UMA DOENÇA NEGLIGENCIADA E RARA: QUAL A RELAÇÃO DA NOMA COM A CIRURGIA PLÁSTICA?

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A NEGLECTED AND RARE DISEASE: WHAT IS NOMA'S RELATIONSHIP WITH PLASTIC SURGERY?

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ABSTRACT

Objective: The aim of the study was to conduct a narrative review, describing the relationship between Noma and plastic surgery.

Methods: The search strategy used databases such as Pubmed, Scielo, and Lilacs to gather information about Noma and its relationship with plastic surgery. Inclusion criteria considered studies on Noma, surgical treatment, clinical outcomes, languages (English, Portuguese, and Spanish), and study designs. Exclusion criteria eliminated studies with non-surgical treatments, non-facial surgical techniques, lack of surgical outcomes, non-human case reports, and treatments for complications from previous surgeries.

Results: It was found that reconstructive plastic surgery results in improvements in patients' quality of life, with advances in both objective and subjective outcomes following interventions. However, the study also highlighted a discrepancy between the number of case reports and the scarcity of prospective studies that use classifications to assess intervention outcomes. Of the reviewed articles, 64% were case reports, highlighting the need to establish more rigorous research to better understand the implications of Noma treatment. The classifications, NOITULP and PRO, proved essential in surgical practice, not only for planning reconstructions but also for assessing the impact on patients' lives.

Conclusion: Noma is a rare and neglected disease, with devastating facial sequelae. The importance of reconstructive plastic surgery lies in restoring physical functionality and providing a more positive self-image, combating suffering and social isolation.

Keywords: Noma, plastic surgery, reconstruction, quality of life, neglected diseases, facial deformities.

INTRODUCTION

Noma is a little-known disease in Brazil, but it was described many years ago. The first description of Noma as a clinical entity in children originated from Carolus Battus, a surgeon in Dordrecht, the Netherlands, in his 'Handboeck der Chirurgijen' *(1,2).* In the nineteenth century, it was well known that malnutrition and measles could lead to Noma. Its incidence decreased in Western Europe at the end of the 19th century. There is good reason to assume that this was related to improved nutritional and hygienic conditions of the poorest population *(2)*. On the other hand, the active treatment of Noma only became possible after the decrease in cases, due to the discovery of penicillin and the development of reconstructive surgery *(2)*. In less developed countries, however, Noma remained what it was: the "true face of poverty," and it was Michael Tempest, who discussed the benefits of penicillin application and reconstructive surgery in his study of Noma in Nigeria *(3)*.

Noma is a rapidly progressive opportunistic ulcer disease with a higher prevalence in sub-Saharan Africa and developing countries *(4)*. Without any treatment, Noma is a deadly disease in 90% of cases *(4).* The condition begins in the oral mucosa and spreads rapidly through the soft and hard tissues of the face. Unlike necrotizing fasciitis, Noma does not

respect the planes of the fasciae: it penetrates and destroys the deeper tissues of the facial muscles, jaw, maxilla, lips and nasal structures, which makes it an extremely aggressive but poorly visible disease *(5,6)*.

Therefore, the study was to conduct a narrative review, describing the relationship between Noma and plastic surgery, and bringing to light this disease so neglected today by countries in which it is not prevalent.

METHOD

The initial bibliographic research was carried out in August 2024 in the Pubmed, Scielo and Lilacs databases.

The search strategy used in Pubmed was: ((((("noma"[MeSH Terms]) OR ("gingivitis, necrotizing ulcerative"[MeSH Terms])) OR ("neglected diseases"[MeSH Terms])) AND ((((("face/surgery"[MeSH Major Topic]) OR ("plastic surgery procedures"[MeSH Terms])) OR ("surgical flaps"[MeSH Terms])) OR ("surgery, plastic"[MeSH Terms]))) AND ((((("noma/complications"[MeSH Major Topic]) OR ("cicatrix"[MeSH Terms])) OR ("quality of healthcare"[MeSH Terms])) OR ("pathologic processes"[MeSH Terms])) from the initial search resulted in a total of 67 articles.

In Scielo it was: ((noma) OR (cancrum oris) OR (gangrenous stomatitis)) resulted in 16 articles.

And in Lilacs it was: (mh:(noma)) OR (mh:("gingivitis, necrotizing ulcerative")) OR (mh:("neglected diseases")) resulted in 9 articles.

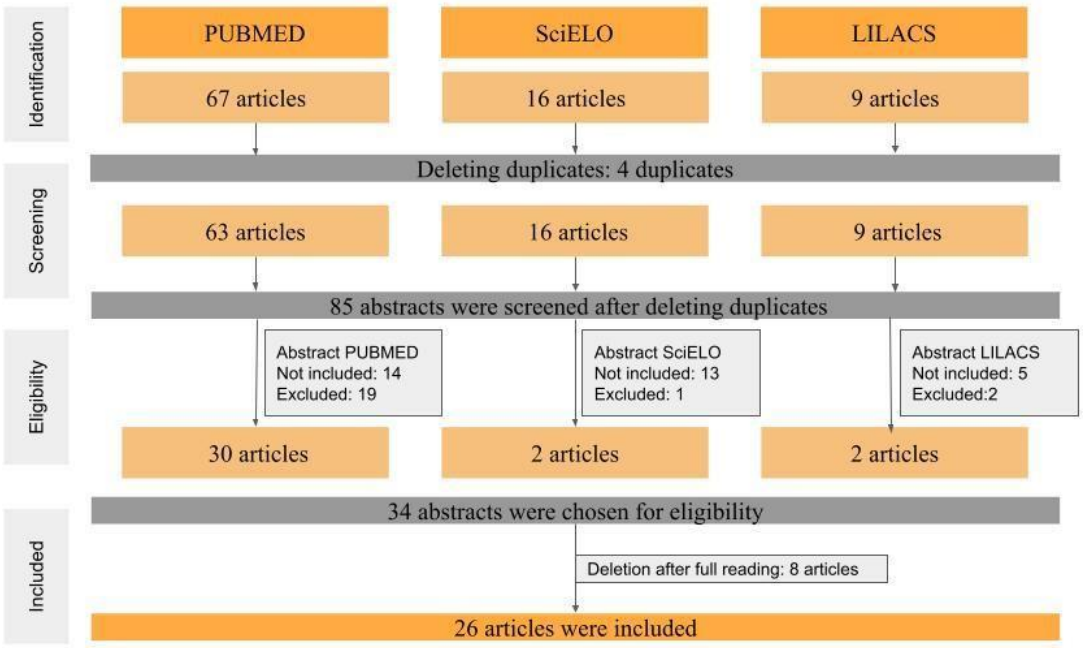
The titles and abstracts of the articles identified by the literature search were manually selected using the following inclusion and exclusion criteria:

Inclusion Criteria

The eligibility for inclusion of the article was (*4)* studies that address Noma; (7*)* presence of surgical treatment of Noma; (*8)* presence of patient clinical outcomes; (*6)* articles in English, Portuguese and Spanish; *(9)* study design: case reports, case cohorts, case-control studies and randomized controlled trials. Prospective and retrospective studies were included; and (*10*) made accessible the reading of abstracts and the full text.

Exclusion Criteria

Studies were excluded if they met the following criteria: (*4*) performed associated non-surgical alternative treatments, such as herbal medicines and medicinal herbs; (*7*) involved non-facial surgical techniques; (*8*) did not mention results of surgical treatment of Noma; (*5*) deals only with surgical technique of head and neck reconstruction; (*6*) case report of Noma in non-humans; *(9)* case report on treatment of complication of previous surgery of Noma.



Source: Elaborated by the authors.
Fig. 1 - Table representing the selection of articles for the preparation of this study.

EPIDEMIOLOGY

The World Health Organization (WHO) estimates an incidence rate of 140 cases of Noma per year *(9)*. These data are not reliable, as there is a precarious notification of diseases in the most incident countries. Most cases occur within the "Noma Belt", that is, in the southern Sahara region and in scattered locations around the world where malnutrition, Human Immunodeficiency Virus (HIV) and Acquired Immunodeficiency Syndrome (AIDS) are endemic *(11, 12, 13)*. Apart from these regions, Noma had notoriety in the Auschwitz concentration camps and Japanese prisons during World War II. The patients reported were all victims of prolonged malnutrition associated with other debilitating diseases *(9, 14)*.

The acute form of Noma is most common in children aged 2 to 6 years *(4)* with extreme malnutrition and is often preceded by immunosuppressive diseases such as measles and malaria. In particular, children weaned from breastfeeding are a vulnerable group *(15)*.

ETIOLOGY

The etiology of Noma is not yet fully understood, but several factors have been indicated as possible causes. It is believed that a compromised immune system, poor oral hygiene, injuries to the gingival mucosal barrier, and the presence of an unknown bacterial agent may contribute to the development of the disease *(16)*. Furthermore, there is evidence that Noma has an infectious origin, involving bacteria such as *Fusiformis fusiformis*, *Borrelia vincentii*, *Bacteroides melanogenicus*, and *Pseudomonas aeruginosa*, which are saprophytic microorganisms of the oral cavity that can cause opportunistic infections in vulnerable populations *(17)*.

The main risk factors associated with Noma include poor socioeconomic conditions, malnutrition, immunosuppression (such as HIV infection), inadequate oral hygiene, and poor sanitation. Furthermore, diseases such as leukemia, measles, and herpes virus infections may also increase susceptibility to the disease. The interaction of these factors facilitates the onset and progression of Noma, making its prevention a significant challenge in vulnerable communities *(16)*.

PATHOPHYSIOLOGY

Necrotizing gingivitis occurs with the rupture of the oral mucosa, extending to the exposure of the alveolar bone, followed by perforation of the cheek or lips, and culminating in gangrene.

Although the cause and pathogenesis of Noma remains confusing, it was never described before the eruption of deciduous teeth. Often, the disease is unilateral and associated with premolar teeth. This suggests that some of the perforating cheek injuries may be caused by pressure necrosis when children rest with their cheek pressed against a mattress or against their mother's back while they are carried. Continuous pressure may be sufficient to devitalize the skin, as occurs in decubitus ulcers, and to cause the rupture of the barriers against bacterial colonization *(15)*.

CLINICAL PICTURE

The initial stage of Noma begins in the oral cavity and spreads to other regions *(3)* and has symptoms such as: localized pain, sialorrhea, bad breath and cheek swelling *(18)*. Usually, the Noma extends to the cheek, infraorbital margin, palate, nose and anthro-mastoid. The masseter, medial and lateral pterygoid, and temporal muscle are involved only occasionally *(19)*. Noma may affect unilaterally or bilaterally *(20)*. There are some cases reported in the medical literature involving scalp, neck, shoulders, perineum and vulva *(21, 22)*. The lesions start with painful, purplish or reddish macules that turn into hardened papules and may be on the gingiva, alveolar margin, and premolars and molars. The lesion progresses rapidly to necrosis and ulcers and reaches the underlying bones. Bone involvement is extensive and may involve most of the mandible or maxilla, or both *(15)*.

A necrotic mass forms with a central portion covered by microorganisms and inflammatory cells surrounded by viable tissue *(21)*. The lesion may be cone-shaped with an intraoral base and the external appearance does not necessarily correlate with the internal extent of the lesion *(14)*. In most cases, the destruction of internal tissue is more extensive than the externally visible lesions. Furthermore, patients with Noma usually seek medical care only in advanced stages of the disease.

CLASSIFICATION

Classification of "NOITULP" *(23)*

The classification of NOITULP described in 1998 *(24)* evaluates the involvement of the tissue according to the anatomical units affected: nose (N-*nose*), outer cheek (O - *outer cheek*), and inner lining of the cheek (I - *inner lining*), presence of trismus (T - *trismus*) that is not an anatomical unit, but a very important functional impairment with severe implication for intubation, upper lip (U - *upper lip*), lower lip (L - *lower lip*) and (P - *Particularities*) that can be used for specific problems (for example, bone loss or loss of an eye) (*24)*. The severity of tissue loss is assessed as 0 for no loss, 1 for up to a quarter of the anatomical unit, 2 for up to half, 3 for up to three quarters, and 4 for almost complete loss of the anatomical unit. For

trismus, the score describes the loss of ability to open the mouth (T0 = normal, T1 = opening less than 4 cm, T2 = less than 3 cm, T3 = opening less than 2 cm, T4 = complete trismus).

		0	1	2	3	4
N	*Nose*	0	¼*	½*	¾*	Complete loss
O	*Outlining*	0	¼*	½*	¾*	Complete loss
I	*Inner lining*	0	¼*	½*	¾*	Complete loss
T	*Trismus*	Full opening of the mouth	< 4 cm	< 3 cm	< 2 cm	< 1 cm or ankylosis
U	*Upper lip*	0	¼*	½*	¾*	Complete loss
L	*Lower lip*	0	¼*	½*	-	Complete loss
P	*Particularities* ♦	0	-	-	-	

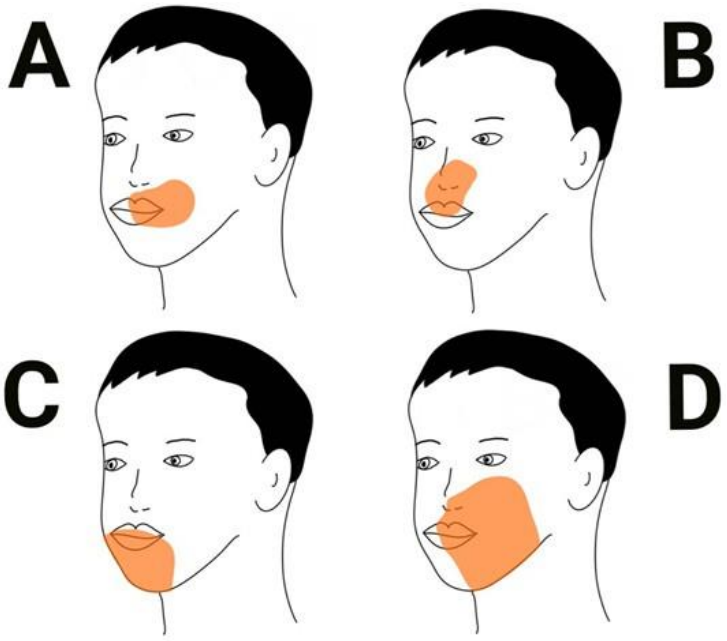
* Fraction corresponding to part of the anatomical unit

♦ Specific impairments (e.g. bone loss or loss of an eye)

Source: adapted from Pistre, 2001[23]
Fig. 2 - Classification of NOITULP.

Noma lesion classification *(25)*

The classification of Noma lesions takes into account the location of soft tissue defects and their extent, as well as the underlying bone defects. It is divided into 4 types (Fig. 3):



Source: Elaborated by the authors.
Fig. 3 - Types of Noma injury.

- Type A: Localized cheek and commissural defect.
- Type B: Includes upper lip and nose, alveolar edge, and palate.
- Type C: Includes lower lip and may include the complete jaw and floor of the mouth.
- Type D: Major defects, which may include the entire cheek, lips, maxilla, palate, and malar bone. It can extend to the eyelid and nose.

SEQUELAE

When a Noma patient survives the acute stage of infection, tissue loss due to necrosis results in a process of contraction of scar tissue accompanied by a remaining defect in the tissue, such as a hole in the cheek. As Noma occurs in younger children, it can also lead to severe facial skeletal growth disorders *(2)*. The sequelae varies in severity and may include severe facial disfigurement, trismus and ankylosis, oral incompetence, speech problems and dental misalignment known as "dental anarchy" *(26)*. In many cases, rehabilitation requires extensive and complex reconstructive surgical procedures *(8)*.

The main sequelae of Noma are facial mutilations and are usually extensive, which causes social isolation and immense suffering for patients *(2)*.

Trismus and ankylosis are seen with some frequency in patients with Noma (close to 40%). False ankylosis, i.e. extra-articular ankylosis are the most common forms of ankylosis in Noma *(2)*. If the disease occurs before the onset of dentition, a bone fusion between the maxilla and mandible may occur, resulting in a bone ankylosis *(27)*.

TREATMENT

Managing the patient

Treatment of acute Noma is simple: hospitalization, intravenous replacement for correction of electrolyte imbalances, nutrition, and wound care. Untreated Noma has a mortality rate of 90% *(28, 29)*. This rate drops to 10% when treated, which happens in the minority of cases *(30)*.

The primary therapeutic approach for trismus and ankylosis in patients with Noma is physical therapy. However, many patients still fail to maintain proper mouth opening. Although coronoidectomy is recommended as a treatment option for trismus in other conditions, it is not indicated for patients with Noma. In this context, the pathophysiology of the disease involves tissue loss and scar contracture between the jaws, rendering coronoidectomy ineffective for treatment. Furthermore, the surgical release of scar contracture or osteotomy of bone ankylosis, when performed without reconstruction of the soft tissue reconstruction, results in the formation of new granulation tissue and scar contracture, leading to high rates of trismus recurrence. *(27)*

Surgical intervention

Correction through plastic surgery aims at removing scar tissue, releasing any ankylosis and facial reconstruction after associated disfigurement. Reconstruction is by a combination of simple skin graft, local or distant flaps, and pedicled or free. *(13,16, 31, 32)*

Reconstructive surgery should begin as soon as the acute phase of infection is over and the patient is stabilized through nutritional support, electrolyte balance, and rehydration. This can usually be achieved, on average, in two to three months of treatment. The principle of facial reconstruction in patients with Noma sequelae is: remove all scar tissue, recreating the original tissue defect, and reconstruct with well-vascularized flaps. The extent of tissue loss is

often underestimated, for this reason, the "Sokoto Law of Noma surgery" was formulated as follows: the required flap extent is always greater than planned *(2)*.

Surgical approaches for the treatment of Noma include different techniques aimed at facial reconstruction and improving the functionality of affected patients. Regarding surgical approaches for Noma treatment, the deltopectoral flap (DP) is the primary type of flap used. The deltopectoral flap is a regional pedicled flap that, although first described in 1917 for nasal reconstruction and later in 1965 for esophageal reconstruction, was replaced half a century ago by free tissue transfer. However, surgical decision-making is influenced by the conditions of each setting, and the lack of intensive care units (ICU) has led to free tissue transfer not being the first-line option for correcting surgical defects in resource-limited environments. Despite being outdated, the deltopectoral flap has many desirable qualities for Noma reconstruction. It is safe, simple, quick to perform, does not require ICU, involves minimal blood loss, and is reliable. Thus, the DP flap is an effective "workhorse flap" for larger Nomas with defects in the cheeks and lips and, most importantly, should be used in resource-limited settings *(33)*.

These approaches are determined according to the patient's condition and the constraints of the surgical environment. The priority is to ensure effective and safe procedures, even in contexts with limited resources. In conclusion, this emphasizes the necessity for facial reconstructive surgeons to adapt their techniques according to resource limitations, ensuring safe care for those in need. *(34)*

PREVENTION

Noma prevention involves promoting national awareness of the disease, reducing poverty, better nutrition, promoting exclusive breastfeeding in the first 3 to 6 months of life, optimal prenatal care, and timely immunizations against common childhood diseases *(35)*.

DISCUSSION

For those who have survived a devastating disease with 90% mortality, correcting facial deformities represents a source of hope — something difficult to measure in metrics such as quality-adjusted life years. When a patient returns to their place of origin after a successful facial reconstruction, it strengthens trust in the community-wide medical system *(33)*. Patients in under-resourced communities can spend their entire lives without traveling more than a day away to their homes. As a consequence, many are unaware of the possibility of surgical treatment. Given this lack of awareness, the dissemination of information becomes a fundamental element for the success of Noma surgery *(33)*. Although it is a disease little noticed in the world, promoting the discussion about Noma is to find better treatments that seek not only to return the physical functionality of individuals living with this defect, but also to offer an opportunity for these patients to enjoy a satisfactory self-image *(36)*.

The main consequence of Noma is facial mutilations, usually of great extent, resulting in deep suffering and social isolation for patients *(2)*, for this reason, the importance of facial reconstructive surgery in improving patients' quality of life. Among the main approaches to facial reconstructive surgery in the treatment of Noma discussed so far, the deltopectoral flap is the main technique, being a safe, simple and effective regional pedicled flap, especially useful in resource-limited environments, where free tissue transfer is not feasible. In addition, other locoregional flaps may be included, such as temporal parietal fascia flap, cervicofacial rotation, Estlander, Abbe, and submental island. Staged procedures, such as pre-lamination and pedicle division, are indicated for complex cases. The release of trismus is necessary when scars prevent the opening of the mouth, which may involve the excision of scar tissue

and reconstruction with flaps. In cases of bone fusion, corticectomy is performed, and interpositional arthroplasty is used for ankylosis of the temporomandibular joint. The techniques are adapted to the patient's conditions and the limitations of the environment, ensuring safe and effective reconstructions even in situations with few resources *(34)*.

A scoping review conducted in 2021, found that out of the 147 articles published between 1843 and 2021, that mentioned Noma, 94 (64%) were case reports, and no treatment trials were identified *(37)*. An example of this can be observed in studies such as those by Marck (1999), Hartman (2006), Daya (2009), Bonilla (2004), Adolph (1996) and Adeyemo (2011) that consists of case reports and series with short-term follow-up. The first prospective study was published in March 2024 in the journal Facial Plastic Surgery & Aesthetic Medicine, which analyzed the treatment outcomes of screened patients based on the NOITULP and PRO (Patient-Reported Outcomes) classifications and showed an improvement in objective and subjective outcome scores after patients underwent Noma surgery, however the NOITULP score did not normalize. After surgery to treat Noma, several improvements in patients' quality of life were observed. The study results indicate that NOITULP and PRO scores demonstrate a more positive experience after surgical intervention in several areas of life. These findings underscore that reconstructive surgery not only corrected physical deformities, but also provided significant benefits to patients' quality of life and overall well-being *(33)*. Therefore, despite the severity of the disease, there is a significant discrepancy between the large volume of case reports and series with short-term follow-up and the scarcity of prospective studies using objective classifications to assess outcomes.

The risk factors for Noma include low nutrition status, lower socioeconomic status, and poor oral hygiene *(38)*. The treatment of Noma requires more precise measurements, both surgical and quality of life, as well as the development of better tools to improve outcome research. In addition, the reliability of assessments can be strengthened through specific training on the use of scoring, including discussion of cases among professionals. Finally, despite the severity of the disease, there remains a shortage of prospective studies evaluating long-term postoperative follow-up *(39)*.

CONCLUSIONS

Noma is a rare and neglected disease, leaving facial sequelae of great variability and often devastating consequences for its survivors. Score such as NOITULP and PRO demonstrate that categorizing these defects is essential for more accurate planning of surgical reconstructions, as well as for assessing the impact of interventions on patients' quality of life. The use of these tools aligns with the practice of plastic surgery, where patient satisfaction and aesthetic improvement are fundamental criteria for success. Furthermore, it is crucial for reconstructive plastic surgery to foster debate aimed at developing more effective treatments and ensuring the success of Noma reconstructive surgeries. These treatments restore the physical functionality of affected individuals, providing a more positive and satisfying self-image. Additionally, interventions aim to restore dignity, well-being, and quality of life, combating the suffering and social isolation often caused by the disease. In this way, reconstructive plastic surgery becomes a source of hope in the fight against Noma, offering a real chance for physical and emotional rehabilitation for patients.

CONFLICT OF INTEREST

Nothing to declare.

AUTHOR`S CONTRIBUTIONS

Creator: Araújo RS. **Conception and design of the study:** Paek CEJK. **Acquisition and interpretation of data:** Paek CEJK; Burle MIBDM; Saad FA. **Manuscript preparation:** Burle MIBDM; Cheberle PHM. **Manuscript writing:** Paek CEJK; Burle MIBDM; Saad FA; Cheberle PHM. **Critical revision:** Ferreira LM.

DATA AVAILABILITY STATEMENT

All data sets were generated or analyzed in the current study.

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ATTACHMENT 1 – README

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This README file was generated on 2024-08-20 by Pedro Henrique Moreira Cheberle

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GENERAL INFORMATION

Dataset Title: A NEGLECTED AND RARE DISEASE: WHAT IS NOMA'S RELATIONSHIP WITH PLASTIC SURGERY?
Dataset DOI:

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Data collection date: 2024-08-18

Keywords: Noma, plastic surgery, reconstruction, quality of life, neglected diseases, facil deformities

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DATA & FILE OVERVIEW

List of Files:
1. noma_article.pdf - Full review of Noma's narrative review
2. figure_1.csv - Table representing the selection of articles for the preparation of this study
3. figure_2.csv - Classification of NOITULP
4. figure_3.pdf - Types of Noma injury.

Relationship between files: CSV files complement the tables presented in the main manuscript.

File sizes and formats:
- PDF: 525KB
- CSV: 1KB each (UTF-8)

Are there multiple versions of the dataset? No

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SHARING/ACCESS INFORMATION

Licenses/restrictions placed on the data: CC-BY 4.0

Access conditions: Free access

Links to other publicly accessible locations of the data:

Links/relationships to auxiliary datasets: Exclusive to SciELO Data

Were the data derived from another source?
- SciELO, Pubmed and Lilacs

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PRIVACY & ETHICS INFORMATION

Privacy considerations: All data is in public domain

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PARTICIPANT CONSENT

Not applicable

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METHODOLOGICAL INFORMATION

Description of methods used for data collection/generation: Narrative review with systematic search in PubMed, SciELO and Lilacs databases using MeSH terms and strategies detailed in the manuscript.

Methods for data processing: Extraction and qualitative synthesis of data from selected articles.

Specific instrument or software information needed to interpret the data:
- Zotero 6.0.30 for reference management
- Excel 365 for data tabulation

Standards and calibration information, if appropriate: Checking according to the inclusion/exclusion criteria present in the manuscript

Environmental/experimental conditions: Not applicable

Describe any quality assurance procedures performed on the data: Pubmed, Scielo e Lilacs

People involved with sample collection, processing, analysis, and/or submission: Anonymous

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DATA-SPECIFIC INFORMATION FOR: noma_article.pdf

Variables:

Missing data codes: "NA"

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SPECIFIC INFORMATION FOR THE DATA: figure_1.csv

Variables:
1. Database (text): PubMed, SciELO or Lilacs
2. Search_terms (text): Search terms used
3. Articles_identified (integer): Number of articles found
4. Articles_included (integer): Number of articles selected

==========

SPECIFIC INFORMATION FOR THE DATA: figure_2.csv

Variables:
1. Anatomy (text): N-Nose, O-Outer cheek, I-Inner cheek, T-Trismus, U-Lip Upper, L-Lower lip, P-Particularities
2. Score_0 (text): Description for score 0
3. Score_1 (text): Description for score 1
4. Score_2 (text): Description for score 2
5. Score_3 (text): Description for score 3
6. Score_4 (text): Description for score 4

Missing data codes: "NA"

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SPECIFIC INFORMATION FOR DATA: figure_3.pdf

Variables:
1. Image (text): A, B, C, D
2. Anatomical area (text): Lateral perioral region (A), Central perinasal region (B), Lower lip region (C), Left lower hemiface (D)

Missing data codes: "NA"

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FILE SIZE AND FORMAT

File size: 527KB

File format: PDF, CSV

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DOCUMENTATION & SUPPORT

Links to additional documentation: Not applicable

Support information: Contact the principal investigator via email

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