



## Dracunculiasis

Dracunculiasis is an infection caused by the nematode *Dracunculus medinensis*, also known as the guinea fire worm.

*D medinensis*

is in the order Spirurida, an order of parasites that includes the filariae

*Wuchereria bancrofti*, *Brugia malayi*,

and

*Loa loa*.

During the last 25 years, concerted efforts to eradicate the guinea worm have been undertaken and these have resulted in a reduction of more than 99% of worldwide cases of dracunculiasis.

Current disease incidence is low and is limited specifically to sub-Saharan Africa. The Centers for Disease Control and Prevention (CDC) proposed a global campaign for eradication of dracunculiasis in 1980, and, in 1988, numerous African ministers of health set a target date of 1995 for total eradication. After that target was missed due to slow mobilization in countries with endemic disease, a target date of 2009 was set. Unfortunately, despite considerable progress, that date was also not met. By the end of 2008, dracunculiasis was endemic in 6 countries

(Ethiopia, Ghana, Mali, Niger, Nigeria, and Sudan), and the number of cases decreased 52% (from 9,585 in 2007 to 4,619 in 2008).<sup>1</sup> Sporadic violence and civil unrest in Sudan and Mali poses the greatest threat to the final eradication of dracunculiasis.

During ancient times, the presence of dracunculiasis can be inferred by the universally recognized symbol of medicine, the Greek *asklepios* (ie, Roman *aesculapius*), which consists of a one-headed snake wrapped around a stick. Dead female worms have also been found in Egyptian mummies older than 3000 years, and writings in ancient Sanskrit, Greek, and Hebrew refer to *Dracuncul*  
*us*

infection. To this day, the most effective method dracunculiasis treatment involves extraction by wrapping the worm around a stick

The term *dracunculus* is Latin for "little dragon," a misnomer and reference to the symbol. Thus, when the guinea worm disappears, one of the original inspirations for the discipline of medicine will also disappear. Currently, the infection persists and, although uncommon, can cause significant morbidity.

Ingestion of water that contains infective *Dracunculus* larvae causes the infection. The larvae reside in an intermediate host, a tiny fresh-water crustacean or copepod of the genus *Cyclops*

. The acidic environment of the stomach and duodenum kills the copepods. The larvae are subsequently released in the stomach or small intestine and penetrate the mucosa to mate and mature in the abdomen or retroperitoneal space approximately 60-90 days after initial infection. The maturation stage can last for up to 1 year, and, during this time, the adult male probably dies because only the female worm is recovered from symptomatic patients.

After maturation is complete, the female *Dracunculus* reaches a length of up to 1 m (with a thickness of only 1-2 mm) and slowly migrates from the GI tract into subcutaneous tissue, usually to a location in the lower extremity. The actual route of migration is unknown. In this subcutaneous location, one or more females prepare larval exit sites through the skin, from whence larvae may be released into another water supply. Free-living larvae can survive only 3

days without a host; they become infective after 2 weeks (2 molts) within the host copepod.

Death due to dracunculiasis is not caused by the primary infection and occurs only in cases in which secondary infection of the worm's exit site leads to sepsis . The mortality rate is quite low; however, morbidity is a major concern, with secondary infection being the most common complication. Cellulitis or the formation of an abscess requires prompt attention, and pain from the exit sites often can incapacitate patients for weeks. This is usually observed in individuals who have multiple worms and rely on their ability to stand or walk for their livelihood. Farmers with untreated dracunculiasis in Nigeria have been found to miss work for up to 3 months. Infected schoolchildren may miss up to 25% of the school year. Therefore, *Dracunculus* infection can cause significant socioeconomic burden for individuals and communities.

Another, more chronic, complication of dracunculiasis is encapsulation of the adult worm, which occurs when the calcified remains of the worm persist in the extremity of the patient. This can result in chronic pain and intermittent swelling of the extremity. In a small percentage of individuals who have permanent scarring or deformity of the lower extremity, even after the worm has been extracted, chronic pain may persist for as long as 18 months. Notably, on average, infected individuals have multiple worm extrusions at the same time (1.8 worms per person, on average). Rarely, dracunculiasis can present with worms located in anomalous locations, including the lungs, pancreas, testes, spinal cord, or periorbital tissue.

- Travel to or residence in endemic countries is invariably part of the history in patients with dracunculiasis.
- Recollection of ingestion of unfiltered or untreated water, ingestion of fresh fruits or vegetables washed with such water, or bathing or swimming in potentially contaminated water are all possibly elicited in the patient's history. The transmission of the disease has seasonal variation. In arid areas, the rainy season, with increased availability of surface water, coincides with most cases. In wet areas, the dry season, when sources of drinking water are limited, is associated with most cases.
- History tends to be useful only to confirm the diagnosis after it has been presumed based on physical examination findings.
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- A blister forms in the epidermis at a site chosen by the female worm, usually in the lower extremity.
- Immediately before blister formation, allergic-type symptoms, such as mild respiratory distress with wheezing, urticaria, periorbital edema, and pruritus, are often present.

- Patients may also be febrile during this period.
  - With the emergence of the worm's head, the blister grows and becomes erythematous at its periphery.
  - Edema occurs around the site, and inflammation of the papule causes further pruritus and burning pain.
  - Usually, after a few days, but possibly as long as 2 weeks, the blister erupts, and the worm releases a collection of larvae-containing fluid.
  - The swelling and pain often are markedly decreased after the blister is opened.
  - At this point, an ulcer forms around the blister site as the adult worm continues to emerge.
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- Definitive diagnosis is made when the head of the worm is identified within the ulcer.
  - As noted, the ulcer tends to become secondarily infected.
  - No other particular physical findings are commonly noted, although some degree of lymphadenopathy may be found at any stage of the illness.

## Causes

- Dracunculiasis is an infection caused by the nematode *D medinensis*.
- The larvae from *D medinensis* are not infective unless a molting process within the copepods occurs. This requires a fresh-water environment; thus, water ingestion is the only identified mode of transmission.

The following studies are indicated in dracunculiasis:

- CBC count with differential
  - The WBC count is likely elevated, even if only slightly.
  - The differential commonly indicates eosinophilia.
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- Serum immunoglobulin levels
  - Immunoglobulin E (IgE), immunoglobulin G1 (IgG1), and immunoglobulin G4 (IgG4) levels are usually elevated, with variability depending on the stage of disease.
  - Patent infections (immediately following blister eruption but before ulcer formation) cause the greatest elevation of the 2 IgG subclasses, whereas both are relatively less elevated with postpatent (ulcerated) or prepatent (blister in formative stage) infections.

- A radiologic examination (plain-film roentgenography) of the lower extremity may prove useful in the identification of calcified worms in the rare case when surgery is considered. Incidental identification of calcified lesions from dracunculiasis has also been reported after radiographic evaluation of a painful lower extremity

## **Medical Care**

- The most common practice to treat dracunculiasis still involves wrapping the worm around a stick.

- The adult worm is extracted from the patient using a stick at the skin surface and wrapping or winding the worm a few centimeters per day.

- This slow process can take many days and, in some cases, up to a few weeks, but it is required to avoid breakage and leaving behind a portion of the worm.

- Metronidazole or thiabendazole (in adults) is usually adjunctive to stick therapy and somewhat facilitates the extraction process. However, one study found that antihelminthic therapy was associated with aberrant migration of worms, resulting in infection in areas other than the lower extremity. Therefore, such medications should be used with caution.

- The worm also can be excised surgically where such facilities are available

## **Antiparasitic agents**

These agents are used to speed the pace of worm extraction.

### **Metronidazole (Flagyl, Protostat)**

DOC as therapy adjunctive to extraction. Active against various anaerobic bacteria and protozoa. Intermediate-metabolized compounds formed bind DNA and inhibit protein synthesis, causing cell death.

**Adult**

250 mg PO tid for 10 d

**Pediatric**

25 mg/kg/d PO divided tid for 10 d; not to exceed 750 mg/d

Cimetidine may increase toxicity of metronidazole; may increase effects of anticoagulants; may increase toxicity of lithium and phenytoin; disulfiramlike reaction may occur with orally ingested ethanol

Documented hypersensitivity; first trimester of pregnancy

**Pregnancy**

B - Fetal risk not confirmed in studies in humans but has been shown in some studies in animals

**Precautions**

Do not use during first trimester of pregnancy; use caution with history of hepatic disease or concurrent hepatotoxic drugs; use cautiously with coagulopathies, history of retinal or visual changes, or CNS dysfunction

**Thiabendazole (Mintezol)**

Acceptable for use in adults only.

Inhibits helminth-specific mitochondrial fumarate reductase; alleviates symptoms of trichinosis during invasive phase.

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**Adult**

50-75 mg/kg/d PO divided bid for 3 d; not to exceed 3 g/d

**Pediatric**

Not recommended