**KT Frequently Asked Questions**

**How do I know if I really have KT?**

Since KT is a rare disorder, many times a diagnosis is not given to patients. They are often told they have a “birthmark” or a capillary malformation. Normally when KT occurs, it will appear on the leg, most often in a “hip to toe” pattern. Sometimes it is patchy, sometimes it can appear on the arms or body.   The criteria for diagnosing KT is that you must have at least two of the following:

* Hypertrophy (enlargement of the limb)
* Absence or duplication of a venous structure
* Port Wine Birthmark

**KT - What is it?**

Klippel - Trenaunay Syndrome is a congenital( present at birth) vascular disorder of unknown cause. Klippel-Trenaunay (KT) is characterized by a triad of signs: Port Wine Birthmark (capillary malformations) covering one or more limbs, vascular anomalies, usually venous varicosities or malformation and hypertrophy (enlargement of the limb) or atrophy (withering or smaller limb). KT normally involves the lower limbs in about 90% of the patients. In rare instances, there is an absence of Port Wine Birthmark and not all three abnormalities need always be present for the syndrome to exist

**What symptoms are associated with Klippel-Trénaunay Syndrome?**

Each case of KT is different. Patients may have symptoms including anemia from occult blood loss, coagulation (clotting) problems, blood clots and platelet trapping in the affected limb. Clotting problems are often first noticed with surgery, trauma or hormonal changes. Any KT patient considering surgery should alert the medical team to the possibility of a clotting problem.

**What are the possible complications of Klippel-Trénaunay Syndrome?**

Complications of the capillary malformation ( Port Wine Birthmark) include skin breakdown and ulceration, bleeding and secondary infection.

* Varicosities may affect the superficial and deep venous systems. Pain and [lymphedema](http://www.cincinnatichildrens.org/health/info/vascular/diagnose/lymphedema.htm) (swelling of extremities due to stoppage of lymph flow caused by malformed lymphatic vessels) are common. Complications due to varicosities include paresthesias (abnormal skin sensations such as burning or tingling), skin ulcers, pulmonary emboli (blood clots in the lungs), inflammation and clots of blood vessels in the legs, and cellulitis (skin and soft tissue infection).
* Hypertrophy( enlargement) of a limb can lead to scoliosis, gait abnormalities and compromise of function. Leg length differences are common.

**How is Klippel-Trénaunay Syndrome managed?**

Management of Klippel-Trénaunay Syndrome is dependent upon the person. Although both nonoperative and surgical procedures are used, treatment is primarily non-operative and supportive.

* Supportive care

Compression garments are often advised for chronic venous insufficiency, lymphedema, recurrent cellulitis and recurrent bleeding from capillary or venous malformations of the extremity. They also protect the limb from trauma. Intermittent pneumatic compression pumps may also be used.

* Pain medication, antibiotics, and limb elevation. These treatments are all used to manage cellulitis.
* Anticoagulant therapy (the use of substances that prevent blood clotting). This approach is indicated in cases of acute thrombosis (clotting) and is also used as a preventive measure prior to surgical procedures.
* Heel inserts. These are sometimes used to manage limb length discrepancies that are less than 1 inch. For greater discrepancies, orthopaedic surgery may be considered.
* Surgical interventions

Laser therapy. Using the flash lamp pulsed-dye yellow laser is often quite effective in lightening the color of the port wine stain. Many treatments are typically required to achieve a desirable result. Laser treatment is also indicated when there is ulceration, since it tends to effect quicker healing.

* Surgery. Depending on the individual case and degree of involvement, a number of surgical options are occasionally used. These include vein ligation, vein stripping, vein resection, and in rare cases, amputation. Vein ligation is a procedure that clamps off a section of veins. The clamp prevents blood flow through the damaged section of veins and promotes blood flow through veins that are not damaged. Vein stripping uses a metal wire to remove varicosities from within the damaged vein. Vein resection is a procedure that removes a section of veins from the body. Amputation is a procedure that removes a portion or all of a limb.
* Other treatment

Sclerotherapy. This therapy is the injection of a chemical into the vein causing inflammation. As the inner wall of the vein becomes inflamed, blood is not permitted to flow through it. The vein then collapses and forms scar tissue. There have been isolated cases where children have experienced a severe reaction to the chemical used to sclerose the veins. Please mention this to your doctor if you are considering this procedure and demand that they ensure that there is no risk of allergy.

**What is Klippel-Trenaunay-Weber?**

Klippel Trenaunay Weber is a misnomer. KT used to be called "KTW Syndrome,” but Weber has been dropped to avoid confusion with the Parkes Weber Syndrome. Parkes Weber Syndrome is characterized by arteriovenous malformations. It also includes at least one fistula. A fistula is an abnormal connection between a vein and an artery. Having this bad connection can sometimes lead to high flow within the lesion, this can cause extremity overgrowth similar to KT but the appearance of the limb is usually pinker and sometimes warmer. This syndrome can lead to high output cardiac failure in some individuals. Sometimes physicians will order an angiogram if they suspect Parkes Weber. In KT the angiogram is normal. With Parkes Weber there would be the appearance of the fistula(s).

Parkes Weber Syndrome is more likely than KT to affect an upper limb but will still affect the lower limb. Parkes Weber Syndrome is the more rare of the two syndromes.