Klippel-Trenaunay syndrome

This leaflet provides information about Klippel-Trenaunay syndrome and what to expect when your child comes to Great Ormond Street Hospital (GOSH) for assessment, review and treatment.

What is Klippel-Trenaunay syndrome and what causes it?

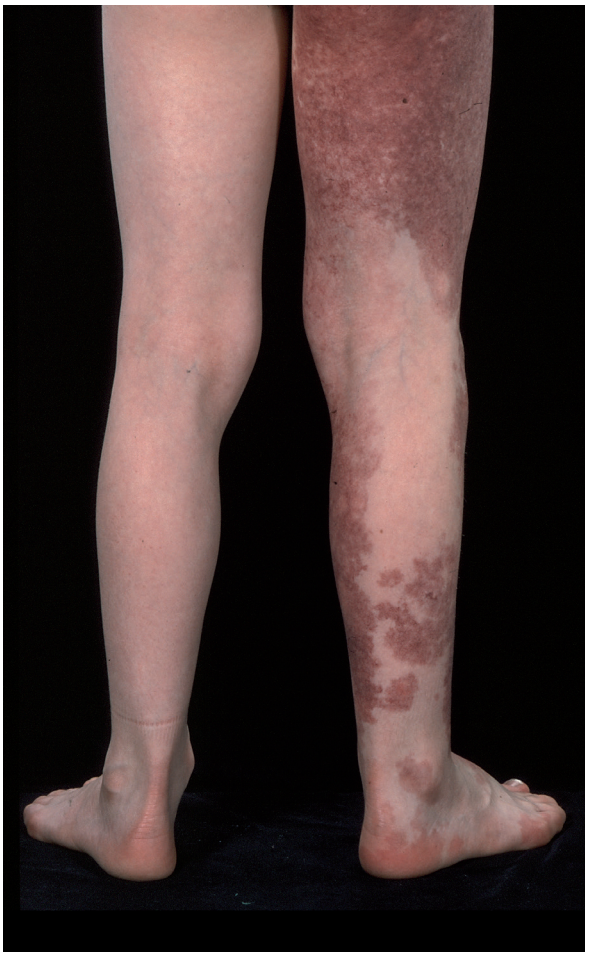
A syndrome is a collection of features that often appear together. The features associated with Klippel-Trenaunay syndrome are port wine stains, varicose veins and hypertrophy (extra growth) of one limb. It is named after the two French doctors who described the condition in 1900.

Klippel-Trenaunay syndrome is present at birth (congenital) but often the only visible sign in babies is the port wine stain. The diagnosis may not be confirmed until the varicose veins and limb hypertrophy become more noticeable.

We do not really know what causes Klippel-Trenaunay syndrome. There are various theories about possible causes, including problems with how the blood vessel system develops, but more research is needed to confirm the cause. It is clear that it is not passed on from parent to child in any direct way.

How common is Klippel-Trenaunay syndrome and whom does it affect?

Klippel-Trenaunay syndrome is a rare condition, affecting about one in every 20,000 to 40,000 children. Children of all ethnic groups can be born with Klippel-Trenaunay syndrome, and it affects males and females in equal numbers.



What are the signs of Klippel-Trenaunay syndrome?

Klippel-Trenaunay syndrome consists of three signs often seen together: port wine stains, varicose veins and limb hypertrophy.

The port wine stain is a flat, red or purple mark on the skin that is often the only noticeable sign of Klippel-Trenaunay syndrome at birth. The shape of the port wine stain varies depending on the part of the body

affected. Port wine stains on the leg are usually patchy. Those on the chest and abdomen tend to affect only one side of the body and have a definite edge. In most cases, the port wine stain is on the same side of the body as the limb affected by the varicose veins and limb hypertrophy. Children with Klippel-Trenaunay syndrome may only have one port wine stain or they may have smaller ones elsewhere on the body, often on the other limb, or head and neck.

Varicose veins are another feature of Klippel-Trenaunay syndrome, and are larger and cover a wider area than normal varicose veins. While the whole leg is usually involved, the varicose vein might only be visible in certain areas, and may become more visible as the child grows older. Varicose veins and the problems associated with them, can be reduced by wearing a pressure garment from an early age.

There is a small risk of blood clots developing in children with Klippel-Trenaunay syndrome.

Limb hypertrophy is the third feature of Klippel-Trenaunay syndrome, although it is not always noticeable at birth and during early childhood. Hypertrophy means 'extra growth'. The affected limb is longer and bigger widthways than the other, although the amount of difference varies from child to child. In most children, one leg is affected, but rarely an arm or an arm and a leg can shows signs of hypertrophy.

It is difficult to predict how much the affected limb will grow. Hypertrophy of a leg can cause an uneven walking style (gait).

Other symptoms have been reported in children with Klippel-Trenaunay syndrome, affecting the skin, skeleton, blood vessels and lymphatic system, but these are less common.

How is Klippel-Trenaunay syndrome diagnosed?

Klippel-Trenaunay syndrome may be suspected in children who have a port wine stain covering an arm or leg, but the diagnosis may not be confirmed until the child is walking and the varicose veins and limb hypertrophy are more obvious. Generally, a child has to have all three features to be diagnosed with Klippel-Trenaunay syndrome.

Can Klippel-Trenaunay syndrome be treated?

Klippel-Trenaunay syndrome itself cannot be treated but some of the features associated with it can be improved to a great extent.

In Klippel-Trenaunay syndrome, the port wine stain is usually on the leg and so may not be as problematic as one affecting the face. Some port wine stains respond to laser treatment, though usually to a limited degree. For more information, please see our leaflet *Port wine stains*.

The varicose veins may become painful, and are often the most troublesome part of Klippel-Trenaunay syndrome. As a child gets older, the veins may occasionally become inflamed (phlebitis), bleed if injured and can very rarely develop ulcers if they are not looked after carefully. If bleeding occurs elevate the limb, apply pressure to the area. Put a gauze dressing over the area, followed by the pressure garment to apply further pressure. There is a chance that deeper blood vessels could be involved, which could affect blood supply to the rest of the leg. In some children, after a detailed scan of the veins in the leg, sclerotherapy might be suggested, which involves injecting a substance into a vein to block it. It can take many sessions of sclerotherapy to improve the varicose veins and is not suitable for everyone.

If hypertrophy leads to one leg being longer than the other, shoe raises might be suggested. These are insoles that go inside shoes, raising up the foot so the difference

in length is corrected. If the affected leg is more than two centimetres longer than the unaffected leg, the growth can be slowed down using an operation called epiphysiodesis. Sometimes it is advised that the shorter leg is lengthened so the legs are more or less the same length. For more information about these options, please see our leaflets Epiphysiodesis and Limb length differences and limb lengthening.

What is the outlook for children with Klippel-Trenaunay syndrome?

As the severity of Klippel-Trenaunay syndrome varies from child to child, a multidisciplinary team often provides the best care. Regular reviews with dermatologists, orthopaedic, vascular and general surgeons may be needed, until the child is a teenager. Klippel-Trenaunay syndrome is a lifelong condition, needing ongoing care and treatment, so transfer to adult services will be needed between 16 to 18 years of age.

Klippel-Trenaunay syndrome is not a life threatening condition and should not interfere with a child's day-to-day activities to any great degree, although some sports may be less easy for some children. The vast majority of people with Klippel-Trenaunay syndrome grow up to lead normal lives, working and raising a family.

Information and support

- **At Great Ormond Street Hospital**

Birthmark Unit
Great Ormond Street Hospital
London WC1N 3JH
Tel: 020 7829 8668

- **Support groups**

Birthmark Support Group
London WC1N 3XX
Tel: 0845 045 4700
Email: info@birthmarksupportgroup.org.uk
Website: www.birthmarksupportgroup.org.uk

Changing Faces
The Squire Centre
33-37 University Street
London WC1E 6JN
Tel: 0845 4500 275
Fax: 0845 4500 276
Web: www.changingfaces.org.uk
www.iface.org.uk

Compiled by the Birthmark Unit with assistance from parents of children with Klippel-Trelaunay syndrome in collaboration with the Child and Family Information Group. All photographs appear in this leaflet with the consent of the child and parent, and must not be reproduced for any other purpose.

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