Amniotic bands, anorectal and bladder agenesis - A unique association in a twin pregnancy with fetus papyraceus -

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Introduction
• Poor cloacal development and inadequate septation - wide range of genitourinary and digestive tract malformations.
• Bladder agenesis - extremely rare congenital anomaly.
• Successful treatment and the long-term prognosis are usually poor because of associated abnormalities

Patient – 20y G2 P2
Previous pregnancy: C-section at term (normal child)
Current pregnancy:
• MC twin pregnancy
• Fetal demise of one twin at 12w, the other fetus continues developing up to 23 weeks.
• NO teratogenic risk factors
• NO screening tests for chromosomal abnormalities in the first trimester

Ultrasound
• Omphalocele
• No bladder in pelvis;
• Hypoechoic structure fixed to the anterior abdominal wall (interpreted as bladder extrophy - UA Doppler imaging (fig 1) and turbulent ureteral jet image (fig 2).
• Amniotic bands.

Case resolution
Spontaneous rupture of membranes at 23 weeks leads to termination of pregnancy.
• Delivery by C-section

Necropsy
• Omphalocele (fig. 3)
• Bladder, vagina, and urethra agenesis (fig. 5)
• Ectopic ureters
• Supernumerary left adrenal gland.
• Agenesis of the sigmoid and rectum
• Imperforate anus
• Fetus papiraceus (fig.4)

Differential DX
• Gastrochisis
• ABS – amniotic band Syndrome
• OEIS complex
• LBWC – Limb body wall complex

Discussions and Take Home Messages
✓ In surviving neonates, surgery is difficult, conducted in multiple sessions by a complex team and is encumbered by early and late complications.
✓ Less severe forms = good outcome with corrective surgery
✓ Early prenatal diagnosis is required, serial scans are necessary and it is also helpful to plan the appropriate perinatal management