



HNF1B: Renal Cysts and Diabetes

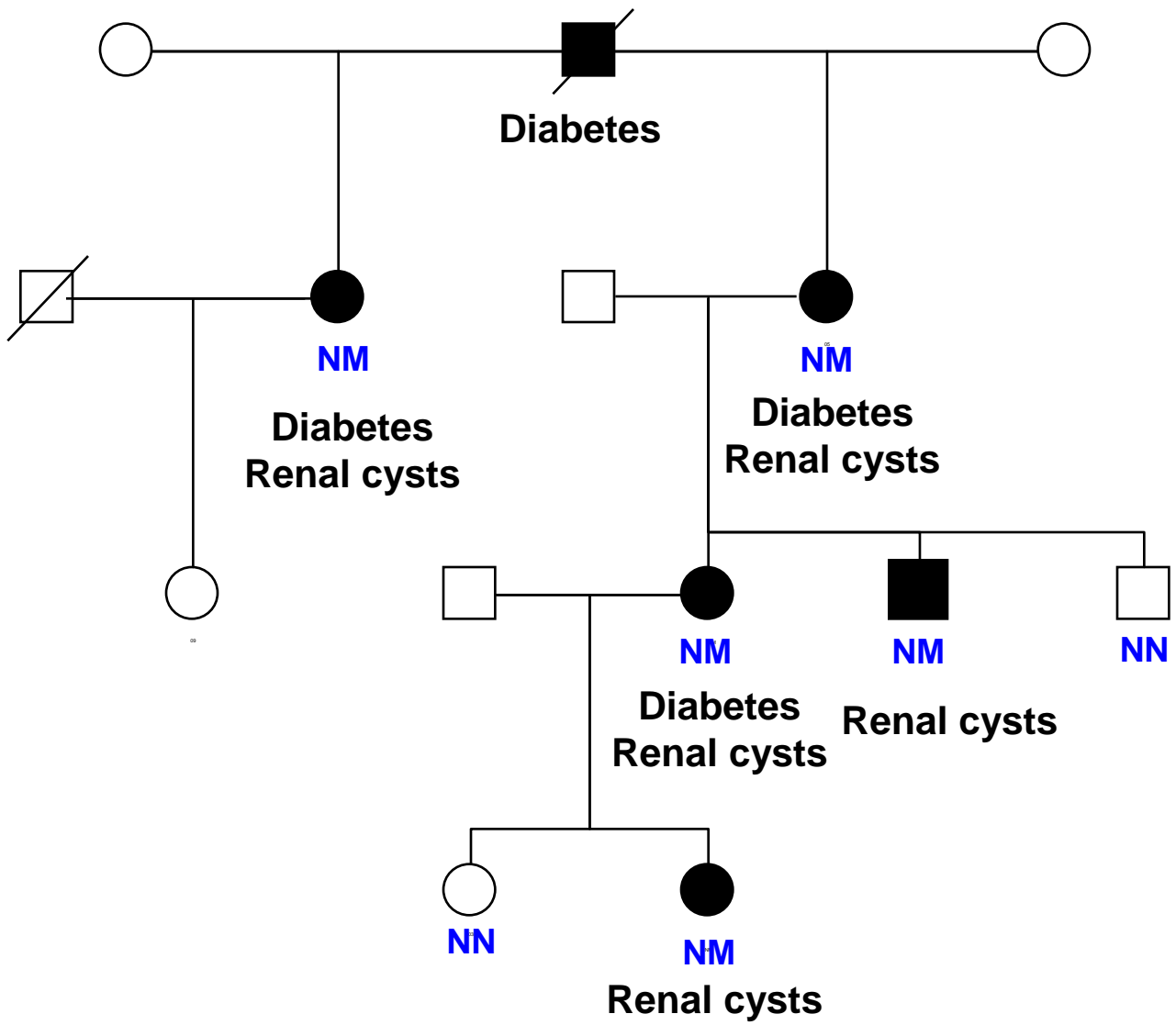
Dr Coralie Bingham

University of Exeter Medical School



Renal Cysts And Diabetes (RCAD)

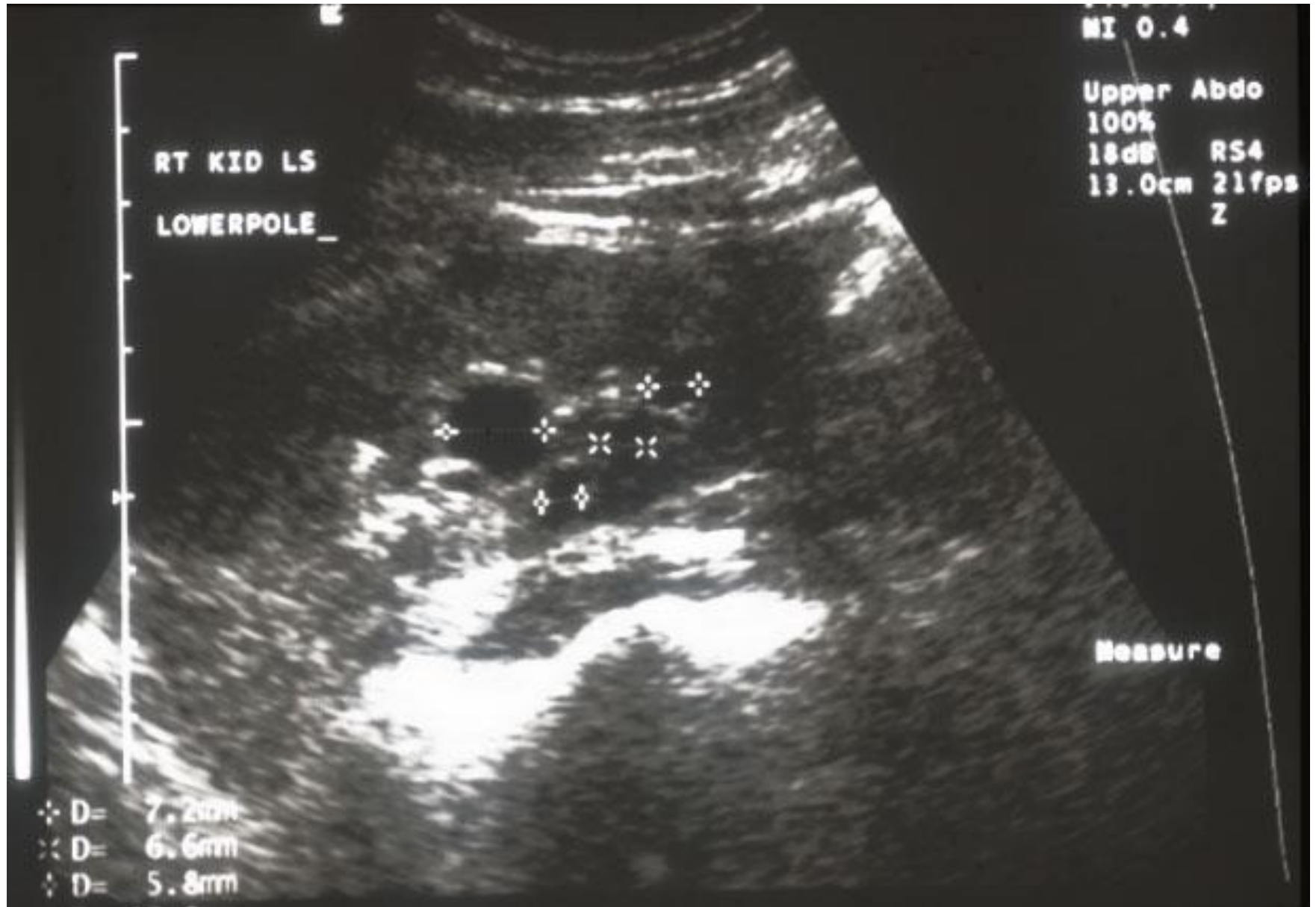
- **Renal cysts** (seen in utero)
- **Renal function variable - mild impairment**
– dialysis dependent
- **Variable renal histology**
- **Diabetes** typically develops after renal disease



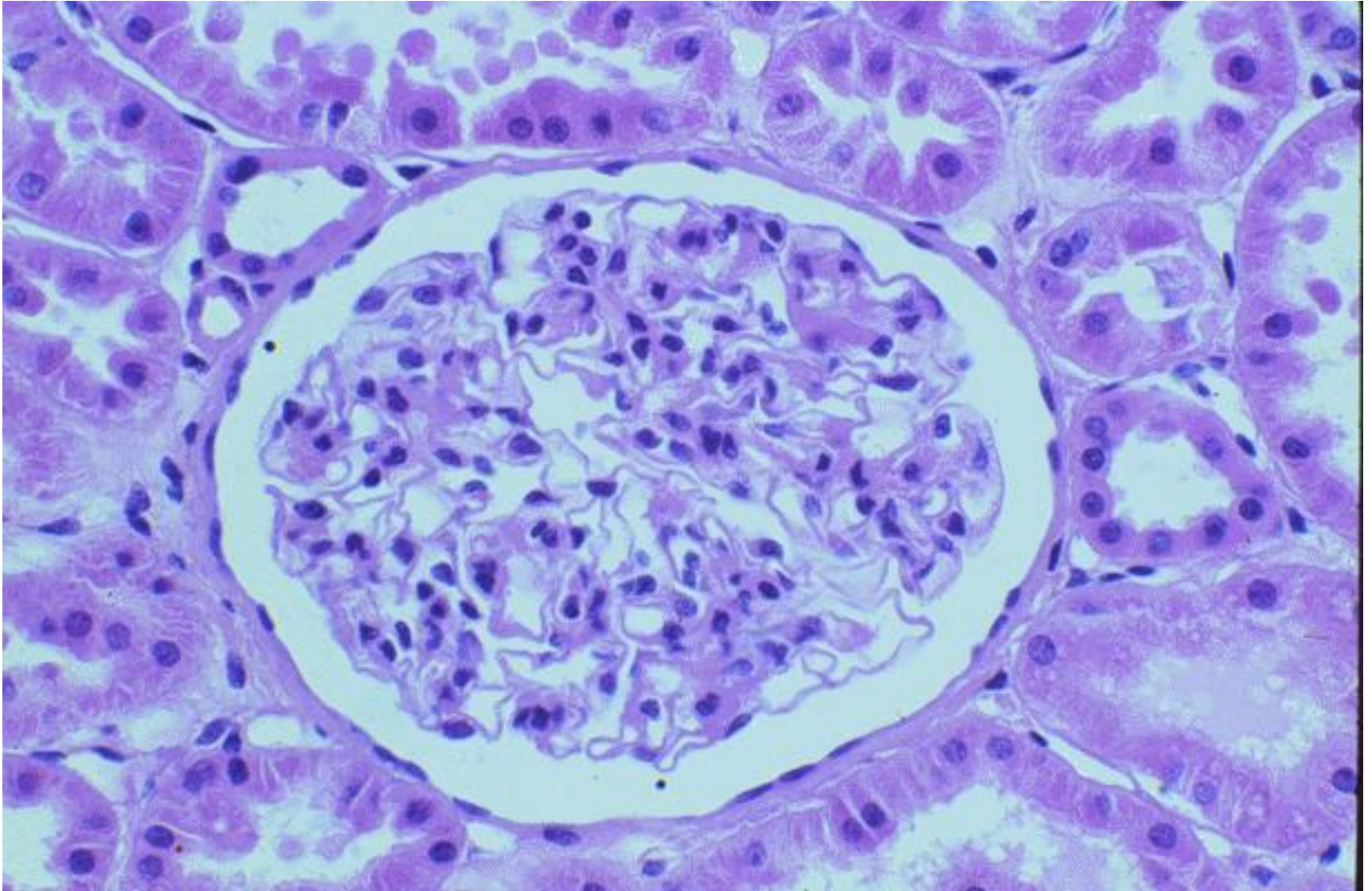
Normal kidney



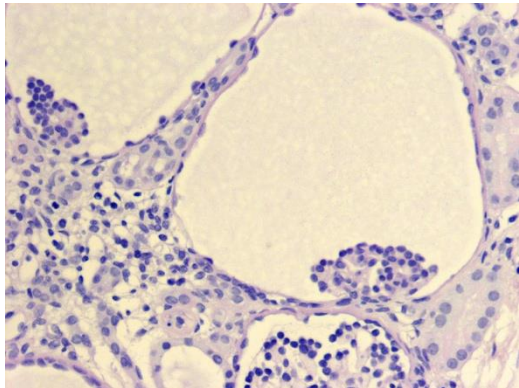
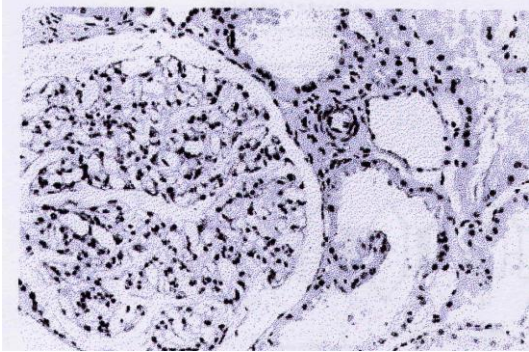
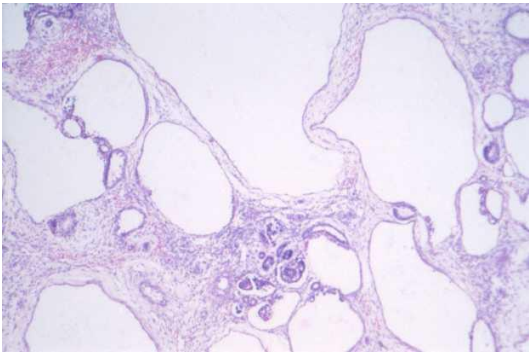
HNF1B kidney



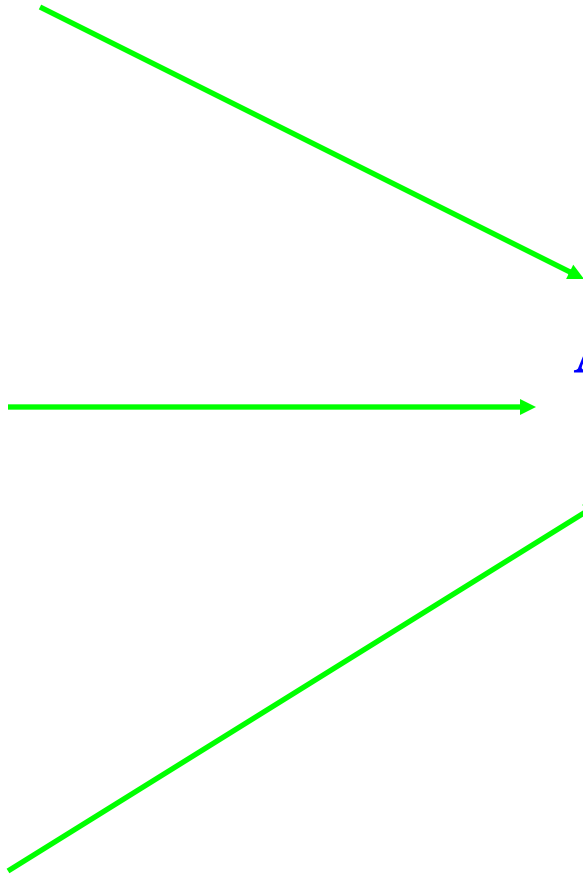
Normal kidney

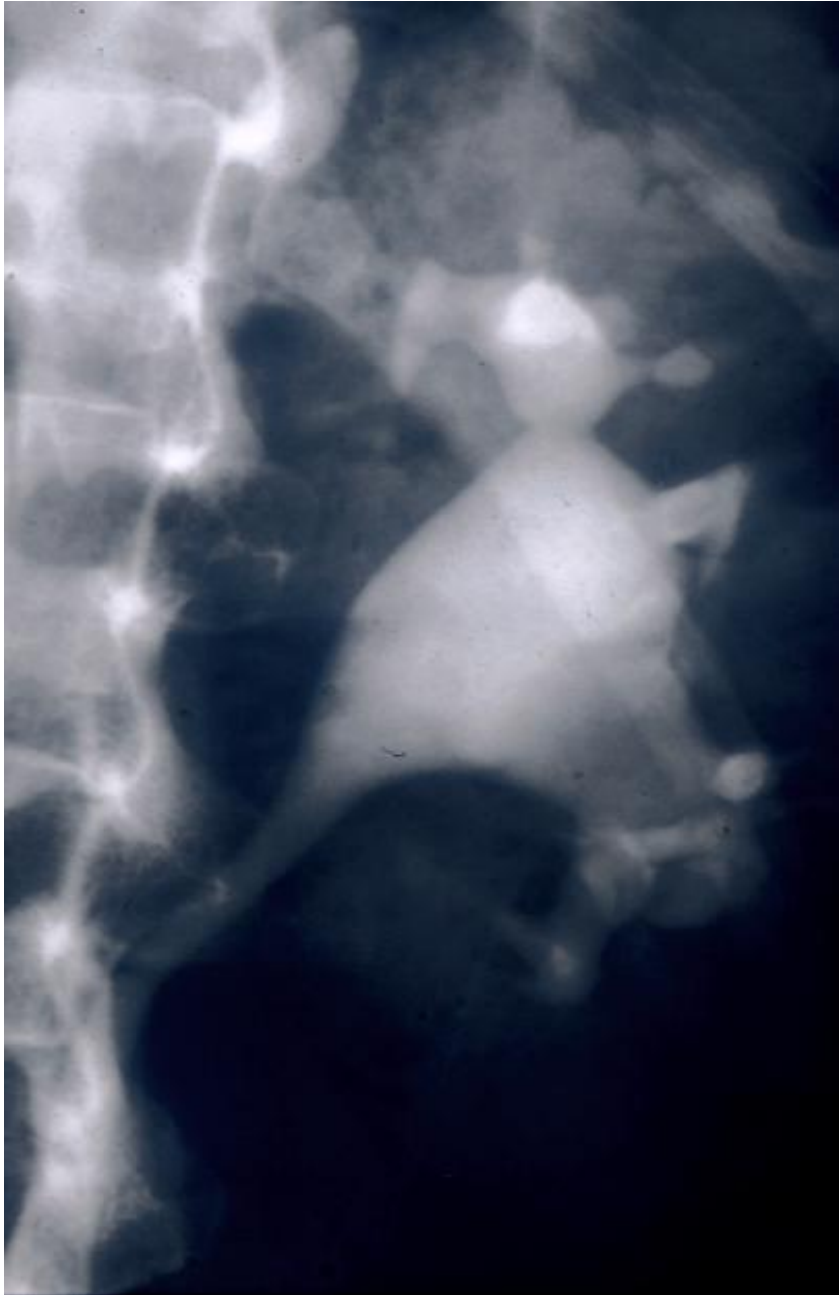


HNF1B kidney



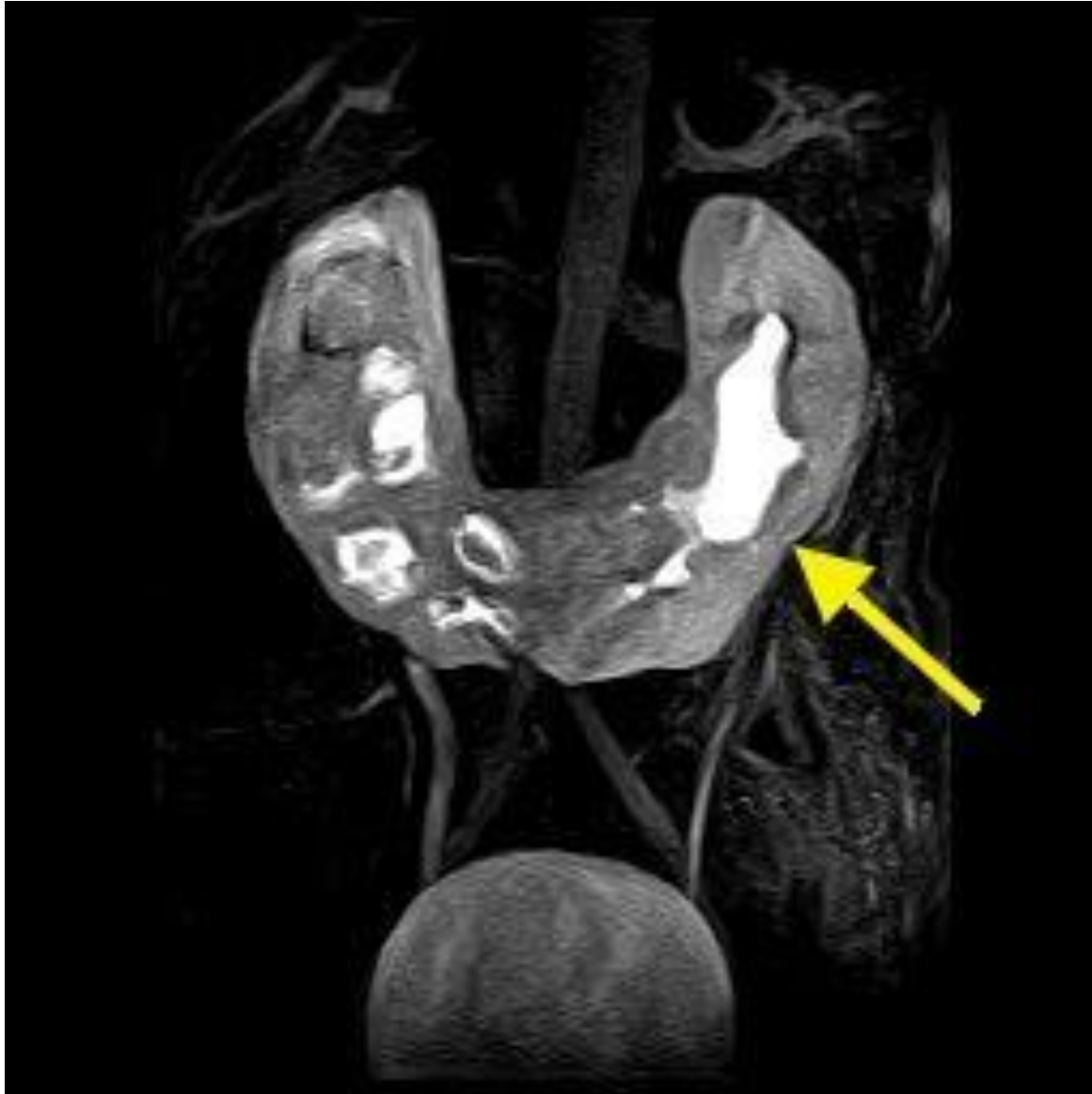
**Abnormal kidney
development**





- **Single left kidney of normal size**
- **Calyces abnormal and blunted**

Horseshoe kidney



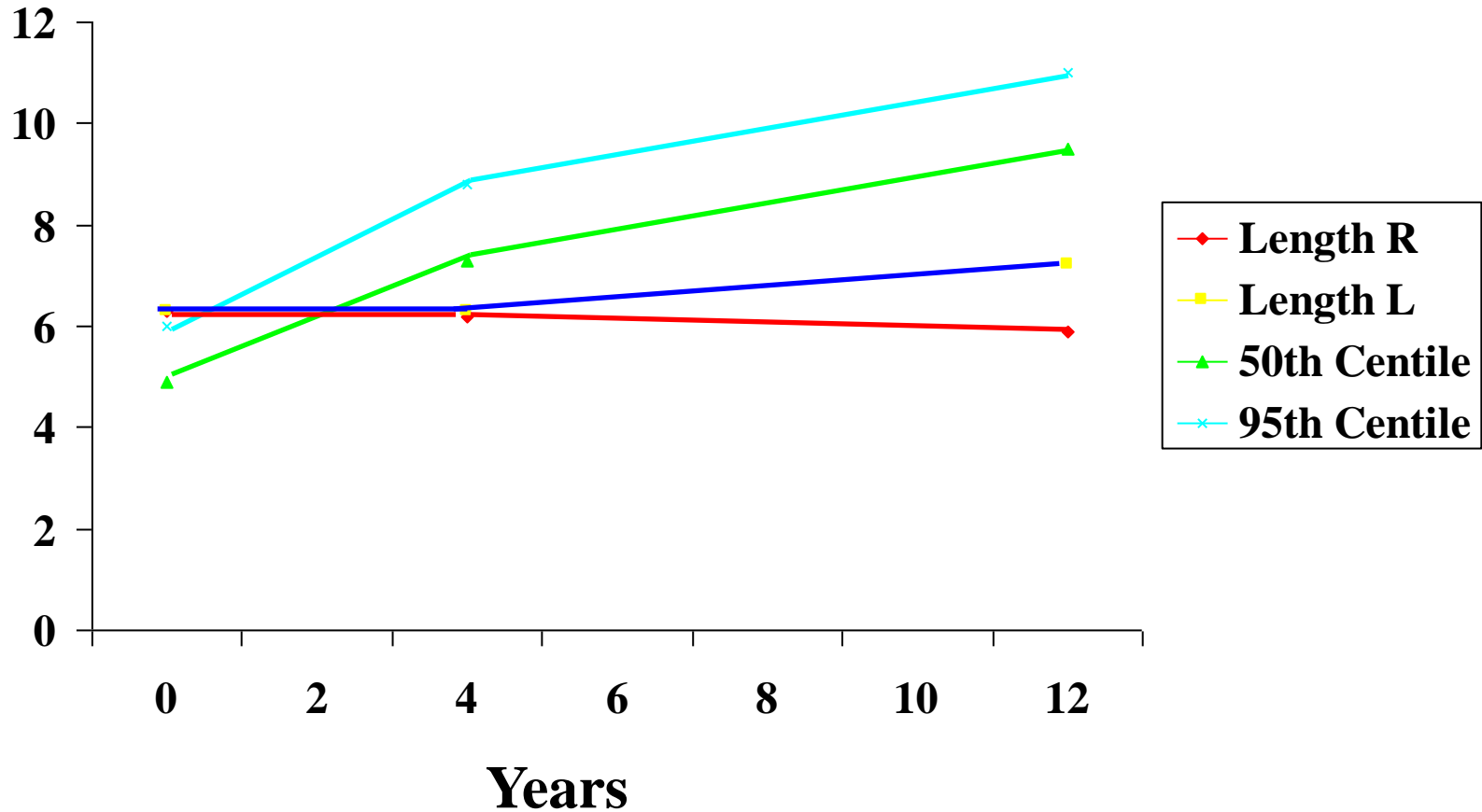
Prune-belly syndrome



**Bilateral
hydronephrosis**

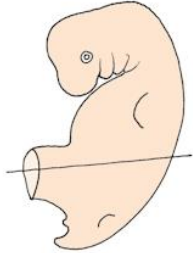
HNF-1B mutations affect renal growth

Length (cm)

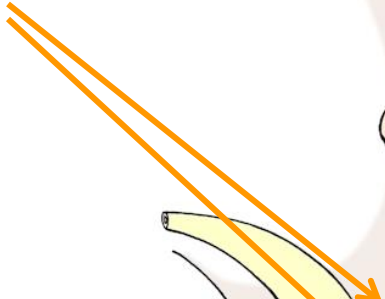


It's not just renal cysts and diabetes

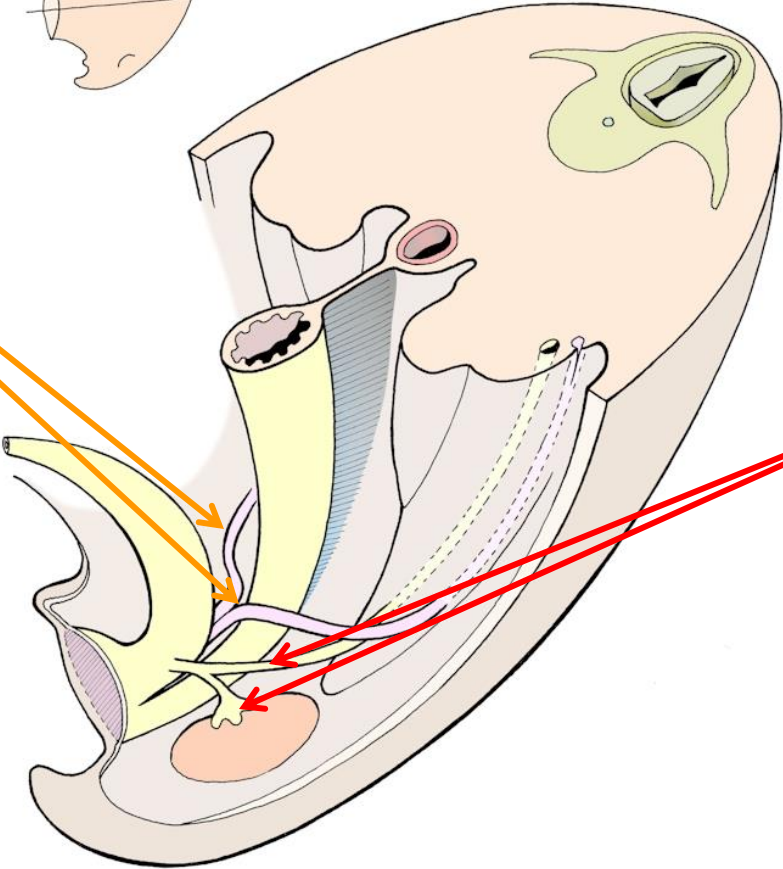
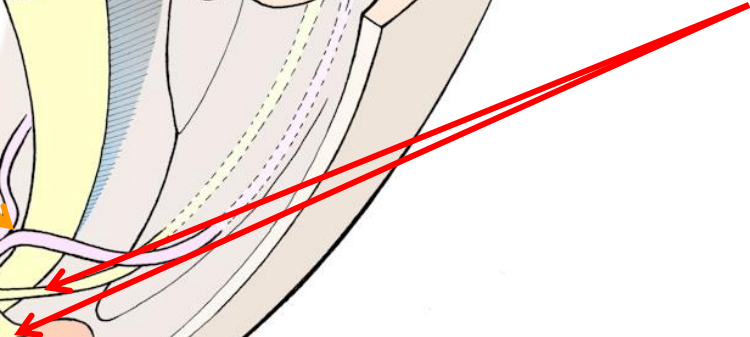
Embryology



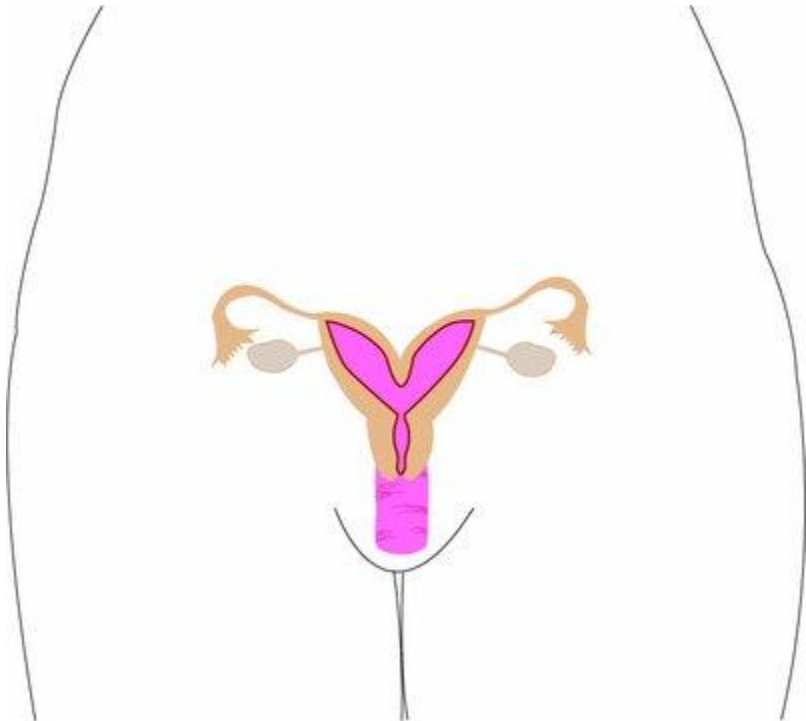
Mullerian duct
(paramesonephros)



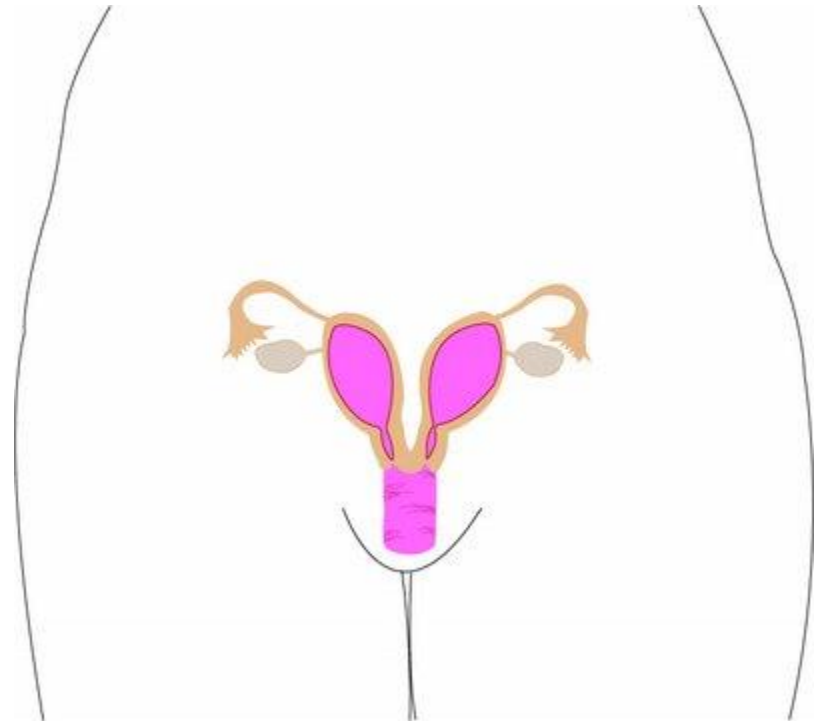
Wolffian (mesonephric)
duct and ureteric bud



Uterine malformations



Bicornuate uterus



Double Uterus

HNF1B-associated disease is a multi-system disorder

