Anesthetic Management for Parturients with Klippel-Trenaunay Syndrome: A Case Series

Amanda Ruiz MD, Emily E. Sharpe MD, Mingchun Liu MD
Mayo Clinic Department of Anesthesiology and Perioperative Medicine – Rochester

Background

Klippel-Trenaunay Syndrome (KTS) triad[1]

- Bone and soft tissue hypertrophy of one or more limbs
- Venous varicosities
- Cutaneous capillary malformations

Cause and incidence

- Sporadic congenital mutation in PIK3CA (angiogenesis)[3]
- 1:30,000 to 1:100,000 live births[3]

KTS complications and comorbidities[1,2]

- Venous thromboembolism
- Bleeding from malformations and chronic anemia

Obstetric anesthesia management of KTS

- Previously: avoid pregnancy[3]
- Data limited to case reports

Study Aims

Review anesthetic and peripartum care of KTS parturients within Mayo Clinic Health System to determine considerations for safe anesthetic care

Methods

- IRB exemption
- Inclusion criteria: pregnant patients with diagnosis of KTS that received anesthetic peripartum care in Mayo Health System
- Review of electronic health record with extraction of demographic, obstetric, and anesthetic details

Results

6 patient with KTS with a combined 10 encounters

Case Type and Anesthetic	No. Cases
Vaginal delivery (VD)	3
Unmedicated/IV	3
Epidural	0
Cesarean Delivery (CD)	6
Spinal	2
Epidural	1
General	3
Other	
Cerclage placement	1

Delivery Method and Neuraxial Considerations

- **CD Indications**: Abdominal cerclage n=2, Repeat n=2, Arrest of descent n=1, and KTS sequelae n=1
- **Vaginal Delivery**: Stillborn at 20 weeks GA with IV analgesia n=1, Unmedicated although offered neuraxial n=1, Unknown n=1
- MRI during pregnancy n=1; MRI prior to pregnancy n=1
- Anticoagulation n=2

Airway Considerations

- No noted difficult airway

Blood Management, Access, and Monitoring

- Estimated blood loss ≥1000 mL, n=4
- Invasive intraarterial monitoring, n=1

Complications

- Acute postoperative pulmonary embolism n=1

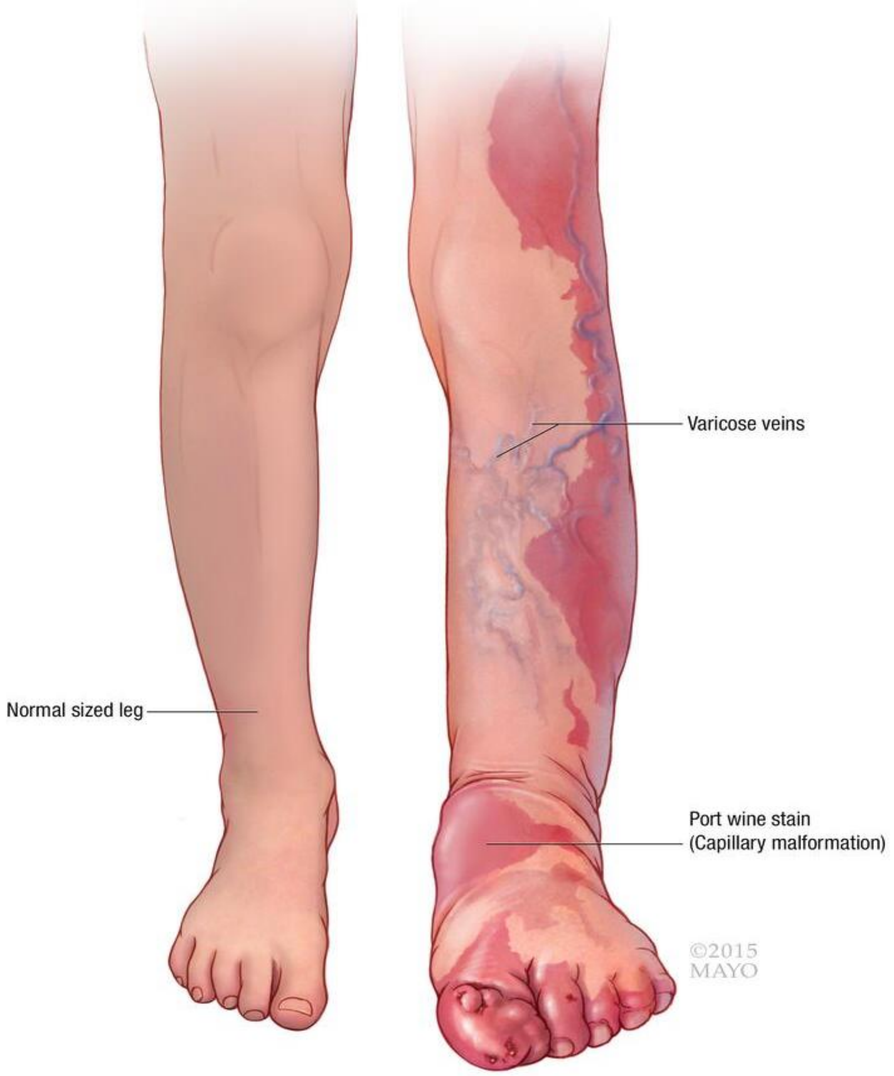
Discussion

Delivery Method[3-5]

- **Concerns**:
  - Venous malformations in pelvis, uterus, perineum, and abdominal wall
  - High-risk vaginal birth or cesarean delivery
- **Recommendations**:
  - Multidisciplinary planning
  - Recent pelvic MRI
  - Documented plan for emergent cesarean delivery

Neuraxial Considerations[3-5]

- **Concerns**:
  - Venous malformations in or near spine
  - Scoliosis
  - Prophylactic anticoagulation use
- **Recommendations**:
  - Physical exam
  - Recent MRI of lumbar spine
  - Review anticoagulation



Klippel-Trenaunay Syndrome – example of signs in lower leg.

Discussion

Blood Management, Access, and Monitoring[3,4]

- **Concerns**:
  - Potential large volume blood loss from abnormal venous plexus in uterus, vagina, and subcutaneous tissue
  - Chronic anemia
- **Recommendations**:
  - Type and screen or crossmatch
  - Cell-salvage
  - Delivery location and surgeon availability
  - Blood product availability
  - Large bore intravenous or central access
  - Invasive arterial hemodynamic monitoring

Airway Considerations[3,5]

- **Concern**:
  - Venous malformations in airway
- **Recommendation**:
  - Airway exam and MRI

Postoperative Cares

- **Concerns**:
  - Risk of coagulopathy and venous thromboembolism
  - Chronic pain in affect limb
- **Recommendations:**
  - Postoperative monitoring of blood loss, coagulation studies, and blood count
  - Multidisciplinary assessment of anticoagulation
  - Shared decision making for multimodal pain control

References

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5. Teixeira CEFA. *Braz J Anesthesiol*. 2018;68(6):641-644.