading rapidly throughout the tissues of the mouth and face. The infection can result in necrosis of tissue and bone in the face which, combined with sepsis, is fatal in most patients. NOMA not only disfigures the patient but also causes dysfunction in eating and speaking, resulting in malnutrition and social isolation (2). If NOMA is untreated, 70-90% of patients will die.

The incidence of NOMA among children is estimated at 140,000 per year and the prevalence at 770,000 cases worldwide (2, 3). Lower estimates are 100,000 children per year who are affected by NOMA, of whom 20,000 survive (www.nonoma.org). However these figures are likely gross underestimates;

NOMA is underreported since it occurs in remote areas, people are not eager to let the world know there is a disfigured member of the family, and the disease progresses rapidly to death. Most cases of NOMA (80%) occur in countries in the SAHEL belt, such as Chad, Nigeria, Niger, but also in Asia and South America. In the past, NOMA occurred in Europe too, where it was associated with poverty and the presence of other infectious diseases such as measles or tuberculosis.

The precise causes of NOMA are unknown, but the disease is thought to be related to immune dysfunction. Reduced immune function is in turn associated with poverty, the presence of other diseases (measles, malaria, pneumonia and HIV/AIDS (4)), malnutrition (5), poor hygiene and sanitation (no clean water, contact with animal waste), as well as lack of primary health care and health promoting activities like vaccination. Lack of oral hygiene is also a risk factor for NOMA; one of the early stages of NOMA is gingivitis and other infections in the mouth.

Treatment of NOMA

The early stages, such as simple gingivitis, should be treated with mouth washes of salted water and general oral hygiene. A complicated gingivitis (with necrosis, spontaneous gingival bleeding and pain) requires professional dental hygiene and follow up (if available). If dental hygiene and follow up cannot be achieved, antibiotics are needed. When there is a necrotising gingivitis/stomatitis with oedema of the corresponding facial region, antibiotics are mandatory. The later stages should be managed by an intense antibiotic regime in order to stop the spread of the infection and to avoid the deadly complications (such as septicaemia). Once the infection is over, and depending on the localisation of the NOMA, physiotherapy aimed to avoid complete trismus (jaws constriction) must be started for those patients

developing this complication. In many cases, specialist reconstructive surgery is needed and can only be planned once the scarring process is over and no earlier than one year after the acute NOMA. Treatment to improve function, counselling and actions to maintain dignity of the patient must always be present from the beginning of the lesion.

Simple gingivitis can be treated in an ambulatory therapeutic feeding centre (ATFC), the clinic and at home by rinsing with salted water for 14 days. In inpatient settings, such as an inpatient therapeutic feeding centre (ITFC) or hospital, mouth washes with 0.5% Betadine 4 times per day for 5 days (maximum) can also be used. Then, application of a solution consisting of 2 parts 1.4% bicarbonates at to 1 part nystatine 4 times per day for at least 10 days, or possibly over the total duration of the hospitalisation, can be applied with the aid of a compress rolled up on a tongue compressor (the caregiver can be taught to do this). In the case of necrotising gingivitis, the above local treatment should be completed with antibiotics (amoxicillin-clavulanate combination or amoxicillin plus metronidazole).

The treatment should be complemented by active nutrition support (e.g. supplementation with lipid nutrition supplements (LNS) or therapeutic foods where severely mal-

Screening for gingivitis:

- Gums: redness, pain, bleeding
- Hyper salivation, drooling
- Bad breath
- Anorexia
- Gingival ulceration
- Facial swelling / oedema.
- Dry necrosis, loss of tissue, possible bone sequestrum

NOMA SYSTEMATIC ORAL SCREENING OF ALL HOSPITALIZED CHILDREN

Noma is an acute gangrenous infection mainly affecting the mouth and face. It has the potential to cause massive tissue destruction.

Noma occurs primarily in malnourished or immunosuppressed children with poor oral hygiene.

Left untreated, noma has a mortality rate of up to 90%.

In the absence of adequate treatment in the acute phase, complex plastic surgery will be necessary to reconstruct the face.

FAMILIARIZE YOURSELF WITH STAGES AND MANAGEMENT OF NOMA!!

| STAGE | DESCRIPTION | MANAGEMENT |
|---------|--|--|
| STAGE 0 | THE SIMPLE GINGIVITIS • Erythra and swollen gingival margins both red and edematous • Pain on bleeding on touch and when brushing teeth • Bad breath • Hyper-salivation • Anorexia | MANAGEMENT • Rinse the mouth with warm salt water 4-6 times per day • Continued dental hygiene (regular brushing, mouthwashes and local application of chlorhexidine 0.1%) • Follow-up (pay careful attention to the speed with which the disease develops) • Multivitamins: Zinc, Vitamin A, Retinolsin • Do not use methylene blue or geriatric violet • Increase general awareness and follow-up on the child if problems persist more than 8 days |
| STAGE 1 | THE HEMORRHAGIC OR NECROTIZING GINGIVITIS / STOMATITIS • Significant inflammation (redness and pain) • Gingival edema • Spontaneous gingival bleeding • Maxillary ulceration and/or gingival necrosis diffuse and/or localized to one or several teeth • Bad breath • Hyper-salivation • Anorexia, fever | MANAGEMENT • Analgesics and desludging + antiseptic mouthwash (0.1% chlorhexidine) 4-6 times per day • Co-Amoxiclav (or amoxicillin) + Metronidazole PO 14 days (if accompanied by malnutrition) • Multi-vitamins: zinc, vitamin A and Retinolsin + Associated Care (other diseases) • Monitor closely for 24 hours (pay careful attention to the speed at which the disease develops) • Test the wound but DO NOT perform surgery on the tissue |
| STAGE 2 | THE SWELLING – EDEMA (EARLY ACUTE) • Same symptoms as the ones described in stage 1 • Facial edema • Bad breath • Pain, anorexia, fever | MANAGEMENT • Co-Amoxiclav (or Amoxicillin) + Metronidazole + Clindamycin IV 14 days • Multi-vitamins: Zinc, Vitamin A and Retinolsin + Major analgesics and antiseptic Mouthwash (chlorhexidine 0.1%) 4-6 times per day • Associated care (other pathologies, especially infection debilitation and shock) • Monitoring vital signs in intensive care (if threatened) • Test the wound but DO NOT perform surgery on the tissue |
| STAGE 3 | THE NECROSIS +/- TISSUE LOSS (LATE ACUTE) • Sometimes persistence of symptoms described in stage 2 • ACCELERATED BY: • Eschar (layer of necrotic tissue) indicating the extent of the tissue loss or wound (area to occur) • Sometimes bone sequestra • Fetid odor • Pain, anorexia, fever | MANAGEMENT • Co-Amoxiclav (or Amoxicillin) + Metronidazole + Clindamycin IV 14 days • Multi-vitamins: Zinc, Vitamin A and Retinolsin + Major analgesics and antiseptic Mouthwash (chlorhexidine 0.1%) 4-6 times per day • Associated care (other pathologies, especially infection debilitation and shock) • Monitoring vital signs in intensive care (if threatened) • Test the wound but DO NOT perform surgery on the tissue |
| STAGE 4 | THE SEQUELA LESION WITH OR WITHOUT TISSUE / BONE LOSS • Very often loss of tissue / bone loss • Significant malposition of teeth (not present before disease) • Sometimes inability to open the mouth • No change for at least 12 months after the acute phase | MANAGEMENT • Hygiene and oral care + Retinolsin if necessary • As soon as possible, contact an NGO capable of reconstruction for specialized surgical advice, immediate reconstruction (removal of bone sequestra, loose teeth...) and a nutritional analysis • Prevent avoidable complications: intensive physiotherapy to avoid contractures and malocclusion • No emergency surgery except cleaning/removal of sequestra and replacement of missing • No urgency to consider reconstruction; minimum period of 1 year after the acute phase |

FOR ALL STAGES, PROPOSE SYSTEMATIC COUNSELING AND HIV TESTING.



SYSTEMATIC ORAL SCREENING OF ALL AMBULATORY CHILDREN NOMA

Noma is an acute gangrenous infection mainly affecting the mouth and face. It has the potential to cause massive tissue destruction.

Noma occurs primarily in malnourished or immunosuppressed children with poor oral hygiene.

Left untreated, noma has a mortality rate of up to 90%.

In the absence of adequate treatment in the acute phase, complex plastic surgery will be necessary to reconstruct the face.

| STAGES | CLINICAL SIGNS | STRATEGY AND STRUCTURE OF TREATMENT | MEDICAL TREATMENT PROTOCOL ACCORDING TO GIVEN STRUCTURE | MESSAGE GIVEN TO FAMILY AND CHILD |
|---|--|--|---|---|
| HEALTHY MOUTH | • Firm gum • No edema • No inflammation (red edge) of the gums around the teeth • No bad breath • No pain • No bleeding | • Community Health Care • Board of Health Education | • Treatment of associated diseases and malnutrition if necessary • Therapeutic education of the child's caregiver | • Nutritional education • Education in oral hygiene • Education in general hygiene (personal and household) |
| STAGE 0 SIMPLE GINGIVITIS | • Swelling of the gums • Inflammation of the gums (red edge) • Occasional bad breath • Pain when touch and brushing • Or -bleeding of the gums when touching or brushing | • Community Health Care & Hygiene • Committee on health education | • Wash mouth with salted boiled water daily • Dental hygiene care • Treatment of associated diseases and malnutrition if necessary • Therapeutic education of the child's caregiver • If clinical signs persist for more than 8 days → Report dental hygiene and general surveillance, strengthen education of patient's caregiver | • Explanation of the disease • Nutritional education • Education in oral hygiene • Education in general hygiene (personal and household) • When no improvement after 8 days → return the child to the health center for verification of progress and reinforce and repeat messages and explanations mentioned above |
| STAGE 1 ACUTE NECROTIZING GINGIVITIS | • Bad smells • Gum pain • Inflammation of the gums (pervet redness) • Swelling of the gums • Spontaneous bleeding of the gums • Or - gum necrosis (located to one or a few teeth) • Sometimes part of the bone visible below the gum | • Immediate referral to the health facility for medical care | If the patient is not malnourished: • Whenever possible refer the child to a health center capable of cleaning gums and teeth • Dental hygiene care and close monitoring • Child analgesics according to the levels of pain • Treatment of associated diseases and nutritional supplements if necessary • Therapeutic education of the child's caregiver If the patient is malnourished: • As above but treat malnutrition and add oral antibiotics for 14 days • Mouthwash with antiseptic | • Explanation of the disease • Nutritional education • Education in oral hygiene • Education in general hygiene (personal and household) • Identify a person in the family that takes care of the child |
| STAGE 2 NOMA STAGE ACUTE EDEMA | | | | REFER FOR EMERGENCY ADMISSION TO HOSPITAL |
| STAGE 3 NOMA STAGE ACUTE NECROSIS | | | | REFER FOR EMERGENCY ADMISSION TO HOSPITAL |
| STAGE 4 NOMA STAGE SEQUELAE | | | | REFER AS SOON AS POSSIBLE TO AN NGO EXPERIENCED IN NOMA OR TO A HOSPITAL FOR CLEANING, REMOVING SEQUESTRA AND ASSESSMENT OF LESIONS |

FOR ALL STAGES, PROPOSE SYSTEMATIC COUNSELING AND HIV TESTING.



nourished), treatment of any other existing infections, and updating of vaccination status. Prompt recognition of the early stage of NOMA (gingivitis) and treatment at this stage can prevent subsequent tissue destruction and disfigurement. This implies early recognition and active screening for NOMA.

Malnutrition and NOMA

Malnutrition (moderate and severe) is the most important risk factor for NOMA (5). Therefore prevention of malnutrition (along with treating underlying diseases, improving vaccination coverage and HIV testing) is an important step in the prevention of NOMA. This means that all moderately or severely malnourished individuals should be screened for signs of gingivitis. In addition, every patient in inpatient and outpatient nutrition treatment centres should be screened for gingivitis (simple and severe) and other mouth abnormalities. Once the NOMA infection is treated, many patients still have severe lesions in the mouth and face that can hamper eating, chewing, swallowing, talking, and sometimes even vision or breathing. Good nutritional support and guidance, with possible physiotherapy, can help to return to an acceptable nutritional status. Prior to any surgery, the lesion must no longer be active and the patient should be well nourished; close monitoring of their nutrient status and supplementary feeding is often necessary before surgery can be performed.

Active screening in feeding programme

Feeding programmes treating moderate and severe malnutrition have a

concentration of children at risk for NOMA and are therefore excellent places to target these children; they can play an important role in controlling NOMA in an area. Activities to include in a feeding centre are:

- Systematic screening of patients on admission for gingivitis
- Nutritional rehabilitation
- Vaccination
- Systematic HIV counselling and testing
- Screening of siblings and mothers,
- Improvement of water quality, sanitation and hygiene
- Education of patients and caretakers on mouth hygiene and NOMA
- Reporting of cases of NOMA in the village by caretakers
- Referral of NOMA patients to specialised institutes (where available)

Accessing guidance and support

Sentinelles, Winds of Hope, GESNOMA (all members of the International NoNoma Federation) and Médecins Sans Frontières (MSF) have created a working group to collaboratively develop several kinds of support:

- Posters for the consultation room in French and English for inpatient (hospital, ITFC) and outpatient facilities (outpatient medical clinic and outpatient/ambulatory therapeutic feeding centres (ATFC)) (included *Field Exchange 53*)
- Guidelines on treatment of NOMA
- Support to specialised centers for surgery
- Background information

- Guidelines on management of moderate and severe acute malnutrition
- Research

Nonoma (FR): www.nonoma.org
 Winds of hope (EN, FR, GE): www.windsofhope.org
 Sentinelles (FR, EN): www.sentinelles.org

Study of the Human Rights Council Advisory Committee on severe malnutrition and childhood diseases with children affected by noma as an example. UN General Assembly. 24th February 2012. Human Rights Council Nineteenth session Agenda item 5. www.righttofood.org/wp-content/uploads/2012/09/A-HRC-19-73.pdf
www.righttofood.org/work-of-jean-ziegler-at-the-un/noma/

References

1. Bourgeois DM, Leclercq MH. The World Health Organization initiative on noma. *Oral Dis.* 1999; 5:172-74
2. Ashok N, Tarakji B, Darwish S, Rodrigues JC, Altamimi MA; A Review on Noma: A Recent Update. *Global Journal of Health Science.* 2016;4 (53-59)
3. Baratti-Mayer D, Pittet B, Montandon D, Bolivar I, Bornand JE, Hugonnet S, Jaquetin A, Schrenzel J, Pittet D; Noma: an "infectious" disease of unknown aetiology. *Lancet infect Dis.* 2003; 3: 419-31
4. Masipa JN, Baloyi AM, Khammissa RAG, Altini M, Lemmer J, Feller L; Noma (Cancerus Oris): A Report of a Case in a Young AIDS Patient with a Review of Pathogenesis; *Head and Neck Pathol.* 2013;7:188-192
5. Baratti-Mayer D, Gayet-Ageron A, Hugonnet S, François P, Pittet-Cuénod B, Huyghe A, Bornand J, Gervais A, Montandon D, Schrenzel J, Mombelli A, Pittet D. Risk factors for NOMA disease: a 6-year, prospective, matched case-control study in Niger. *Lancet Global Health* 2013; 1: e87-96