Charcot's neuroarthropathy

When a timely diagnosis matters

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Charcot's neuroarthropathy is a diabetes complication that is important to recognise in order to prevent onset or progression of foot deformity. Patients with suspected Charcot's neuroarthropathy require urgent referral to a specialist high-risk foot service for intensive management.

Key points

- **Charcot's neuroarthropathy (CN) is an uncommon complication of diabetes requiring a timely diagnosis to prevent foot deformity and optimise outcomes.**
- **The diagnosis of CN relies mainly on clinical features, combined with targeted imaging such as plain x-rays and sometimes MRI.**
- **CN pathogenesis remains unclear, with marked foot inflammatory changes in the early phases, usually on a background of dense peripheral neuropathy.**
- **Immediate treatment involves avoidance of weight bearing and urgent referral to a specialist high-risk foot service, usually for total contact casting.**
- **Management of chronic CN usually requires custom-made footwear and orthotics; plantar osteotomy may have a role to prevent recurrent ulceration.**

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Charcot's neuroarthropathy (CN) is an uncommon but important complication of diabetic foot disease in which appropriate care can have a major impact on whether patients achieve a functional outcome. Considered an inflammatory condition in its acute presentation, CN is characterised by varying degrees of bone and joint disorganisation. A delay in diagnosis of CN can lead to increasing foot fractures, dislocations and the need for amputation, or severe long-term deformity and poor foot function, with an increased risk of recurrent foot ulceration. In contrast, a timely diagnosis and appropriate therapy can prevent or minimise long-term deformity.

There is general consensus that effective management of patients with CN requires:[1]

- early identification of CN
- immobilisation through minimising the weight-bearing load (or 'off-loading') on the affected joint
- management of resulting complications.

The prevalence of CN in people with diabetes is reported to be approximately 1.2 to 1.4%; even with likely under-reporting, CN is uncommon.[2] As a result, many doctors may go through their entire career without seeing a patient with CN. In this article, we provide key information for health professionals to help in the recognition of patients with acute CN and the initiation of management and urgent referral.

Presenting features of CN

Patients with acute CN typically present with a unilateral red, hot, swollen foot. On examination, there are readily palpable pedal pulses

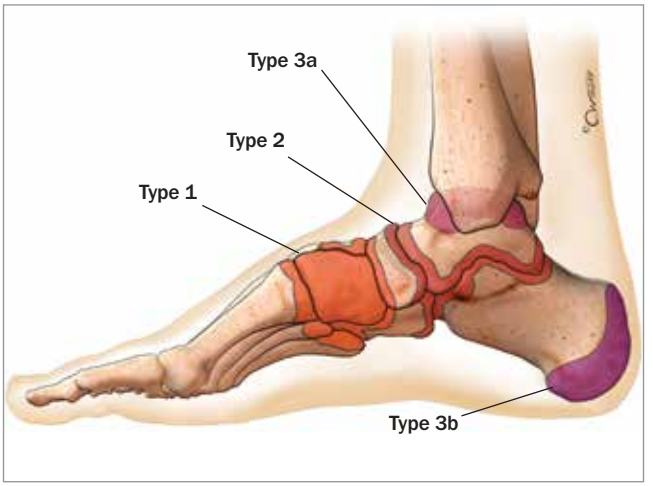
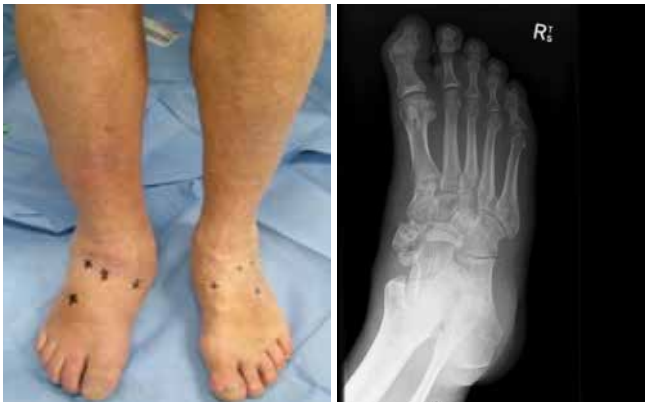


Figure 1. Brodsky classification of the distribution of joints affected by Charcot's neuroarthropathy. Type 1 (midfoot) is most common.



Figures 2a and b. Acute presentation of Charcot's neuroarthropathy in the right foot. a (left). The foot and lower leg show erythema and swelling extending to the knee. b (right). An x-ray shows disorganisation of the midfoot, with fragmentation and medial displacement of the navicular.

on a background of insensate diabetic peripheral neuropathy. The affected foot is usually warmer by two to three degrees Celsius than the unaffected foot (a difference which can be detected by palpation). CN occurs in patients with either type 1 or type 2 diabetes, most often aged in their 40s or 50s.

The area of the foot involved usually reflects the bones affected by CN. CN can affect a number of sites in the foot (Figure 1). Most commonly it involves the tarsal and metatarsal bones; rarely it involves the ankle, where it has a much poorer prognosis.[3] Multiple regions may be involved concurrently.

Foot deformity may or may not be present, depending on the stage of CN at presentation (Figures 2a and 3a). Pain is often present but is usually much less severe than would be expected from the degree of foot deformity. The presence of pain implies significant pathology.

Patients sometimes remember an initiating trauma, but the relationship with trauma may be masked by dense peripheral neuropathy.[2] Recent foot surgery and revascularisation may also sometimes precipitate CN. The duration of symptoms at presentation varies; some patients with CN present with acute foot inflammation of several days' duration whereas others present with persistent inflammation many weeks after they first noticed nonresolving swelling or deformity. Thus CN should be considered in the presence of foot inflammation with or without foot deformity.

Pathogenesis and course

The pathogenesis of CN remains unclear.[2] Insensate peripheral neuropathy is nearly universal in CN. Repeated foot trauma and localised inflammation possibly link to upregulated proinflammatory cytokines. Prominent or bounding pulses are often present. Autonomic neuropathy may coexist, including in the feet.

CN occurs in a series of time-based phases. In the early acute phase, which can last from weeks to months, inflammation occurs and is thought to contribute to osteolysis, ligamentous rupture and fracture and dislocation. During the subsequent subacute phase, signs of inflammation settle, the temperature elevation resolves and healing is apparent on x-ray and MRI. In the chronic phase, the foot bones have moulded, and the foot is usually clinically stable.

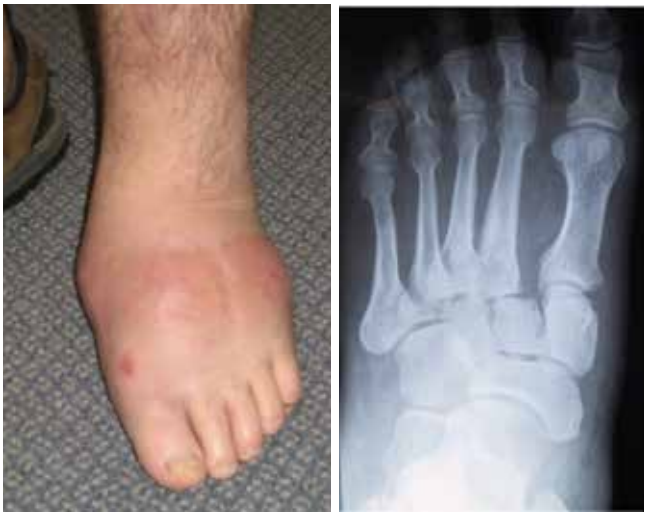
Diagnosis

A survey of patients with CN referred to the High Risk Foot Service at the Royal Prince Alfred Hospital Diabetes Centre in Sydney in 2011 found that most had seen an average of three to four health professionals for their foot problem before being diagnosed with CN. As CN symptoms often mimic a range of other diagnoses, CN is in our experience often misdiagnosed and thus mismanaged during the crucial onset period, potentially resulting in progressive irreversible deformity.

What are the differential diagnoses?

The swollen leg and foot in patients with CN may be attributed to a deep venous thrombosis, infection (cellulitis and/or osteomyelitis), stress fractures, soft tissue injuries or arthropathies such as gout. Excluding these differential diagnoses can be difficult, but there are a number of factors we can consider, as follows.

- Idiopathic cellulitis can occur but is unlikely. If a careful examination of the affected foot, including between the toes, shows no visible portal of entry for infection such as a break in the skin or ulcer then infection is a less likely diagnosis.
- In the later stages of CN the foot is often deformed, usually with evidence of tarsal or metatarsal fracture and/or dislocation seen on x-ray, as described above (Figures 2b and 3b).
- Levels of inflammatory markers can also help in differentiating the cause; in CN, levels of inflammatory markers are elevated either minimally or not at all, whereas in infections such as cellulitis or osteomyelitis they are usually increased markedly.[4]
- Swelling is one of the most consistent signs of CN and often involves both the affected foot and leg. In our experience, in the



Figures 3a and b. Acute presentation of Charcot's neuroarthropathy. a (left). The left foot shows erythema and swelling. On examination, the foot temperature was elevated and bounding foot pulses were present. b (right). An x-ray of the foot shows widening of the space between the first and second metatarsals and Lisfranc subluxation of metatarsals.

presence of CN and general fractures oedema does not resolve completely following elevation or bed rest. Diuretics and compression have minimal effect. If DVT is suspected, it may be excluded by performing a venous ultrasound examination.

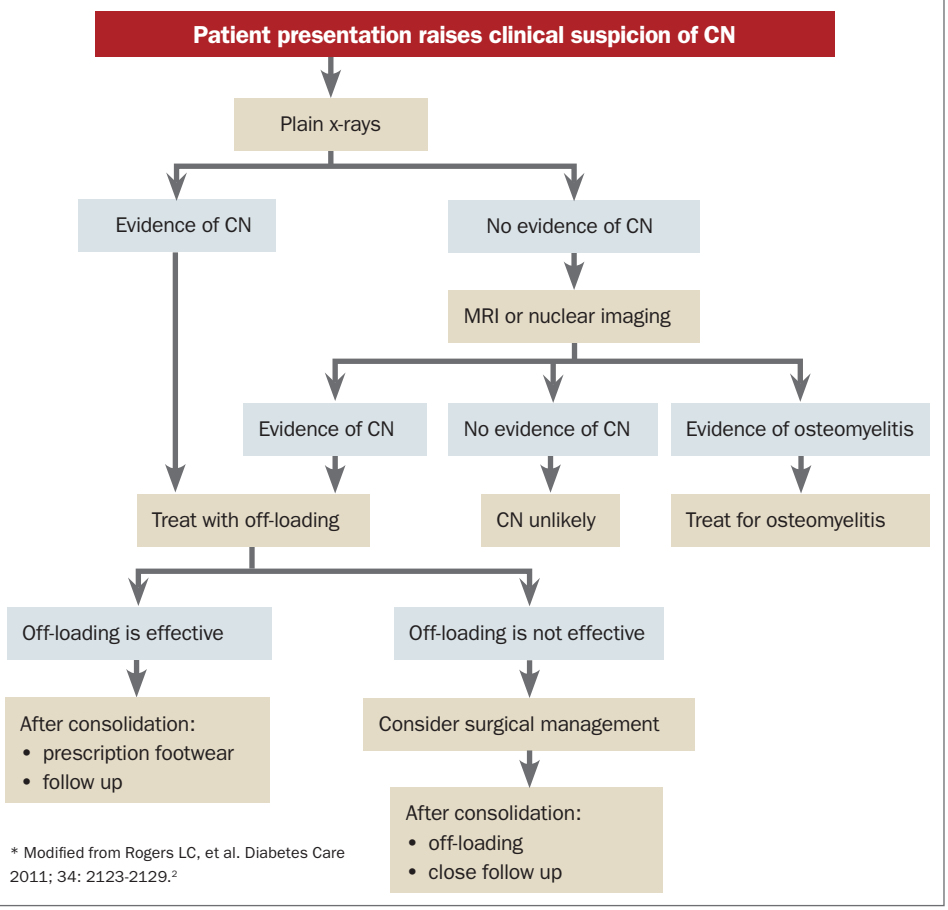
- Acute arthropathies such as gout are often considered. However, their presentation is usually focal to the affected joint, in contrast to the presentation of CN. Arthropathies may be excluded by radiological investigation (which often gives normal results in acute gout) and measurement of blood urate and C-reactive protein levels.

If a patient presents with diabetic peripheral neuropathy and symptoms that raise the possibility of CN as a differential diagnosis then we encourage prompt patient assessment and referral.

What investigations should be performed?

A suggested clinical pathway for assessment and management of patients with suspected CN is outlined in the flowchart. Plain radiographs are the primary imaging method for assessing underlying pathologies in the feet of patients with diabetes. They can also help exclude common differential diagnoses, such as infection, stress fracture and foreign bodies.

Suggested pathway for assessment and management of patients with suspected Charcot's neuroarthropathy (CN)*



* Modified from Rogers LC, et al. Diabetes Care 2011; 34: 2123-2129.[2]

When CN is suspected, a baseline x-ray should be performed as soon as possible as it is useful to identify changes and track progress. Suggested views are anteroposterior, lateral and oblique. Unfortunately, x-ray reports do not always formally describe the subtle changes that occur in acute CN, and we therefore recommend that clinicians check the x-rays themselves for fractures, subluxation, dislocations or bone fragments consistent with a diagnosis of CN (see Figures 4a to c). It is worth noting that although the presence of fractures alone is most often associated with CN, we have observed an increasing number of patients presenting with complete dislocation, as seen in a Lisfranc (midfoot-type) fracture.

If no abnormalities are apparent on the initial x-ray and there is still a high level of suspicion for CN, we suggest proceeding with an MRI scan as it can identify more subtle, early pathology in both the soft tissue and bone in great detail. However, if MRI is not easily accessible then a repeat plain x-ray in four to six weeks is advisable as it may show new

changes; the patient's foot should be immobilised in the interim.

Gallium bone scans are also sometimes ordered but are not specific for CN and will highlight any 'hot spot', such as an old fracture or arthritis. Instead, if nuclear medicine scans are to be used, we suggest a three-phase technetium bone scan followed by a white blood cell scan. A diffuse result on a white blood cell scan is more consistent with CN, whereas a focal hot spot at the site of pathology identified by a contemporaneous bone scan is highly suggestive of underlying osteomyelitis.

Management

What can GPs do?

Referral to a high-risk foot service

The NHMRC diabetes foot disease guidelines (2011) state that suspected acute CN is an emergency and that the patient should be immediately referred to a specialist high-risk foot service for intensive management.[1]

Advice to avoid weight-bearing

While the patient waits for a rapid assessment appointment at a specialist high-risk foot service, we strongly recommend advising them to reduce all unnecessary weight bearing, to help protect the foot from further trauma and to preserve foot shape. When there is any suspicion of injury to the foot – soft tissue or otherwise – avoidance of weight bearing on the foot is of prime importance. Bed rest is optimal; however, as this is rarely feasible the use of a wheelchair, crutches or a walking stick for support is an acceptable alternative.

To further temporarily immobilise the foot, a prefabricated cast boot is an option and can be fitted by a podiatrist, orthotist or physiotherapist. The patient should be encouraged to avoid walking and standing as much as possible, as this will help minimise trauma to the foot.

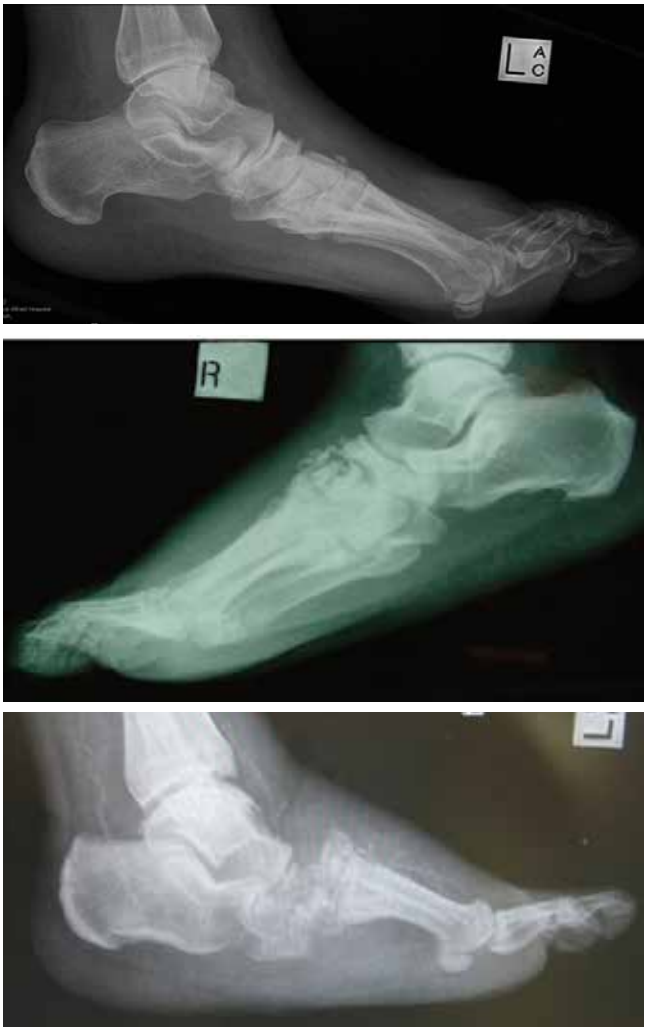
Medical therapy

There is no clear evidence that medical therapy targeting bone resorption (e.g. antiresorptive medication such as bisphosphonates) aids important clinical outcomes in patients with CN. The NHMRC guidelines state: 'Bisphosphonates have also been studied for their potential to decrease bone resorption. However, further studies are required to determine the role of these agents in the management of [CN]'.[1] For this reason, we do not recommend antiresorptive medication as a therapy for patients with CN outside the context of formal clinical trials. CN is not a TGA-approved clinical indication for antiresorptive medication in Australia.

What will a specialist high-risk foot service do?

Intensive immobilisation

Once the diagnosis of CN is confirmed, intensive immobilisation is instituted. A total contact cast is considered the 'gold standard' treatment and is our preferred treatment option. The cast is reviewed and usually replaced weekly to fortnightly until oedema has stabilised.



Figures 4a to c. Features of Charcot's neuroarthropathy seen on x-ray depend on the stage of presentation. a (top). In an early presenting patient, an x-ray shows a subtle fracture and subluxation. b (centre). In a later presenting patient, the changes are more indicative of overt fracture and dislocations. c (bottom). In a late presenting patient, major disruption of the foot bones is seen.

Once oedema has settled, the cast may be bivalved, but the patient must wear it at all times. It is replaced as required.

More recently, prefabricated casts offer an alternative to total contact casting but their use depends on the individual patient presentation, and they are not our preferred treatment. Prefabricated casts act similarly to total contact casts but have limited use when significant deformity is present, in which case custom-made devices are preferable. However, a prefabricated cast can be fitted with an orthotic to improve skin contact and is a cost-effective alternative. The patient is advised to wear the cast at all times, including during sleep. There is an option where the prefabricated cast can be bound with fibreglass casting material to discourage removal, but this is not our usual practice.

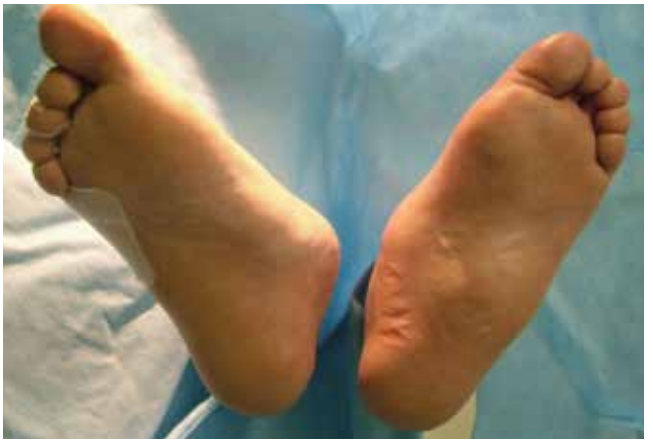


Figure 5. Bilateral chronic Charcot's neuroarthropathy showing the classic 'rocker bottom' deformity when the midfoot is affected.

If the patient's foot is in the later stages of the disease process at the time of presentation, where it is stable but still requires protection until healing is fully achieved, then a prefabricated cast boot may be used until medical-grade footwear and orthoses are fitted.

For both off-loading options (total contact and prefabricated casts), patients are encouraged to limit activity to that required for activities of daily living. Long periods of standing or walking are discouraged. If patients are working then we encourage them to take leave or if this not possible to seek shorter working hours or light/restricted duties. Driving can also be a problem even in an automatic car, and patients are encouraged to contact their insurance companies for advice.

Support and orthopaedic referral

Median duration of treatment for acute CN is nine months, although this can vary markedly and should be individualised.[2] The lengthy duration of treatment is understandably confronting for patients, who will require both psychological and physical support, including help at home from family members or arranged by the social work department. In our service, consultation with an orthopaedic surgeon is usually also arranged.

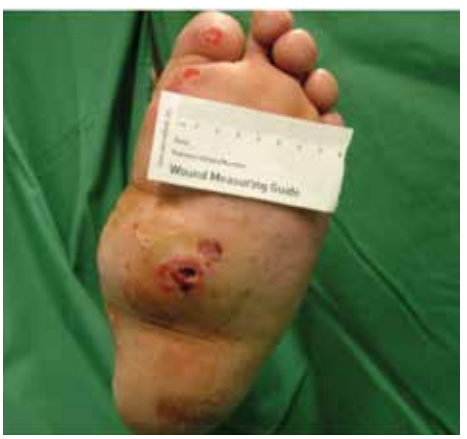


Figure 6. Charcot's neuroarthropathy deformity with a secondary midfoot ulcer.

Monitoring

During treatment the foot is monitored by:

- clinical assessment, including measurement of temperature and foot circumference
- assessment of foot stability
- six-weekly x-ray to detect evidence of healing such as callus formation and sclerosis.

In the later stages of treatment when the foot is close to healing, a prefabricated cast boot may be fitted. This can be removed for showering and sleeping. It is often used to protect the foot until medical-grade footwear and orthoses are fitted.

In the chronic phase of CN, the disease process in the foot is no longer active but marked foot deformity is common. For example, the classic 'rocker bottom' foot is often seen in chronic CN of the midfoot (Figure 5). Patients will require ongoing input from podiatry and pedorthic services to reduce the likelihood of ulceration and secondary infection, potentially leading to amputation. Patients will also continue to need monitoring to ensure use of appropriate footwear, skin integrity and education. Furthermore, as the opposite foot can be involved in 9 to 40% of cases, ongoing surveillance is required.

When is surgical management an option?

First-line treatment

Surgery is generally not the first-line treatment to stabilise the foot in patients with acute CN, as the foot is in an inflammatory state and the bone is mechanically unstable, increasing the risk that surgery will fail. Nevertheless, based on low level (level IV) evidence, surgery may be the first-line treatment for highly selected patients.

For example, in patients with acute CN who have major foot changes such as dislocation or subluxation and will not be able to achieve a stable foot with total contact casting, internal fixation is an option. In addition, CN of the ankle is notoriously unstable and usually requires some kind of surgical fixation.[3]

After surgery, patients need to avoid any weight bearing for a minimum of three to six months. This is very difficult for many patients as it requires a large amount of family support. In some instances where surgery is the only option, the patient may opt, by informed consent, to have a below-knee amputation.

Ulcer management

For management of plantar forefoot ulcers, Achilles tendon lengthening can reduce forefoot pressure and equinus and often allow the ulcer to heal. Following this surgery, patients need off-loading in a prefabricated cast for six weeks.

Many patients present with recurrent ulceration as a result of chronic foot deformity caused by CN (Figure 6). When conservative management with footwear and orthoses has failed, corrective surgery such as removal of plantar bone (plantar osteotomy) may be considered (Figure 7a). The indications for surgery and type of surgery depend on the individual patient. For instance, in patients with significant plantar midfoot deformity such as a 'rocker bottom',

osteotomy is a relatively simple procedure that has good outcomes (Figures 7b and c). After two weeks' off-loading, patients should resume wearing medical-grade footwear and orthoses.

Figures 7a to c. A patient with chronic Charcot's neuropathy before and after surgical treatment.
a (top left). Radiograph showing a midfoot bony prominence before surgery.
b (top right). Radiograph after surgical removal of the bony prominence (plantar osteotomy).
c (left). Healed foot after plantar osteotomy.

Long-term management: footwear, orthoses and podiatry

Because patients with chronic CN often have a markedly deformed foot, footwear and orthoses are key in their long-term management. Footwear and orthoses can protect and accommodate the deformity, thereby reducing the risk of ulceration. In many patients, medical-grade footwear and accommodative orthoses are sufficient. However, when foot deformity is severe, custom-made footwear will be needed (Figure 8).

Medical-grade and custom-made footwear can be obtained through referral of the patient to a specialist footwear manufacturer with specific training in the provision of these devices. Private health funds may cover part of the cost of specialist footwear. If appropriate, funding can usually be obtained through a state or territory government scheme. Specialist footwear can range in price from $950 to $3000.

On discharge from a high-risk foot service, patients with CN should be followed up by a podiatry service, either in the public system or privately. Patients with CN will need ongoing review to maintain foot health and reduce the risk of complications associated with abnormal foot shape. Podiatrists are also able to organise footwear referrals as the need arises. Patients should receive ongoing education about methods to protect their feet and the need to report promptly (within 48 hours) to their health care professional if they detect a foot ulcer developing, for example during their daily foot inspection.

Figure 8. Custom-made footwear and orthotics to accommodate a deformity of the right foot in a patient with Charcot's neuroarthropathy.

Conclusion

CN is uncommon but when it occurs a timely diagnosis, which is mainly clinical, can improve the chance of a good long-term foot outcome. Despite the uncertainty about its pathogenesis, intensive off-loading combined with vigilant follow up remain the cornerstones of care. Patients with chronic CN require ongoing podiatric care and review and referral for the provision of medical-grade or custom-made footwear and orthoses to reduce the risk of ulceration. In some individuals with CN, judicious use of surgery can help achieve a functional foot, reduce the risk of ulceration and provide a good quality of life. **ET**

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