Cancrum oris — a 35-year retrospective study

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Objectives. To determine the factors predisposing to cancrum oris and its frequency, clinical features, treatment and outcome. Cancrum oris is a gangrenous condition of the face usually occurring in poorly nourished children in a Third-World setting.

Design. Retrospective review of clinical data on children with cancrum oris.

Setting. All children with cancrum oris seen over a 35-year period, from 1960 to 1995, at Red Cross War Memorial Children's Hospital (RXH) in Cape Town.

Subjects. Twenty-six patients (16 female and 10 male) with cancrum oris. Their average age at presentation was 4 years and 4 months (range 1 - 15 years).

Results. The peak incidence was between 1971 and 1975 (11 cases); subsequently only 1 new case was seen at RXH in every 5-year period. Most children were referred from rural areas. Associated conditions could only be determined in 11 of the 26 patients. In order of frequency, these were malnutrition, gastro-enteritis, measles and anaemia. Twenty-three of the 26 children had soft-tissue involvement affecting the lips, cheek, chin, nose or other structures. Eighteen had bony or cartilaginous involvement; the maxilla was affected in 15, the palate in 7, the vomer in 5, the mandible in 3 and the nasal septum in 7. Three children had bony involvement only, the soft tissues being spared. Operative records were available for 18 children. These 18 children had a total of 84 operations, with an average of 4.7 per child (range 0 - 12).

Conclusions. Cancrum oris is a devastating condition affecting malnourished children. Reconstruction is complex and demanding, involving both soft tissue (23 of 26 cases) and bone (18 of 26 cases). Most children require multiple procedures. Prevention is best effected by comprehensive primary health care.

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Cancrum oris is a gangrenous condition of the face, usually occurring in poorly nourished children in a Third-World setting. It is also known as noma, from the Greek meaning 'to devour'. The gangrenous process usually affects the mouth,[1] but it may also involve the jaw, nose, cheek and maxilla, destroying both soft tissue and bones (Fig. 1), and

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even extend up to the orbits and mastoid.[1] The condition is devastating for the child and his or her family. In children who survive, the gangrenous destruction of the face poses a reconstructive challenge to the plastic surgeon.

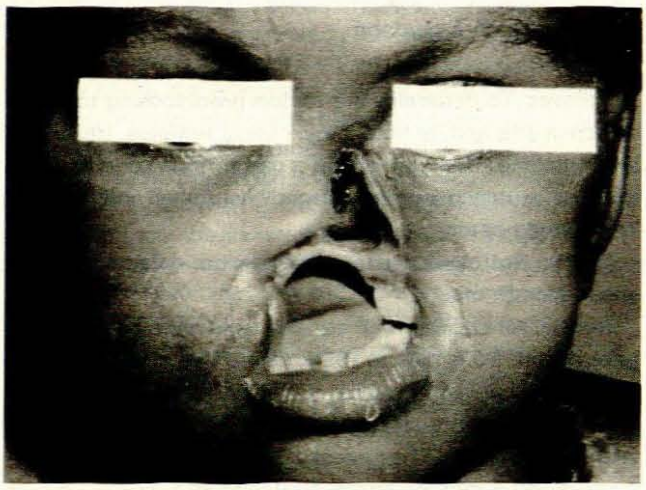

Fig. 1. Child with gross facial destruction as a consequence of cancrum oris.

Noma was known to Hippocrates, Galen and other ancient practitioners. Tourdes (1848) (cited by Tempest[1]) recognised it as a gangrenous affectation of the mouth which attacked especially those children whose constitution was altered by poor hygiene and serious illness such as exanthematous rashes. He noted that noma began as an ulcer of the mucous membrane with oedema of the face and extended from inside outwards, rapidly destroying the soft tissue and bone; it was almost always rapidly fatal.[1] Although cancrum oris is primarily a disease of children, it has also been described in adults. Dawson (1945) (cited by Tempest[1]) described the disease in victims of the Nazi concentration camps. It has also been reported in immunosuppressed adults.[2-4]

This study reports on the management and outcome of 26 children seen at a major paediatric referral hospital between 1960 and 1995.

Patients and methods

A retrospective study of 26 patients with cancrum oris seen at Red Cross War Memorial Children's Hospital (RXH) during the 35 years from 1960 to 1995 was undertaken. The hospital records, clinical photographs and radiographs, where available, were studied. Demographic data, the temporal and geographical referral pattern, associated conditions, the anatomical site and type of lesion and the number and type of surgical operations performed on these children were analysed.

Results

Age and sex distribution

There were 16 girls and 10 boys. The average age at presentation was 4 years and 4 months (range 1 - 15 years). Most patients were referred for reconstruction, the active disease having been treated by the primary referral hospital. Twenty out of 24 patients (83%) were under the age of 7 years at the time of presentation.

Referral pattern with time and geographical location

The number of cases referred to RXH per 5-year period is represented in Fig. 2. It can be seen that the peak incidence occurred between 1971 and 1975 (11 cases). The incidence declined so that subsequently only 1 new case was referred for reconstruction every 5 years.

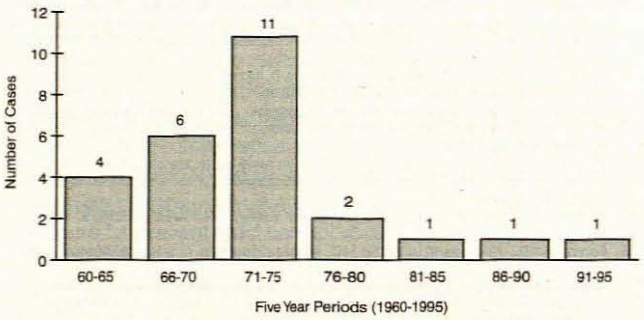

Fig. 2. Number of new cases per 5-year period.

Three children lived locally and another 3 within 300 km of Cape Town. Eleven children had to travel over 600 km to get to RXH; of these, 6 had to travel over 1 000 km. Referral was predominantly from rural communities.

Associated conditions

In 11 cases the records were adequate to determine associated conditions. The most significant were malnutrition (< 80% of expected weight), gastro-enteritis (frequently complicated by dehydration and acidosis) and antecedent measles (Table I). Multiple associated conditions often coexisted in a single patient.

Table I. Associated conditions in 11 cases of cancrum oris

Associated conditions	No. affected
Underweight for age	7
Gastro-enteritis	4
Measles	3
Anaemia (microcytic, hypochromic)	2
Pneumonia	1
Mucormycosis	1
Osteogenesis imperfecta	1

Anatomical areas involved (Table II)

Cancrum oris affects the face, usually causing extensive tissue destruction (Fig. 1). Of the 26 children, 23 (88%) had soft-tissue involvement and 18 (69%) bone and/or cartilage destruction. Eight patients (31%) had only soft-tissue lesions, the bones and cartilages being spared, while 3 children (12%) had destruction of bone only (the maxilla). Fifteen of the 26 children (58%) had combined soft-tissue and bone or cartilage involvement.

Table II. Tissue destruction in 26 cases of cancrum oris

Soft tissue (23 patients)			Bone and cartilage (18 patients)					
Lips	Cheek	Nose	Nasal septum	Lateral maxilla	Pre-maxilla	Palate	Vomer	Mandible
17 (65.4%)	9 (34.6%)	8 (30.7%)	7 (26.9%)	9 (34.6%)	8 (30.8%)	7 (26.9%)	5 (19.2%)	3 (11.5%)

Treatment

Therapy was usually commenced by the original referring hospital. This consisted of nutritional support, measures needed to combat any associated conditions, correction of anaemia, treatment of any infections with antibiotics, and the cleansing and sometimes debridement of wounds.

Operative records were available for 18 patients. These 18 patients required a total of 84 procedures, an average of 4.7 operations per patient. One patient healed without surgery while, at the other extreme, another patient required 12 operations. The number of operations performed per patient is reflected graphically in Fig. 3.

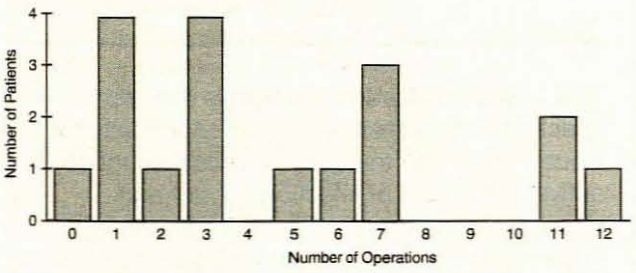

Fig. 3. Number of operations per patient.

The type and number of operations done is shown in Table III. Of the 21 flaps performed, 10 were loco-regional flaps and 11 were distant pedicled flaps. The loco-regional flaps used were two Estlander flaps, two nasolabial flaps, two gull-wing alar flaps, a forehead flap, a pharyngeal flap, a Limberg flap and a triangular skin flap.

Table III. Type and number of operations done*

Type of operation	No. done
Debridements and sequestrectomies	11
Examination under anaesthesia	2
Flap delays	8
Flaps	21
Insetting, division of pedicle and flap refinement	18
Bone grafts	4
Split skin grafts	4
Revisions	19
Other (drainage of abscess; tarrsorrhaphy)	2

* On occasion more than one procedure were done under a single anaesthetic.

Of the 11 pedicled flaps used, 6 originated from the neck. The remaining donor areas were abdomen, back, chest (deltopectoral flap), arm and head. The most recent patient's reconstruction was done using a pedicled composite temporoparietal flap as a two-stage procedure. This successfully closed a large palatal fistula and improved her mid-face projection.

Discussion

Cancrum oris starts in the oral cavity, probably as a gingivitis in the pre-molar or molar area.[1] The lesion may then ulcerate and spread to the buccal or lingual sulcus and to the mucosa of the lip or cheek. Progression of the destructive process will result in extension through to the skin. The greater involvement of the interior of the oral cavity with the lesser involvement of the overlying skin has resulted in the lesion being described as a '*cone gangreneux*'.[1] Once the slough separates, sequestration of exposed bone and teeth occurs.[1] The process is rapid and the defect formed is typically well-defined. Microbiological organisms, *Fusiformis fusiformis* and *Borrelia vincenti*, are believed to play a role in the development of cancrum oris. Anaerobic organisms are also thought to be of importance.[5] The rapid onset of secondary infection and the delayed presentation of cases makes bacteriological studies difficult.[1]

Twenty six cases of cancrum oris were seen at RXH over the 35-year period 1960 - 1995. Like other authors,[1,5] we found the disease to be slightly more prevalent in girls than in boys (1.6:1). Young children are primarily affected: at the time of presentation, 83% were under the age of 7 years. This is in accordance with the findings of others.[1,5-7]

Malnutrition was the most important associated condition, documented in 64% of children studied. Other authors have found malnutrition to be the most important predisposing factor.[1,6] Cancrum oris has been found to be associated with malnutrition that occurs around the time of weaning, and the condition therefore occurs infrequently in children under the age of 1 year. Measles was also an important condition associated with cancrum oris, occurring in 3 out of 11 cases. Others[1,6] have documented the importance of measles as a predisposing factor. Measles, like malnutrition, depresses T-cell immunity and infection may thus arise from opportunistic overgrowth in an immunosuppressed patient.[8] Other associated conditions were gastro-enteritis and anaemia.

The soft-tissue defects in our series affected the lips in about two-thirds of cases (17 out of 23), the cheek in about a third (9 out of 23) and the nose in another third (8 out of 23). Of the bones involved, the maxilla was commonly affected (15 out of 18 cases), whereas the mandible was rarely diseased (3 out of 18 patients, 2 of whom also had maxillary destruction). Most children in our series had severe functional and cosmetic deformities. Because of soft-tissue and bony destruction, oral incompetence, salivary leak and maceration of skin were frequent. Fistulas were both orocutaneous and oronasal, with 7 children having palatal destruction. Erosion into the maxilla frequently resulted in

exposure of the maxillary sinus. Pain, scarring and contracture can result in trismus and bony ankylosis of the temporomandibular joint with resultant limitation of mouth opening.[9] Cosmetic problems seen were destruction of the mouth and lips, cheek and even the whole nose.

Early management should involve particular attention to the correction of malnutrition, the treatment of associated conditions, antibiotics where indicated, and initial debridement and cleansing of wounds. The aim of initial treatment is to minimise the destructive process and return the child to a condition of fitness to allow reconstruction. Reconstruction can only be undertaken once the disease is quiescent, infection eradicated and the child well nourished and fit for surgery.

The reconstructive requirements are complex. Soft- and bony tissue loss, fistulas (orocutaneous, palatal), trismus and the cosmetic deformity all need to be addressed. No single 'standard' procedure is sufficient for all. Each reconstruction must be carefully planned and individualised. Usually all three layers — skin, skeletal support (bone, cartilage) and mucosal lining — are deficient and need to be provided. Local flaps should be used wherever possible, because they provide better colour and texture match. If the defects are too extensive, distant flaps may have to be considered. Newer plastic surgery techniques such as tissue expansion, prefabrication of flaps and the use of microsurgical tissue transfers may facilitate reconstruction. Anaesthetic difficulties, especially those related to poor mouth opening, may be overcome by the use of nasal or endoscopic intubation. Children frequently require multiple procedures:[6,7] our patients had a total of 84 operations between them (4.7 operations per patient).

Preventive measures, as elucidated by authors working in Nigeria, where the disease is prevalent, may be applicable to South Africa.[6] They suggest that the provision of adequate nutrition, a comprehensive vaccination programme, education and better dental and general health care is the crux of prevention. In addition, better primary health care, especially in rural areas, is of importance in the prevention of this disease.

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