# A Case Report of Klippel Trenaunay Syndrome in Pregnancy

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## Introduction

Klippel Trenaunay Syndrome (KTS) is a rare congenital disorder characterised by the triad of cutaneous hemangiomas, varicose veins, and bony or soft tissue hypertrophy affecting one or more limbs[1]. Common sequelae include cellulitis, thrombophlebitis, venous ulcers, pain and thromboembolism. KTS poses unique obstetric and anaesthetic challenges in pregnancy due to the risk of vascular malformations and haemorrhage and requires a personalised approach to management, particularly mode of delivery.

## Aim

To demonstrate the implications of KTS in pregnancy, and the importance of a multidisciplinary team approach.

## Case

A 32-year-old multigravida presented to a regional referral hospital for antenatal care, with a background of KTS diagnosed 12 years prior. She experienced recurrent right lower limb venous ulcers, varicose veins, superficial thrombophlebitis, and deep vein thrombosis, the most recent episode being in the first trimester of the current pregnancy.

She was managed by a multidisciplinary team involving obstetrics, anaesthetics, vascular surgery and community nursing. Antenatally she was managed with prophylactic enoxaparin, compression stockings and wound care of her chronic lower limb venous ulcers. Following counselling regarding the risks of vaginal and caesarean delivery in the context of KTS and previous caesarean delivery, she opted for elective repeat caesarean section.

She was referred for MRI abdomen and lumbar spine at 34 weeks gestation, which excluded vascular lesions at the lumbar spine or posterior soft tissues, or in the anterior abdominal wall at site of expected caesarean section. She was noted to have prominent vessels around the bladder, cervix and vulva.  Antenatal ultrasound at 34+1 demonstrated suspected macrosomia with estimated fetal weight of 97%.

She gave birth via emergency caesarean section due to prelabour rupture of membranes at 37+2 weeks. The procedure was uncomplicated with estimated blood loss of 300mL. The postpartum course was unremarkable, and she received 6 weeks of prophylactic enoxaparin postpartum.

## Discussion

Owing to its rarity, there is a lack of evidence-based guidelines regarding the management of KTS in pregnancy. Physiological changes in pregnancy including increased blood volume and vena caval compression typically worsen symptoms of venous stasis, vulval and limb varicosities, limb pain, and predispose to venous thromboembolism.

A key consideration is mode of delivery; vaginal delivery poses the risk of massive intrapartum haemorrhage due to rupture of vulval, vaginal or cervical varicosities, yet caesarean section poses anaesthetic risks associated with spinal or epidural arteriovascular malformation or haemangioma, or airway vascular malformation and soft tissue hypertrophy.

Imaging studies including ultrasound and MRI are a key element of antenatal assessment by mapping any vascular malformations and guiding mode of delivery and anaesthesia. A multidisciplinary approach allows for thorough investigation of venous malformation and birth planning to achieve optimal outcomes.

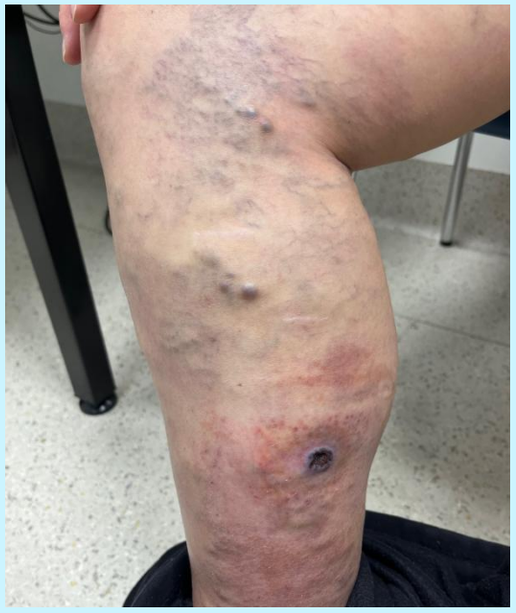

Figure 1: The patient's extensive varicose veins and chronic venous ulcer

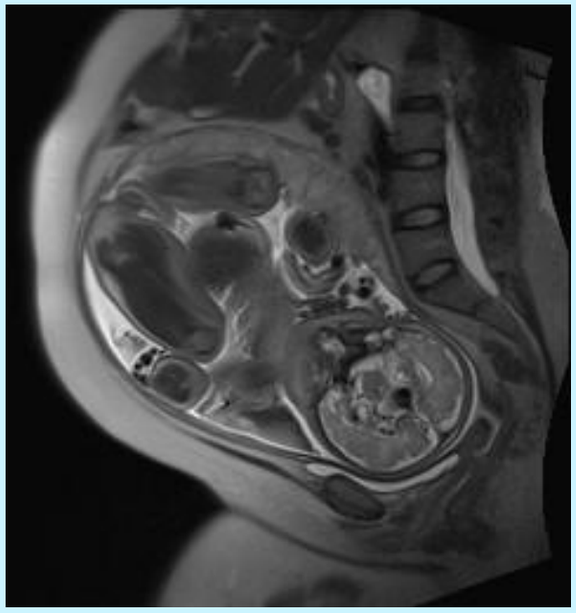

Figure 2: MRI abdomen in pregnancy[4]

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