We report a case of 10p11-p12 microdeletion diagnosed prenatally. 31 year old woman with no medical history underwent ICSI for severe oligospermia with unremarkable ongoing pregnancy.

The ultrasound at 22 w showed a female foetus with horseshoe kidney and heart congenital anomaly type double outlet right ventricle (DORV). There is no evidence of dysmorphic face and other fetal abnormalities.

Chromosomal microarray showed a pathogenic deletion within cytobands 10p11.22-p12.1, which overlapped for four protein-coding genes: ARMC4, MPP7, WAC and BAMBI.

The 10p11-p12 deletion syndrome is rare with an incidence of less 1 in a million.

To date, only 10 postnatal and one prenatal cases with microdeletion of this region have been described, and all patients shared a common phenotype, including intellectual disability, abnormal behavior, distinct dysmorphic features, visual impairment, cryptorchidism and cardiac malformations.

Microarray analysis is a powerful diagnostic tool and can add new phenotypic undescribed before feature to this syndrome like, horseshoe kidney.