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Perspective

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# Noma — The Ulcer of Extreme Poverty

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In 2001, my research team was visiting a farming community in northwestern Nigeria, where word quickly spread to neighboring villages that foreign doctors were examining the mouths of sick children. Soon, a farmer arrived with one of his three wives and their two-year-old daughter. The mother told us that the girl was her fifth child and that three of the previous four had died before five years of age — two from "high fever" (presumably caused by malaria or measles) and one from noma, or cancrum oris, a noncommunicable infectious disease that destroys the hard and soft tissues of the mouth and may involve the face.

The girl had had fever and rash (presumably measles) a few weeks before she began complaining about itching in her mouth. She had been taken to a traditional healer, who had diagnosed "rashes" or "worms" and applied an herbal remedy. Nevertheless, the lesion soon perforated the cheek, and now the girl had localized gangrene — a yellowish groove around the perimeter of a blackened necrotic center. Intraoral examination revealed extensive destruction of the right posterior quadrants of the maxilla and the mandible. The child had markedly stunted growth and wasting, with edema of the limbs, characteristic signs of malnutrition.

As is common in rural sub-Saharan Africa, the child had been breast-fed exclusively for only six weeks, after which her diet had been supplemented with herbal tea, glucose water, unprocessed cow's milk, and indigenous cereal-based foods (no doubt prepared under less-than-hygienic conditions). The father blamed his wife's carelessness for the child's illness and for the deaths of their children. He refused to accept any suggestion that the child was malnourished. The mother confided that one of her husband's other wives had lost a child to noma.

We referred the girl to a hospital for treatment and the parents to a public health nurse for counseling on nutritional and health practices. The lesson we learned from this and many similar cases was that poverty-reduction efforts must go hand in hand with appropriate nutrition and health education if noma is to be eliminated.

"Noma" derives from the Greek word *nemo*, meaning "to graze"

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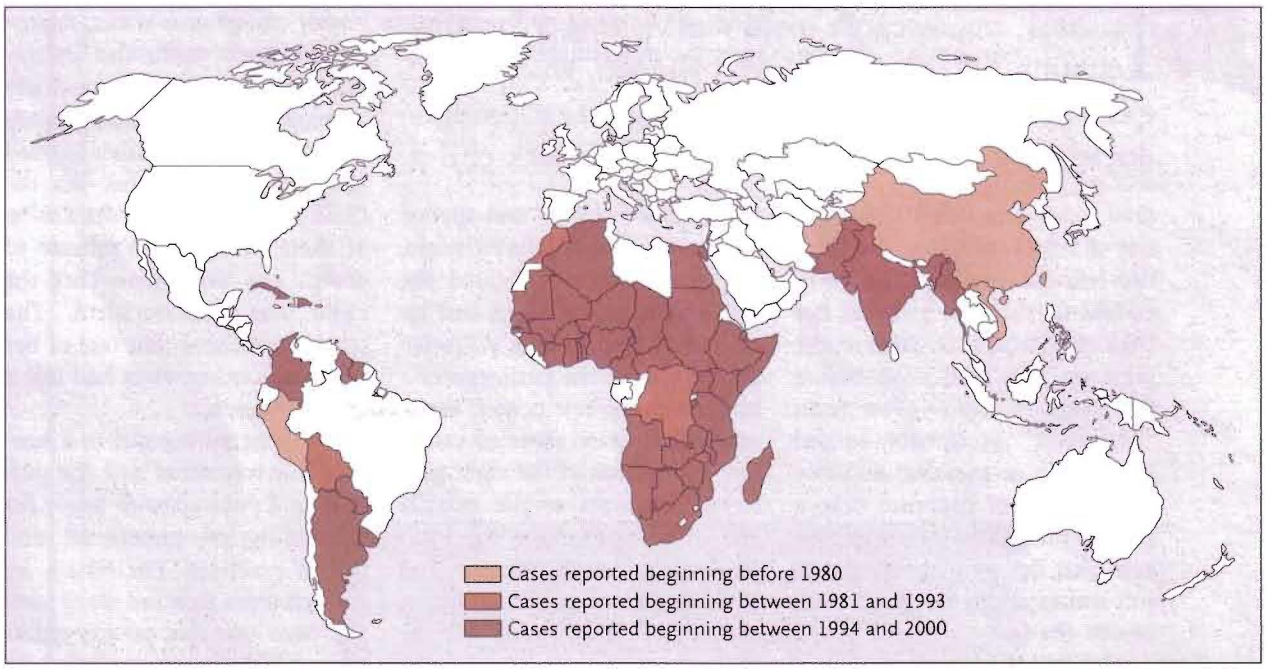
or "to devour." The disease dates back to antiquity, but its name was first used by a Danish physician in 1680 and was meant to underscore its astonishingly rapid development.[1] To Hausa communities in Nigeria, it is known as *ciwon iska* — an unexplained, sudden illness linked to the spirits. The incidence of acute noma peaks at one to four years of age, although late stages occur in adolescents and adults.[2]

Noma was common in Europe and North America until the early 20th century, when it essentially disappeared from developed countries, except for cases found in the concentration camps of Bergen-Belsen and Auschwitz and, more recently, in association with intensive immunosuppressive therapy, in some patients with human immunodeficiency virus (HIV) infection, and in Native American children with severe combined immunodeficiency syndrome. Yet the disease is still prevalent in developing countries. The World Health Organization (WHO) has compiled a global picture of reported cases (see map); although African countries are the most affected, Asia and Latin America are not exempt.

Noma thrives in communities characterized by extreme poverty, severe malnutrition (particularly micronutrient deficiencies), unsafe drinking water, poor sanitation, poor oral health practices, high infant mortality, limited access to high-quality health care, and a high prevalence of low birth weight, attributable primarily to intrauterine growth retardation. In these communities, families may share their overcrowded, poorly ventilated living quarters and sources of drinking water with domestic animals, and foods for infants may be prepared under unhygienic conditions. All these factors promote chronic and recurrent infection-induced immunostimulation by environmental antigens. Noma is usually preceded by a debilitating illness, such as measles, malaria, diarrhea, tuberculosis, or necrotizing ulcerative gingivitis.

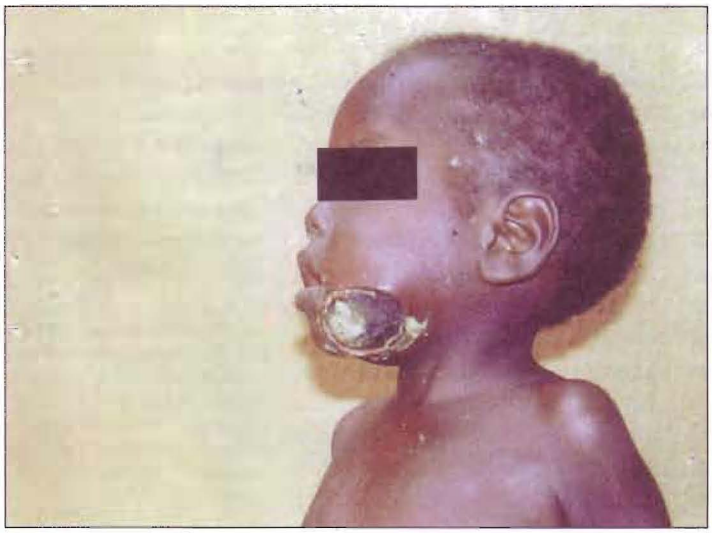
Accurate epidemiologic data are lacking because noma occurs predominantly in poor communities that do not keep health records and, frequently, have a nomadic lifestyle. Also, the disease is often hidden by families who consider it an evil omen. In 1998, the WHO estimated the annual global incidence at 140,000



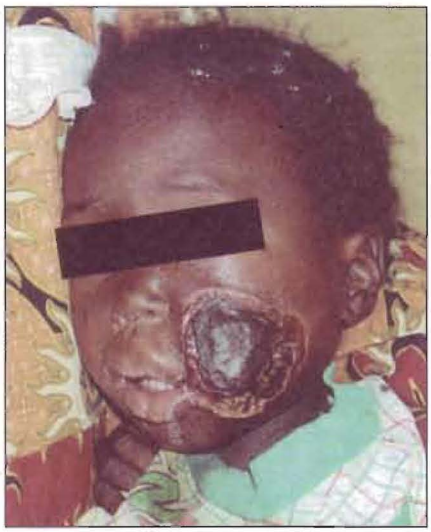

**Worldwide Distribution of Reported Cases of Noma.**

Before 1980, many sub-Saharan countries had underdeveloped health-reporting systems. Since the 1990s, awareness of noma has increased, and many countries have developed control plans. Sporadic cases have also been reported in higher-income countries, including the United States and some Western European countries, where noma-like lesions may be associated with HIV infection or AIDS.

Adapted from the World Health Organization.



Noma Lesion with a Well-Demarcated Perimeter in a Malnourished Child with a Recent History of Measles.



Child with Severe Noma.

Noma Children Hospital, Sokoto, Nigeria.

and the associated mortality at 70 to 80 percent among persons who are not treated promptly. A more recent report estimates an annual incidence of 25,000 in the developing countries bordering the Sahara.[3] But these estimates reflect the tip of the iceberg, since it is believed that no more than 10 percent of affected persons seek medical care.

Patients with acute noma usually present with malodorous breath, fluctuating fever (temperatures of 101 to 105°F [38 to 41°C]), marked anemia, a high white-cell count, severe growth failure, and other manifestations of malnutrition and poor general health. The lesion, usually unilateral, is often well established by the time medical help is sought. Noma generally starts as a gingival ulceration that can be treated easily at the early stage with local disinfection, antibiotics, and nutritional rehabilitation. If the ulcer is left untreated, it progresses rapidly to involve the cheek or lip; swelling is often the earliest externally visible sign of disease. The swelling increases, and within days, a blackish furrow appears where intraoral tissue is being lost. The lesion finally establishes itself with a well-demarcated perimeter surrounding a blackened necrotic center. Sequestration of exposed bone and teeth occurs rapidly after the separation of the soft-tissue slough. A hole remains after the scab is removed.

The sequelae depend on the anatomical sites affected, the extent and severity of tissue destruction, and the stage of development of the oral tissues. Survivors of acute noma often have severe disfigurement and functional impairment. They may therefore be shunned by society, and although reconstructive surgery is possible, it rarely restores normal facial appearance.[1]

Noma neonatorum, which is considered a distinct entity, affects newborns and resembles noma in older children. Preterm birth and severe intrauterine growth retardation are considered important predisposing factors. Necrotizing diseases of the oral tissues associated with HIV infection may be confused with early signs of noma, so serologic testing for HIV should be performed when noma is suspected.

Noma is a polymicrobial opportunistic infection, but there is no consensus regarding the causative microorganisms. Studies point to *Fusobacterium necrophorum* and *Prevotella intermedia*, which may enter children's mouths through water and food contaminated with animal feces.[4] The evidence suggests that the pathogenesis of noma involves a complex interaction among malnutrition, immune dysfunction, and infection with endemic viruses that creates a "staging period" characterized by impaired oral mucosal immunity, defective structural integrity of the oral mucosa, selective overgrowth of pathogenic microorganisms, increased oxidative stress, and a shift from an inflammatory to an anti-inflammatory cytokine profile, among other changes. An intraoral ulcer develops and probably

provides a site of entry for a triggering microorganism. In a final invasive phase, the lesion spreads rapidly, probably owing to the presence of necrotizing toxins or tissue-destroying enzymes and inflammatory mediators.

Still puzzling is the observation that in the same households, oral mucosal ulcers progress to noma in only a small subgroup of severely malnourished children. At least 85 percent of Nigerian children under three years of age who have noma also have severely stunted growth.[2] Stunting (failure to grow) in early infancy is considered in some cases to be a continuation of intrauterine growth retardation, which is prevalent in underprivileged African communities. It is possible that children with noma are victims of intrauterine growth retardation, which impairs development of immune function. This hypothesis would suggest that there is a close pathogenic similarity between noma in children and the histopathologically similar noma neonatorum.

Management of acute noma requires correction of dehydration and electrolyte imbalance, treatment of associated diseases (such as malaria and measles), testing for HIV infection and appropriate referral, the administration of antibiotics (penicillin and metronidazole are generally effective), local wound care with antiseptics, and removal of any remaining tissue slough and sequestra. No major surgery is performed until the acute stage has been controlled.

Recommendations for the prevention of noma include the inculcation of good nutritional practices, the promotion of exclusive breast-feeding during the first three to six months of life, immunization against endemic communicable diseases, proper oral hygiene practices, the segregation of livestock from human living areas, and education about noma. Yet it is clear that the elimination of the root causes would require improvement in living conditions through the eradication of poverty.

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