

Case 3: a complex lesion

- 3-month-old female presents to clinic to discuss a “hemangioma” noted at birth.



What is this child's diagnosis?

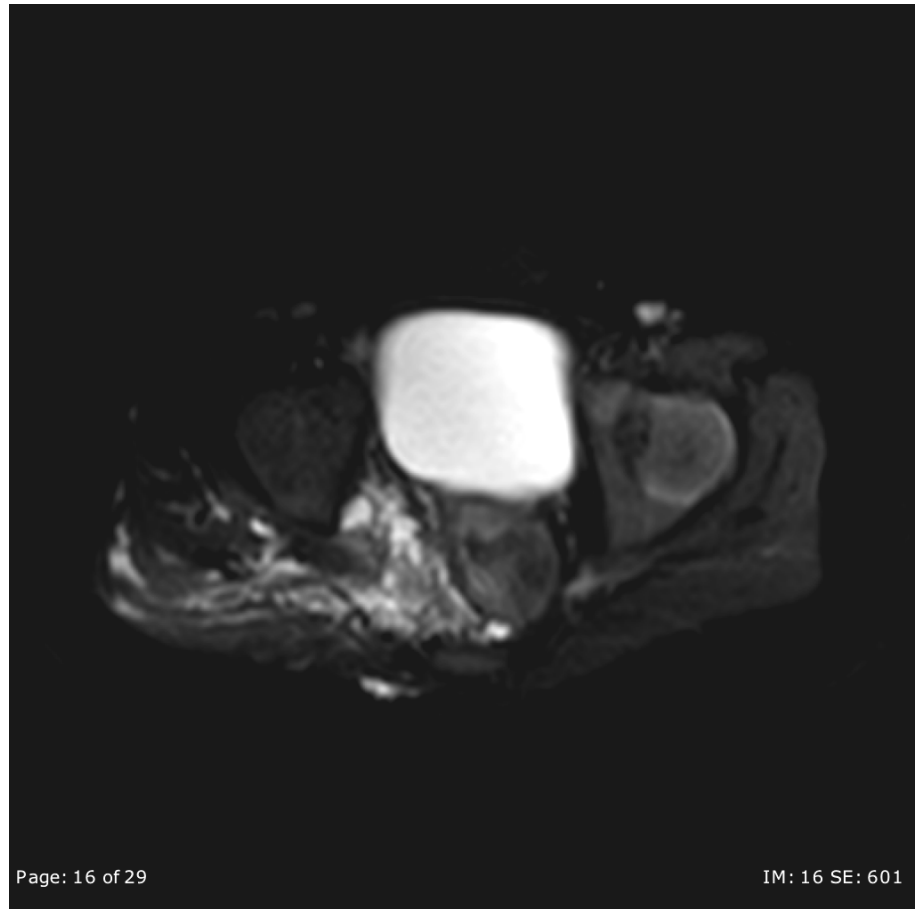
Case 3: a complex lesion

- 3-month-old female presents to clinic to discuss a “hemangioma” noted at birth.
- Klippel-Trenaunay syndrome
 - Capillary Venolymphatic Malformation
 - Overgrowth of the affected limb
 - Limb length discrepancy
 - Bleeding
 - Pain



Case 3: a complex lesion

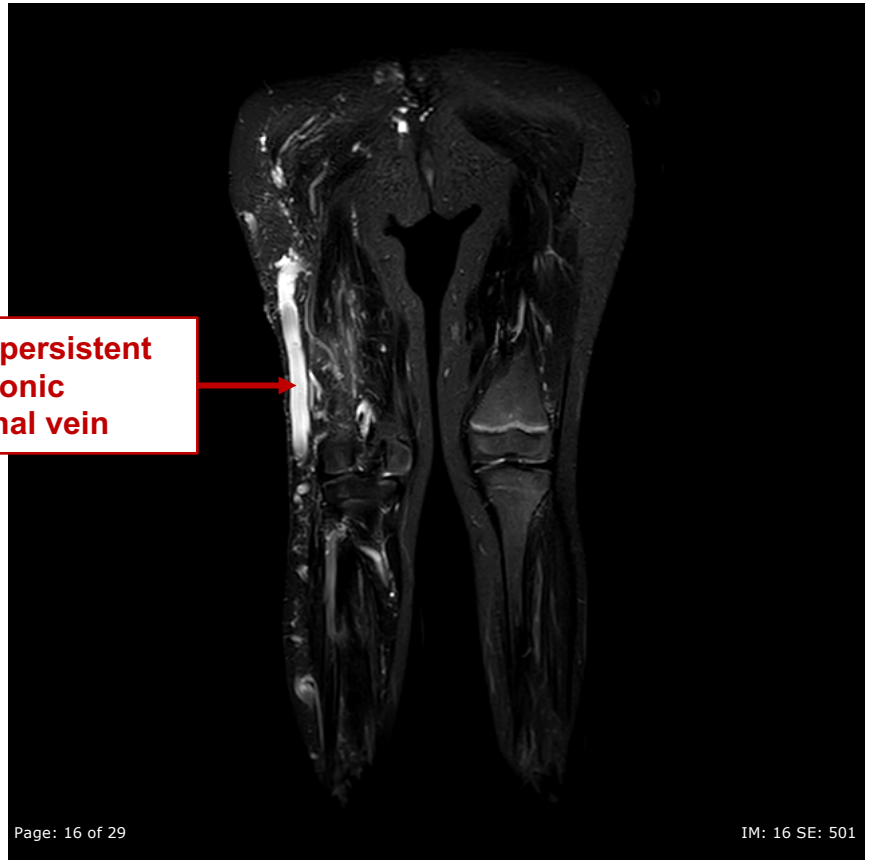
- 3-month-old female presents to clinic to discuss a “hemangioma” present at birth.
- Klippel-Trenaunay syndrome
 - Capillary Venolymphatic Malformation
 - Overgrowth of the affected limb
 - Limb length discrepancy
 - Bleeding
 - Pain
- MRI: Slow-flow malformation of right pelvic soft tissue extending into RLE consistent with mixed venous and lymphatic malformation.



Case 3: a complex lesion

- 3-month-old female presents to clinic to discuss a “hemangioma” present at birth.
- Klippel-Trenaunay syndrome
 - Capillary Venolymphatic Malformation
 - Overgrowth of the affected limb
 - Limb length discrepancy
 - Bleeding
 - Pain
- MRI: Slow-flow malformation of right pelvic soft tissue extending into RLE consistent with mixed venous and lymphatic malformation.

Large persistent embryonic marginal vein



Page: 16 of 29

IM: 16 SE: 501

Case 3: a complex lesion

- 3-month-old female presents to clinic to discuss a “hemangioma” present at birth.
- Klippel-Trenaunay syndrome
 - Capillary Venolymphatic Malformation
 - Overgrowth of the affected limb
 - Limb length discrepancy
 - Bleeding
 - Pain
- MRI: Slow-flow malformation of right pelvic soft tissue extending into RLE consistent with mixed venous and lymphatic malformation.

LAB EVALUATION	RESULTS	
D-dimer	0.27	Normal
PT	13.1	Normal
aPTT	29.8	Normal
Fibrinogen	249	Normal

What are this child's therapeutic options?

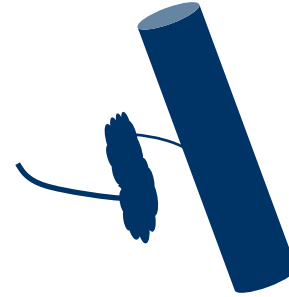


Should this patient receive periprocedural anticoagulation?

Venous Ectasia and Embolic Risk



TYPE 1



TYPE 2



TYPE 3



TYPE 4

High Risk Diagnosis

Extensive or multifocal venous malformations (VM)
Combined slow-flow lesions with VM (LVM, CLVM)
CLOVES

LVM: lymphatic venous malformation, CLVM: capillary lymphatic venous malformation = Klippel-Trenaunay syndrome, CLOVES: congenital lipomatous with vascular anomalies, epidermal nevus and spinal/skeletal/scoliosis

Obtain laboratory investigation:

- D-dimer
- PT / aPTT
- Fibrinogen
- CBC with smear

Low Risk

- Normal PT / aPTT &
- Normal fibrinogen &
- No venous ectasia &
- Normal platelet count &
- D-dimer normal or < 5x ULN &
- Negative personal medical history of thrombosis &
- No family history of thrombosis in 1st degree family members

- **LMWH likely not indicated**
- Encourage early ambulation
- Encourage compliant use of mechanical thromboprophylaxis

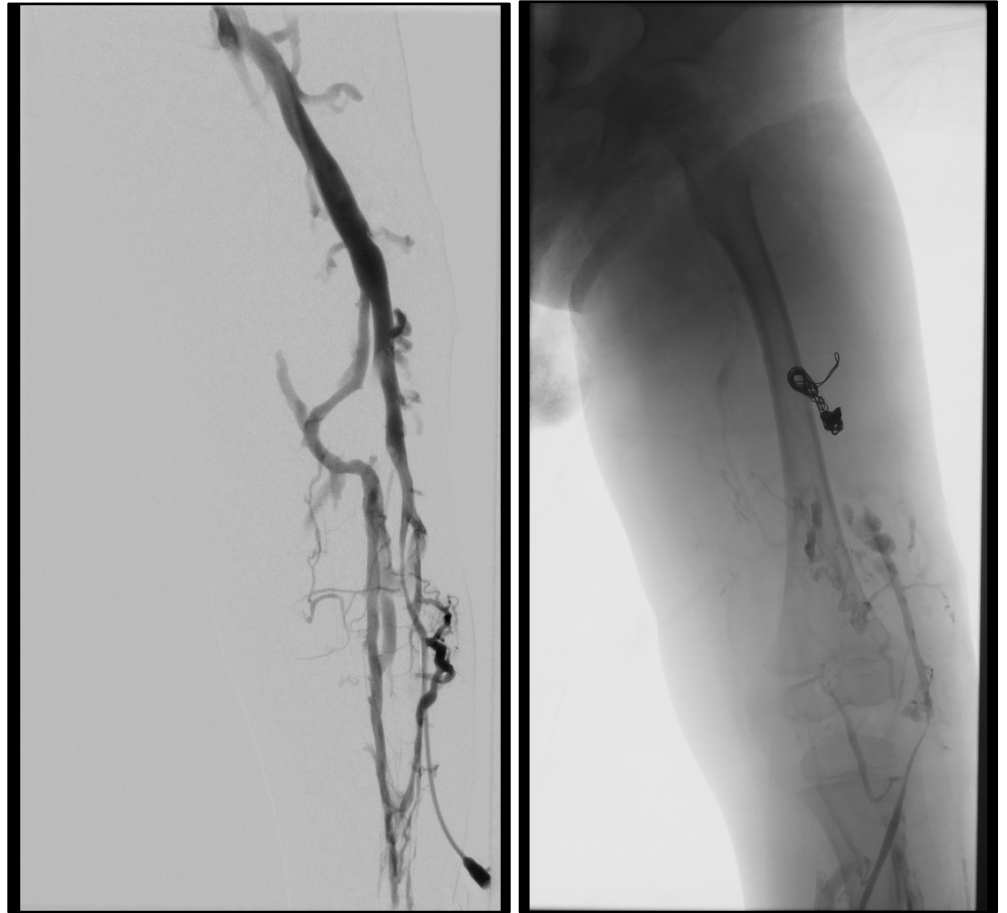
High Risk

- Low fibrinogen **OR**
- Venous ectasia **OR**
- D-dimer >5x normal **OR**
- Thrombocytopenia (without other explanation) **OR**
- Positive personal or family history of thrombosis

- **Two weeks prior to procedure, start LMWH 0.5mg/kg SC BID for 1 week, recheck labs.**
- **If abnormality still present, increase to 1mg/kg SC BID, recheck labs in pre-op.**
- **Post-procedure, continue pre-procedure dose for 2 weeks or baseline ambulation, whichever is longer**
- Encourage early ambulation
- Encourage compliant use of mechanical thromboprophylaxis

Case 3: a complex lesion

- 3-month-old female presents to clinic to discuss a “hemangioma” present at birth.
- Klippel-Trenaunay syndrome
 - Capillary Venolymphatic Malformation
 - Overgrowth of the affected limb
 - Limb length discrepancy
 - Bleeding
 - Pain
- MRI: Slow-flow malformation of right pelvic soft tissue extending into RLE consistent with mixed venous and lymphatic malformation.



What additional supportive care would you consider for this patient?

Supportive Care

- Compression therapy
- Leg length discrepancy
- Wound care
- PIK3CA testing
- Cancer screening
- Physical therapy
- Psychosocial support