

VASCULAR BIRTHMARKS: SALMON PATCHES, PORT WINE STAINS AND STRAWBERRY NAEVI

What are the aims of this leaflet?

This leaflet has been written to help you understand more about three of the most common vascular birthmarks. It tells you what they are, why they develop, what can be done about them, and where you can find out more about them.

What are vascular birthmarks?

They are marks made up of excess or abnormal blood vessels in the skin. There are many different types, and only three are described in this leaflet: salmon patches (naevus simplex); port wine stains (naevus flammeus); and strawberry naevi (infantile haemangiomas).

SALMON PATCHES

These are the commonest type of vascular birthmark, occurring in up to 40% of newborn babies. They are present at birth and are flat, dull-red areas that can be present on the eyelids, bridge of the nose, the upper lip and on the nape of the neck (where they are sometimes referred to as 'stork bites'). They are more noticeable when the child is crying. Most fade during infancy, though 50% of those present on the nape of the neck persist into adult life. No treatment is required.

PORT WINE STAINS

What are port wine stains?

A port wine stain is a pink or red mark or patch on the skin that is usually present at birth.

What causes a port wine stain?

The cause of port wine stains is not fully understood. However, they are thought to develop in areas of skin lacking the small nerves that control the constriction of small blood vessels. The blood vessels stay open, causing a permanent blush in the affected area of skin. Port wine stains occur in approximately 3 per 1000 births, affecting males and females and all racial groups equally.

Are they hereditary?

Usually they are not.

What are the symptoms of port wine stains?

Port wine stains usually occur on the face and neck and less often on the trunk and limbs. They do not usually cause any symptoms but they can produce long-term psychological trauma and problems with self-image and self-esteem if they are present on visible areas such as the face. A lot can be done to reduce their psychological impact, including the treatments discussed below. Cosmetic camouflage can also be helpful as can be advice from patient support groups.

Later in life port wine stains can become deeper red or purple in colour and become raised or lumpy and more difficult to cover with makeup. The raised areas can bleed easily if they are scratched.

Port wine stains of the eyelid area and upper jaw sometimes lead to increased pressure within the eye (glaucoma). Rarely patients with facial port wine stains are prone to seizures and require further assessment and investigation. An extensive port wine stain of a limb may be associated with an increase in growth of that limb.

What do port wine stains look like?

Port wine stains cause pink, red or purple areas or patches of discoloured skin. They usually affect only one side of the body. They can develop on any area of the skin, but the face and upper trunk are the most common sites. The pink, smooth and flat port wine stains of babies eventually become red and then purple, darkening and even thickening with age. Port wine stains persist, enlarging in proportion as the body grows. In adult life, their surface becomes uneven and bumpy, and bleeds easily if scratched.

How are port wine stains diagnosed?

Port wine stains are recognised by their typical appearance.

Can port wine stains be cured?

They can be made much less obvious by the treatments listed below.

How can port wine stains be treated?

Laser treatment. Pulsed dye laser (PDL) is the treatment of choice and is available in specialist dermatology centres in the U.K. It helps most patients, particularly if the mark is on the face, but may not clear the port wine stain completely. Port wine stains in older children can be treated using a local anaesthetic e.g. an anaesthetic cream. Young children, and those having very extensive areas treated, especially around the eyes, will need to have laser treatment under a general anaesthetic or with sedation.

Depending on the size and site of the birthmark, up to 10 treatment sessions may be required at intervals of approximately 8 weeks. Port wine stains on the limbs respond less well than those on the face. Treatments given early in life, before the birthmark becomes thickened, are more successful than those used later on.

Cosmetic camouflage. Cosmetic or camouflage creams are often very helpful. Expert advice is available in special Cosmetic Dermatology clinics provided at some dermatology centres. Changing Faces and the Disfigurement Guidance Centre also offer a comprehensive service for patients and their families with all types of birthmark. The details are given at the end of this leaflet.

STRAWBERRY NAEVI

What are strawberry naevi?

These are soft raised vascular swellings on the skin, often with a bright red surface, that may have the appearance of a strawberry. They are also known as 'infantile haemangiomas'. They usually appear after birth, often within the first month, and may occur anywhere on the skin. They may be more troublesome when they affect the face or nappy area.

What causes strawberry naevi?

This cause is not fully understood. They are a benign overgrowth of blood vessels in the skin, and are made up of cells that usually form the inner lining of blood vessels (endothelial cells). They are thought to occur as a result of a localised imbalance in factors controlling the development of blood vessels.

Strawberry naevi occur in about 5% of the population. They are commoner in females, premature babies and multiple births. Strawberry naevi are not a sign of ill health, or associated with cancer. There are many myths about their cause that should be discounted and parents should not feel responsible if their child develops one.

Are they hereditary?

No they are not hereditary.

What are the symptoms of strawberry naevi?

Usually there are no symptoms but they can be visible and be unsightly. A few grow quite rapidly and can become large and may bleed or become infected. Sometimes they can affect an organ or body function and will need special treatment early in life (see below).

What do strawberry naevi look like?

At birth, strawberry naevi are usually absent or appear as a red or bruise-like patch. After birth there is a rapid growth phase usually in the first 5 to 8 weeks. Most strawberry naevi (80%) reach their final size by 3 months. The early growth phase is followed by a slower growth phase. Finally the strawberry naevus regresses or becomes smaller in size and resolves

(involutes) over a period of years (usually 3 years). Most haemangiomas cease to improve after 3.5 years of age.

Usually there is only one strawberry naevus, but sometimes several may occur. They can appear on any area of skin but a majority (60%) do so on the face and neck. If the haemangioma is near the surface of the skin, it will be bright red, like a strawberry, whereas if it is located deeper, it may appear blue in colour. When located deep beneath the skin, the haemangioma may be skin coloured.

The result following involution is often very good. Sometimes the skin stretched by the strawberry naevus will be thin, loose or scarred after the haemangioma has shrunk. Plastic surgery reconstruction can help when the child is older. It is not possible to accurately predict how fast or how large strawberry marks will grow, how long they will take to shrink, or whether they will leave a mark behind.

How will a strawberry naevus be diagnosed?

The diagnosis will be based on the appearance of the haemangioma and on the way it grows.

How can strawberry naevi be treated?

Most strawberry naevi do not require treatment as they resolve spontaneously. For most "active non-intervention" is the best option and consists of education, reassurance that the naevus will resolve, observation and good skin care of the overlying skin e.g. with an emollient.

Bleeding may occur but, even with accidental trauma, this is likely to be minor. It can be stopped easily and quickly with gentle pressure followed by a sterile dressing to cover the area that bled.

A small number of strawberry naevi require treatment usually for one of three reasons: because of ulceration (breakage of the surface of the skin); because it may cause disfigurement (e.g. if present on the centre of the face); or because it is causing or may cause impairment of function of a vital organ e.g. impair development of hearing or vision. A few haemangiomas grow so quickly that they threaten to interfere with vision, breathing, or feeding (if located on the face), or passing urine or stools (if located in the nappy area). These are best treated early to avoid complications. The treatment is aimed at inducing rapid shrinkage.

Current treatment options

- *β*-blockers:
 - Propranolol is a β-blocker used to treat high blood pressure. Its use has dramatically altered the treatment of strawberry naevi over the past 5 years since it was discovered to be an effective treatment for strawberry naevi. Oral propranolol is now considered by most experts to be the first-line treatment for strawberry naevi requiring treatment and has been successfully used in babies with large complicated haemangiomas. It is effective in halting the growth and diminishing the size of strawberry naevi.
 - **Timolol** solution (a topical β-blocker) can be useful to treat small surface strawberry naevi that do not need oral treatment.
- **Steroids** have been used by injection and by mouth for the rare haemangiomas that are serious or dangerous because of their size or location. The use of steroids has mostly been taken over by oral propanolol when treatment is required.
- Laser treatment (pulsed dye laser therapy) available in specialist dermatology centres can be used to stop bleeding in ulcerated haemangiomas, and can speed up healing and shrinkage of the haemangioma.

Where can I get more information about vascular birthmarks?

www.dermnetnz.org/dna.haemangioma/haeman.html www.patient.co.uk/health/port-wine-stain-leaflet www.birthmarksupportgroup.org.uk www.skincamouflagenetwork.org.uk

British Association of Skin Camouflage (NHS and private practice) Tel: 01254 703 107 Email: <u>info@skin-camouflage.net</u> Web: <u>www.skin-camouflage.net</u>

Changing Faces Tel: 0300 012 0276 (for the Skin Camouflage Service) Email: <u>skincam@changingfaces.org.uk</u> Web: <u>www.changingfaces.org.uk</u>

Skin Camouflage Network (NHS and private practice)

Helpline: 0785 1073795 Email: <u>enquiries@skincamouflagenetwork.org.uk</u> Web: <u>www.skincamouflagenetwork.org.uk</u>

Disfigurement Guidance Centre PO Box 7 Cupar Fife KY15 4PF

For details of source materials used please contact the Clinical Standards Unit (<u>clinicalstandards@bad.org.uk</u>).

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: its contents, however, may occasionally differ from the advice given to you by your doctor.

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

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