# **Buruli ulcer disease**



Epidemiology and transmission Clinical manifestations and diagnosis Treatment



## **Buruli ulcer disease**

### <u>KEY POINTS</u>

- Buruli ulcer is an infection caused by an organism called *Mycobacterium ulcerans* that lives in the environment: in lakes, swamps, and rivers.
- The disease is most frequent in children (>50% of cases) and others living or working close to water.
- The mode of transmission is unknown, although evidence suggests that trauma to contaminated skin may be one of the means by which the organism enters the body.
- The disease manifests itself on the skin as papules, nodules, plaques, oedematous areas, or ulcers.
- In general, early cases present with nodules, while ulcers occur later in the course of the disease.
- A course of antibiotics (rifampicin and streptomycin/amikacin) given for 8 weeks cures small lesions and reduces the area of the larger lesions prior to surgical excision.

he following information is summarized from the manual *Buruli* ulcer – management of *Mycobacterium* ulcerans disease, published by the World Health Organization (WHO) in 2001. For more detailed information, please read that manual, or contact WHO by e-mail at *buruli@who.int*, or visit the WHO web site at *www.who.int/gtb-buruli* 

#### **Epidemiology and transmission**

The prevalence of the disease is not accurately known. The disease exists or is suspected in over 30 countries, but the majority of cases occur in West Africa. Buruli ulcer is caused by a slow-growing, environmental mycobacterium called *Mycobacterium ulcerans*, whose toxin causes tissue necrosis.

The disease is more severe in impoverished people living in remote rural areas. Children under the age of 15 years make up more than 50% of those affected. Mortality due to the disease is low but morbidity (disability and deformity) is high. Some of the complications of the disease are secondary bacterial infection, extensive scarring, contractures, deformities to the limbs, amputations, and involvement of the eye, breast, and genitalia. In some areas, 20–25% of those with healed lesions are reported to have disability causing long-term socioeconomic burdens.

The disease occurs most frequently among people who live or work close to rivers and slow-moving bodies of water. The construction of irrigation systems and dams seems to have influenced the resurgence of the disease. The mode of transmission is not known, although some evidence suggests that aquatic insects (*Naucoris* and *Dyplonychus*) may be involved, and that trauma to contaminated skin may be one of the means by which the organism enters the body. Health education programmes encourage communities to identify the disease early by the detection of nodules, and to care for wounds and injuries by washing and dressing them. In addition, individuals are encouraged to protect their bodies against injury by wearing appropriate clothing and shoes or boots.

#### **Clinical manifestations and diagnosis**

The disease manifests itself as **papules**, **nodules**, **plaques**, **ulcers**, and **oedematous** areas of the skin (*Figure 2.1*). A new case is a person with no previous history of treatment for Buruli ulcer. A recurrent case is a person presenting within one year with a further lesion at the same site or at a different site. Recurrence rates after surgical treatment are 16% for persons presenting early and 28% for persons presenting late. Recurrence at the same site may be due to inadequate excision. Recurrence at a different site may be due to haematogenous or lymphatic spread.

Initially, there is a non-ulcerative phase which, if untreated, will usually progress to an ulcerated phase. Both phases end with a scar – the complications of which will depend on the location and extent of the lesion, and wound-management techniques. In addition, individual healing factors and the surgical technique used for the removal of necrotic tissues and skin grafting will influence the type and extent of scar formation (*Figure 2.2*).

#### **Non-ulcerative lesions: early diagnosis**





Nodule 🔺

Oedema with beginning of ulceration

#### **Ulcerative lesions: late diagnosis**





Ulcer with osteomyelitis

Figure 2.1 Non-ulcerative and ulcerative Buruli ulcer lesions

Figure 2.2 Example of scars limiting function and causing deformity

#### **Scaring following ulcerated lesions**



Limited eye closure (lid gap) 🔺



Limited elbow extension and flexion 🔺



Limited knee extension and foot dorsiflexion



Deviation of the trunk



Deviation of the hand



Deviation of the foot 🔺

In a known endemic area, an experienced health worker can make a diagnosis of BU based on clinical observation that considers the following clinicoepidemiological features:

- the person lives in or has travelled to a known endemic area;
- most cases are in children under 15 years of age;
- about 85% of lesions are on the limbs; lower-limb lesions are twice as common as upper-limb lesions.

In addition to the clinical diagnosis, at least one of the following laboratory findings is required to confirm the diagnosis of BU:

- acid-fast bacilli (AFB) in a smear stained by the Ziehl-Neelsen technique;
- histopathological study of a biopsy specimen showing typical necrosis and acid-fast bacilli;
- positive polymerase chain reaction (PCR) test for *M. ulcerans*; and/or
- positive culture of *M. ulcerans.*

#### Treatment

Recent research and clinical experience have shown that a combination of rifampicin and an aminoglycoside (streptomycin or amikacin) given for eight weeks is promising in the management of *M. ulcerans* disease. In early cases, it may be curative. In more advanced cases, such treatment probably reduces the area that will require subsequent surgical excision.

Prior to surgical management of the lesions, POD interventions such as correct antideformity positioning, splinting, controlling oedema, movement, and active exercising should be started – these interventions must not be delayed until surgical excision of the lesions.

Limited supplies of drugs, lack of surgical experience, inadequate surgical facilities, poor wound management, delays in skin grafting, prolonged hospitalization, high treatment costs, and the risk of recurrence after surgical treatment are the practical difficulties that may affect the provision and uptake of treatment.

#### **Review questions**

- **1.** What causes Buruli ulcer?
- 2. In which countries is Buruli ulcer found?
- **3.** How is Buruli ulcer transmitted?
- 4. How is Buruli ulcer diagnosed and classified?
- 5. What are the early signs of Buruli ulcer?
- 6. What is the treatment for Buruli ulcer?