

A young man with dark hair is standing on a green lawn. He is wearing a white short-sleeved polo shirt with red accents on the collar and sleeves. The shirt has several logos: 'O'NEILLS' at the top, a circular logo on the left chest, a shield logo on the right chest, and 'W.J. DOLAN CONSTRUCTION' in a large, stylized font across the center. He is also wearing white trousers and pink shoes. He is holding a yellow and red ball in his left hand, which is slightly blurred. The background shows a wooden fence and some greenery.

Klippel-Trenaunay syndrome

Information for families

Great Ormond Street Hospital
for Children NHS Trust

This leaflet explains about Klippel-Trenaunay syndrome and what to expect when your child comes to Great Ormond Street Children's Hospital for assessment, review and treatment.

What is Klippel-Trenaunay syndrome and what causes it?

A syndrome is a collection of symptoms that often appear together. The symptoms 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 during pregnancy, 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 symptoms of Klippel-Trenaunay syndrome?

Klippel-Trenaunay syndrome consists of three symptoms 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 symptom 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 but 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 much larger and cover a wider area than normal varicose veins. Generally, they cover the whole leg from foot to thigh, and sometimes the buttocks and abdomen; 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 support stockings from an early age.

If the arm is affected the veins may appear prominent. There is a small risk of blood clots developing in children with Klippel-Trenaunay syndrome. Your specialist may suggest aspirin or other anticoagulant (anti-clotting) medicine if appropriate.

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' and the affected limb, usually one leg, 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 show signs of hypertrophy too. The average difference in length between the affected leg and the unaffected one is about two centimetres, although in some children the difference is greater or smaller.

The affected leg is bigger widthways because the skin and muscles grow more and become thicker. Often, the difference in size is more noticeable towards the ankle than in the thigh. It is difficult to predict how quickly the limb will grow or what its eventual size will be as the hypertrophy develops in irregular phases. The limb hypertrophy can cause an uneven walking style (gait) but this can be improved. A child may also appear as if they have a condition called scoliosis, where the spine is curved, but this is also caused by the limb hypertrophy that puts the hip out of line, which in turn affects the straightness of the spine.

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 symptoms to be diagnosed with Klippel-Trenaunay syndrome.

Can Klippel-Trenaunay syndrome be treated?

Klippel-Trenaunay syndrome itself cannot be treated but the symptoms 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 noticeable as one affecting the face. Port wine

stains can be treated; for more information, please see our leaflet *Port wine stains*.

The varicose veins in the affected leg may become painful, and are often the most troublesome part of Klippel-Trenaunay syndrome. As a child gets older, the veins become inflamed (phlebitis), bleed if injured and can occasionally develop ulcers if they are not looked after carefully. The usual way of treating varicose veins, by tying them off or removing them is not advised in children with Klippel-Trenaunay syndrome. If bleeding occurs, apply pressure to the area and smear on some Vaseline® to seal the bleeding. Put a gauze dressing over the area, followed by the support stocking 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. The treatment usually suggested includes resting the affected leg in a raised position and using compression stockings when standing or walking. 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 several months apart to improve the varicose veins so is not suitable for everyone.

Limb hypertrophy can be treated in various ways, depending on the difference in size between the affected and unaffected leg. If the affected leg is only slightly longer than the unaffected one shoe raises might be suggested. These are insoles that go inside a pair of shoes, raising up the foot so the child can walk more easily and the difference in length is not so noticeable. If the affected leg is more than two centimetres longer than the unaffected leg, the growth in the affected leg can be slowed down using an operation called epiphysiodesis or the shorter leg could be 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*. Physiotherapy and compression can

help reduce any puffiness in the leg and make it more comfortable, but not all children will need this treatment.

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

As the severity of symptoms 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 will be needed, often until the child is a teenager. Klippel-Trenaunay syndrome is a lifelong condition, needing ongoing care and treatment, so a child will need to transfer to adult services when he or she reaches 16 to 18 years old.

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. Sports may be less easy for some children, so a review appointment could be useful.

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

The Proteus Family Network UK may be able to offer help and supports. Proteus syndrome has similar symptoms to Klippel-Trenaunay syndrome, as Proteus syndrome has similar symptoms to Klippel-Trenaunay syndrome.

Proteus Family Network UK

31 Baswich Lane
Weeping Cross
Staffs ST17 0BH
Tel: 01785 254 953
Email: info@proteus-uk.org
Website: www.proteus-uk.org

Birthmark Support Group

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

Changing Faces

1 & 2 Junction Mews
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London W2 1PN
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Notes

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Compiled by the Birthmark Unit with assistance from parents of children with Klippel-Trenaunay syndrome in collaboration with the Child and Family Information Group.

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www.goshfamilies.nhs.uk www.childrenfirst.nhs.uk