BLUE RUBBER BLEB NAEVUS SYNDROME

What are the aims of this leaflet?

This leaflet has been written to help you understand more about blue rubber bleb naevus syndrome. It tells you what this condition is, what it is caused by, what can be done about it, and where you can find out more about it.

What is blue rubber bleb naevus syndrome?

Blue rubber bleb naevus syndrome has also been called ‘Bean syndrome’ after Dr William Bean, who described it. It is caused by the congenital development of multiple venous malformations in the skin, soft tissues and gastrointestinal tract (intestine).

A ‘syndrome’ is a group of signs and symptoms that occur together. ‘Naevus’ is Latin for ‘birthmark’. The ‘blue rubber blebs’ are small areas of blue mis-shaped veins with a rubber-like feel.

In some people, blebs are only seen in the skin. They can also occur in any other part of the body, most commonly the intestine.

What causes blue rubber bleb naevus syndrome?

The cause of blue rubber bleb naevus syndrome is not known. During the development of an affected baby in the womb, clusters of abnormal veins are formed. This is not related to any known event during pregnancy.

Is blue rubber bleb naevus hereditary?

Most people with blue rubber bleb naevus syndrome do not have affected family members, although there are some reports of this syndrome running in a family.
Blue rubber bleb naevus syndrome is not contagious or cancerous.

**What are the symptoms of blue rubber bleb naevus syndrome?**

Apart from their appearance, the skin lesions often do not cause any symptoms. However, they may be tender to touch, painful, show increased sweating or bleed.

Other symptoms depend on the involvement of other organs. This does not usually become apparent until adulthood. If the intestine is involved, blood may be passed when opening the bowels and the faeces may be black. Blood loss may result in anaemia.

Symptoms need to be investigated as they arise, and referral to different specialists may be required.

**What does blue rubber bleb naevus syndrome look like?**

Groups of small to medium sized purple to bluish-black rubbery lumps can be seen in the skin either at birth, or may appear in childhood or later in life. The number of blue rubber blebs usually increases during life and can range from a few to over a hundred. Pressing onto a bleb will cause it to blanche (turn whiter). On releasing the pressure, the blood will return and the bleb will recover its bluish colour.

**How is blue rubber bleb naevus syndrome diagnosed?**

A Dermatologist may suspect blue rubber bleb naevus syndrome after examining the skin. It may be necessary to confirm the diagnosis by taking a sample of the abnormal skin (a biopsy) to be examined under the microscope. A local anaesthetic injection will usually be given beforehand to numb the skin.

**Can blue rubber bleb naevus syndrome be cured?**

Unfortunately, there is currently no cure for blue rubber bleb naevus syndrome.

**How can blue rubber bleb naevus syndrome be treated?**
Some people require no treatment other than an explanation of the condition. Treatment is directed at the symptoms, which may vary from patient to patient and may be different depending on which parts of the body are affected.

For blebs of the skin, different surgical and laser treatments can be used if the areas are causing problems. These require a local anaesthetic injection to numb the areas beforehand and leave a scar:

- Excising of lesions or scraping them off (‘curettage’) and treating the bleeding base with heat (cautery)
- Using an ‘ablative’ laser to remove the bleb layer by layer, such as the carbon dioxide (CO2) laser, or the Erbium Yag laser

Other treatment options include freezing the lesions with liquid nitrogen (‘cryotherapy’) and sclerotherapy. Sclerotherapy is the injection of a solution into the veins of the bleb that causes inflammation and shrinks the veins.

Skin camouflage can be useful to cover the bluish discolouration. Skin camouflage is water resistant and matched to the colour of the patient’s skin (see leaflet on skin camouflage).

Where can I get more information about blue rubber bleb naevus syndrome?

Web links to detailed leaflets:


http://rarediseases.info.nih.gov/gard/5940/blue-rubber-bleb-nevus-syndrome/resources/1
For details of source materials used please contact the Clinical Standards Unit (clinicalstandards@bad.org.uk).

This leaflet aims to provide accurate information about the subject and is a consensus of the views held by representatives of the British Association of Dermatologists: individual patient circumstances may differ, which might alter both the advice and course of therapy given to you by your doctor.

This leaflet has been assessed for readability by the British Association of Dermatologists’ Patient Information Lay Review Panel

BRITISH ASSOCIATION OF DERMATOLOGISTS
PATIENT INFORMATION LEAFLET
PRODUCED JANUARY 2015
UPDATED MAY 2018
REVIEW DATE MAY 2021