

Neonatal Transfemoral Amputation Following Congenital Limb Deficiency Secondary to Twin-to-Twin Transfusion Syndrome

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INTRODUCTION:

Congenital limb deficiency are birth defects with aplasia or hypoplasia of bones over limbs, occurred in 7/10000 newborns. Causes varied from chromosomal abnormalities, teratogens, genetic disorders and intra uterine abnormalities for instances Twin-to-twin transfusion syndrome (TTTS), oligohydramnios, amniotic band syndrome may cause isolated limb deficiencies. Limited reported cases in the region.

REPORT:

A second twin baby boy of monochorionic diamniotic twin pregnancy, born via emergency caesarean section for TTTS (Quintero stage 3) with abnormal doppler at 27 weeks of gestation, weighted 1kg as a recipient twin. Antenatally mother had hyperthyroidism in pregnancy on treatment. Delivery was uncomplicated, however, noted gross deformity over right lower limb. Tibia and fibula were protruded with under-developed soft tissue. Multiple constriction bands at level of knee and leg with dangling part which was pale and discolored distal to the bands. Vascular examination revealed PTA and DPA pulse were not palpable. Radiographs showed under-developed femur, tibia and fibula, dislocation of ankle joint with dysplastic tarsal bone.

Detailed scan on 23 weeks of gestation noted abnormal doppler with discrepancy up to 40%. No abnormalities detected on the lower extremities of either twin. The baby underwent a transfemoral amputation of right lower limb at proximal thigh after definitive demarcation. The operation was uneventful with controlled hemorrhage. Excellent anesthetic and pediatric support in preparing the patient perioperatively are vital.



Figure 1: Photograph of right lower limb deformity



Figure 2: Transfemorally amputated right lower limb

Varies causes of intra-uterine gangrene of the limb. Prenatal ultrasound able to detect TTTS using flow velocity doppler signals. Incidence of in-utero acquired limb ischemia higher in monochorionic twins. Prenatal ultrasound may not detect limb defects especially in twin pregnancy.

CONCLUSION:

Congenital limb deficiency is uncommon. The ultimate treatment maybe higher-level amputation of affected extremity, until definitive demarcation awaited.

REFERENCES:

1. Vakhshori V, et al. Congenital limb deficiency requiring transfemoral amputation. *BMJ Case Rep.* 2018 May 30;2018:bcr2017223980. doi: 10.1136/bcr-2017-223980. PMID: 29848527; PMCID: PMC5990066.