

Outcomes at 18 mo of 37 noma (cancrum oris) cases surgically treated at the Noma Children's Hospital, Sokoto, Nigeria

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Background: Noma is a rapidly progressing infection of the oral cavity frequently resulting in severe facial disfigurement. We present a case series of noma patients surgically treated in northwest Nigeria.

Methods: A retrospective analysis of routinely collected data (demographics, diagnosis and surgical procedures undergone) and in-person follow-up assessments (anthropometry, mouth opening and quality of life measurements) were conducted with patients who had surgery >6 mo prior to data collection.

Results: Of the 37 patients included, 21 (56.8%) were male and 22 (62.9%) were aged >6 y. The median number of months between last surgery and follow-up was 18 (IQR 13, 25) mo. At admission, the most severely affected anatomical area was the outer cheek (n = 9; 36.0% of patients had lost between 26% and 50%). The most frequent surgical procedures were the deltopectoral flap (n = 16; 43.2%) and trismus release (n = 12; 32.4%). For the eight trismus-release patients where mouth opening was documented at admission, all had a mouth opening of 0–20 mm at follow-up. All patients reported that the surgery had improved their quality of life.

Conclusions: Following their last surgical intervention, noma patients do experience some improvements in their quality of life, but debilitating long-term sequelae persist.

Keywords: cancrum oris, noma, outcomes

Introduction

Noma (cancrum oris) is a poorly understood, rapidly progressing infection of the oral cavity with a reported 90% mortality rate within 2 wk from the onset of symptoms if untreated.¹ Noma begins as a mouth ulcer and, within days, progresses to oedema of the cheek followed by necrosis and the rapid destruction of the hard and soft tissues of the face.² Treatment with antibiotics, wound debridement and nutritional support in the early reversible stages of the disease greatly reduce mortality and morbidity.³ Noma is thought to be multifactorial in nature.³ The aetiology of noma is currently debated; organisms such as *Fusobacterium necrophorum* and *Prevotella intermedia*^{4,5} have been identified but not consistently.³

If the patient survives the acute stages, the disease can become inactive, after which patients often need complex surgical reconstruction to restore function and improve aesthetics.² Reconstruction often entails rebuilding the lips and cheeks and, in some cases, the eyelids and nose.^{6,7} However, each noma case is unique and, as such, the surgical procedures used to treat noma differ.⁶

Long-term physical sequelae of noma include displacement of the teeth and intense scarring and bony fusion between the maxilla and mandible.^{2,3,6,8–10} Sequelae around daily functioning may include difficulty eating, seeing, talking and breathing.^{2,10,11} The social isolation and mental health sequelae of noma patients have not been documented in the literature but should not be underestimated.

Trismus (restriction of mouth opening) is one of the most disabling sequelae of noma¹² and can lead to complications such as aspiration pneumonia, malnutrition, poor oral hygiene, speech deficits, airway compromise and pain.¹³ Trismus associated with noma can be caused by scarring around the temporomandibular joint capsule (extra-articular ankylosis) or by destruction and scarring of the temporomandibular joint itself (intra-articular ankylosis).^{12,14–17}

Reported surgical techniques to address the spectrum of noma defects include pedicled supraclavicular flaps for the treatment of large unilateral facial defects,¹⁸ abbe, estlander and fan flaps for the reconstruction of the lips and corner of the mouth,^{6,19} forehead, deltopectoral, radial forearm and free flaps for the reconstruction of the cheek^{6,15} and abbe, radial forearm, free, medial forehead and local turnover flaps for the reconstruction of central defects (upper lip and nose).^{6,8,15} Mouth opening is improved by performing bone-bridge excision, sometimes associated with contralateral coronoidectomy.¹⁵

Outcomes of treatment are difficult to ascertain due to inconsistent patient follow-up often due to the remote locations of the home villages of patients. One of the few studies on long-term outcomes of trismus release on 36 noma patients was based in northwest Nigeria and showed that, after a mean follow-up time of 43 mo, results were poor with only 39% of patients showing an improvement in mouth opening.¹²

The Noma Children's Hospital (NCH), northwest Nigeria, run by the Nigerian Ministry of Health and supported by Médecins Sans Frontières (MSF), offers care to noma patients including reconstructive surgical interventions. We present a description of the follow-up of a series of patients treated surgically at the NCH to inform ongoing clinical treatment.

Our study adds to the existing noma literature and is unique as we have followed up with the patients over a longer period (18 mo) in comparison with other studies, which had follow-up periods of 2–6 wk.^{8,20,21}

Materials and methods

We conducted a case series study with patients who had been surgically treated for noma at the NCH more than 6 mo before data collection and who lived in Sokoto or Kebbi states, northwest Nigeria. Data collection took place from April to June 2018.

Data collection

Data collection was performed in two stages. The first stage was based on data collected at admission and for the duration of patients' stay at the NCH (routine data). The second stage of data collection occurred during in-person follow-up visits conducted in the home villages of the patients.

Routine data

The routinely collected data were stored in a bespoke database at the NCH. Information gathered on each of the patients included demographics, diagnosis upon admission (chronic is defined as the absence of ongoing acute infection) and nutritional status.

The NOITULP (nose, outer and inner cheek lining, upper and lower lip) classification system¹⁴ was used by a surgeon with

experience in the treatment of noma to grade patients upon admission. This classification system delineates the extent of orofacial damage and related functional compromise, according to fractional loss of anatomical units ranging from no loss to 100% loss. Trismus categories range from normal mouth opening (>40 mm) to no mouth opening.¹⁴

Surgical data collected included the number and type of surgical procedures and surgical complications (infection, dehiscence, flap necrosis, flap failure). American Society of Anaesthesiologists (ASA) scores were assigned to patients prior to surgery (indicating the fitness of patients before surgery): ASA I, a normal healthy patient; ASA II, a patient with mild systemic disease; ASA III, a patient with severe systemic disease; ASA IV, a patient with severe systemic disease that is a constant threat to life; ASA V, a moribund patient who is not expected to survive without the operation; and ASA VI, a declared brain-dead patient whose organs are being removed for donor purposes. Anaesthesia complications during and after surgery were recorded (difficult airway, hypothermia, equipment failure).

Follow-up data

Data collected included the consenting patients' weight (kg), height (cm), age (y), middle upper arm circumference (MUAC; for children aged 6 mo to 5 y) and maximum mouth opening (mm).

Height was measured using a height board for those aged ≤5 y and a tape measure for those aged >5 y. A manual floor scale was used for weighing all participants. Age estimated to the closest year was self-reported by either the patient (if ≥18 y) or their caregiver. Mouth opening was measured by trained research assistants using a ruler. The maximum mouth-opening measurements were recorded as the mouth wide open minus the mouth closed (incisor-to-incisor where possible or alveolar ridge to alveolar ridge).

We asked questions related to the ability of each patient to eat and drink, their self-reported changes in appearance and how they currently experienced social inclusion in their community. These questions were based on tools used in prior reconstructive surgical studies.^{22–26} Quality of life questions were asked to adult respondents directly and to child respondents if they felt comfortable discussing these issues with the interviewer. If children were aged <7 y or felt uncomfortable talking with the interviewers, the children's caregivers were asked the questions. As part of this quality of life assessment, open-ended qualitative questions were asked to participants about how and if the surgery had changed their lives. These responses were analysed thematically.

Data analysis

Data analysis routine data

A descriptive analysis was conducted. Median and interquartile range (IQR) were reported for non-normally distributed continuous values; means and standard deviations (SD) were reported for values with normal distributions.

For children aged 6 mo to 5 y, MUAC measurements were used to classify the nutritional status of children upon admission as having severe (SAM; MUAC <115 mm), moderate (MAM;

MUAC \geq 115–<125 mm) or global (GAM; MUAC<125 mm) acute malnutrition.²⁷

For children aged 6–15 y, body mass index (BMI) was calculated and gender-specific World Health Organisation (WHO) BMI charts were used to categorise children according to BMI for age. Children were classified as either underweight (BMI<5th percentile), normal (6th–84th percentile), overweight (85th–95th percentile) or obese (>95th percentile).²⁸

For each individual aged \geq 16 y, weight and height were used to classify their BMI range as either underweight <18.5 kg/m², normal 18.5–25 kg/m², overweight 25–30 kg/m² or obese >30 kg/m².²⁹

Data analysis follow-up

Nutritional status at follow-up was assessed by the same means as at admission: SAM, MAM, GAM and BMI were calculated.

Mouth-opening measurements were used to grade patients according to the NOITULP scale trismus categories at follow-up by the research team in conjunction with a consultant-level surgeon.

All analyses were conducted in Stata 15 (StataCorp LP, College Station, TX, USA).

Results

Demographics

We included 37 (82.2%) of the 45 eligible patients. The other eight patients could not be located either due to inaccessible roads or because the individual had moved from the listed village of residence. All patients were alive at follow-up. Of these 37 patients, 21 (56.8%) were male, 34 (91.9%) were from Sokoto state and 12 (34.3%) were aged >15 y at admission, as were 17 (46.0%) at follow-up (the patients age range was between 4–50 y at follow-up). The majority of patients (n = 35, 94.6%) were diagnosed as having chronic noma upon admission. The main reported reasons for seeking care were cosmetic (n = 25; 67.6%) and the related stigmatisation (n = 24; 64.9%) (Table 1). The median number of months between last surgery and follow-up was 18 (IQR 13, 25) mo.

NOITULP classification at admission

At admission, the most severely affected anatomical area was the outer cheek (n = 9; 36.0% of patients had lost between 26% and 50%) (Table 1).

Surgical procedures

Of the 37 patients included in our study, 12 (32.4%) had one surgery, 15 (40.5%) had two to three surgeries and the other 10 (27.0%) had four or more surgeries. In total, 92 surgeries were conducted, during which 125 procedures were performed. The mean duration of each surgery was 90 (SD 49) min. The most frequently used surgical procedure was a deltopectoral flap (n = 16 patients; 43.2%) followed by trismus release (n = 12

patients; 32.4%) (Table 1). No blood transfusions were required for any patients during their surgeries.

Eight surgical complications were noted in seven patients (18.9%); one patient had a superficial infection and an abscess. Complete dehiscence was reported in two patients (5.4%). One (2.7%) of each of the following complications were reported: flap failure, flap necrosis, flap detachment, neck pain needing physiotherapy and an infection on a corner of the mouth. There were no donor site complications and no deaths.

Anaesthesia information

All patients who had data available on the type of anaesthesia received (n = 35) had undergone general anaesthesia. ASA scores were assigned for the 87 surgeries for these 35 patients: 69 (79.3%) surgeries had an ASA score of I, 17 surgeries (19.5%) had a score of II and one surgery (1.15%) had a score of III. Four (10.8%) patients had anaesthesia complications noted during five surgeries. Three surgeries had unanticipated difficult airways (difficulty with facemask ventilation, difficulty with endotracheal intubation); there was one case of hypothermia and one case of anaesthesia equipment failure.

Nutritional status

Of the 37 patients included in the study, 5 (13.5%) had an improved nutritional status at follow-up and the nutritional status of 4 (10.8%) had deteriorated. The other patients had an unchanged nutritional status.

Of the three patients aged between 6 mo and <5 y at admission, two patients (66.7%) were classified as having SAM and one (33.3%) was classified as having GAM. At follow-up, all three patients (100%) were within the normal range, indicating that these three children had an improved nutritional status at follow-up.

In the 6–15 y age group (n = 17), 12 children (70.6%) were categorised as normal weight at admission, 4 (23.5%) of these children were underweight at follow-up and the others (47.1%) were normal. Two patients (11.8%) were underweight at admission: one (5.9%) was classified as normal and the other was still underweight (5.9%) at follow-up. One patient (5.9%) was overweight at admission and normal at follow-up.

For those aged \geq 16 y (n = 17), mean BMI was 19.0 (SD 3.3) upon admission and 18.7 (SD 3.5) at follow-up. The difference between these points is minimal and both measurements fall within the WHO-classified normal range.

Trismus

There were 17 trismus-release procedures conducted on 12 patients (8 patients =1 procedure, 3 patients =2 procedures and 1 patient =3 procedures). At follow-up, the median maximum mouth opening for those aged \leq 15 y (n = 7) was 15.3 mm (IQR 7, 18 mm) and 10 mm (IQR 2, 20 mm) for those aged \geq 16 y (n = 5).

Although we had mouth-opening measurements at follow-up that could be translated into the NOITULP mouth-opening score for all patients, at admission we only had the allocated NOITULP score and not the actual mouth-opening measurements for

Table 1. Respondent characteristics at admission, surgical procedures performed, self-reported quality of life responses and NOITULP scores upon admission, noma case series, n = 37

		n (37)	%
Gender	Female	16	43.2
	Male	21	56.8
State	Kebbi state	3	8.1
	Sokoto state	34	91.9
Patient age upon admission (y)	0–5	13	37.1
	6–15	10	28.6
	>15	12	34.3
Patient education	None	1	2.8
	Arabic studies	34	94.4
	Primary school	1	2.8
Noma diagnosis on admission	Missing	1	
	Acute noma	2	5.4
	Chronic noma	35	94.6
Had treatment for noma prior to coming to NCH	No	22	59.5
	Yes	15	40.5
Kinds of previous noma treatment	Antibiotics	11	73.3
	Traditional	4	26.7
Self-reported comorbidities reported upon admission at the NCH	Malaria	10	27.0
	HIV	0	0.0
Any vaccination before hospital admission	TB	0	0.0
	Measles	14	37.8
	No	17	46.0
Healthcare-seeking reason	Yes	20	54.1
	Cosmetic	25	67.6
NOITULP classification upon admission	Stigmatisation	24	64.9
	Functional disability	14	37.8
Nose	0: no loss	12	50.0
	1: 25% lost	5	20.8
	2: 26–50% lost	5	20.8
	3: 51–75% lost	1	4.2
	4: 76–100% lost	1	4.2
	Missing	13	
Outer cheek lining	0: no loss	5	20.0
	1: 25% lost	5	20.0
	2: 26–50% lost	9	36.0
	3: 51–75% lost	6	24.0
	4: 76–100% lost	0	0.0
	Missing	12	
Inner cheek lining	0: no loss	6	24.0
	1: 25% lost	5	20.0
	2: 26–50% lost	8	32.0
	3: 51–75% lost	6	24.0
	4: 76–100% lost	0	0.0
	Missing	12	
Upper lip	0: no loss	8	36.4
	1: 25% lost	4	18.2
	2: 26–50% lost	5	22.7
	3: 51–75% lost	2	9.1
	4: 76–100% lost	3	13.6
	Missing	15	

Table 1. Continued

		n (37)	%	
Lower lip	0: no loss	10	47.6	
	1: 25% lost	8	38.1	
	2: 26–50% lost	1	4.8	
	3: 51–75% lost	1	4.8	
	4: 76–100% lost	1	4.8	
	Missing	16		
Trismus	Normal mouth opening (>40 mm)	12	54.6	
	Mouth opening 21–39 mm	3	13.6	
	Mouth opening 0–20 mm	2	9.1	
	No mouth opening	5	22.7	
	Missing	15		
Surgical procedures performed on study cohort	Deltpectoral flap	16	43.2	
	Release of trismus	12	32.4	
	Commissuroplasty and lip reconstruction	11	29.7	
	Estlander flap	10	27.0	
	Forehead flap	6	16.2	
	Nasal reconstruction	5	13.5	
	Fan flap	5	13.5	
	Cheek rotation flap	2	5.4	
	Abbe flap	1	2.7	
	Submentalisland flap	1	2.7	
	Other procedures	12	32.4	
	Self-reported quality of life assessment at long-term follow-up	At this point in time, go to school	23	62.2
		At this point in time, I have friends	34	91.9
I am now included in the community		32	86.5	
I can now get married		7	18.9	
I can eat more easily than before the surgery		32	86.5	
I can drink more easily than before the surgery		31	83.8	
People can now understand what I am saying more easily than before the surgery		32	86.5	
I feel more happy with the way I look than before the surgery		32	86.5	

eight patients. As such, a comparison of the preoperative and postoperative mouth-opening NOITULP classification for these eight patients is provided in Table 2. No patient had a normal mouth-opening status at the follow-up visit (all classified as T2, 0–20 mm) (Table 2).

Quality of life at follow-up

All respondents reported that the surgery had improved their quality of life in one way or another. Respondents reported that the surgery led to decreased social isolation (having friends, $n = 34$, 91.1%; being included in communal activities, $n = 32$, 86.5%) and functional improvements (eating more easily, $n = 32$, 86.5%; improvements in speaking, $n = 32$, 86.5%).

At the follow-up visits, patients and caregivers were asked if and how the surgery had changed their lives. Some feedback from patient caregivers was negative and they did not want any further care:

The surgery was not successful and I do not want to come back to the hospital (8-y-old patient).

Other caregivers reported difficulties with restricted mouth opening and related functional issues:

The opening of the mouth is very small making it difficult to eat or drink (29-y-old patient).

There was some mixed feedback, showing improvements in quality of life but continued difficulty with mouth opening:

The mouth opening is a bit difficult because the side stitches are tight. However, she can eat and talk well (28-y-old patient).

And some other patients reported positive functional changes and social acceptance:

Table 2. Mouth-opening categories upon admission and follow-up by age group

Patient	Age at follow-up, y	NOITULP score admission	Maximum mouth opening at follow-up, mm	NOITULP score at follow-up
1	7	T2	13	T2
2	7	T3	7	T2
3	8	T1	18	T2
4	8	T0	15	T2
5	10	T3	5	T2
6	18	T3	20	T2
7	20	T3	2	T2
8	22	T2	10	T2

T0, normal mouth opening: >40 mm; T1, mouth opening: 20–40 mm; T2, mouth opening: 0–20 mm; T3, no mouth opening.

He was shy and angry before the surgery and he is now able to eat and go to school. He used to not be audible but now he talks loud and clear (10-y-old patient).

He is very happy to have the treatment and he can meet different people since the wound is closed and healed (10-y-old patient).

Discussion

Our findings suggest that surgical care for noma patients improved their quality of life, despite minimal evidence that trismus had improved. This corroborates findings from an Ethiopian study, which showed that post-operative follow-up revealed significant improvement in the lives of noma patients³⁰. Most of the patients had more than one surgery, and the most commonly used procedure in our cohort was the deltopectoral flap, which is utilised in the reconstruction of the cheek, the most severely affected anatomical area.

Our study confirms the complexity and unique manifestation of noma and the need for numerous surgical procedures to obtain an acceptable functional result. The most commonly performed procedures were the deltopectoral flap, release of trismus, commissuroplasty and lip reconstruction, estlander flap, forehead flap and nasal reconstruction. The procedures performed rely on regional flaps and local tissue and are in line with other reported surgical techniques used to treat noma.^{6,8,15,18,19,31,32} Other providers have described the use of free flaps^{33,34} however, given the technical and resource demands of microsurgery, these techniques are not currently utilised at the NCH. The surgical programme instead enlists older but reliable reconstructive techniques that are less risky in this context.

Noma cases are at a high risk of developing trismus resulting in difficulties in speech, chewing, and maintaining healthy oral hygiene practices.¹² This study has highlighted that despite surgical intervention, none of the patients in this cohort regained a normal mouth opening. Respondents reported ongoing concerns with their restricted mouth opening and the impact this restriction had upon their lives. Similar studies with noma patients

noted that the results of trismus release in noma patients was extremely poor.^{12,16} A northwest Nigerian study reported that 43 mo after surgery, the mean mouth opening of 36 patients was 10 mm.¹² A similar study with 95 patients from Niger and Burkina Faso reported that after 3 y, the mean mouth opening was 21 mm.¹⁵ Our results were similar to these studies and showed that all patients had a mouth opening of between 0–20 mm at follow-up (median mouth opening of our cohort: 15 mm for those aged ≤15 y; 10 mm for those aged >16 y).

This outcome is not unique to noma. The overall success rates in curing trismus in inflammatory processes other than noma (oral submucous fibrosis, chronic non-bacterial osteomyelitis) in the paediatric population can be low.^{35,36} In the majority of noma patients (as well as in patients with other inflammatory causes of trismus), the trismus is extra-articular.¹⁵ Noma can disrupt the peri-articular bony tissues of the subcondylar area, which is the location of the growth plate of the mandible. Other researchers in mandibular trauma have also reported that this disruption of the growth plate has resulted in increased fibrosis and trismus,^{13,36–38} and we can hypothesise that this is also a contributory factor in noma. Similarly, this area of the mandible can be disrupted by surgery to correct trismus (such as gap- or inter-positional arthroplasty).^{20,35} This disruption is mitigated by delaying trismus release until the mid-teen years, to avoid, or at least decrease, the need for revision trismus surgery. Delaying trismus-release surgery until skeletal maturity is likely to be effective for noma patients, to reduce the postsurgical reduction in mouth opening. This delay should only occur if a child with noma can drink and consume sufficient calories to continue to grow and gain weight. While there are no specific rules or recommendations for trismus release and ankylosis repair in the literature, in studies and meta-analyses demonstrating high success rates for inflammatory causes, the patients are typically in their teenage years.³⁵ The success rates for paediatric trismus surgery are far higher in congenital and traumatic cases, where the surrounding capsule and other structures are unaffected.³⁵ A further step that has been shown to maintain adequate mouth opening and resulting quality of life after trismus-release surgery is longer term postoperative physiotherapy which can enable patients to have

an improved mouth opening.^{15,35} We do not have physiotherapy treatment or uptake information for this study; however, the NCH program currently includes rigorous physiotherapy following trismus release while at the hospital, patients are also provided with an exercise list to complete when at home after discharge and they are assessed by the physiotherapist at the follow-up appointments at the hospital.

Our study has illuminated three potential changes for program planning and potential interventions. Firstly, as the anaesthesia complications were notable, it is important that surgical teams should include anaesthesia professionals who have specialised in difficult airway management. Secondly, as some patients remained malnourished, it is important to conduct nutritional follow-up with all patients. Lastly, vaccine preventable diseases and a lack of vaccines are a risk factor for the development of noma³. The inadequate vaccination status of the patients included in this study shows that these populations should be the target for public health interventions, which could reduce the number of noma cases along with a host of other diseases.

There were several limitations to this study. Most patients reported improvements in their quality of life; however, these questions were only asked at follow-up. To improve our assessment of quality of life changes, it would be beneficial to use a standardised, validated assessment tool at admission and follow-up.³⁹ Social desirability bias could have influenced these answers, changing our understanding of patient-reported outcomes. The addition of preoperative and postoperative photographs to this study would have been beneficial. Furthermore, the retrospective review of routinely collected data limited the type and quality of data available for analysis. We have implemented a prospective study to assess the outcomes of patients that will address many of the weaknesses of the current study.

Following their last surgical intervention, noma patients do experience some improvements in their quality of life, but debilitating long-term sequelae persist. Reconstructive surgery does appear to restore form and function in some patients. However, noma is a preventable condition that, if detected early, can be effectively treated with antibiotics before the devastating consequences described in this cohort of patients occur. Therefore, public health interventions should prioritise strategies which address known risk factors for noma through a community-based health systems approach that targets prevention, early detection and the rapid treatment of acute noma.

Authors' contributions: EF, MA, AL and KB were responsible for the study concept and design. LT conducted the retrospective chart review with assistance from EF and MJO. EF and MJO collected the remaining data. EF conducted the statistical analysis. EF, MA, RW, AOT and AL interpreted the findings. EF, MA, RW, AOT and AL drafted the manuscript. All authors critically reviewed the manuscript.

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Competing interests: All authors declare there are no competing interests.

Ethical approval: The MSF, Usmanu Danfodiyo University Teaching Hospital and Sokoto and Kebbi State Ministries of Health Ethics Review Boards approved the study protocol. All participants were treated in accordance with the ethical principles of the Helsinki Declaration. Adult participants and caregivers of children participants provided written consent for participation in the study, and children aged between 7 and 18 years provided written assent. For illiterate individuals, a thumbprint was requested.

Data availability: MSF has a managed access system for data sharing that respects MSF's legal and ethical obligations to its patients to collect, manage and protect their data responsibility. Ethical risks include, but are not limited to, the nature of MSF operations and target populations, being such that data collected often involve highly sensitive data. The dataset supporting the conclusions of this article is available on request in accordance with MSF's data-sharing policy (available at: <http://fieldresearch.msf.org/msf/handle/10144/306501>). Requests for access to data should be made to data.sharing@msf.org.

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