



**The National Alliance
for Thrombosis
and Thrombophilia**

Postthrombotic Syndrome (Venous Stress Disorder)

An In-Depth Guide for Patients and Health Care Providers

AWARENESS • PREVENTION • TREATMENT • SUPPORT

ABBREVIATIONS USED:

DVT = deep vein thrombosis; a blood clot in the deep veins of the legs and arms.

WHAT IS VENOUS STRESS DISORDER?

Normally, blood flows through veins out of the legs and arms back to the heart. Small valves in the veins enable the blood to flow in the right direction and prevent blood from flowing backwards and pooling in the veins of the legs and arms. A clot (thrombosis) in the deep veins of legs or arms (deep vein thrombosis, DVT) leads to an obstruction of blood outflow from the extremities back to the heart. Acute leg or arm swelling and pain, therefore, result. This is called an acute DVT. When the body tries to heal from these clots the valves in the veins are often damaged. The obstruction of the veins and the destruction of valves lead to impaired blood flow.

If a vein is completely blocked, neighboring smaller veins may enlarge to bypass the obstruction. These bypassing veins are called collaterals and can get quite large, particularly in the pelvis and abdomen in patients who have thrombosis of the big veins in the abdomen (vena cava) or pelvis (iliac veins). Such collaterals can sometimes be seen as prominent veins underneath the skin. If good collaterals have formed, symptoms of leg swelling and pain may not occur or may only be mild. However, in some people collaterals do not get very large and can, therefore, not carry all the blood needed to drain the legs or arms; this then leads to chronic leg or arm swelling, pressure and pain.

Several different terms are used for the chronic symptoms that can occur after a deep vein thrombosis (Table 1). These terms all describe the same symptom complex. It is noteworthy that not all people who have

Table 1: TERMINOLOGY

Medical terms used:

Postthrombotic syndrome (PTS)
Postphlebitic syndrome (PPS)
Venous stasis syndrome (VSS)
Chronic venous insufficiency (CVI)

Patient terms used:

Venous stress disorder
Chronic venous limb disorder

these symptoms have had blood clots. Actually, the majority of people (88 %) have not had documented blood clots. The same symptoms occur in people with dysfunction of the valves in the veins, heart failure, obesity and other, often not clearly identifiable causes. The best, most accurate, and most widely used medical term for this condition is “venous stasis syndrome”. If one refers to venous stasis syndrome occurring after a DVT, an appropriate term is “postthrombotic syndrome”. A good non-medical term is “venous stress disorder”. “Lymphedema” also refers to swelling of one or both legs or arms, but is usually not painful and does not lead to skin ulcers. However, its causes are different: it is due to obstruction of lymph vessels (and not the veins) that leads to a lack of drainage of fluid from the extremities, and, thus, to swelling.

SYMPTOMS

While some people who have had a DVT recover completely, others may be left with some symptoms in legs or arms (Table 2): leg or arm swelling, pain, aching, heaviness, and cramping are some of the symptoms.

Table 2: SYMPTOMS AND FINDINGS OF POSTTHROMBOTIC SYNDROME

- chronic extremity swelling
- chronic (or waxing and waning) pain
- unspecific discomfort of the extremity
- diffuse aching
- heaviness, tiredness and cramping of extremity
- dark skin pigmentation (=postthrombotic pigmentation)
- bluish discoloration of toes/fingers, foot/hand or diffusely of leg/arm
- skin dryness
- eczema
- hardening of the skin
- formation of varicose veins
- skin ulcer (stasis ulcer)
- “atrophie blanche” or “white atrophy (description in text)
- “dermatoliposclerosis” (description in text)



Postthrombotic pigmentation



Healed skin ulcer and postthrombotic pigmentation



Chronic (left) leg swelling, skin hardening, and postthrombotic pigmentation

Figure 1: FINDINGS IN PATIENTS WITH POSTTHROMBOTIC SYNDROME

The pain and swelling can be disabling. Symptoms in the legs are typically worse after standing for a long time, and may be worse at the end of the day. They are improved in the morning or after elevating the limb. Dark pigmentation (Figure 1) may occur. It is due to the leakage of miniscule amounts of blood out of the veins into the soft tissues. Blood contains red cells (which are our oxygen carriers) and red cells contain iron. The iron from the red cells that leaked into the soft tissues is bound to a storage protein, and the complex is called hemosiderin, which is brown. The postthrombotic pigmentation may be unsightly; however, it does not cause symptoms and is not dangerous – it does not harm the skin. If a person has significant chronic leg swelling, the skin may become hard, dry, and scaly.

If the chronic swelling is severe, skin breakdown may occur and an ulcer may form (venous stasis ulcer). Some people will develop what the physicians call “dermatoliposclerosis”: the tightness of the swollen leg leads to destruction of the skin and fat tissues. The person experiences this as hot, red, thickened, painful and tender areas of the skin, typically in the lower legs. This problem is not infrequently misdiagnosed as cellulitis (inflammation of the skin due to infection). If scar tissue forms in the skin without the development of ulcers, areas of white skin may arise. This is referred to as “atrophie blanche” or “white atrophy.”

Doctors may, subjectively, classify postthrombotic syndrome as mild, moderate or severe. For clinical studies, more objective classification systems exist, the so-called Villalta and Ginsberg scales.

WHO DEVELOPS IT?

An estimated 330,000 people in the United States have the postthrombotic syndrome. Typically, the more extensive the DVT, the more severe the symptoms of postthrombotic syndrome will be. However, this is not always so: even people who have had very extensive acute DVTs with severe symptoms may recover completely and may not be left with any long-term symptoms. Approximately 60 % of patients will recover from a leg DVT without any residual symptoms, 40 % will have some degree of postthrombotic syndrome, and 4 % will have severe symptoms. The symptoms of postthrombotic syndrome usually occur within the first 6 months, but can occur up 2 years after the clot. If a patient has done well for ½ - 2 years after the clotting event it is highly unlikely that he/she will develop the postthrombotic syndrome.

In people with arm DVT, postthrombotic syndrome develops in approximately 15 % of patients. People with DVT of larger veins, i.e. those in the shoulder and upper chest area (in medical terms “axillary or subclavian vein thrombosis”) and people who still have left-over clot after the acute event (residual thrombosis) appear to be at particular risk for postthrombotic syndrome.

Little is known as to who will develop chronic symptoms and who will not. However, it is known, that people with DVT who wear daily compression stockings (see below) for several month after the acute DVT will develop significantly less postthrombotic syndrome.

PREVENTION AND TREATMENT

Prevention is the key issue. If a person has leg swelling after an acute DVT, the person should wear a **compression stocking** to decrease the swelling. The stocking should be custom fitted, i.e. a person’s leg should be measured to find a stocking that fits well. It needs to have a certain compression pressure, 35 mm Hg (mercury) at the ankle, 25 mm Hg at the mid-calf, and 18 mm Hg just below the knee. This is also sometimes called a “grade 2” stocking. If the leg swelling is below the knee, then a below-knee stocking is appropriate, but if swelling also involves the thigh, then an above-knee stocking should probably be worn. However, research on the benefit of compression stockings and prevention of the postthrombotic syndrome is ongoing. Sleeves (“gauntlets”) for

postthrombotic syndrome in the arm also exist and should be worn if there is arm swelling or pain.

Unfortunately, compression stockings are often not worn because they are deemed unsightly or are uncomfortable. People should know that stockings come in various skin-tone and fashion colors, different shapes, sizes and materials, and from a variety of companies. It is worthwhile to make inquiries to find the right stocking that fits well, is relatively comfortable, and is acceptable in appearance. If stockings tend to roll down, you may want to choose a stocking that has a rubber strip at the upper end or wear a garter belt or compression pantyhose. Stockings should be worn during the day, while standing; stockings do not need to be worn at night. They should be worn for weeks, months, or years to control symptoms. For example, if swelling has disappeared a few weeks or months after the acute DVT with the use of the stockings, you may stop wearing the stocking. If swelling recurs then the stocking should be worn again. If there is no more swelling, then stockings are not needed any more.

So-called “*Anti Embolism Stockings*” or “TED hose” are often given to people who are hospitalized and have had surgery. They put mild pressure on the legs to prevent blood from clotting and can, to some degree, prevent blood clots in the legs (DVT). However, due to their low compression pressures they are not useful to prevent or treat the postthrombotic syndrome.

Table 3: TREATMENT

- Elevation of extremity at rest and at night
- Compression stockings, grade 2
- Weight loss
- Increased exercise with strengthening of extremity muscles
- Pain management
- Compression pump
- Vascular interventional radiology procedure: balloon opening and stenting of narrowed vein

Table 4: SUPPLIERS OF STOCKINGS AND PUMPS

The major compression stockings companies:

Jobst -----	www.jobst.com
Juzo -----	www.juzo.com
Medi USA-----	www.mediusa.com
Sigvaris -----	www.sigvaris.com
Venosan -----	www.venosan.com

The major compression pump companies:

Bio Compression-----	www.biocompression.com
Huntleigh -----	www.huntleigh-healthcare.com
KCI -----	www.kci1.com
Mego Afek -----	www.lymphapress.com
Tyco Healthcare-----	www.tycohealthcare.com

** NATT does not endorse specific medical products. Other products may also be available.*

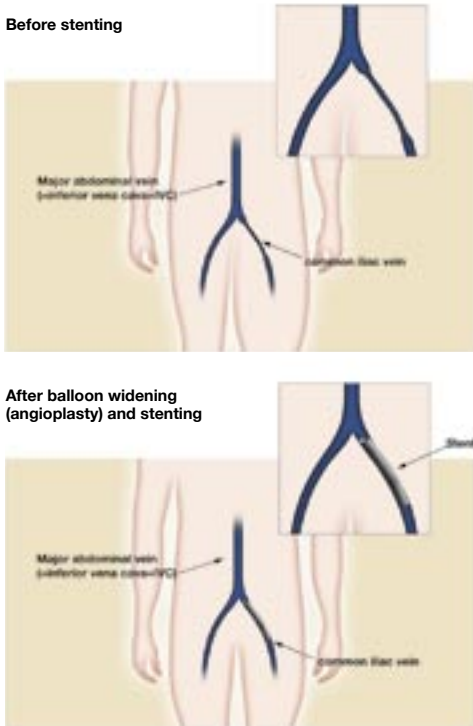
Elevation of the extremity above the level of the heart while resting or sleeping is also appropriate, if there is leg or arm swelling. ***Normalization of weight*** may also improve the symptoms. It is well possible that ***physical exercise*** and strengthening of the extremity muscles may improve the postthrombotic syndrome. However, the role of physical exercise still needs to be investigated in clinical studies. In cases of pronounced swelling that does not improve with compression stockings, ***a compression pump*** should be tried (table 4). A battery-powered, transportable device is available (SCD EXPRESS by Tyco Healthcare), suitable for people who travel.

Pain management is important and needs to be individualized. Since many people with postthrombotic syndrome are on warfarin, pain medications that increase the risk for bleeding when taken regularly should not be used. Drugs that contain aspirin or the so-called non-steroidal anti-inflammatory drugs (NSAIDS), such as ibuprofen (Motrin®), naprosyn (Aleve®, Naproxen®, etc.) and others, should not be used. Drugs that can be considered are (a) the non-narcotic drugs, such as acetaminophen (Tylenol®), Celebrex®, tramadol (Ultram®), and others, or (b) the narcotic drugs, such as acetaminophen with codeine or oxycodone (Tylenol® #2, 3 or 4, Tylox®, Percocet®,

etc.), hydromorphone (Dilaudid[®], etc.), fentanyl patch (Duragesic[®]), and others. Pain management can be complex and input from a specialized Pain Clinic may be helpful. Neurontin[®] (Gabapentin) is a pain modifier that has been used for so-called neuropathic pain associated with diabetic neuropathy and other pain syndromes. It has not been studied in the pain of postthrombotic syndrome, but could be tried for some time to see whether a person's pain improves.

Figure 2: BALLOON DILATATION AND STENTING PROCEDURE

Scarring of veins after a blood clot (DVT) in the left pelvic veins has led to narrowing of the major pelvic vein. This results in an obstruction of blood outflow from the leg, and, thus, swelling and pain of the leg. A vascular radiologist can balloon open the narrowing (a procedure called angioplasty) and place one or more stents to keep the vein open. Symptom improvement or even resolution may result.



Balloon widening and stenting: Sometimes, people with postthrombotic syndrome have a narrowing of one of the major veins in the pelvic area (iliac vein) or the abdomen (vena cava). This may be present from birth (called May-Thurner syndrome) or due to scarring of the blood vessel from a healed blood clot (stricture). If such a narrowing is present, it may be helpful to undergo a vascular radiology procedure during which the narrowing is ballooned open and stented (figure 2). This should only be undertaken in a center that is experienced in doing these procedures.

Venous skin ulcers may be difficult to heal. Visits with a vein or wound care specialist may be helpful to get expert care. Elastic bandages (Unna boots: bandages that contain a combination of calamine lotion, glycerin, zinc oxide, and gelatin), or foam dressings (Profore™; <http://wound.smith-nephew.com/us/Product.asp?NodeId=857>) often lead to wound healing. However, this is a slow process. Diligent wound care is necessary.

Psychological and social aspects: The short and long-term impairment of physical functions may be frustrating, especially to people who were physically very active before their postthrombotic syndrome. Oftentimes, such people will have to adjust their level of expectations, at least for the time being, and allow for their extremity function to slowly recover. However, the impairment of extremity function may also lead to permanent inability to work in the previous occupation and the need for retraining or for disability application. A visit with a social worker to discuss these issues may be helpful. It may also have a big impact on a person's abilities to continue to pursue his or her hobbies, such as athletic activities. And last but not least, it may negatively influence a person's self esteem, family and interpersonal interactions.

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RESOURCES:*

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Acknowledgement:

Review of manuscript: Elizabeth Varga, Columbus, OH;
Graphic Design: Jeff Harrison, Wilmington, NC.



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08.31.06