



# **Clinical Case**

## **Klippel – Trenaunay Syndrome**



This is a 46 year-old male with past medical history of Klippel-Trenaunay Syndrome (KTS) who presents with **left leg swelling, venous claudication, port wine stains, prominent varicosities, and intolerance to cold temperatures** especially in left forefoot/toes partially alleviated with massage and elastic compression stockings.

## History

**Medical history:** KTS diagnosed in 1993, Obesity, Superficial thrombophlebitis (SVT). No previous DVT, PE or Thrombophilia.

**Surgical history:** Denies

**Social history:** Non-smoker, EtOH socially

**Family history:** Non-contributory

**Allergies:** None

**Medications:** Coumadin 15 mg daily

**Integumentary: Port wine stain (nevus flammeus)** extending from left lateral thigh to the calf and toes.

**Vascular:**

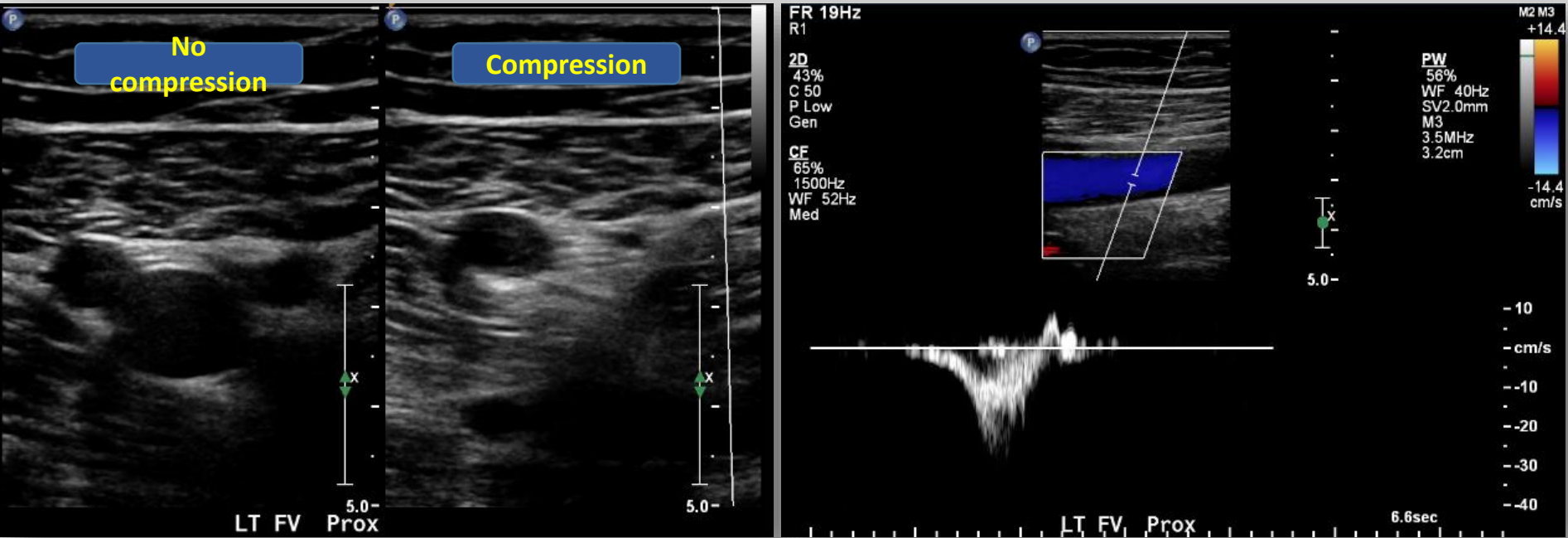
Bilateral lower extremities warm and well perfused

2+ distal pulses bilaterally

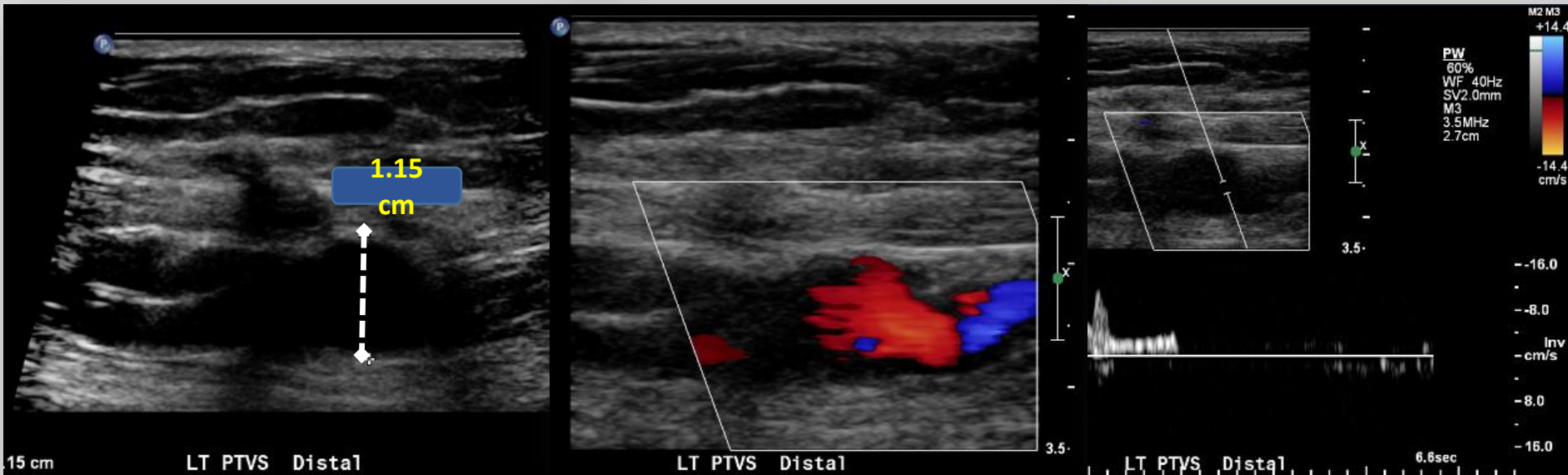
**2+ pitting edema** involving left thigh and calf.

**Prominent varicosities** of the left thigh, medial calf and shin.

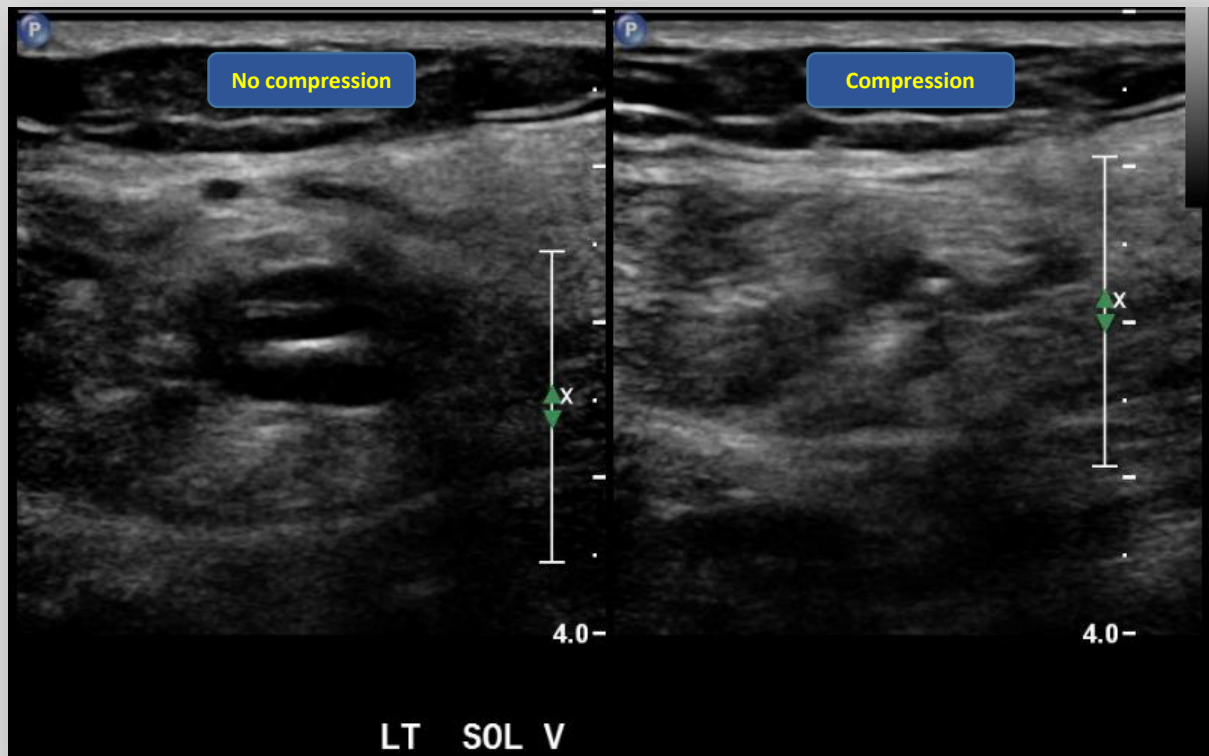




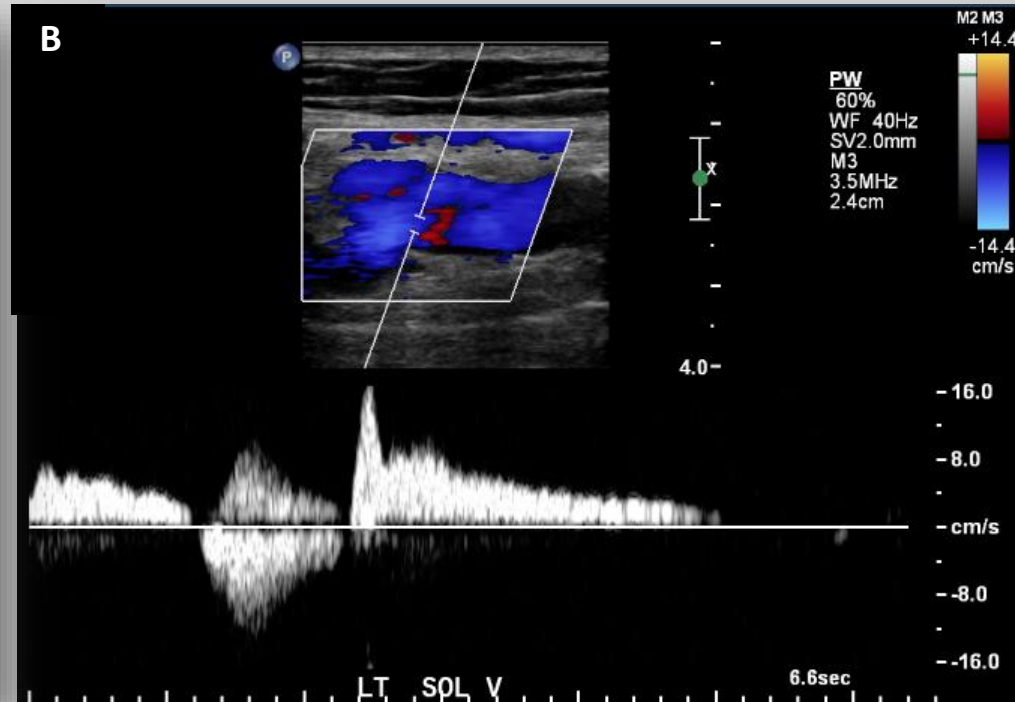
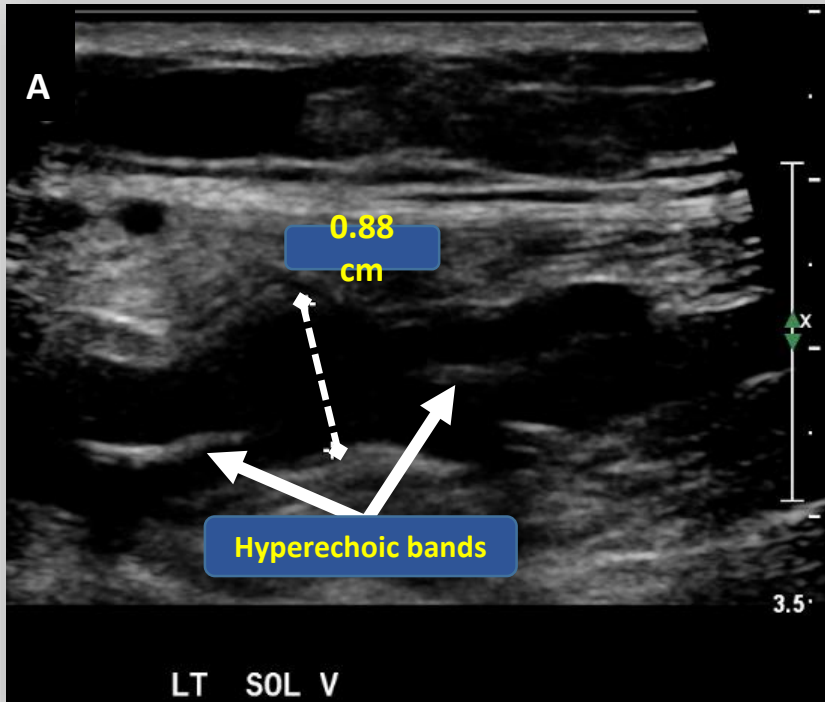
**Compressible left Femoral Vein (FV). Absence of filling defect or pathological reflux.**



Dilated left Posterior Tibial Vein (PTV). No filling defect. **Pulse Wave Doppler shows > 0.5 seconds reflux.**

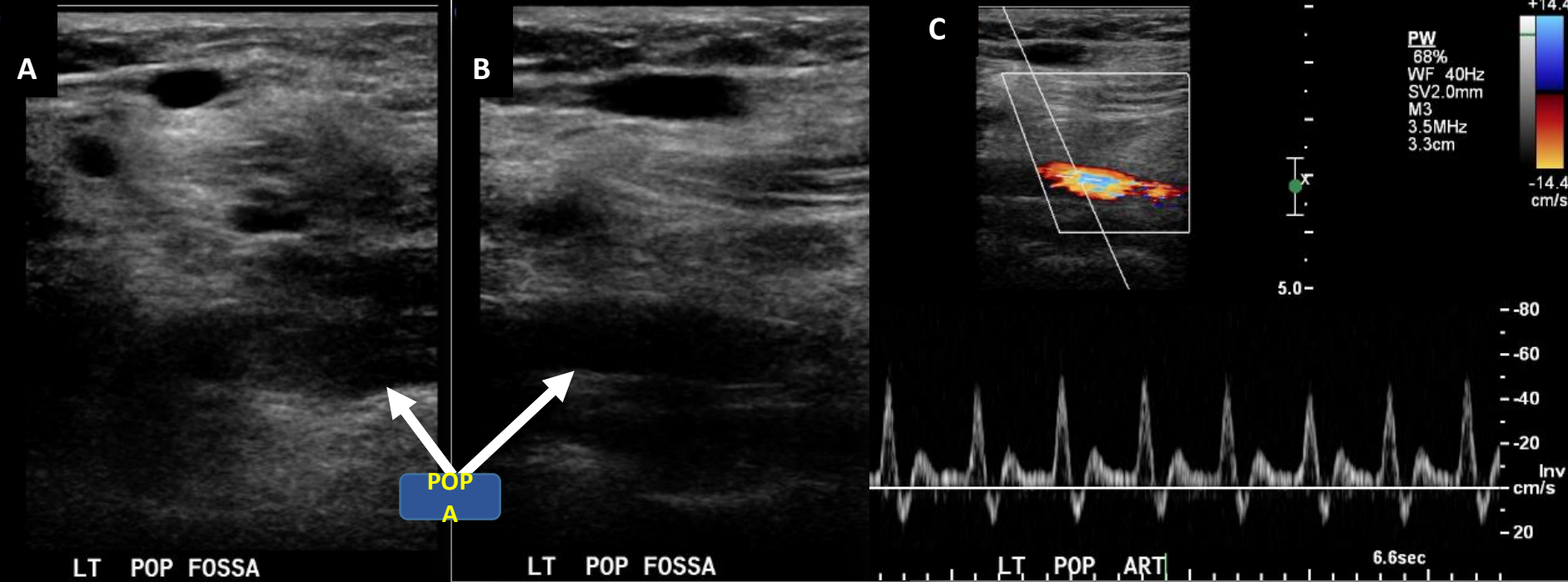


**Lack of compressibility of the left Soleal vein.**

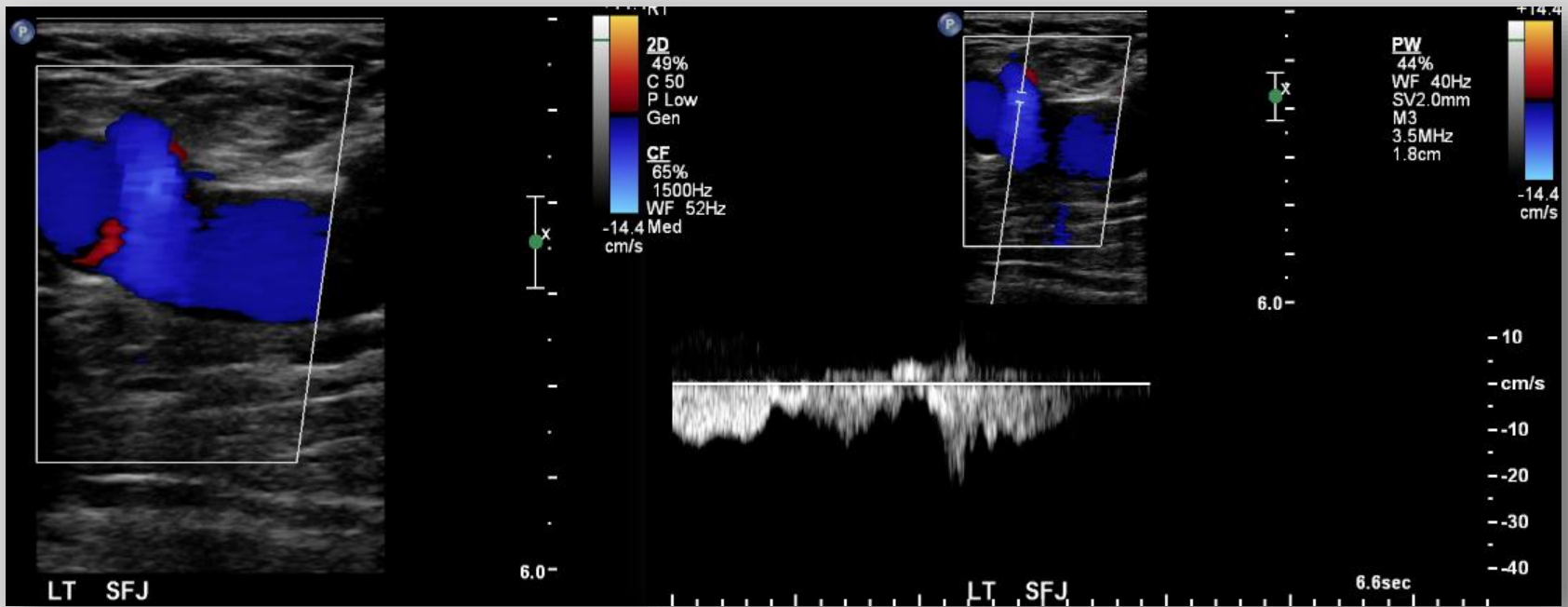


- A. Post-thrombotic luminal changes of the left Soleal vein.**
- B. Pulse Wave Doppler shows > 0.5 seconds reflux.**



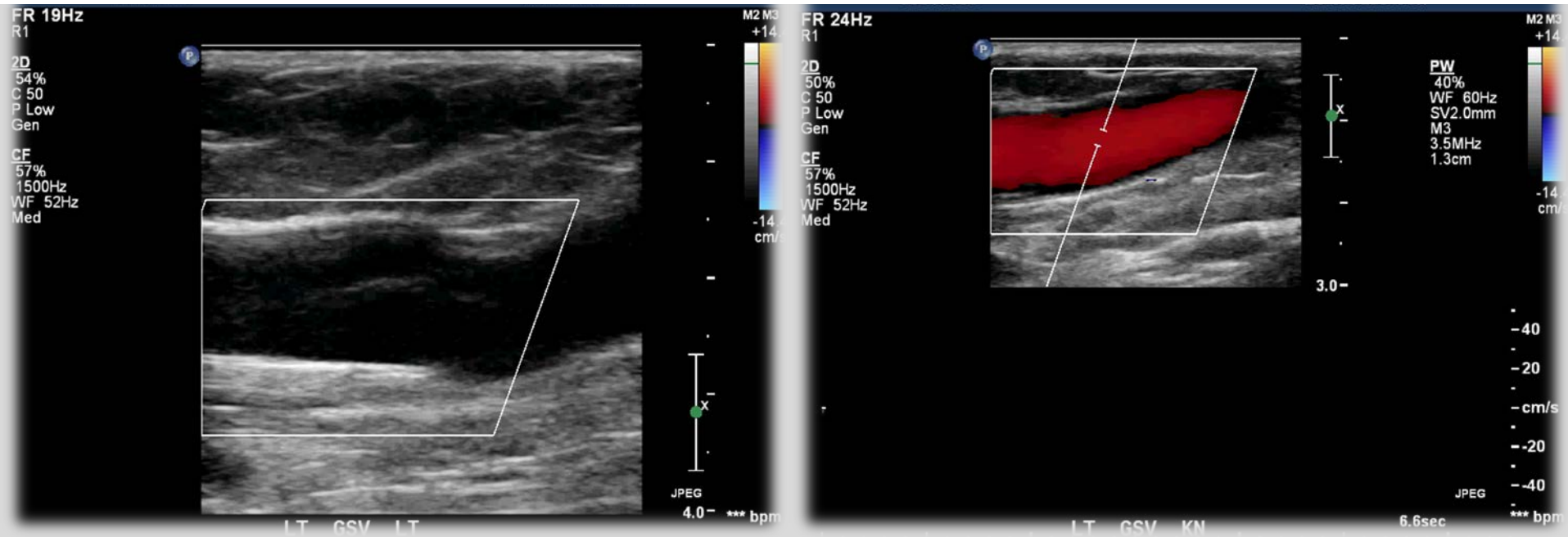


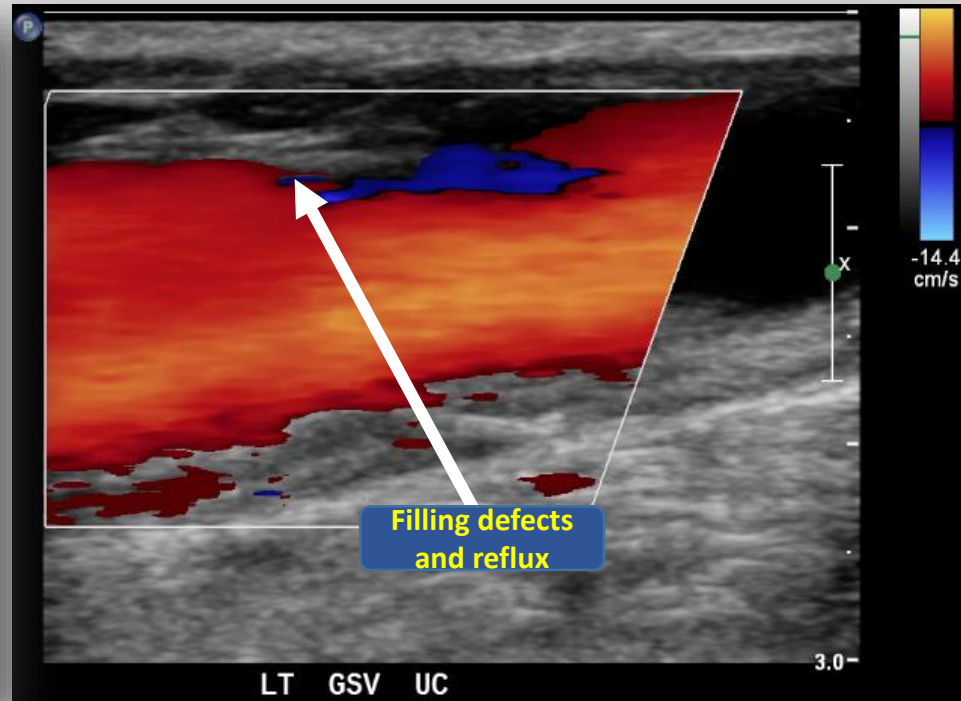
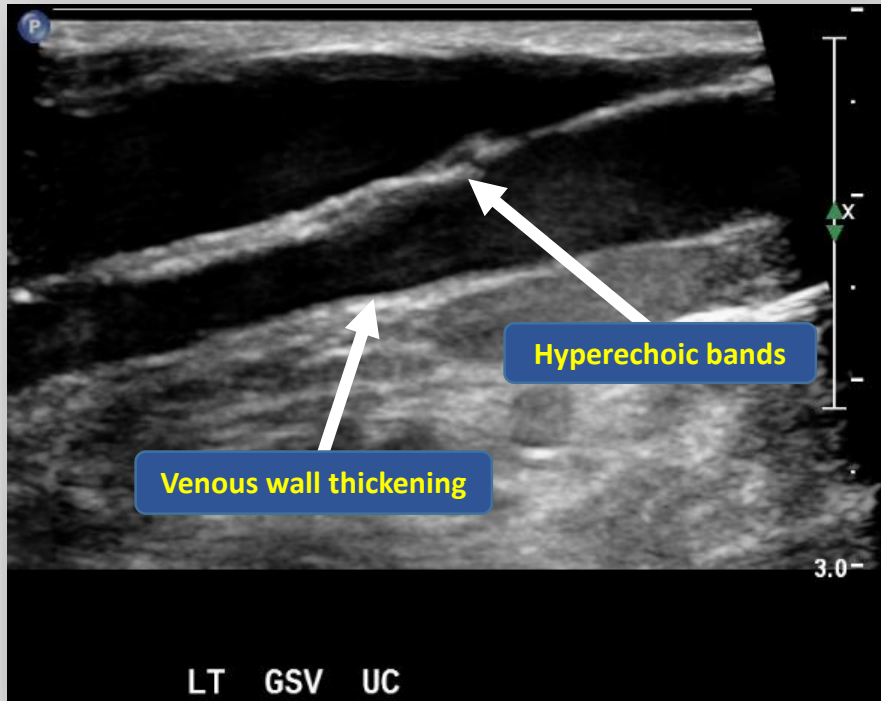
**A, B. Agenesis of the left Popliteal vein (POP V). B. Normal flow in the left popliteal artery (POP A)**



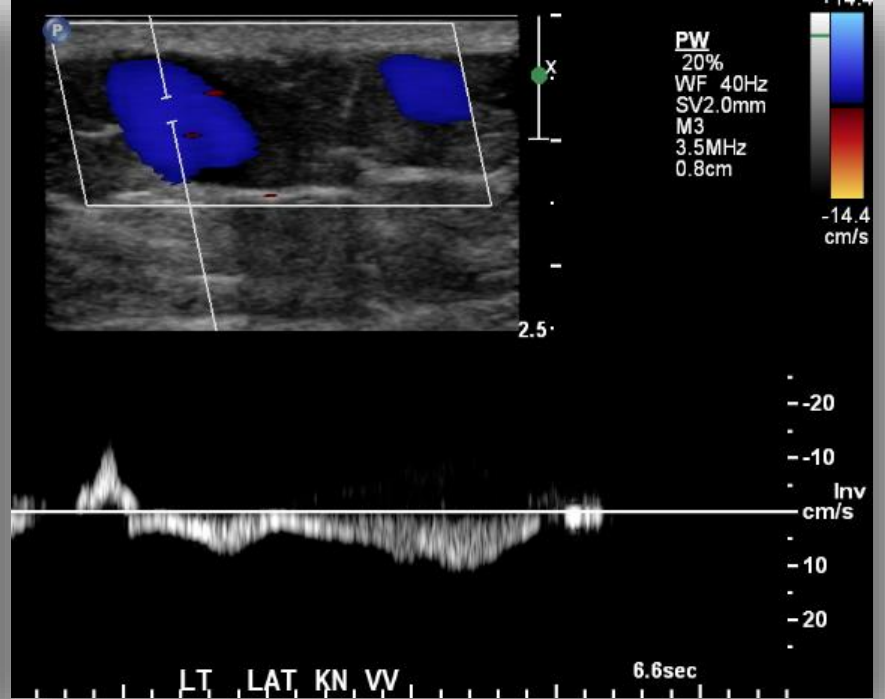
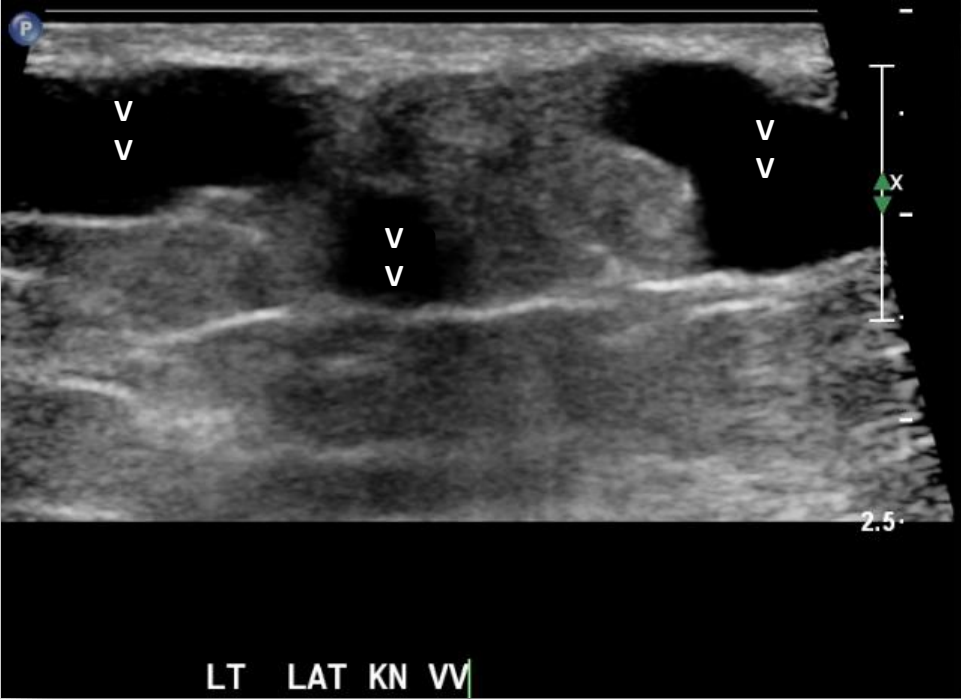
**Left Saphenofemoral junction (SFJ) reflux > 4.0 seconds.**

# Proximal Great Saphenous Vein (GSV) dilated with > 5 sec retrograde flow.





**Chronic luminal post-thrombotic changes of the GSV in the upper calf.**



Multiple refluxing varicose veins (VV) in the left lateral knee.

Based on our patient's clinical presentation and Duplex ultrasound findings, how would you categorize this patient using the CEAP classification?

1. C<sub>5s</sub> E<sub>s</sub> A<sub>p</sub> P<sub>o</sub>

2. C<sub>3s</sub> E<sub>c</sub> A<sub>sd</sub> P<sub>ro</sub>

3. C<sub>2s</sub> E<sub>p</sub> A<sub>s</sub> P<sub>r</sub>

4. C<sub>3s</sub> E<sub>p</sub> A<sub>d</sub> P<sub>ro</sub>

5. C<sub>4s</sub> E<sub>p</sub> A<sub>s</sub> P<sub>r</sub>

Based on our patient's clinical presentation and Duplex ultrasound findings, how would you categorize this patient using the CEAP classification?

1. C<sub>5s</sub> E<sub>s</sub> A<sub>p</sub> P<sub>o</sub>

2. C<sub>3s</sub> E<sub>c</sub> A<sub>sd</sub> P<sub>ro</sub>

3. C<sub>2s</sub> E<sub>p</sub> A<sub>s</sub> P<sub>r</sub>

4. C<sub>3s</sub> E<sub>p</sub> A<sub>d</sub> P<sub>ro</sub>

5. C<sub>4s</sub> E<sub>p</sub> A<sub>s</sub> P<sub>r</sub>

C<sub>3s</sub>: Leg edema

E<sub>c</sub>: Congenital (Klippel-Trenaunay Syndrome)

A<sub>sd</sub>: Superficial and deep venous system

P<sub>ro</sub>: Reflux and Obstruction

The patient had no skin damage. The discoloration was typical of KTS.

Based on our patient's clinical presentation and Duplex ultrasound findings, how would you treat this patient?

1. Continue anticoagulation and compression therapy with no procedural intervention.
2. Continue current management plus endovenous ablation of the GSV.
3. Deep venous valve transposition or transplantation if structural damage exist should be the first treatment option.
4. Add a venoactive drug to current therapy.



Based on our patient's clinical presentation and Duplex ultrasound findings, how would you treat this patient?

1. Continue current management with anticoagulation and compression therapy with no procedural intervention.
2. **Continue current management plus endovenous ablation of the GSV.**
3. Deep venous valve transposition or transplantation if structural damage exist should be the first treatment option.
4. Add a venoactive drug to current therapy.

## Conclusion

- ❖ Our patient presented with **venous claudication** which could be due to **popliteal vein aplasia and extensive venous reflux.**
- ❖ Duplex ultrasound demonstrated **superficial and deep venous reflux and popliteal vein aplasia.**
- ❖ Treatment recommendations included **EVA of the GSV.**
- ❖ He will continue with **lifelong anticoagulation and elastic compression stockings.**

**GSV was dilated with high velocity long duration reflux (>5s).  
Such vein is not a collateral and can be safely ablated.**

## Discussion

- ❖ **Klippel-Trenaunay Syndrome** is a complex congenital disorder characterized by the **classic triad of nevus flammeus, varicosities and venous malformation (VM), and limb overgrowth, with or without lymphatic malformation.**

Wassef M, et al. Vascular Anomalies Classification: Recommendations From the International Society for the Study of Vascular Anomalies. Pediatrics. 2015 Jul;136(1):e203-14.

- ❖ **Varicosities and other VM** are present in the majority of patients (**72%**).

Jacob AG, et al. Klippel-Trénaunay syndrome: spectrum and management. Mayo Clin Proc. 1998 Jan;73(1):28-36.

## Discussion

- ❖ Most common deep venous aplasia concerned the **POP V (51%), FV (16%), both POPV and FV (25%)**. The iliac and the inferior vena cava are rarely involved.

Servellw M, Babillot J. Deep vein malformations in the Klippel-Trenaunay syndrome. *Phlebologie*. 1980 Jan-Mar;33(1):31-6.

- ❖ Symptomatic varicose veins and VM can be treated using different procedures when the deep system is patent, however it encompasses a **high recurrence rate**.

Jacob AG, et al. Klippel-Trénaunay syndrome: spectrum and management. *Mayo Clin Proc*. 1998 Jan;73(1):28-36.

Noel AA, et al. Surgical treatment of venous malformations in Klippel-Trénaunay syndrome. *J Vasc Surg*. 2000 Nov;32(5):840-7.