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OBJECTIVE:

Klippel Trenaunay Weber syndrome (KTS) is a congenital triad of capillary malformations, varicose veins, and limb overgrowth. Varicose veins in these patients are often extensive, debilitating and psychologically distressing. This study reviews endovascular varicose vein treatment pearls and pitfalls in KTS patients.

MATERIALS AND METHODS:

This study retrospectively reviewed three KTS patients with debilitating and distressing varicose veins, treated at our institution.

DIAGNOSIS:

The diagnosis of KTS can be made clinically, with observation of limb hypertrophy, capillary malformation and extensive varicose veins. Patients should then be evaluated for possible coagulopathy, including laboratory assay (e.g., CBC, INR, PTT, Fibrinogen, D-Dimer). Clinical history and physical exam should assess for additional vascular malformations. KTS patients have high-rates of venous malformations. Targeted follow-up imaging including ultrasound and MRI can be obtained.



Patient 3, a 27 year old male presenting to our vascular anomalies clinic with progressively worsening pain with walking, cramping and burning. Exam demonstrated limb hypertrophy, "port wine stain" capillary malformation of the affected limb and extensive varicose vein formation. The patient had been previously treated with GSV excision at outside institution.

FOLLOW-UP:

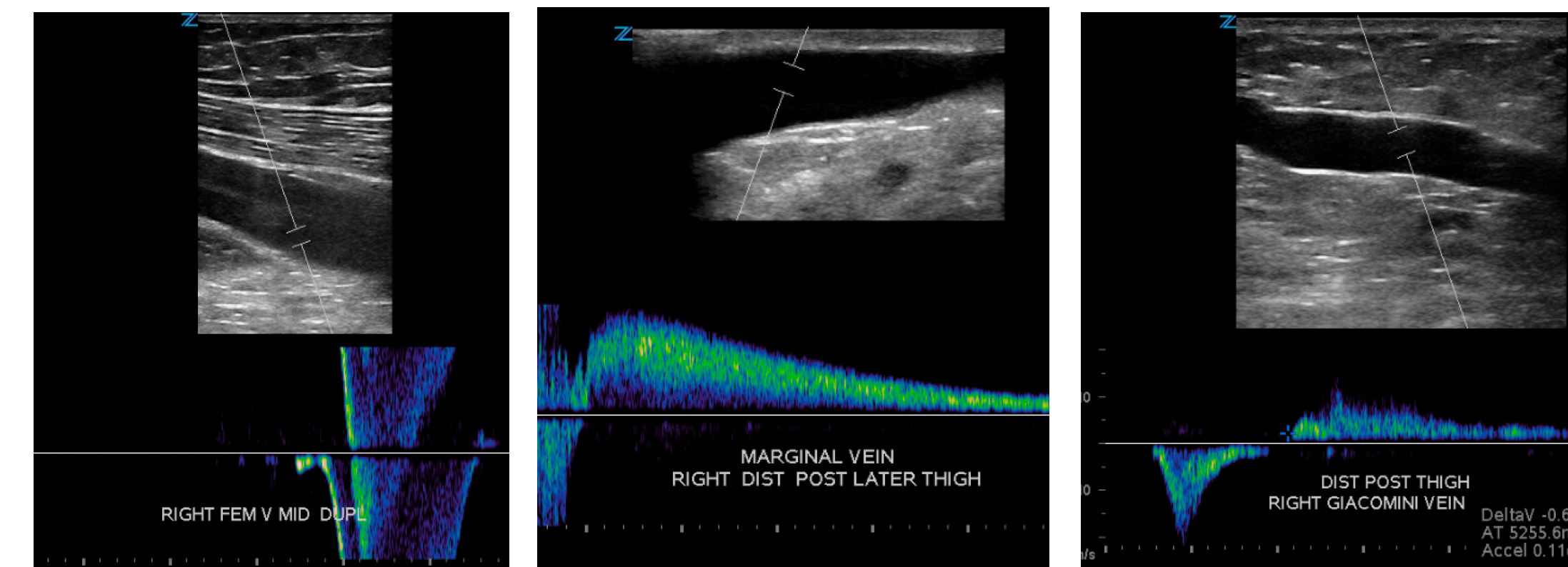


Patient 3 (pre-treatment imaging shown above), underwent 15 endovascular treatments including repeat endovenous ablation of the lateral marginal vein and vein of Giacomini, with intervening check-ups, physical exams and ultrasound assessments, until the patient ultimately achieved satisfactory cosmetic and clinical improvements.

Following treatment of varicose veins patients should be followed in the immediate post-procedural time period to assess for complications, with a focus on neurovascular injury. Follow-up is then again performed in the one to three month post-treatment period to assess for treatment response. Incomplete responses, which are to be expected, should prompt repeat evaluation of the embryological veins. In this case series, significant clinical and cosmetic improvements were achieved in all cases. Recurrence invariably occurs and can be repeated the same treatment regimen.

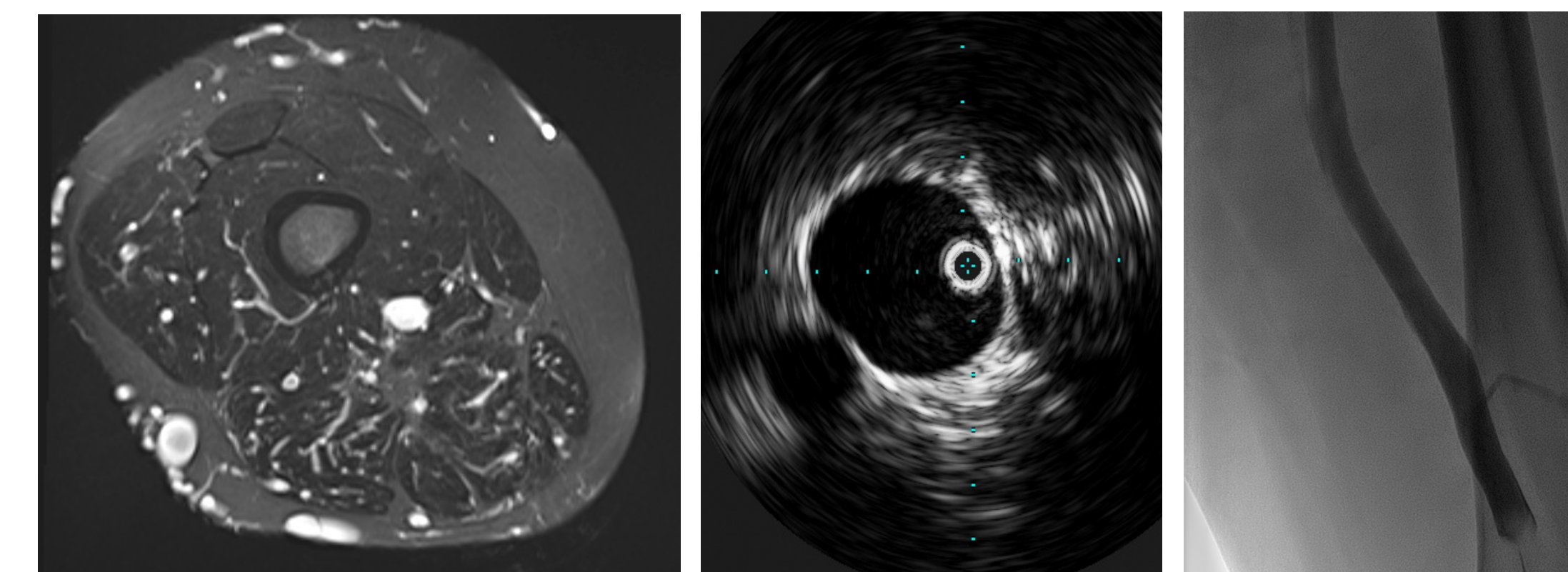
WORK-UP

Treatment of varicose veins in KTS patients is made challenging by the extensiveness of the varicose veins and the underlying embryological veins which cause reflux and are difficult to close. Management of these patients is exacerbated by surgical excision or ablation of the great saphenous vein, which redirects blood flow into the embryological veins. The work-up should begin with MRI and Duplex US to evaluate for the presence of embryological veins and assess the deep venous system.



Ultrasound assessment demonstrated a patent deep venous system, a lateral marginal vein 8-10mm in diameter with 6s of reflux, and a vein of Giacomini measuring 5-7mm with 6s of reflux.

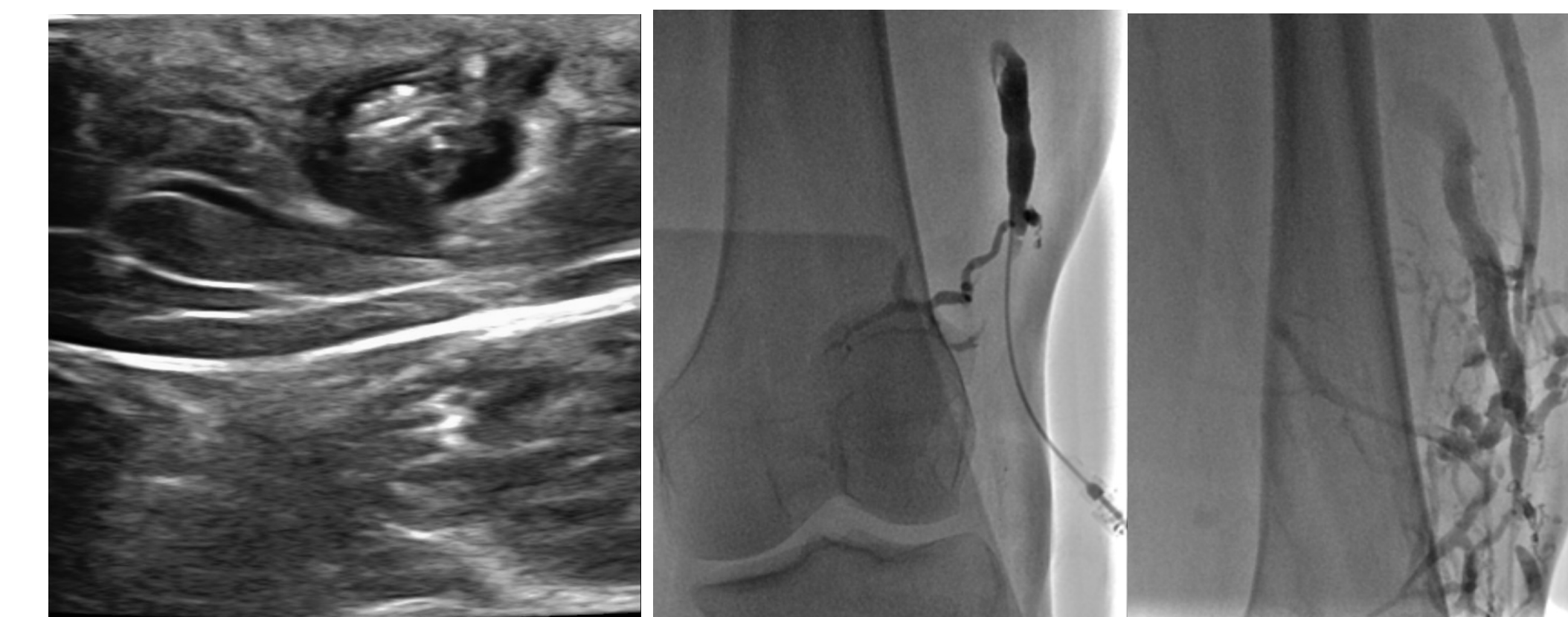
In cases where there is concern for deep venous stenosis, intravascular ultrasound (IVUS) can be performed to confirm patency. Once the deep venous system has been confirmed to be patent. Mapping of the embryological veins and communication with the deep system and varicose veins should be performed through a combination of Duplex, MRI and Venography.



From left to right: MRI demonstrated a large lateral marginal vein which drained into the deep femoral and common femoral veins, consistent with a Type III lateral marginal vein. Intravascular ultrasound and venography showing a patent deep venous system without narrowing.

ENDOVASCULAR TREATMENT

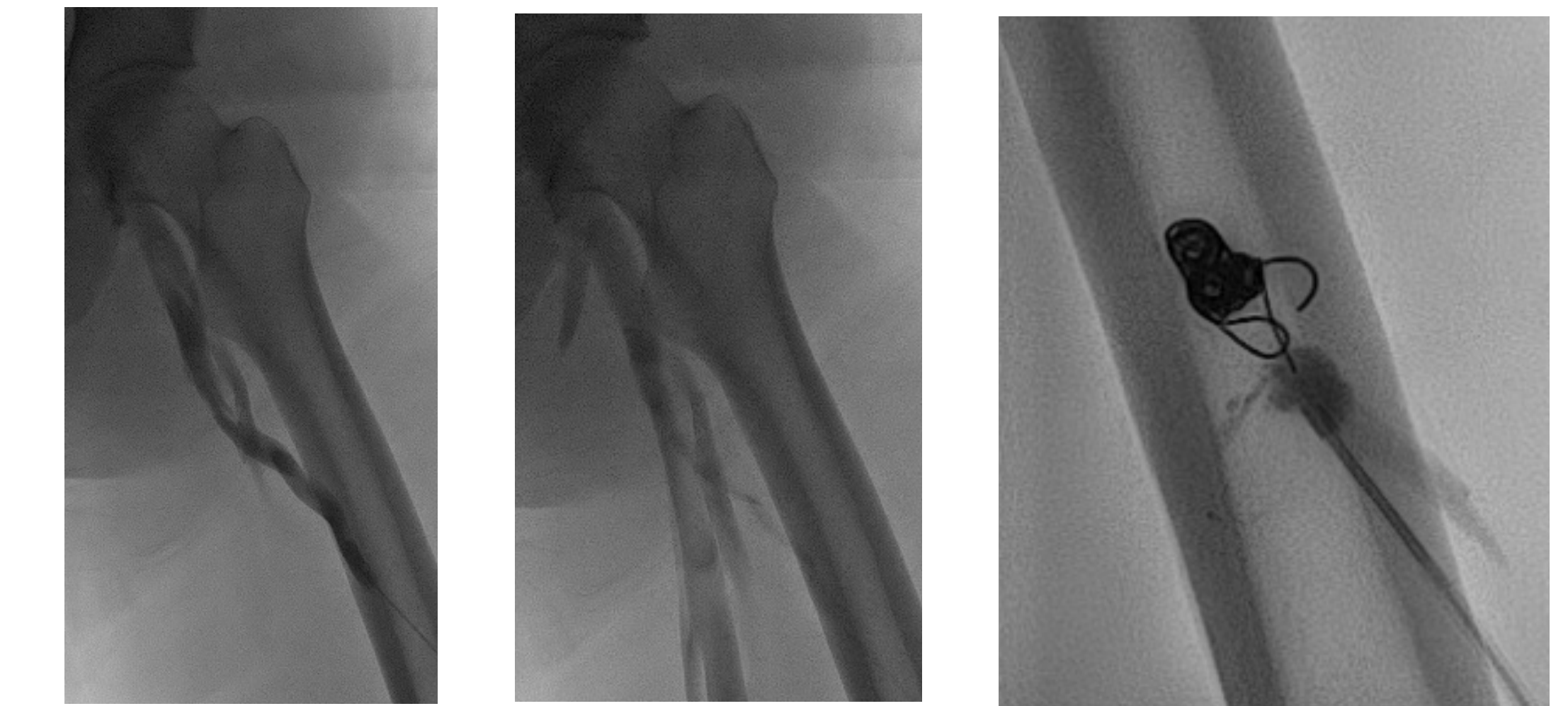
Treatment should focus on occluding the embryological veins, followed by occlusion of the superficial varicosities. Common options include endovenous ablation, sclerotherapy and mechanochemical ablation. General anesthesia can be used for pediatric patients. In our institution, embryological veins were first treated with endovenous ablation, followed by sclerotherapy of the varicose veins and residual embryological veins. Endovenous ablation is typically performed with 500cc of saline, 25 cc of 2% lidocaine, and 10 cc of sodium bicarbonate. Multiple access points are necessary due to the tortuosity of the vessels. Post-treatment compression stockings are recommended. Ultrasound follow-up and sclerotherapy is then repeated until 100% occlusion is achieved. A lateral marginal vein was the primary culprit in two cases, with an accessory great saphenous vein in the other.



From left to right: Ultrasound showing thrombus of the lateral marginal vein following endovenous ablation; direct percutaneous venogram of the lateral marginal vein showing severe reflux into numerous varicose veins.

EMBOLIZATION

Occlusion of embryological veins is challenging in itself. However, this can be exacerbated in patients with excised or ablated great saphenous veins (GSV). In the absence of the GSV, more blood is drained via embryological veins and thus may increase the propensity for recanalization. In patients with recurrent recanalization of embryological veins, coil embolization at the insertion to the deep venous system can be considered. In one case, a patient with with recanalization of a 8-10mm lateral marginal vein was coil embolized.



Direct stick venogram of the femoral vein demonstrates reflux into the embryological lateral marginal vein. This is followed by percutaneous coil embolization of the lateral marginal insertion into the femoral vein.

ADVERSE EVENTS

One patient developed temporary malleolar ulceration which was treated with topical anti-biotics. No major adverse events occurred.

Patient Number	Age & Gender	Embryological vein	Great Saphenous vein	Ablation treatments	Sclerotherapy treatments	Additional procedures	Clinical outcome
1	24 F	Lateral Marginal	Surgically excised	0	3	None	Near complete. Good satisfaction.
2	53 F	Anterior accessory	Intact	2	4	None	Near complete. Good satisfaction.
3	27 M	Lateral Marginal and Giacomini	Surgically excised	3	12	Coil embolization of marginal	Near complete. Good satisfaction.

REFERENCES

Lambert G, Teplisky D, Cabezas M, et al. Mechanochemical Endovenous Ablation of Varicose Veins in Pediatric Patients with Klippel-Trénaunay Syndrome: Feasibility, Safety, and Initial Results. *J Vasc Interv Radiol.* 2021;32(1):80-86. doi:10.1016/j.jvir.2020.08.019
 Bittles M, Jodeh DS, Mayer JLR, Gallant M, Rottgers SA. Laser ablation of embryonic veins in children. *Pediatr Int.* 2019;61(4):358-363. doi:10.1111/ped.13804
 Frasier K, Giangola G, Rosen R, Ginat DT. Endovascular radiofrequency ablation: a novel treatment of venous insufficiency in Klippel-Trenaunay patients. *J Vasc Surg.* 2008;47(6):1339-1345. doi:10.1016/j.jvs.2008.01.040