

Case 1

35yo F presents to CHCO VAC 11/2018

- Purple stain to right lateral thigh, RLE overgrowth, blebbing to foot, right buttock, groin
- Evaluated at multiple facilities throughout lifetime
 - Has been given several different diagnoses
 - Has undergone several surgical excisions of bothersome “hemangiomas”
- DVT at R knee @ 18yo, Coumadin since that time
- Main complaints
 - Syncopal unless wearing compression
 - Painful blebbing in groin





What is the proper diagnosis?

Klippel-Trenaunay Syndrome

First described ~1900 by French physicians Dr. Marice Klippel and Dr. Paul Trénaunay

Constellation of:

- Lateral capillary malformation
- Underlying venolymphatic malformations
 - Lateral marginal vein (vein of Seville)
 - +/- superficial and deep lymphatic malformation
- Overgrowth of tissue and bone
- Driven by somatic PIK3CA mutation

Goals of Therapy

- Supportive
 - Improve function
 - Optimize quality of life
- Preventative
 - Early imaging
 - Surgical, Interventional, Pharmacotherapy
- Reactive
 - Complications
 - Surgical, Interventional, Pharmacotherapy

Evaluation: Capillary Malformation

- Typically laterally placed on the extremity

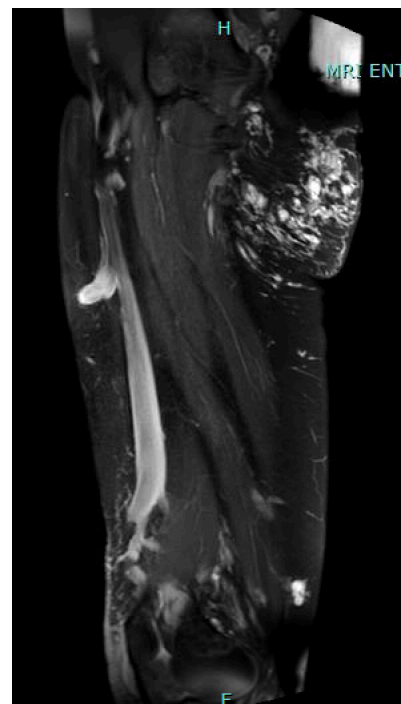
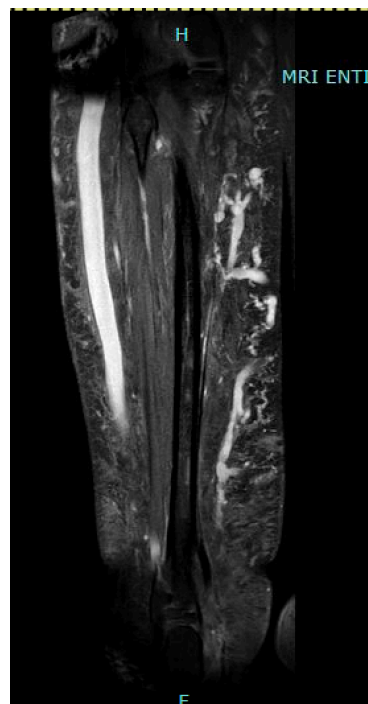


Evaluation: Venous Malformation

MRI abdomen/pelvis & MRI RLE:

- Lateral marginal vein that empties into greater saphenous
- Ectasias around R knee
- Perforators draining into R internal iliac
- Large infrarenal inferior vena cava





Remnant of Embryonic Vessels

- Failure to regress or involute
- Defect in regression of lateral fibular vein results in the inappropriate persistence of a marginal vein
 - Valveless
 - Increased reflux
 - Chronic venous hypertension / stasis
- High risk for venous thromboembolism and pulmonary embolus

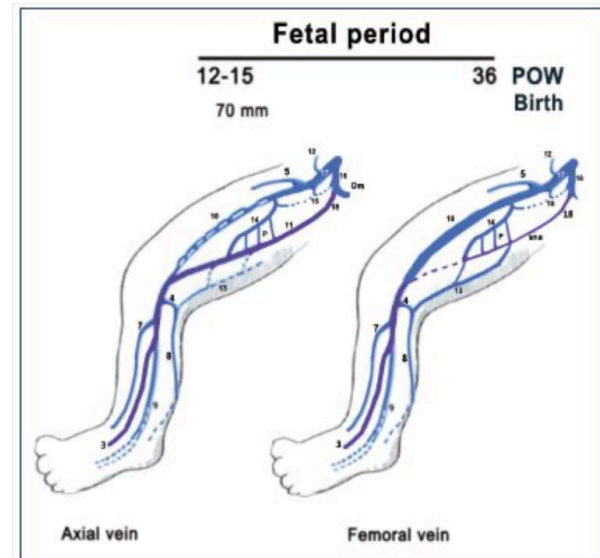


Figure 8. Summary of the fetal period of development (after week 8).

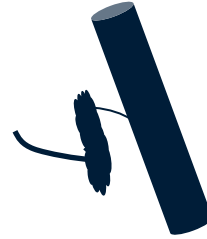
The primitive veins are colored in purple. Notice that the fetus (weeks 9 to 15) commonly has a big axial vein while the femoral vein is smaller and plexus shaped. Later, the axial vein becomes a small arcade along the sciatic nerve, and the femoral vein becomes the main trunk of the thigh in the majority of cases at birth, commonly with a collateral canal.

Abbreviations: 3, fibular veins; 4, arch of the SSV; 5, great saphenous vein; 7, anterior tibial veins; 8, SSV; 9, posterior tibial; 10, femoral vein; 11, axial vein; 12, epigastric vein; 13, thigh extension of SSV; 14, deep femoral vein; 15, obturator vein; 16, hypogastric vein; 17, external iliac vein; 18, inferior gluteal vein; cc, collateral canal; P, perforating branches of the femoral vein; POW, postovulatory week; sna, axial arcade along the sciatic nerve; SSV, small saphenous vein.

Venous Malformation & Venous Ectasia



TYPE 1



TYPE 2

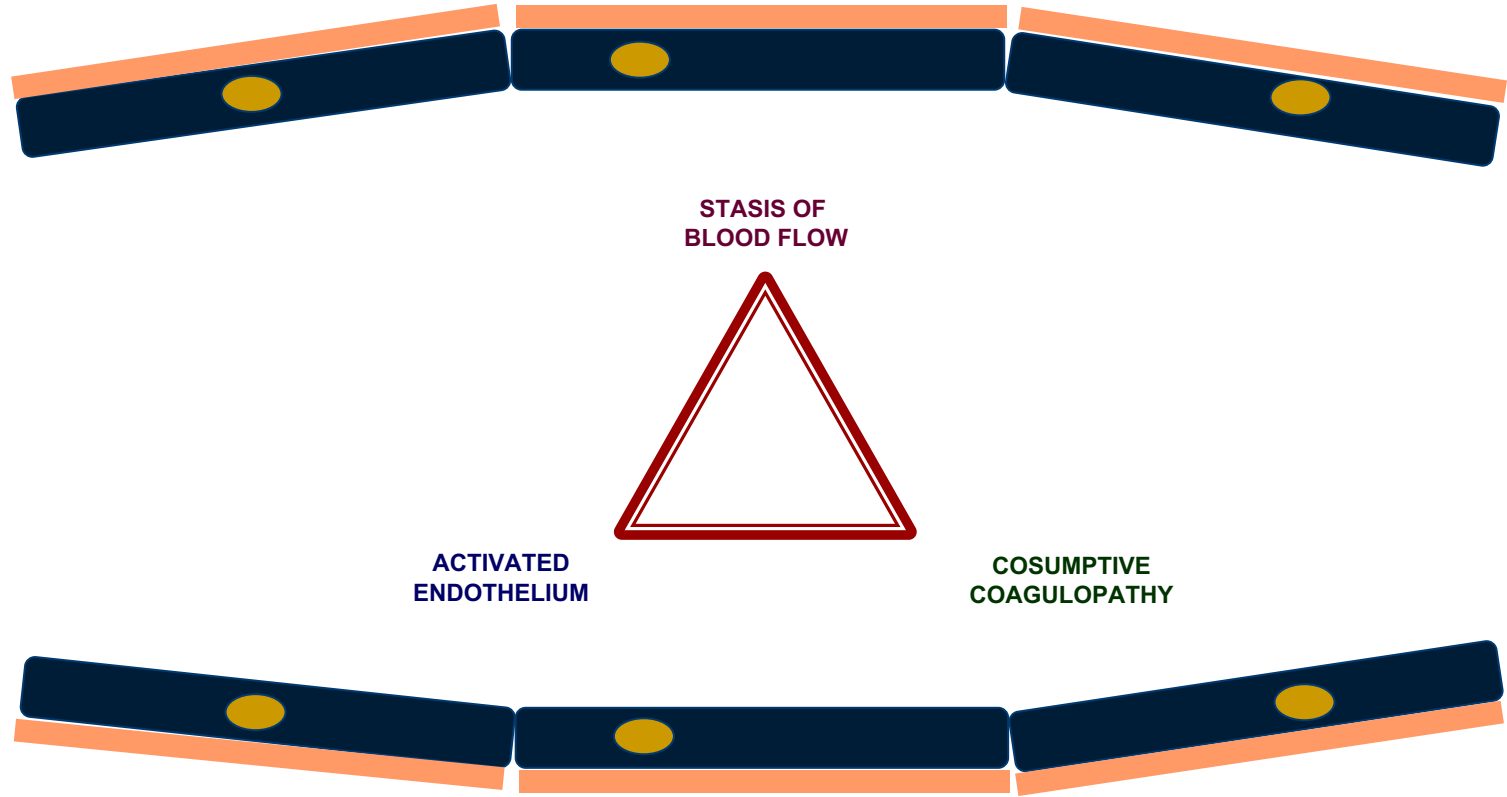


TYPE 3



TYPE 4

Venous Malformation

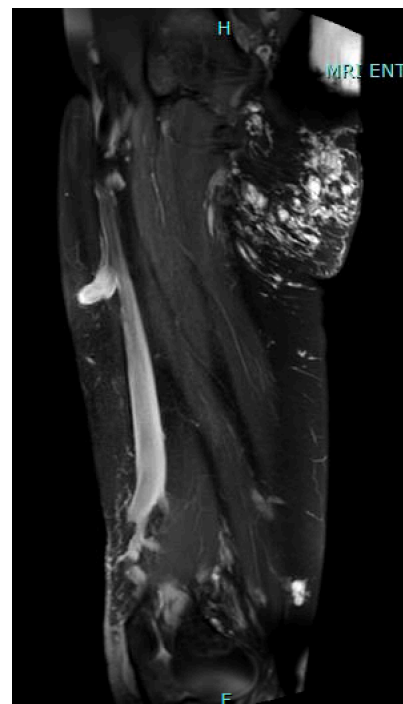
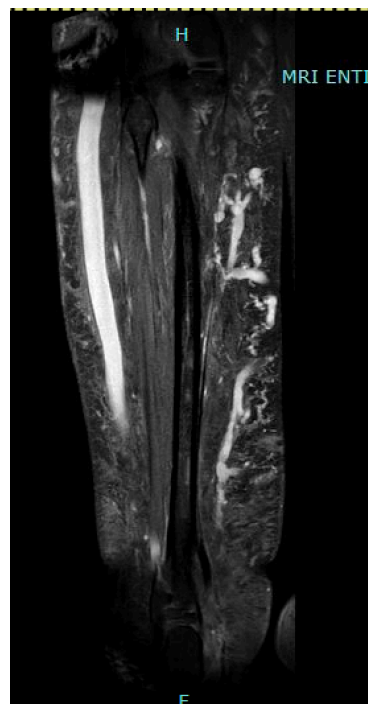


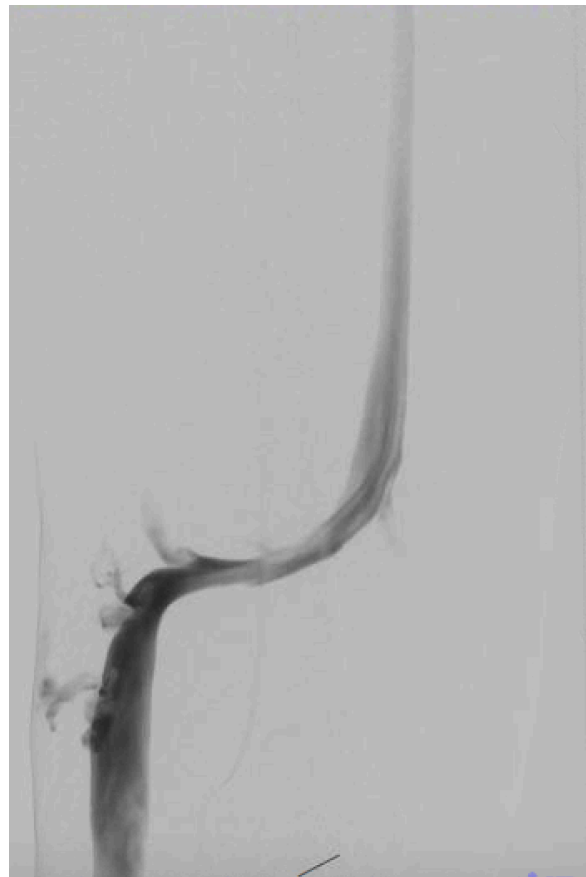
Evaluation: Localized Intravascular Coagulopathy

- Intralesional thrombophilia causes local consumption of clotting factors.
- Transient thrombi and chronic phleboliths are present, may be palpable, and can cause discomfort.
- Demonstrates a decrease in plasma fibrinogen, factor V, factor VIII, factor XIII and increased D-dimer
- Increased peri-procedural (surgical or interventional) risk of severe hemorrhage

Coagulopathies of Pediatric Vascular Anomalies

VASCULAR ANOMALY	PLATELETS	FIBRINOGEN	D-Dimer	PT/PTT	IMPACT
RICH	↓↓	NORMAL	↑	↑	Low risk of Hemorrhage
KHE with KMP	↓↓↓↓	↓↓↓↓	↑↑	↑	Life-threatening Hemorrhage
VM with LIC	NORMAL/↓	↓	↑↑	↑↑	Risk of Thrombosis & Hemorrhage
HHT	NORMAL	NORMAL	NORMAL	NORMAL	AVMs risk life-threatening hemorrhage





Treatment options?

- Patient progressed to syncopal episodes while wearing compression, likely due to large size of marginal vein
- Concern regarding presence of deep venous system
 - Would marginal vein ablation lead to venous congestion?
- Referral to vascular surgery due to patient age:
 - Resection of vein
 - Surgical ligation
 - Endovenous laser ablation

Outcome

Patient received venogram and ascending phlebography 12/2019, which demonstrated presence of deep venous system

Endovenous ablation to RLE marginal vein at the knee

Patient has demonstrated a drastic improvement in orthostasis

- No syncopal episodes while wearing compression
- Able to walk short distances without a compression garment