Vascular UPDATE

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Lymphoedema

What is Lymphoedema?

Lymphoedema is a chronic and progressive condition where excess fluid accumulates in the tissues, causing a protein rich swelling. Lymphoedema can affect any part of the body, and is most often seen in the legs, arms, face, neck and genitals. It cannot be cured, but can be controlled with appropriate treatment.

Primary lymphoedema is when the lymphatic vessels or nodes have not completely developed before birth. Specific forms include:

- Milroy's disease (congenital lymphedema). This disorder begins in infancy and causes lymph nodes to form abnormally.
- Meige's disease (lymphedema praecox). This disorder often causes lymphedema around puberty or during pregnancy, though it can occur later, until age 35.
- Late-onset lymphedema (lymphedema tarda). This occurs rarely and usually begins after age 35.

Secondary lymphoedema (Table 1) develops as a result of damage to the lymphatic vessels. It may also be the result of high output failure of the lymphatic circulation, eg in chronic oedema due to venous insufficiency or post-thrombotic syndrome, when the function of the overloaded lymphatic system eventually deteriorates.

Table 1: Causes of Secondary Lymphoedema

Trauma and tissue damage	lymph node excision, radiotherapy, burns varicose vein surgery/harvesting, large/circumferential wounds, scarring, joint replacements
Malignant disease	lymph node metastases, infiltrative carcinoma, lymphoma, pressure from large tumours
Venous disease	chronic venous insufficiency, venous ulceration, post-thrombotic syndrome, intravenous drug use
Infection	cellulitis/erysipelas, lymphadenitis, tuberculosis, filariasis
Inflammation	rheumatoid arthritis, dermatitis, psoriasis, sarcoidosis, dermatosis with epidermal involvement
Endocrine disease	pretibial myxoedema
Immobility and dependency	dependency oedema, paralysis
Factitious	self harm

Diagnosis

Clinical signs include pitting oedema, positive Stemmer sign, skin changes such as fibrosis, dermatitis, papilloma's, lymphorrhea, hyperkeratosis and limb distortions.

Tests are available to establish the diagnosis, to investigate co-existing conditions or to exclude other pathologies. These include lymphoscintigraphy, duplex doppler ultrasound to exclude venous pathology, antigen card tests, genetic testing, ICG lymphography or CT/MRI scans.

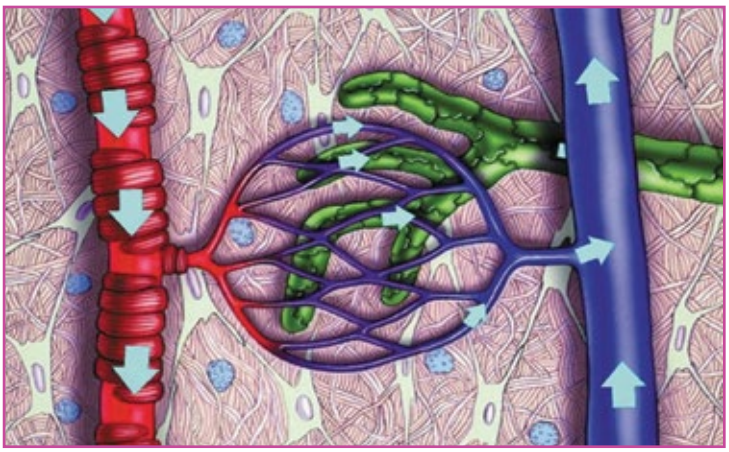
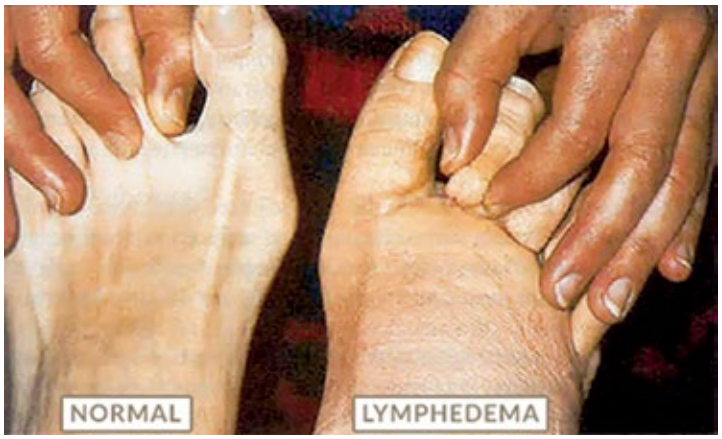


Fig 1: Forces of Lymphoedema



Stemmers Sign: try to pinch and lift a skinfold at the base of the second toe or middle finger. If you can pinch and lift the skin, Stemmer's sign is negative. If you can't, the sign is positive.

Table 2: **Stages of lymphoedema**	Skin texture	Pitting	Effect of elevation on oedema
Stage 0 Latent stage	"Normal" tissue consistency	Slight	Completely reduced
Stage 1 Reversible stage	Soft and spongy	Mild to moderate	Marked improvement **or** complete reduction
Stage 2 Spontaneously irreversible stage	May be firm	None **or** Pitting when pressure applied	No reduction
Stage 3 Lymphostatic elephantiasis	Increase in volume and size and Significant changes to skin including fibrosis, papilloma's, skin folds and hyperkeratosis	None **or** Pitting when pressure applied	No reduction

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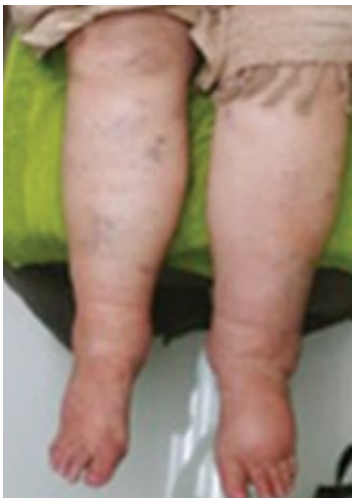
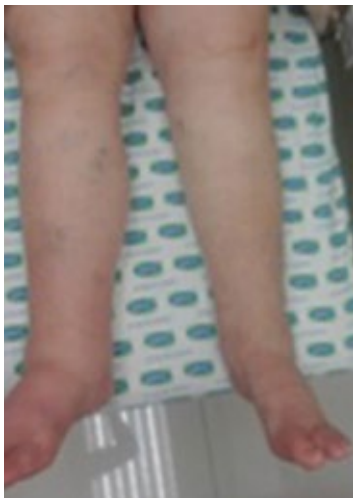
Treatment:

The international best practice for the treatment of lymphoedema is complete decongestive therapy (CDT). CDT makes use of manual lymph drainage (MLD), multi-layer bandaging systems, compression hosiery and night time garments. Education is a key therapy tool and focuses on self-maintenance, nutrition, meticulous skin care and exercise.



Before



After

Some methods of risk reduction of the affected limb besides meticulous skin care include:

- Not drawing blood or taking blood pressure on the at-risk limb
- Avoid high impact sports, carrying heavy items or prolonged movements
- Avoid exposure to heat such as sauna's, steam room, sunburn
- Avoid tight clothing, shoes & jewellery
- Wearing protective clothing, gloves or shoes e.g. when gardening or washing dishes
- Wearing compression garments prophylactically on flights or when exercising

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LAOSA Lymphoedema Association of South Africa

Lymphatic THERAPY CENTRE

Post Thrombotic Syndrome

PTS is a clinical condition with pain and disability resulting from chronic venous insufficiency following a DVT. It is the most frequent complication of DVT and develops in 20-50% of patients within 2 years of DVT diagnosis, even with appropriate anticoagulation.

The total cost per patient following DVT is 50% higher in patients who develop PTS. This cost, especially in patients who develop ulcers is related to surgery, dressings, lost work days and loss of employment.

Patients with PTS often report a poorer quality of life than those with diabetes, osteoarthritis and chronic lung disease.

Clinical manifestations

The manifestations are similar to those of chronic venous insufficiency. Symptoms include leg pain, leg heaviness, pulling or fatigue, and leg swelling. Signs include leg oedema, redness, dusky cyanosis when in a dependent position, telangiectasia, new varicose veins, hyperpigmentation, skin thickening and leg ulcers.

Diagnosis

PTS is a clinical diagnosis, based on symptoms and signs in a patient with a previous DVT. A diagnosis of PTS should be deferred until after the acute phase (i.e. 3–6 months) has passed.

The Villalta PTS scale is used to diagnose and grade the severity of PTS. A score of >4 denotes PTS *(Table 3)*.

Risk factors for development of PTS

The risk factors for PTS include: increased age, raised BMI, proximal deep vein thrombosis (2-3x increased risk), pre-existing venous insufficiency, inadequate anticoagulation (2x increase), recurrent ipsilateral DVT (4-6x), persistent symptoms >1 month post DVT, residual thrombus on ultrasound (1.5-2x), persistently elevated d-dimer.

Prevention

The best way to prevent PTS is to prevent the occurrence of DVT.

The evidence is unclear whether or not stockings prevent PTS. Class II below knee stockings are thus are not routinely required, but are helpful in patients with swelling and symptoms.

Patients with extensive ilio-femoral deep vein thrombosis benefit from pharmaco-mechanical thrombolysis to prevent PTS.

Treatment of Established PTS

Class II or class III compression stockings are the mainstay of treatment, especially in patients with leg symptoms and swelling. Exercise is beneficial.

Surgical or endovascular procedures such as venous valve repair, venous bypass and venous stents to treat appropriately selected PTS patients have potential to decrease post-thrombotic manifestations due to deep vein obstruction or reflux.

Table 3: **Villalta PTS Scale**

Symptoms	Signs
Pain	Oedema
Cramps	Skin Induration
Heaviness	Hyperpigmentation
Pruritis	Venous Ectasia
Paraesthesia	-Redness -Pain during calf compression

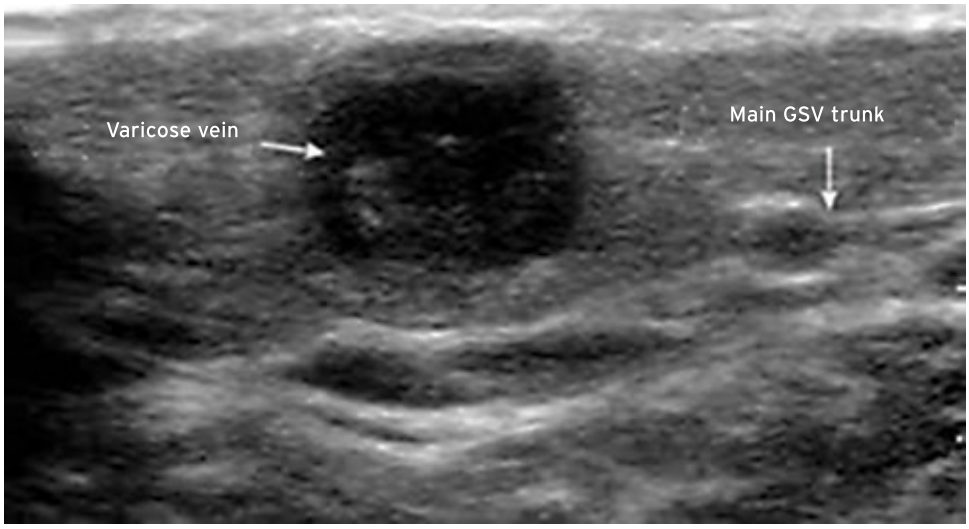
Severity of Each Item rated:
0 (absent) 1 (mild) 2 (moderate) 3 (severe)

Points are summed:	
0-4	No PTS
5-9	Mild PTS
10-15	Moderate PTS
>15 or Ulcer	Severe PTS

Superficial Thrombophlebitis

Superficial Thrombophlebitis (ST) is a relatively common inflammatory process associated with a blood clot (thrombus) that affects the superficial veins, most commonly in the lower extremity.

The condition is generally benign and self-limiting; however, when the axial veins - the great saphenous (GSV) or small saphenous veins (SSV) - are involved propagation into the deep vein system (DVT) and even pulmonary embolism (PE) can occur.



Typical duplex appearance of superficial thrombophlebitis involving a varicosity arising from the great saphenous vein (GSV)

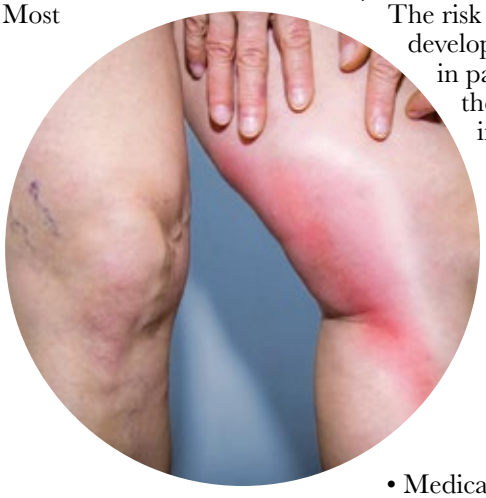
Risk Factors:

The risk of ST is associated with conditions that increase the risk of clotting, including those that lead to venous stasis, abnormalities of coagulation or fibrinolysis, and endothelial dysfunction, as is true with deep venous thrombosis. The most common factors associated with lower extremity superficial thrombophlebitis are varicose veins, post-vein excision/ablation procedures, pregnancy, oestrogen therapy, prior vein thrombosis, malignancy, hypercoagulable states and intravenous catheter use.

Clinical Presentation & Diagnosis:

The clinical presentation is that of pain, tenderness, induration, and/or erythema along the course of a superficial vein. Sometimes the thrombosed vein is palapable as a nodular cord-like structure.

- **Uncomplicated:** Most commonly present with the signs and symptoms listed above. These patient can have a low grade temperature.
- **Complicated:** Patients with clinical features of recurrent ST, infected ST, and those with associated features suggestive of DVT or PE.



Patient with uncomplicated ST where axial vein (GSV or SSV) involvement is not suspected, the diagnosis can be made clinically and a repeat clinical assessment should be repeated a week after the initial diagnosis to look for resolution or progression.

For all other patients with ST (where axial vein involvement is suspected or complicted clinical presentation) a duplex assessment upon initial presentation should be undertaken to rule out the presence of coexistent DVT.

Management:

For all patients diagnosed with ST of the lower extremity superficial veins, supportive measures should be instituted. These include limb elevation, warm or cool compresses, compression stockings, and pain management.

In addition to instituting supportive measures, patients are risk assessed for the likelyhood of developing a DVT.

The risk factors for developing DVT in patients with ST of the lower limb veins includes:

- Extensive thrombosis ≥5 cm
- Proximity of thrombus to the deep venous system (≤5 cm from the saphenofemoral or saphenopopliteal junction)
- Medical risk factors for DVT (prior DVT, thrombophilia, malignancy, estrogen therapy)

In patients who are low risk for DVT development - oral nonsteroidal anti-inflammatory drugs (NSAIDs) and supportive measures are adequate therapy.

In patients who are at increased risk for DVT - anticoagulation for up to 45 days in conjunction with supportive care is indicated. Fondaparinux (most evidence), low-molecular-weight and unfractionated heparin, NOAC's and warfarin appear to be effective.

The exception is in patients who develop ST after radiofrequency or laser vein ablation. These patients are not at high risk for DVT and are managed initially with supportive care and repeat duplex ultrasound assessment.

For patients with ST and DVT at the time of intial diagnosis, the treatment should be according to standard protocols for DVT.

WHAT'S NEW at Matley&Partners

Congratulations to Bhavesh, Claudia and Avi who celebrated the arrival of precious Maya on 17 October 2018.

Vascular Disease and the Anti-Phospholipid Syndrome

Anti-phospholipid syndrome (APS) is an acquired pro-thrombotic state in which recurrent arterial or venous thrombosis may coexist with other pathology and positive anti-phospholipid antibodies. The disease is rare, occurring either as a primary condition or secondary to other auto-immune conditions as systemic lupus erythematosus (SLE).

In APS, a heterogeneous family of anti-phospholipid antibodies (aPL) are directed against phospholipid-binding plasma proteins such as 2-glycoprotein I. According to the test used to detect these auto-antibodies, they are termed either anti-cardiolipin antibodies (aCL) or lupus anticoagulants (LA).

We have recently treated a 14-year-old male with idiopathic juvenile arthritis who presented with a critically ischaemic left foot and a two-year history of claudication symptoms involving both lower limbs, in addition to well established sacro-iliitis and ankylosing spondylitis. On clinical examination, the left popliteal and ankle pulses were absent and the right sided pulses diminished. Duplex Doppler demonstrated a high-grade stenosis in the proximal left SFA and subsequent angiography showed that in addition to this high-grade SFA lesion, there was a complete occlusion of the left popliteal artery (with a generous collateral network around the knee) as well as an intermediate-grade stenosis of the right popliteal *(Figures 1 and 2)*.

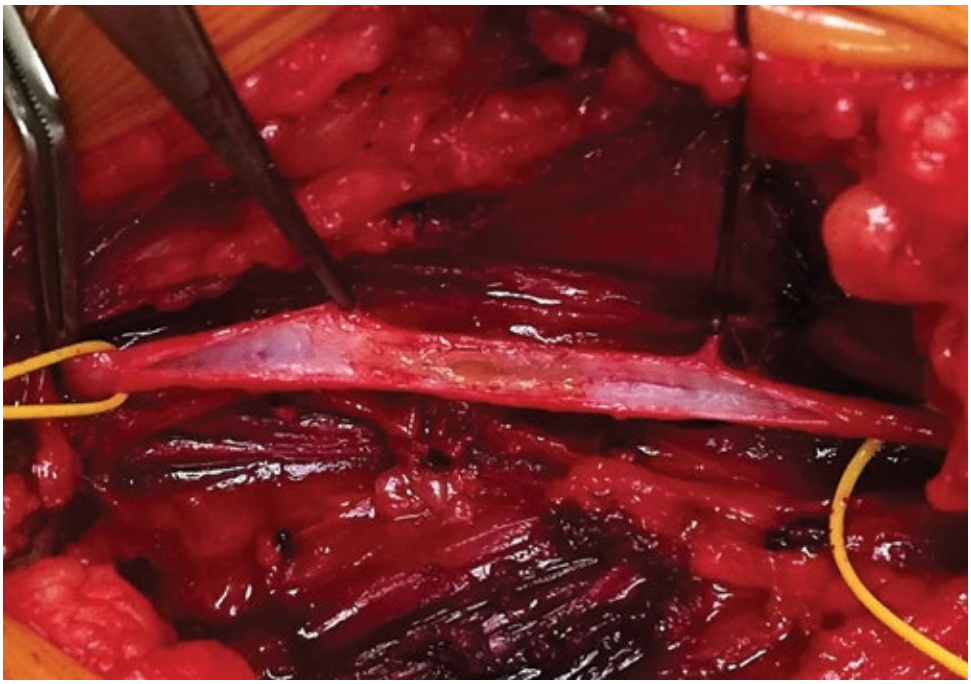


Fig 3: Left SFA at operation

At surgery, a focal 3cm obstruction of the SFA was found with thick firm tissue obstructing the lumen *(Fig 3)*. The diseased section of SFA was resected with placement of an interposition saphenous vein graft. Histological examination of the resected artery showed aggressive intimal hyperplasia as the cause of the stenosis with no evidence of vasculitis or thrombosis. *(See Fig 3)*

Tests for anti-nuclear factor (ANF) and rheumatoid factor were negative as was the test for HLA-B27. A very high titre of anti-phospholipid antibodies was found. Antiphospholipid syndrome has been demonstrated to be an important cause of intimal hyperplasia and is almost certainly the cause of the arterial obstruction in our recent case.

APS is associated with an increased risk for coronary artery and peripheral arterial disease. In addition to overt clinical conditions such as peripheral ischaemia and stroke, patient with APS may have sub-clinical endothelial dysfunction, increased arterial stiffness, increased arterial intima-media thickness, and a high prevalence of asymptomatic atherosclerotic plaques.

An increase in intima-medial thickness can often be detected non-invasively using a carotid duplex scan.

Treatment of APS involves anti-coagulation, usually with vitamin K antagonists and treatment of any associated auto-immune disease. Low dose aspirin is of uncertain benefit and statins are reserved for proven hyperlipidaemia. APS should be considered as a possible cause of coronary, cerebrovascular and peripheral artery disease as well as recurrent venous thrombo-embolism, particularly in patients with other systemic and auto-immune conditions.

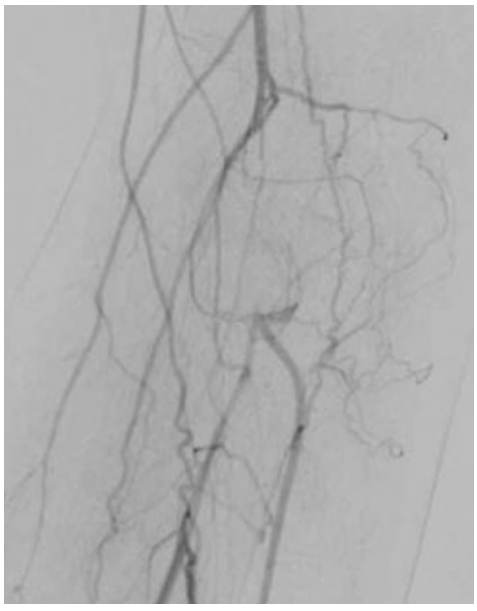


Fig 1: Left Popliteal artery

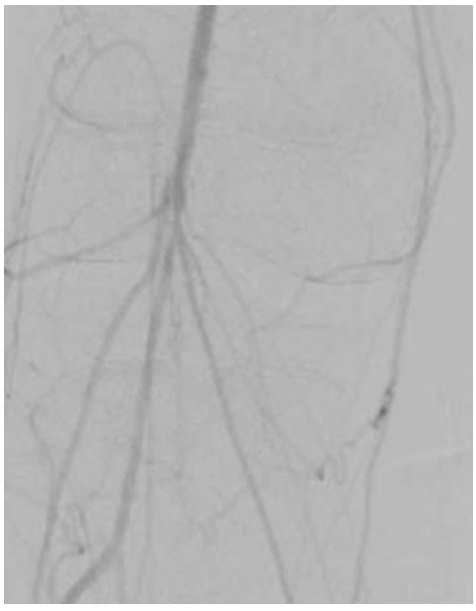


Fig 2: Right Popliteal artery