

Podoconiosis and endemic non-filarial tropical elephantiasis – tropical lymphoedemas can be managed effectively in community settings

Podoconiosis is a significant public health burden in tropical areas in Africa, India and Central and South America.

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Podoconiosis is a non-infectious lower leg lymphoedema, not to be confused with filariasis. It occurs following long-term barefoot exposure to volcanic red soils found in tropical highland areas with heavy annual rainfall. It leads to a significant public health burden in at least 10 countries across tropical Africa, North India and Central and South America. In Ethiopia alone there are thought to be at least 1 million cases.^[1] Although the condition has been known for more than a millennium, it has been neglected and under-researched. It has recently been designated neglected tropical disease status by the WHO.^[2] Podoconiosis does not occur in all barefoot farmers in such red clay soils, only in a genetically susceptible group.^[3] Management of established cases can be achieved with simple low-cost intervention and prevention, achieved when genetically at-risk individuals avoid prolonged contact with the triggering soil type. If genetically susceptible children regularly wore shoes, the disease would be eradicated.

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Clinical features

This geochemical obliterative endolymphangitis typically presents with onset of leg swelling in individuals in their early twenties, although it may start as early as the age of ten.

There are three common symptoms noted at this early stage:

1. The burning limb – an episodic complaint of nocturnal intense burning or itching of one foot and the lower leg is reported and may typically follow a period of intense activity (long walking journey the day before) and/or alcohol ingestion. Pain may extend into the thigh and be associated with tender femoral lymph nodes or fever and is therefore known as adenolymphangitis (ADL). Subsequent episodes typically affect the same limb. Onset in the other limb may not occur for many months or years. These attacks resolve spontaneously or after a few days of rest and elevation of the offending limb. Medical input is seldom sought at this stage, although the limb gradually increases in size during this phase.
2. Itching of foot – persistent or intermittent itching of the dorsum of the foot is noted, often over the first or second web space. Constant scratching of this area leads to a reactive thickening of the

skin known as ‘lichenification’, as seen in stasis or venous eczema. Repetitive scratching may lead to cracking of the skin, allowing entry of cutaneous pathogens. Secondary infection further complicates matters.

3. Knocking together of the big toes during walking is noted, due to splaying of the forefoot – one of the early signs of lymphoedema of the foot. In the early stages the clinical signs are subtle, but recognising them enables timely intervention and prevention of progression.

These features could occur following any cause of lymphoedema. The following all point towards podoconiosis rather than lymphatic filariasis: the onset on one side many months before the other side is affected; the persisting lack of symmetry, with one leg being affected significantly more than the other; and the lack of involvement of the scrotum with lymphoedema.

Examination reveals a number of signs:^[4]

- Swelling of the foot: (a) unilateral splaying of the forefoot due to deep oedema between metatarsal heads; (b) block toes – the swelling of the forefoot causes the toes to resemble sausages, lacking their usual curvature; (c) plantar oedema manifested by a doughy, diffuse swelling on the soles of the feet, with one side affected more than the other.
- Skin changes over the foot: (a) lichenification – the skin becomes diffusely thickened secondary to scratching, although a more doughy thickening can occur which renders the skin – particularly that overlying the first toe web space – stiff, so that it cannot be lifted up (Stemmer’s sign); (b) increased skin markings – obvious longitudinal skin markings may be evident and exaggerated by squeezing the toes together; the markings are striking between the first and second toes; (c) damp skin follows the transcutaneous ooze of serum and leads to the persistent presence of flies; (d) mossy foot – because of underlying dilated lymphatics, the surface of the skin produces excess keratin in a rather classic way that visually resembles ‘moss’, although it feels very hard and rough to the touch.

... splaying of the forefoot – one of the early signs of lymphoedema of the foot.

Eventually the increased leg diameter persists and established lymphoedema sets in after a series of burning episodes associated with

Podoconiosis



Impact of podoconiosis on young life.



Simple washing technique using local material.



Podoconiosis patient graduating through microfinance programme as a market trader.



Graduated patient teaching new patients what to do.



Early swelling of foot.



Simple elevation and compression bandaging reduces the size of the limb in a few days.



Large foot with nodules.



Mossy changes on dorsum of foot.



Shoes made by patients for patients.



Woody fixed swelling in advanced disease.

swelling. As with any underlying trigger of lymphoedema, from filariasis to leprosy, the clinical features can vary from a predominantly soft, squashy or lymphoedematous variant to a thickened, leathery, rough, predominantly

fibrotic or elephantiasis-like appearance. Most cases show a combination of both, but the predominant type varies little within each case, apart from the degree or extent, unless complications ensue.

Staging

In order to categorise disease severity and measure the impact and response to treatment, a simple field staging system has been developed that classifies patients

according to the extent of reversibility of their swelling (Appendix 1).

Management of the established lymphoedematous patient

This aspect shares many similarities with the management of any lymphoedematous patient. Consequently, sharing of intervention methods across disease management programmes that deal with other causes of lymphoedema seems prudent, such as joining the morbidity management arm of the Global Alliance to Eliminate Lymphatic Filariasis [GALEF].^[5] Approaches are detailed below.^[6]

The swollen limb

Addressing the swollen limb involves a constellation of approaches, all of which, although simple on their own, can be highly effective cumulatively. These considerably improve the quality of life of the patient by reducing the limb size and decreasing the number of episodes of infection or ADL. Sophisticated and expensive techniques are largely unavailable in endemic areas, so the focus is on more practical, low-cost methodologies that are accessible in the resource-poor settings of the majority of podoconiosis patients.

These simple methods include the following:

- Cleansing – washing the affected limb daily with soap and water and, if possible, antiseptic, greatly reduces the

bacterial load on the skin and minimises the presence and influence of infective agents.

- Emollient – moisturising the limb daily after cleansing improves the barrier function of the skin and softens it, preventing it from cracking, which in turn reduces the risk of transcutaneous acquisition of infection.
- Elevation of the limb to above hip height for prolonged periods leads to improved venous and lymphatic return and considerable reduction in limb size, especially in the water bag-type lymphoedema.
- Breathing and exercise – deep breathing aids in emptying the thoracic duct into the large veins above the level of the heart. This enhances lymphatic drainage distally. This central lymphatic clearance is essential before encouraging more peripheral manoeuvres to increase lymphatic drainage. Gentle isotonic exercise further stimulates both peripheral venous and lymphatic drainage of the affected limbs.
- Compression produced by bandages and hosiery (stockings or socks) is important in lymphoedema management. However, in resource-poor settings, these items are often unavailable owing to lack of trained personnel and materials.
- Manual lymphatic drainage – light superficial massage can greatly enhance lymphatic drainage. De Godoy and de Godoy^[7] have produced an accessible training manual, detailing this technique, for use in the field in Brazil.
- Surgery – extensive surgical techniques involve the removal of redundant tissue, skin grafting and then prolonged limb elevation during recovery. Although not simple, this can produce good results in the short term with reduction of limb size; however, this rarely lasts and the consequent scarring can produce further problems of its own. More recently, simple shave excision of hard nodules with secondary intention healing works surprisingly well, even in a resource-poor setting. The technique leads to healing within a relatively short time and is acceptable in terms of pain control and popular with patients in terms of outcome.^[8] The Debra Markos Community Podoconiosis Programme in Ethiopia exemplifies what is possible with basic surgical facilities (personal

observation). Surgical management of the fibrous nodules has recently been reported in Japan.^[9]

- Footwear – the concomitant wearing of appropriate and protective footwear forms an essential part of the approach to the lymphoedema patient. Once limb reduction has been achieved, safeguarding the foot and limb with robust and supportive hosiery and protective footwear forms a key role in preventing and slowing down recrudescence of the condition by avoiding further exposure and providing protection from injury and secondary infection.

Prevention of recurrence

While wearing shoes contributes to preventing relapse of limb swelling in the treated lymphoedematous patient, additional measures include avoiding prolonged contact with the soil responsible for the condition. In a community of barefoot farmers it is important to develop a skill for recovered patients to enable them to earn a living off the land. This arm of management, known as vocational training in some projects, runs training programmes to teach alternative 'trades' such as shoe-making (often the first for a new project so that treated patients can start producing custom-made shoes for the programme), hairdressing, sewing, barbering, bicycle repairing, as well as microfinance projects to enable patients to select their own approach to a non-farming income. So far in Ethiopia these have all been successful in delivering alternative earning schemes for the recovered patient.

Stigmatisation and misunderstanding

One study from Northern Ethiopia recorded that 13% of patients reported stigmatisation at school, work place or market as well as being excluded from marriage or shunned within marriage.^[10] The Mossy Foot Treatment and Prevention Association in Soddo, Southern Ethiopia, has been developing strategies on how to deal with this since their start-up more than 10 years ago. The results are such that members of the public now approach project workers in the street on market days asking if they might have 'podo', whereas 10 years ago project workers were not even allowed to visit the homes of affected patients for fear of tainting the rest of the family by association. This has been achieved through a mixture of public

meetings where project workers and eloquent, treated patients have undertaken educational talks within communities, e.g. at religious gatherings and in schools. Other Ethiopian podo treatment projects have set up patient forums and community leader meetings to develop a locally appropriate strategy for education and stigmatisation reduction.^[11]

Elimination of podoconiosis globally

Podoconiosis can easily be prevented by wearing shoes that protect the feet from the soil at all times. Podoconiosis is thus a completely preventable disease, and with appropriate and simple interventions could be eradicated completely. However, there are several challenges to this. An excellent study exploring public perceptions of podoconiosis in Northern Ethiopia^[10] found that fewer than half of the population of a community with a large number of podoconiosis patients (7.5% of those surveyed were patients and 40% had a family member with podo) thought that the disease could either be prevented or controlled.

Effective community management models in improving quality of life and outcomes for patients

The majority of programmes have begun in Ethiopia, where the condition is especially prevalent and where they have achieved national recognition over the past 10 years of dedicated advocacy programmes. The Mossy Foot Treatment and Prevention Association in Soddo, Ethiopia, has been evaluated using the WHO's Innovative Care for Chronic Conditions Framework: Building Blocks for Action, launched in 2002 for evaluating non-communicable disease projects.^[11] Part of the conclusion of this review was how effective it is to transform previous patients into community agents for podoconiosis. These individuals become channels for information and models for motivation for the community. Additionally, this project has been assessed as effective in stigma reduction.

The Debre Markos Community Podoconiosis Programme has generated and published specific lessons learnt for the establishment of community-led podoconiosis programmes, which are aimed at informing any subsequent set-up for both podoconiosis as well as other tropical lymphoedema programmes. The authors specifically emphasise the need

for baseline data, effective training, local leadership, experience sharing, mass awareness, involvement with other sectors such as water and waste management, and integration with government health systems.^[12]

Conclusion

Podoconiosis is a common but neglected tropical geo-genodermatosis leading to dramatic lymphoedema/elephantiasis in barefoot farmers in the tropics. It can be managed effectively with simple interventions of washing, moisturising, reducing infection, and wearing of compression bandages and suitable shoes. It is completely preventable if the genetically at-risk wear shoes all their lives and do not expose their skin and feet to the geologically triggering soil.

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Appendix 1

Podoconiosis Staging Sheet (podoconiosis-endemic areas)

Staging for field workers^[13]

Instructions

The field worker is expected to look at *and examine* the right and left leg of each patient in turn and give a score to each leg separately.

- 'Swelling' here means a general increase in size of part of the foot or leg.
- 'Reversible swelling' here means a swelling that is not present when the patient first gets up in the morning but becomes more marked as the day advances.
- 'Persistent swelling' here means a swelling that is present all the time.
- 'Knob or bump' here means a discrete, hard lump that can be seen or felt to protrude from the rest of the foot or leg.
- 'Ankle' here means the level of the two ankle bones when the patient is standing.
- 'Knee' here means the level of the top of the knee cap when the patient is standing.

In addition to the numerical stage, the field worker should measure the greatest below-

knee circumference and record the presence (M+) or absence (M) of mossy changes. For example, if a patient's right leg has irreversible below-knee swelling, nodules below the ankle, mossy changes around the heel and a circumference of 48 cm, the staging should be recorded as Stage 2, M+, 48.

Stage 1. Swelling reversible overnight. The swelling is not present when the patient first gets up in the morning.

Stage 2. Below-knee swelling that is not completely reversible overnight; if present, knobs/bumps are below the ankle ONLY. Persistent swelling that does not reach above the knee. If knobs or bumps are seen or felt, they are only present below the ankle, NOT above the ankle.

Stage 3. Below-knee swelling that is not completely reversible overnight; knobs/bumps present above the ankle. Persistent swelling that does not reach above the knee. Knobs or bumps can be seen or felt above and below the ankle.

Stage 4. Above-knee swelling that is not completely reversible overnight; knobs/bumps present at any location. Persistent swelling that is present above the knee. Knobs or bumps can be seen or felt at any place on the foot or leg.

Stage 5. Joint fixation; swelling at any place in the foot or leg. The ankle or toe joints become fixed and difficult to flex or

dorsiflex. This may be accompanied by apparent shortening of the toes.

Description for health professionals

A more detailed and technical description of the changes that may be present at each stage is given, although the definitions for each stage remain the same. The stages represent severity of disease, and do not necessarily represent the disease process. For example, it is possible for an individual to have Stage 5 disease but never to have had above-knee swelling. The following terms are used in the descriptions:

- Dermal nodules: elevated, non-translucent lesions >0.5 cm diameter, with width approximately equal to length
- Dermal ridges: elevated lesions >0.5 cm width, with length greater than width
- Dermal bands: palpable, but non-elevated ridges
- Mossy changes: round or fusiform, either fluid-filled (and hence translucent) lesions, or papillomatous hyperkeratotic horny lesions giving the skin surface a rough velvet-like appearance.

Stage 1. Swelling reversible overnight. The swelling is not present when the patient

first gets up in the morning. Changes such as hyperpigmentation and nail dystrophy are unusual, but may be seen. The swelling is usually confined to beneath the ankle.

Stage 2. Below-knee swelling that is not completely reversible overnight; if present, knobs/bumps are below the ankle ONLY.

Persistent swelling that does not reach above the knee. If present, knobs or bumps do not extend beyond the ankle. The knobs or bumps may take the form of dermal nodules, ridges or bands. Tourniquet-like effects may be observed at this stage or any subsequent stage, depending on the position of dermal ridges and nodules in relation to joints. Mossy changes may be apparent, but their presence depends on a range of factors including the use of plastic footwear. Interdigital maceration and hyperpigmentation are often present at this stage, and nail dystrophy is almost always present.

Stage 3. Below-knee swelling that is not completely reversible overnight; knobs/bumps present above the ankle.

Persistent swelling that does not reach above the knee. Dermal nodules, ridges or bands are seen or felt above the ankle. Tourniquet-like effects are frequently observed at this stage.