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Combined Spinal and Epidural Anesthesia in a multiparous parturient with Klippel-Trenaunay-Weber Syndrome

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Abstract Body: Klippel-Trenaunay-Weber Syndrome (KTWS) is a rare congenital disease involving capillary and venous malformations, limb hypertrophy, and arteriovenous malformations. Reports of pregnancy in KTWS are uncommon, and pregnancy can be associated with thrombotic or hemorrhagic complications because large cutaneous hemangiomas may sequester platelets, leading to a consumptive coagulopathy. Patients may have posterior cutaneous hemangiomas which may be a marker of underlying epidural or subdural vascular malformations that may complicate or contraindicate neuraxial anesthesia.

Case: We describe a case of a 32 y.o. morbidly obese, multiparous patient with KTWS, pregnant with twins, on prophylactic enoxaprin for treatment of a chronic right popliteal deep venous thrombosis (DVT). She has had successful neuraxial anesthesia for spontaneous vaginal and cesarean deliveries in the past. After developing an abdominal rash thought to be a reaction to enoxaprin, she was transitioned to a therapeutic dose of fondaparinux. At 26 weeks gestation she developed worsening symptoms of edema and cellulitis in the right leg and was found to have a new distal femoral DVT. Subsequently, after hematology consult the previous enoxaparin reaction was ruled out, and the fondaparinux was stopped. She was transitioned back to enoxaparin. MRI of the brain was obtained during the third trimester at an outside hospital and was nondiagnostic for venous malformations. MRI of the lumbar spine showed multiple superficial venous structures in the subcutaneous fat without epidural or subdural vascular malformations. The rest of her pregnancy was uncomplicated and she was admitted to labor and delivery at 35 weeks for frequent contractions. Enoxaparin was stopped at that time and she was started on IV heparin. Heparin was held 7 hours prior to a scheduled repeat lower segmental transverse caesarean delivery with combined spinal and epidural anesthesia. She had an uneventful delivery of two healthy neonates.

Conclusions: Parturients with KTWS pose a challenge to obstetrical anesthesiologists. In general, they are at high risk for complications from general anesthesia such as aspiration and difficult intubation, as well as blood loss and coagulopathy. In this case, our patient was morbidly obese and had a difficult airway by examination. Ideally, neuraxial imaging should precede neuraxial anesthesia to identify vascular malformations and minimize rare, but potential catastrophic complications of neuraxial anesthesia. Importantly, prior recommendations lean toward avoiding neuraxial anesthesia without confirmation of the absence of neuraxial anomalies. Our patient had 2 prior neuraxial anesthetics for deliveries in the past without complication, no spinal venous malformations described on MRI, and unclear absence of brain arteriovenous malformations. Therefore, after a careful risk-benefit analysis, a neuraxial anesthetic was deemed safest and subsequently performed.