# Buerger's disease

*Synonyms: thromboangiitis obliterans, endangitis obliterans von Winiwarter-Buerger, Winiwarter-Buerger syndrome, Winiwarter-Manteuffel-Buerger syndrome and Billroth-von Winiwarter disease*

## What is Buerger's disease?[1]

Buerger's disease is a chronic disease characterised by segmental inflammation and thrombosis of the small- and medium-sized arteries and veins of the peripheral upper and lower limbs.

The thrombus leads to arterial ischaemia in the distal extremities and superficial thrombophlebitis, which may progress to gangrene and ulceration. The aetiology is unknown but the use of tobacco is the key factor in the development and progression of the disease.

There is evidence that autoimmune factors may be involved.[2] The pathophysiology is thought to involve endothelial cells, platelets, leukocytes and sensory neurons.

In addition to smoking, male gender, genetic factors, infectious agents and deprivation have all been suggested as possible trigger factors.[3] Cardiovascular risk factors may also be important, especially glucose intolerance.[4]

## How common is Buerger's disease? (Epidemiology)

- The prevalence of the disease is decreasing in developed countries. Interestingly, the rate of decrease is more closely linked with improvements in socio-economic factors than with smoking status.[5]

- It mostly affects men but is increasing in women, due to changes in smoking habits.[6]

- One study reported an age range of 19-55 years with a mean age of 38.[7]
- The disease is found across the world. Its prevalence among all patients with peripheral arterial disease ranges from values as low as 0.5-5.6% in Western Europe to values as high as 45-63% in India, 16-66% in Korea and Japan and 80% among Jews of Ashkenazi ancestry living in Israel.[8]
- In the United States the incidence is estimated as 12.6-20 per 100,000 people.[9]
- The association of Buerger's disease with tobacco use, particularly cigarette smoking, cannot be overemphasised. Most patients with Buerger's disease are heavy smokers but some cases occur in patients who smoke 'moderately'.[4]

## Buerger's disease symptoms

The early symptoms of Buerger's disease include claudication in the feet and/or hands or pain in these areas at rest (about 20% of cases). The pain typically begins in the extremities but may radiate to more central parts of the body.

It may be very intense. As the disease progresses, the resting pain can be severe enough to cause insomnia. Other signs and symptoms may include:

- Two or more limbs being affected.
- Discolouration of the affected limb.
- Pain which may increase with activity such as walking and decrease with rest.
- Symptoms worsening with exposure to cold or with emotional stress.
- Numbness and tingling in the limbs.
- Raynaud's phenomenon.
- Skin ulcerations and gangrene of the digits, which are common.
- Pulses which may be decreased or absent in the affected extremity.
- Later symptoms which include enlarged, red, tender cord-like veins.

- Ocular manifestations, including ischaemic optic neuropathy and retinal artery occlusion.[10]

Assessment should also include consideration of other associated cardiovascular disease and other smoking-related health problems.

## Differential diagnosis

- In the early stages other causes of Raynaud's phenomenon must be considered, including systemic lupus erythematosus (SLE) and scleroderma.
- Compared with other causes of peripheral arterial disease, it tends to be aggressive with early onset of gangrene and ulceration.
- Exclude autoimmune diseases, hypercoagulable states and diabetes mellitus.
- A proximal source of embolism may be sought by echocardiography and arteriography.

## Diagnostic criteria

Diagnosis based on a list of criteria has been suggested, such as those of Shionoya:[8]

- Age under 50 years.
- Current or recent history of tobacco use.
- Presence of infrapopliteal arterial occlusive disease indicated by claudication, pain at rest, and ischaemic ulcers or gangrenes and documented by non-invasive vascular testing.
- Either upper-limb involvement or phlebitis migrans.

- Absence of atherosclerotic risk factors other than smoking, ie exclusion of:
    - Autoimmune diseases.
    - Hypercoagulable states.
    - Diabetes mellitus.
    - A proximal source of emboli by echocardiography or arteriography.

There must also be consistent arteriographic findings in the clinically involved and non-involved limbs. Confident clinical diagnosis of Buerger's disease requires all five features. Strict clinical diagnostic criteria are essential for any study of a disease to ensure the homogeneity of the selected patient population for valid comparisons.

# Investigations

There is no specific diagnostic test.[11]

### Serological

There are no specific serological markers to diagnose Buerger's disease. Recommended tests to rule out other causes of vasculitis include FBC, LFTs, renal function tests, fasting glucose, ESR, CRP, autoantibodies and screening for hypercoagulability.[12]

### Imaging

- **Angiography** - certain angiographic features are typical (but not pathognomonic) of Buerger's disease.[13] These include a 'corkscrew' appearance of arteries that results from vascular damage, particularly the arteries in the region of the wrists and ankles. Angiograms may also show occlusions or stenoses in multiple areas of the arms and legs. It is sometimes necessary to perform angiograms of other parts of the body regions, like a mesenteric angiogram to exclude other forms of vasculitis that involve vascular regions atypical for Buerger's disease.

- **Doppler ultrasound** - this may be helpful. Lately, colour Doppler has been used to distinguish Buerger's disease and other causes of secondary Raynaud's phenomenon from primary disease.[14]
- **Echocardiography** - this may be required to exclude a source of recurrent emboli.

### Other

Skin biopsies of affected extremities are rarely performed because of fear that a biopsy site in an ischaemic area will not heal.

## Buerger's disease treatment and management

### General points

Patients with Buerger's disease must be advised to stop smoking immediately and completely. This is the only treatment known to be effective. Otherwise, there is not yet an agreed consensus on the treatment of choice.[15]

**Supportive measures include:**

- Gentle massage and warmth to increase circulation.
- Avoiding conditions that reduce peripheral circulation, like cold temperatures.
- Avoiding sitting or standing in one position for long periods.
- Not walking barefoot, to avoid injury.
- Avoiding tight or restrictive clothing.
- Aggressive treatment of any injuries (such as ulcers).

Patients who have claudication but not critical ischaemia should be encouraged to walk, whereas those with critical ischaemia should be admitted for bed rest.[11]

### Drug treatment[8]

- Although low-dose aspirin has been used, the drug iloprost (a prostacyclin analogue) has been shown in the latest Cochrane review to be superior to aspirin when given intravenously.[16] However, oral iloprost is not superior to placebo.
- Another treatment sometimes tried is bosentan, an oral dual endothelin receptor antagonist; however, further research is needed to confirm its effectiveness..
- Calcium-channel blockers, steroids, anticoagulants and other antiplatelet drugs are ineffective.
- Some have suggested that, if the disease is due to sensitivity to a component of tobacco other than nicotine, nicotine replacement therapy (NRT) may be used. However, recent findings suggest that there is an association of smokeless tobacco with progressive limb ischaemia and therefore all tobacco products should be stopped.[17]
- Vasoconstricting drugs should be avoided.

There is work being done in the field of stem cell therapy to treat intractable symptoms related to ischaemia, where conventional therapy has failed; however, this is still at the research stage.

Likewise, the use of granulocyte colony-stimulating factor-mobilised autologous mononuclear cells has been shown to reduce the amputation rate in initial clinical trials. However, further work is required.

Therapeutic angiogenesis is under review but lacks strong evidence for efficacy.[9]

### Surgical[8]

- Surgical sympathectomy is more effective than intravenous iloprost, but only with low quality evidence.[18]
- Periarterial sympathectomy has been found useful in some patients with chronic digital ischaemia.[19]
- Bypass surgery has not yielded good results in these patients.[12]
- However, a new technique of extended angioplasty directed at each obstructed tibial and foot artery has reduced the number of amputations and resulted in sustained clinical improvement.[20]

- A stent puncture technique has been described for patients with multilevel disease.[21]
- Spinal cord stimulation has been shown to reduce the number of amputations needed. The exact mechanism of action is poorly understood and further research is needed.
- Distal limb amputation: areas with gangrene must be removed surgically; patients who continue to smoke are likely to require amputation of fingers and toes.

## Complications

Ulceration, infection and gangrene must be treated energetically.

## Prognosis

The disease is progressive in patients who do not stop smoking. The only way to prevent the progression of the disease is to abstain from all tobacco products.

One study identified four forms of the disease: relapsing-remitting (75%), secondary progressive (4.6%), primary progressive (14.2%) and benign (6.2%). Most amputations occurred due to relapses within six years after diagnosis.

Duration of smoking of more than 20 years had a significant relationship with further major amputation.[22]

---

## Further reading


- Chen JY; Thromboangiitis Obliterans. Anatol J Cardiol. 2021 Feb;25(2):E8. doi: 10.14744/AnatolJCardiol.2020.87425.

| | | |
|---|---|---|
| Authored by: | Peer Reviewed by:<br>Dr Toni Hazell | |
| Originally Published:<br>21/11/2023 | Next review date:<br>21/06/2023 | Document ID:<br>doc_1606 |

View this article online at: patient.info/doctor/buergers-disease-pro

Discuss Buerger's disease and find more trusted resources at Patient.