



Challenges during Labor Analgesia in a Patient with Klippel-Trenaunay Syndrome

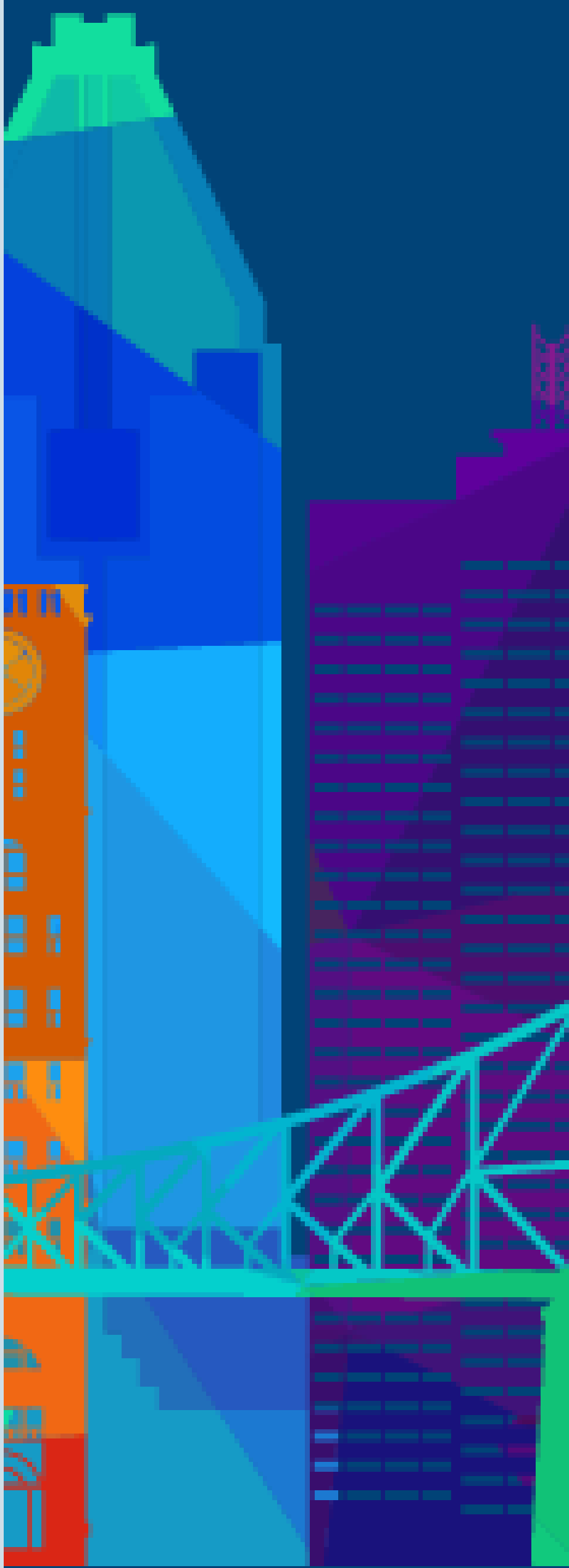
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Andrew Lee, M.D., Daniel Chang, M.D.

BACKGROUND

- **Klippel-Trenaunay (KT) syndrome:** manifests as capillary, venous, and lymphatic malformations predominantly affecting a single lower extremity.
- Presents as **cutaneous** port-wine stains, (90-100%); **venous** (varicosities, aplasia/hypoplasia, valvular incompetence, [70-100%]); and **lymphatic** (lymphedema [15-50%]).^{1,2}
- Patients with KT syndrome present challenges for the obstetric anesthesiologist due to **vascular malformations surrounding the spinal cord**, vascular malformations within the pelvis increasing risk of **post-partum hemorrhage**, and risk of **thrombus formation and embolus** due to physiologic changes of pregnancy causing venous distension and stasis.
- In this case, we highlight the management of a parturient with KT syndrome without complete workup.





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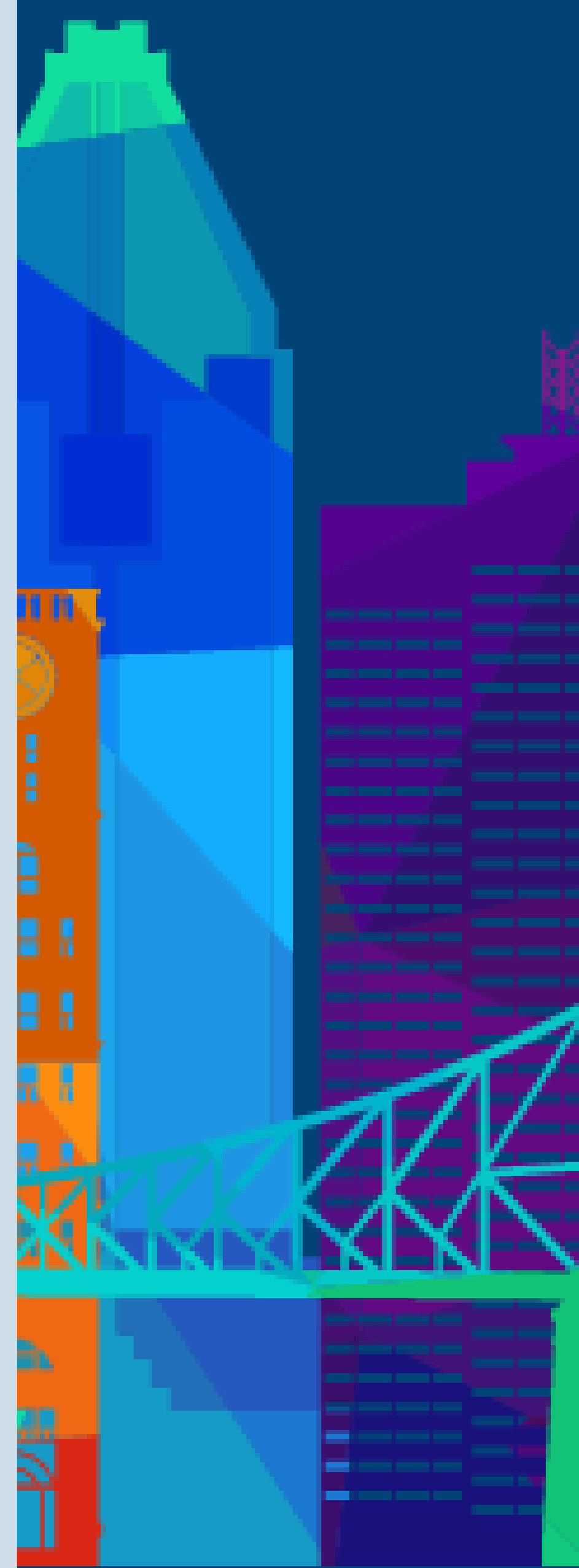
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CASE DESCRIPTION

- **29-y-o G1P0 @39 wks 4 d** in latent labor.
- **KT syndrome affecting primarily the left lower extremity** (varicose veins s/p failed sclerotherapy & separate surgical excisions). Now minimally symptomatic, no significant manifestations of the syndrome.
- MFM advised on **prophylactic LMWH -> heparin at 37 wks**. MRI brain thoracic spinal cord (to L2), **BUT NOT** lumbar spine obtained revealing no vascular malformations. of the visualized anatomy. The thoracic spine imaging only visualized as low as L1-L2. Anticoagulation held >12 hours prior to presentation.
- After discussion of risks, opted for neuraxial analgesia. **Epidural placed at L1-L2** without complication and maintained with Bupi 0.0625%/fent 2mcg/mL at 10 mL/hr with no top offs.
- NSVD 7 hours later. **Methylergonovine & misoprostol given due to brisk bleeding** noted despite good uterine tone. QBL 550mL. LMWH restarted following epidural catheter removal, and the patient was discharged 36 hours after delivery.





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DISCUSSION

- Despite no consensus for the anesthetic management of parturients with KT syndrome, case reports describe the management of these patients.^{3,4}
- Workup includes:
 - MRI abdomen/pelvis to exclude vascular malformations
 - MRI of the brain/spine to assess safety of neuraxial anesthesia
 - prophylactic anticoagulation.
- MFM, hematology, and obstetric anesthesia involvement guide workup and management.

