THE AUSTRALASIAN COLLEGE
OF DERMATOLOGISTS

# Cutaneous Small Vessel Vasculitis

**Also known as** ... Cutaneous Leukocytoclastic Vasculitis, Cutaneous Leukocytoclastic Angiitis, Hypersensitivity Angiitis and Cutaneous Necrotizing Venulitis

## What is Cutaneous Small Vessel Vasculitis?

Cutaneous Small Vessel Vasculitis (CSVV) is a condition caused by inflammation of the skin's blood vessels. It classically presents as areas of red or purple discoloration. This is due to bleeding under the skin and does not disappear when pressure is applied (purpura).

## What causes it?

CSVV may be a reaction to medications, food and food additives, underlying infection, autoimmune connective tissue disease, malignancy or other diseases. However, there is no known cause in approximately one third to half of cases.

In a minority of affected people, cutaneous vasculitis can be part of a more severe vasculitis affecting other organs in the body known as "systemic vasculitis".

Henoch-Schönlein purpura (HSP) is a subtype of CSVV caused by deposition of immunoglobulin A (IgA) within the blood vessel wall. HSP is often preceded by viral respiratory illness. It is more common in children but can also occur in adults.

## What does it look like?

The most common presentation of CSVV is raised red-purple spots (purpura, 0.3 to 1 cm in diameter) and/or petechiae (purpuric lesions less than 3 mm in diameter) which do not disappear with pressure (i.e. non-blanching).

These lesions can join together, become ulcerated, and may be associated with blood filled blisters. The rash of vasculitis is most common on the lower legs, areas of tight-fitting clothing and areas of trauma (Koebner phenomenon). However, any surface can be involved.

People affected with CSVV may experience itching, burning or pain or they may not experience any symptoms at all.

General symptoms such as fevers, weight loss and muscle aches may occur. Five to 25% of people with CSVV can have joint, genitourinary or gastrointestinal symptoms. This raises the possibility of internal involvement.

In HSP, lesions tend to occur on the buttocks and lower limbs. Children may develop systemic disease with gastrointestinal, joint, and/or kidney involvement. In adults with HSP, arthritis and kidney disease occur more frequently.

**How is it diagnosed?**

A detailed medical history and physical examination will be needed, as well as selected laboratory studies including blood and urine tests to look for underlying causes or organ involvement. A skin biopsy is usually taken to exclude other causes. Two samples of skin are taken –one for histology (microscopic examination of the skin) and the second for immunofluorescence (to look for antibodies in the vessel walls).

**How is it treated?**

***General measures and symptomatic treatments*** include:

- Rest and leg elevation
- Avoiding tight clothing
- Pain management with nonsteroidal anti-inflammatory drugs (NSAIDs)
- Oral antihistamines and topical corticosteroids may help relieve itching

***Systemic therapy*** to arrest the disease process is usually reserved for those people who have persistent symptoms or develop skin or systemic complications. These include:

- Systemic corticosteroids (e.g. prednisone)
- Colchicine
- Dapsone
- Hydroxychloroquine
- Systemic immunosuppressive therapy such as azathioprine, methotrexate and mycophenolate mofetil, cyclosporine, cyclophosphamide, rituximab and intravenous immunoglobulin.

It is important to rule out internal organ involvement and a referral to a physician may be necessary in those with systemic signs or symptoms. Any identified trigger or cause must also be removed or treated.

In most people, the condition is self-limiting which means that it resolves spontaneously, usually within 2 to 4 weeks. Sometimes the skin inflammation can leave residual darkening of affected areas known as post-inflammatory hyperpigmentation. This usually fades slowly over many months. Chronic or recurrent disease occurs in about 10% of affected people.

This information has been written by Dr Davin Lim and Dr Heba Jibreal