Introduction: Uterus didelphys with obstructing hemivaginal septum and ipsilateral renal agenesis, also known as Herlyn-Werner-Wunderlich syndrome is a very rare syndrome. Mullerian duct anomalies (MDA) are congenital anatomic abnormalities of the female genital tract arising from non-development or non-fusion of the mullerian ducts or failed resorption of the uterine septum. The overall prevalence of MDAs is 0.4% and among them obstructing hemivaginal septum and ipsilateral renal agenesis (OHVIRA) constitutes 0.16–10%.

Case report: A 32-year old, primigravida as referred to our tertiary centre with echogenic cavity next to the bladder and a small- or pelvic kidney on the left side. First trimester screening NIPT had been performed with a normal result. Detailed scan at 19+5 weeks showed a normal right kidney, however on the left side there was a cystic structure with small light echogenic tissue around it. The couple underwent amniocentesis. QF-PCR for common trisomies and microarray CNV analysis was normal. At 39+5 weeks a daughter was born. She weighed 3808g with no external dysmorphic features. After an ultrasound of the abdomen the girl is diagnosed with Herlyn-Werner-Wunderlich syndrome.

Conclusion: Müllerian duct anomalies are infrequently encountered clinical problems and often present with difficulty in diagnosis. A high level of suspicion is the key to diagnosis, which is usually made soon after menarche. In our case the diagnosis was set short after birth because of a kidney abnormality by the fetus at the 20 week scan.