

CONCEPT NOTE

in support of

THE INCLUSION OF NOMA (CANCERUM ORIS) AMONG THE WHO NEGLECTED TROPICAL DISEASES

Introduction

1. This concept note* presents the case for the inclusion of noma (cancerum oris) among the neglected tropical diseases covered by the World Health Organization (WHO).
2. Noma is an infectious, yet non-contagious disease that begins as a localized ulceration in the gingiva or the mucosa of the cheek or lip, which rapidly becomes necrotic, and spreads to produce extensive destruction of the soft and hard facial tissue.¹ Typically, within a week a gaping hole appears in the face.² Noma occurs predominantly in malnourished children aged 1-6 years, living in conditions of extreme poverty in developing countries.³ If left untreated noma leads to death in 80-90 per cent of cases; survivors face physical hardship, as well as stigmatization and discrimination.⁴ Recognized early and treated correspondingly with oral hygiene, mild antibiotics and nutritious interventions, mortality can decrease to 20 per cent.⁵
3. Following this introduction, the second part of the concept note presents background information concerning noma: incidence and distribution, causes, predisposing factors, and treatment, as well as WHO initiatives related to noma, and the work of the UN Human Rights Council and its Advisory Committee. In the third part, the justification and purpose, noma is introduced as a disease which shares the characteristics of the 17 neglected tropical diseases currently covered by the WHO—it is in fact a “neglected-neglected disease”.⁶ This part will also outline the purpose of listing noma among the neglected tropical diseases. Finally, the note will propose a recommendation for consideration by the World Health Assembly.

Background

Incidence of noma

4. According to WHO data 140,000 individuals contract noma on a yearly basis.⁷ Given the survival rate of 10-20 per cent, at least 110,000 people, the vast majority of which are children, die every year because of noma.⁸ In 1998, 770,000 persons are estimated to have survived the disease with heavy sequelae.⁹
5. Experts consider that cases of noma are severely underreported; less than 10 per cent of the individuals suffering of noma seek medical care.¹⁰ In the evocative words of Dr Cyril Enwonwu, a leading expert on

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¹ C.O. Enwonwu et al, “Noma (cancerum oris): Seminar”, 386 *The Lancet* (2006), 147–156, p. 147; D. Baratti-Mayer et al, “Noma: an “infectious” disease of unknown aetiology”, 3 *The Lancet Infectious Diseases* 7 (July 2003), 419–431, p. 419; *Dorland's Illustrated Medical Dictionary*, 27th ed.

² Ibid.; K. Bos, K. Marck, *The Surgical Treatment of Noma*, (Alphen aan den Rijn: Belvedere/Medidac, 2006), pp. 13–14.

³ D.E. Barmes et al, “The need for action against oro-facial gangrene (noma)”, 2 *Tropical Medicine and International Health* 12 (1997), 1111–1114, p. 1113; M. Tsechkovski, Director, WHO Division of Non-Communicable Diseases, “A disease such as Noma should not exist”, *Noma Contact*, (WHO: Geneva, October 1997), p. 1; P.A. Van Damme, “Essay: noma” 368 *The Lancet*, (2006), S61–62.

⁴ Ibid.

⁵ K. Bos, K. Marck, *supra note 2*, pp. 13–14.

⁶ M. L. Srouf, K. W. Marck and D. Baratti-Mayer, “Noma: Neglected, Forgotten and a Human Rights Issue”, 7 *International Health* 3 (2015), 149-150, p. 150.

⁷ WHO, *The World Health Report 1998 - Life in the 21st Century: A Vision for all* (Geneva, 1998), p. 45.

⁸ Ibid; P.E. Petersen, *The World Oral Health Report 2003: Continuous Improvement of Oral Health in the 21st Century – the Approach of the WHO Global Oral Health Programme* (Geneva: WHO, 2003), pp.7- 8.

⁹ WHO, *supra note 7*, p. 45; A. Fieger et al, “An Estimation of the Incidence of Noma in North-West Nigeria”, 8 *Tropical Medicine and International Health*, 5 (2003), 402-407, p. 402.

noma, these cases represent merely “the tip of the iceberg”.¹¹ A cumulus of biological, administrative and social factors are responsible for the underreporting. Among them are the rapid progression of the disease, which can result in death within a matter of weeks, associated with a high mortality rate and the lack of inclusion of noma as a cause of death in national statistics.¹² The lack of adequate knowledge on noma among health personnel (including in respect to clinical signs and treatment), as well as the lack of appropriate information provided to parents. Because of the latter, noma is often perceived as a curse or as shame on the family whose child contracts it; faced with social stigma, families sometimes hide away or isolate their children with animals, instead of seeking health care.¹³ Provided they survive, these children are left out from any statistics on the disease.

6. A recent estimate of the global burden of disease from noma resulted in “1.1 million daily-adjusted life years, even if a best case scenario was assumed”.¹⁴ This estimate is high when compared with many of the neglected tropical diseases which are covered by the WHO.¹⁵
7. Information concerning the incidence and global distribution of noma can only be improved if the biological, administrative and social factors which hinder the collection of accurate statistics are addressed. Including noma among those neglected tropical diseases covered by the WHO would certainly focus efforts in the area of data collection as well.

Distribution of noma

8. The global distribution of reported cases of noma shows that the disease has been prevalent in Africa, as well as in Asia and Latin America. Low- and middle-income countries, particularly in Africa and Asia, are the most affected by noma.¹⁶ Experts, including non-governmental organizations (NGOs) working on the ground, describe the area that stretches across parts of West Africa, Central Africa towards Sudan as the “noma belt”.¹⁷ A 2008 medical study has also concluded that noma is likely to be “much more frequent in remote Asian rural communities than is likely currently appreciated”.¹⁸ Rural communities in some regions of Latin America are also affected.
9. A more accurate appreciation of noma’s geographical distribution is made difficult by the same biologic, administrative and social factors mentioned above. In addition, the current focus of the WHO, through the Regional Office for Africa, is on noma on the African continent. The latter Office, of course, cannot be expected to keep an overview of statistics from other continents.
10. Historically, noma was present in Europe and North America—economic development and an improved diet is largely credited with the eradication of noma at the beginning of the twentieth century. More recently, sporadic cases have been documented in developed states in persons suffering of HIV/AIDS, severe malnutrition, poor oral hygiene and living in extreme poverty conditions.¹⁹ This data shows that while today noma is prevalent in tropical environments, it is, fundamentally, a poverty-related disease that can be tackled.

¹⁰ C.O. Enwonwu, “Noma-The Ulcer of Extreme Poverty”, 354 *The New England Journal of Medicine* 3, (January 2006), pp. 221–224; L. Srour, K. W. Marck and D. Baratti-Mayer, *supra* note 6, p. 149.

¹¹ C.O. Enwonwu, *supra* note 10, pp. 221–224; Winds of Hope, WHO/AFRO, *Rapport de la 5ème Table Ronde sur le Noma*, Genève, 9 septembre 2006, http://www.nonoma.org/doc_pdf/rapport_table_rond.pdf.

¹² M. L. Srour, K. W. Marck and D. Baratti-Mayer, *supra* note 6, p. 149.

¹³ D.M. Bourgeois, M.H. Leclercq, “The World Health Organization Initiative on Noma”, 5 *Oral Diseases* (1999), 172–174, pp. 153–154, p. 421; J.E. Tonna et al., “A Case and Review of Noma”, 4 *Neglected Tropical Diseases* 12 (December 2010).

¹⁴ M. L. Srour, K. W. Marck and D. Baratti-Mayer, *supra* note 6, p. 150. See also, P.F. Hotez, M. Alvarado, M.G. Basanez et al., “The global burden of disease study 2010: interpretation and implications for the neglected tropical diseases”, *PLoS Negl Trop Dis* (2014).

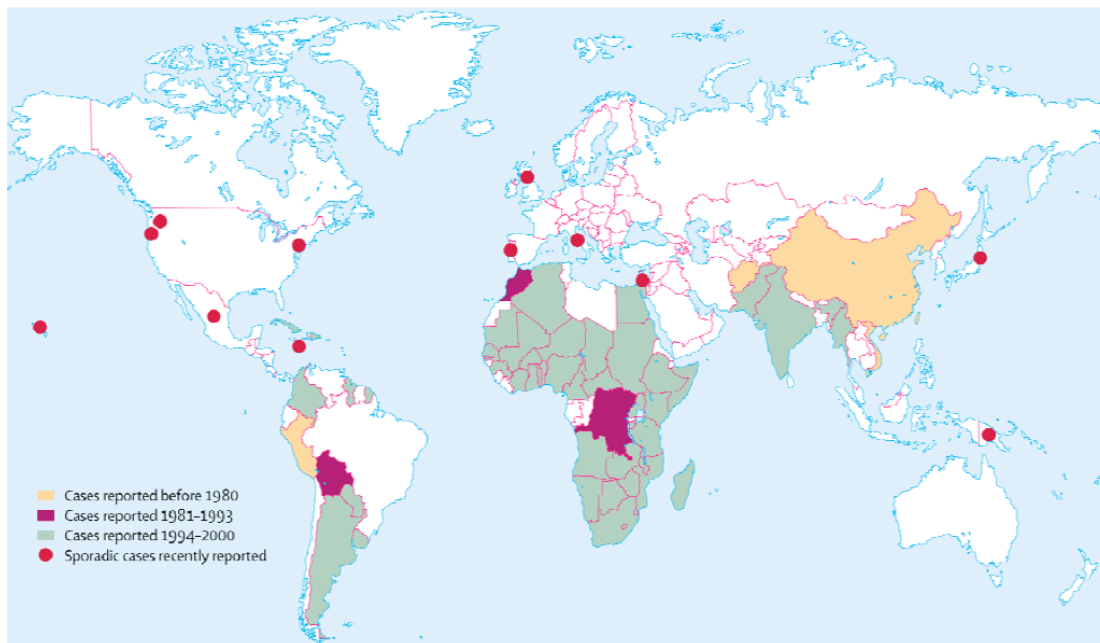
¹⁵ *Ibid.*

¹⁶ P. E. Petersen, “World Health Organization Global Policy for Improvement of Oral Health – World Health Assembly 2007”, 58 *International Dental Journal* (2008), 115–121, p.1 17.

¹⁷ Consultative Meeting on Management of the Noma Programme in the African region, Harare, 19-21 April 2001, Final Report; see also WHO Regional Office for Africa, “Noma”, <http://www.afro.who.int/en/health-topics/topics/4356-noma.html>.

¹⁸ M.L. Srour et al., “Noma in Laos: Stigma of Severe Poverty in Rural Asia”, 78 *American Journal of Tropical Medicine and Hygiene* 4 (2008), 539–542, p. 540.

¹⁹ See A. G. Buchanan et al “Necrotizing stomatitis in the developed world”, *Clinical and Experimental Dermatology* 31 (2006), 372–275, p. 372; A. Maley, M. Desai, and S. Parker, “Noma: A Disease of Poverty Presenting at an Urban Hospital in the United States” 1 *JAAD Case Reports* 1 (2015), 18-20.



Source: Adapted from WHO by C.O. Enwonwu et al, “Noma (cancrum oris): Seminar”, 386 *The Lancet* (2006), p.148.

Causes, predisposing factors, and treatment of noma

11. There is a wide consensus among experts that noma results from the interaction between several main elements: intraoral infections, malnutrition and poor oral hygiene – both consequence of poverty – and compromised immunity.²⁰
12. First, although investigations have not identified a specific microorganism/bacteriological agent responsible for causing noma,²¹ they concluded that a high bacterial load of normal microorganisms from the mouth breaks the resistance of a failing immune system.²² Acute necrotizing gingivitis is the precursor of noma.²³ *Fusobacterium necrophorum* and *Prevotella intermedia* are considered key elements in this process in which components of the normal oral flora become pathogenic.²⁴
13. Second, malnutrition is considered to be the major predisposing factor for noma. Experts describe malnutrition as “mandatory” for noma to appear.²⁵ The absence of noma cases in well-nourished African children, strongly supports the evidence that malnutrition plays a significant role in the development of this disease.²⁶ The NGO Sentinelles has observed the effects of food crises in countries in Africa: a number of years after such a crisis the number of small children with Noma had doubled. This increase in number of Noma cases is likely explainable by the fact that malnourished babies were born to

²⁰ A. Fieger et al, “An Estimation of the Incidence of Noma in North-West Nigeria”, 8 *Tropical Medicine and International Health*, 5, May 2003, 402-407, p. 402; C.O. Enwonwu et al, *supra* note 1, p. 151; C.O. Enwonwu, *supra* note 10; Interview de Bertrand Piccard, «Notre nouveau but: mettre sur pied une Journée mondiale contre le noma», *Tribune Medicale*, 29 septembre 2006.

²¹ D. Baratti-Mayer, A. Gayet-Ageron et al “Risk factors for noma disease: a 6-year, prospective, matched case-control study in Niger”, 1 *The Lancet Global Health* 2 (2013) 87-96; R. S. Phillips, C. O. Enwonwu, W. A. Falkler, “Pro- versus anti-inflammatory cytokine profile in African children with acute oro-facial noma (*cancrum oris*, noma)”, 16 *European Cytokine Network* 1 (2005), 69–176, p. 70; D. Baratti-Mayer et al, “Noma: an “infectious” disease of unknown aetiology”, 3 *The Lancet Infectious Diseases* 7 (2003), 419–431, p. 419; B.J. Paster et al, “Bacterial Species and Novel Phylotypes in Advanced Noma Lesions”, 40 *Journal of Clinical Microbiology* 6 (2002), 2187–2191.

²² A. Fieger et al, *supra* note 20, p. 402.

²³ D. Baratti-Mayer et al, *supra* note 21, p. 421; P. Moynihan, P. E. Petersen, “Diet, Nutrition and the Prevention of Dental Diseases”, 7 *Public Health Nutrition* 1A (2004), 201–206, p. 203.

²⁴ B. Neville et al, *Oral and Maxillofacial Pathology*, 3rd Edition, (Elsevier Health Sciences, 2008), p. 201; see also M.L. Srour et al, *supra* note 18, p. 539.

²⁵ D.E. Barmes et al, *supra* note 3, p. 1111.

²⁶ C.O. Enwonwu et al, *supra* note 1, p. 151.

malnourished mothers, the latter victims of food crises.²⁷ In the words of researchers: “The global distribution pattern of the disease reflects the worldwide distribution of malnutrition”.²⁸

14. Third, poor oral hygiene is an essential element in the development of noma. Experts consider that noma is less likely to occur in malnourished children provided that oral care is undertaken regularly and thoroughly.²⁹ Preventive oral examinations and providing education to parents on good practices in respect to oral hygiene are key interventions to prevent noma in malnourished children. Unsafe drinking water and dehydration, poor sanitation including due to unsanitary housing conditions and close proximity to unkempt livestock are considered to also be important risk factors for the development of noma.³⁰
15. Fourth, the weakening of the immune system due to malnutrition and infections, such as measles, malaria, tuberculosis, HIV can contribute to the development of noma in children.³¹
16. Noma can reach its terminal phase in the extremely short time span of three weeks.³² The WHO identifies four stages of the disease and the treatment of noma differs accordingly. In the early stage, when the gingiva is bleeding and has lesions, impending noma can be treated in a manner which is “simple, effective, low-cost”³³ with disinfecting mouth-rinses and daily food with vitamins.³⁴ During the next phase, involving the swelling of the face and fever, mouth-rinses, administration of antibiotics and nutrients supplementation is essential.³⁵ These have been shown to prevent the progression from the initial ulceration to the extensive gangrene, which presuppose emergency care, and in the late stage reconstructive surgery.³⁶
17. Survivors suffer disfigurement and functional impairment. Doctors describe the restriction of the jaw movement and the loss of part of the maxilla, mandible or other facial bones as the usual consequences of noma.³⁷ Without reconstructive surgery “[a] child who survives is unlikely ever to be able to speak or eat normally again.”³⁸ Survivors also suffer severe social stigma and discrimination.
18. As was noted in the introduction, the mortality rate associated to noma is very high. However, if recognized early and treated correspondingly with oral hygiene, antibiotics and nutritious feeding, mortality can decrease from 70–90 per cent to approximately 20 per cent.³⁹ Hence, early recognition of the clinical signs of noma and timely treatment is critical for saving the lives of children affected by it.

WHO initiatives concerning noma

19. WHO organized the first information session on noma at the World Health Assembly in 1989; a five-point program to address noma was adopted in 1994. The program comprised prevention, epidemiology and surveillance, etiological research, primary health care and surgery rehabilitation.⁴⁰ The WHO Regional Committee declared noma as a priority on the African continent in 1998; two years later, the Noma Program activities were transferred from the WHO headquarters to the WHO Regional Office for Africa. Some progress has been achieved in the African region, with the important support and involvement of NGOs, charities and private individuals and the cooperation of Governments. However, as noted, currently the WHO does not cover other regions with prevalence of noma, beyond Africa. Increased attention in Africa to the disease would also benefit those most vulnerable.

²⁷ P. Joly, Sentinelles, Feedback on UN. Doc. A/HRC/AC/3/CRP, 27 July 2009.

²⁸ C.O. Enwonwu et al, *supra* note 1, p. 151.

²⁹ Submission by MSF on A/HRC/AC/7/CRP.229, November 2011.

³⁰ P.E. Petersen, *supra* note 16, p. 117.

³¹ K. Bos, K. Marck, *supra* note 2, p. 13.

³² WHO, “Acting against Disease, Open the Mouth of Your Children. Acting Against Noma”, Geneva.

³³ K. Bos, K. Marck, *supra* note 2, p. 18.

³⁴ WHO, *supra* note 32.

³⁵ *Ibid*; K. Bos, K. Marck, *supra* note 2, p. 18; M.L. Srour et al, *supra* note 18, p. 539.

³⁶ *Ibid*.

³⁷ D.E. Barmes et al, *supra* note 3, p. 1113.

³⁸ *Ibid*.

³⁹ K. Bos, K. Marck, *supra* note 2, p. 18

⁴⁰ D.M. Bourgeois, M.-H. Leclercq, *supra* note 13, pp. 153-154.

20. In 2009, the UN Human Rights Council Advisory Committee on the initiative of Committee member Jean Ziegler has commenced research on noma in relation to malnutrition, childhood diseases and human rights. The Committee has consulted extensively with states, UN bodies, including the WHO and the UN Secretary General's High Level Task Force on Food Security, NGOs working in the field of noma, human rights and humanitarian organizations,⁴¹ and medical doctors. The research has revealed the multiple links between noma and the right to adequate health care, the right to food, the right to water and sanitation, and not least the right to life of the most vulnerable members of society, children living in poverty. It has underlined the imperative of addressing noma, given that states hold obligations which are correlative to the aforementioned rights. It has also underlined the responsibilities of international organizations in their response to noma and malnutrition.⁴²
21. In 2012, the final Study of the Human Rights Council Advisory Committee on severe malnutrition and childhood diseases with children affected by noma as an example (A/HRC/19/73) together with the thereto annexed Human Rights Principles and Guidelines to Improve the Protection of Children at Risk or Affected by Malnutrition, Specifically at Risk of or Affected by Noma was submitted to the Council. In its resolution of March 2012 (A/HRC/19/L.21), the Human Rights Council encouraged States to implement the aforementioned Human Rights Principles and Guidelines.⁴³ One of the principal recommendations of Study and the Human Rights Principles and Guidelines was the formal recognition of noma as a neglected disease by the WHO in order to raise awareness, and trigger a **global approach** including worldwide surveillance, prevention, primary health care, surgical and rehabilitation treatment and etiological study of noma.

Justification and Purpose

22. Noma shares all the relevant characteristics with the 17 neglected tropical diseases currently covered by the WHO. As the 2013, Report on the Neglected Tropical Diseases prepared by the WHO Secretariat emphasizes: “[d]espite their medical diversity, neglected tropical diseases form a group characterized by their association with poverty and their proliferation in tropical environments where multiple infections in a single individual are common.”⁴⁴ Indeed, noma affects malnourished children living in extreme poverty, predominately in tropical climates—it has been termed “the face of poverty”. Noma’s global burden of disease is higher than many of the other neglected tropical diseases. Moreover, noma, is a complication of the major killers such as malaria, diarrheal diseases, HIV-infection/AIDS, measles, tuberculosis, and severe chronic malnutrition.⁴⁵
23. Furthermore, noma fits squarely within the WHO framework on neglected diseases outlined in the Global Plan to Combat Neglected Tropical Diseases 2008–2015, the WHO Roadmap to accelerate the work to overcome the global impact of neglected tropical diseases, the 2013 Report on Neglected Tropical Diseases by the WHO Secretariat,⁴⁶ and the resolution 66/12 adopted during the 66th session of World Health Assembly.⁴⁷
24. Without a doubt, as the background information demonstrates and the consensus among medical doctors forcefully emphasizes, noma is a neglected disease. As stakeholders deplore, noma is in fact a “neglected-neglected” disease:⁴⁸ while having all the characteristics of a neglected disease, it has not received the required recognition as a neglected disease.

⁴¹ Including (in alphabetical order): CARE International, Dutch Noma Foundation, Ecumenical Advocacy Alliance, Facing Africa, GESNOMA, Hilfsaktion Noma, Label Vert, Medico International, Noma-Hilfe Schweiz, Médecins sans Frontières, Sentinelles, Winds of Hope Foundation, No-Noma International Federation.

⁴² See A/HRC/19/73.

⁴³ A/HRC/19/L.21, para. 51.

⁴⁴ A66/20, para. 2.

⁴⁵ C. O. Enwonwu, “Ruminations on the causation of noma”, 104 *Stomatologie* 1 (2007), 43–48, pp. 43.

⁴⁶ A66/20.

⁴⁷ WHA66.12.

⁴⁸ M. L. Srour, K. W. Marck and D. Baratti-Mayer, *supra* note 6, p. 150.

25. The purpose of including noma among the WHO-covered neglected diseases is:
- a. To reduce human suffering, morbidity, mortality and stigmatization;
 - b. Through improvement in socioeconomic status, to reduce the conditions that exacerbate poverty;
 - c. To highlight the public health importance of these diseases in affected communities.

These are the very goals of the existing national programmes for the control of neglected tropical disease.

26. Targets for control of noma could be realistically set at a level at which the cost and resources for continued control measures are affordable and sustainable. As was pointed out, preventative action and early detection of noma is key. If caught early on noma requires a relatively cost-effective intervention: oral hygiene, mild antibiotics and nutritious interventions, with a spectacular impact which decreases the mortality rate by up to 70 per cent.
27. Given the nexus which noma entertains with poverty, hunger and malnutrition, access to health, water, and sanitation, the inclusion of noma among the WHO-covered neglected tropical diseases can contribute directly to various targets of the Sustainable Development Goals under goals 1 (No poverty), 2 (Zero Hunger), 3 (Good Health and Well-Being), 6 (Clean Water and Sanitation), 16 (Peace, Justice and Strong Institutions).

Recommendation

For the above reasons,

It is recommended that the World Health Assembly, by resolution, includes noma (cancrum oris) among the neglected tropical diseases formally covered by the WHO.