



uterus and vagina normally developed



vagina subseptum



arcuate uterus



uterus subseptum



septated uterus

# Vývojové poruchy rodidel



uterus bicornis



uterus bicornis



Lenka Krobová

uterus didelphys



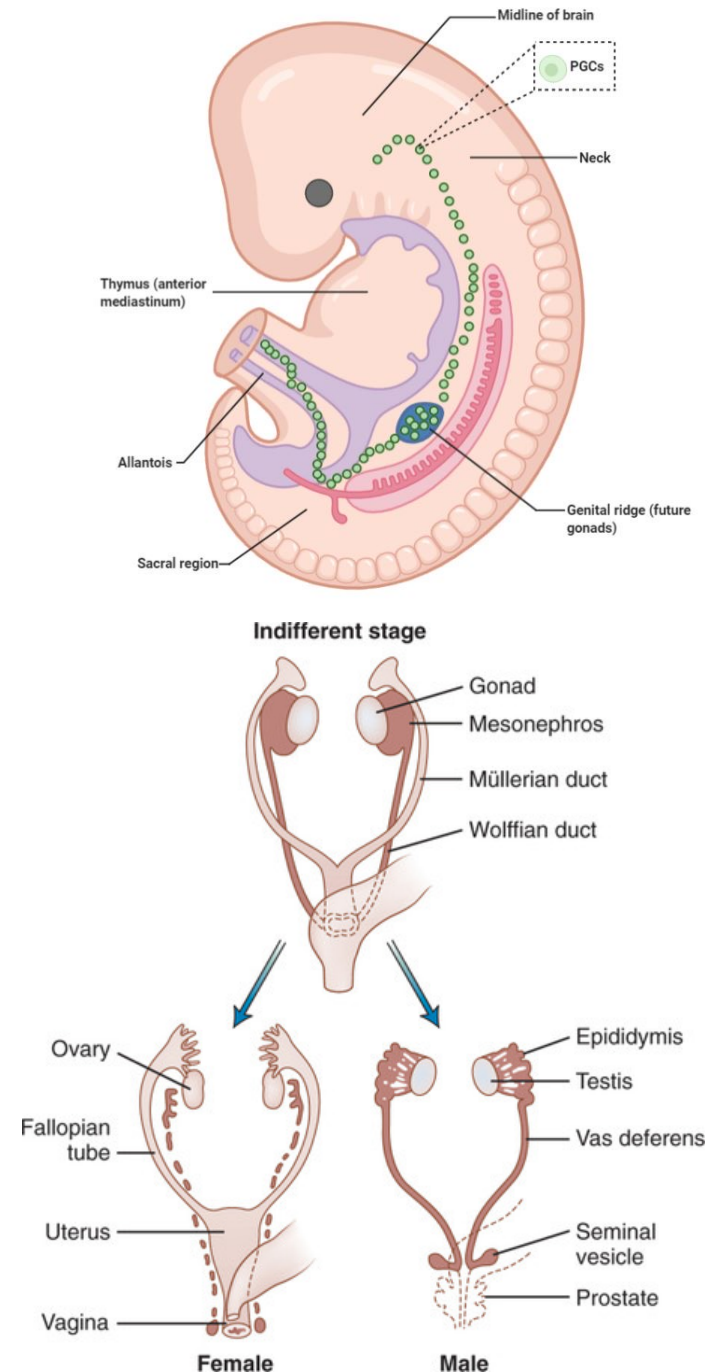
asymmetrical

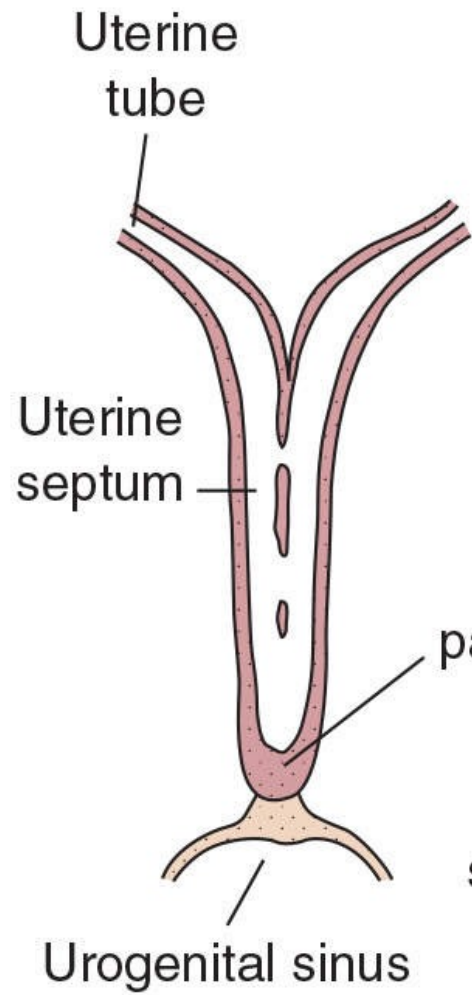


unicornuate

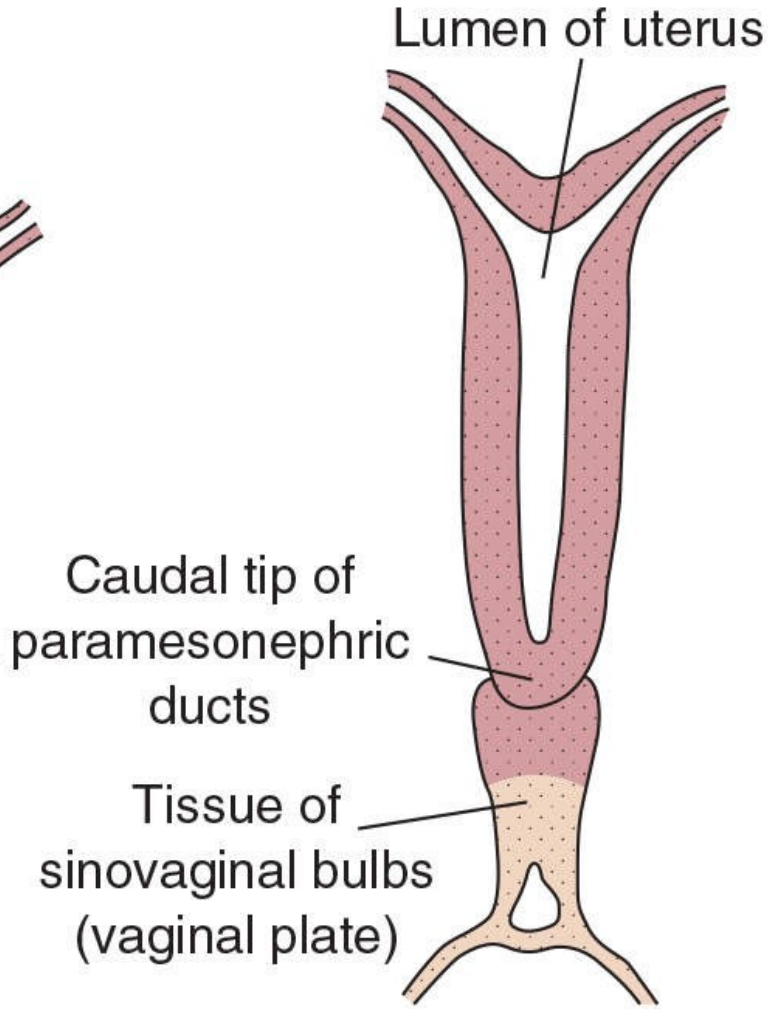
# Vývoj pohlavního systému

- SRY (*sex-determining region on Y*)
- Wollfův a Müllerův vývod
- Uterovaginální kanál
- Spojení se sinus urogenitalis

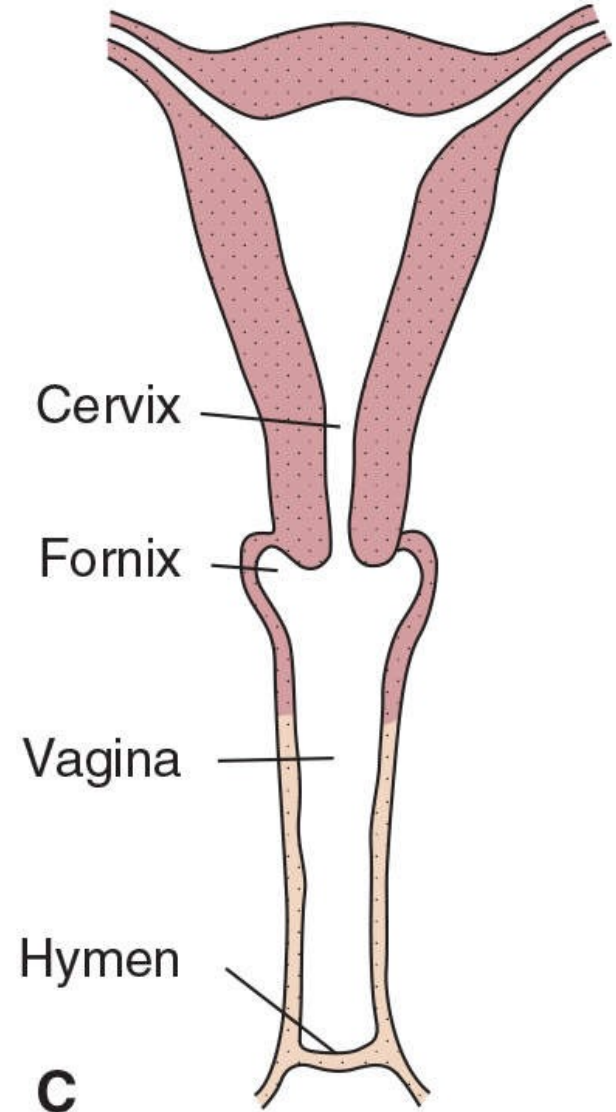




**A**



**B**



**C**

Ageneze a dysgeneze

Intersexuální malformace

Poruchy vývodů

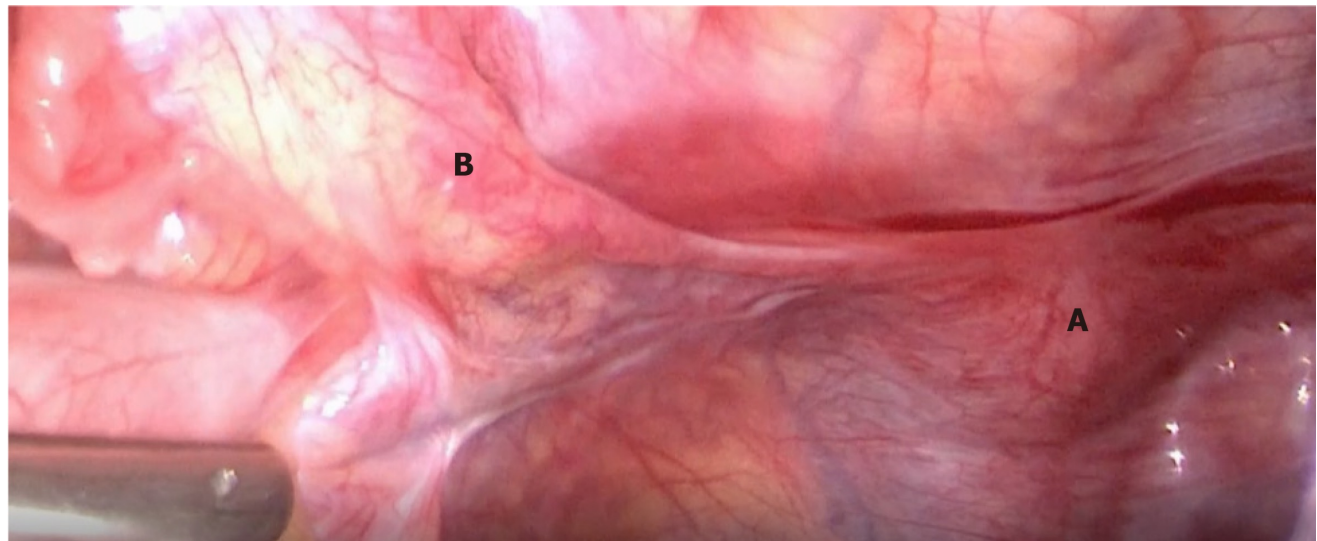
Sdružené vady

# Ageneze a dysgeneze gonád

**Ageneze** – gonády nejsou vytvořeny

**Dysgeneze** – vazivové proužky

- nejsou folikuly → není hormonální produkce



**46 XX**

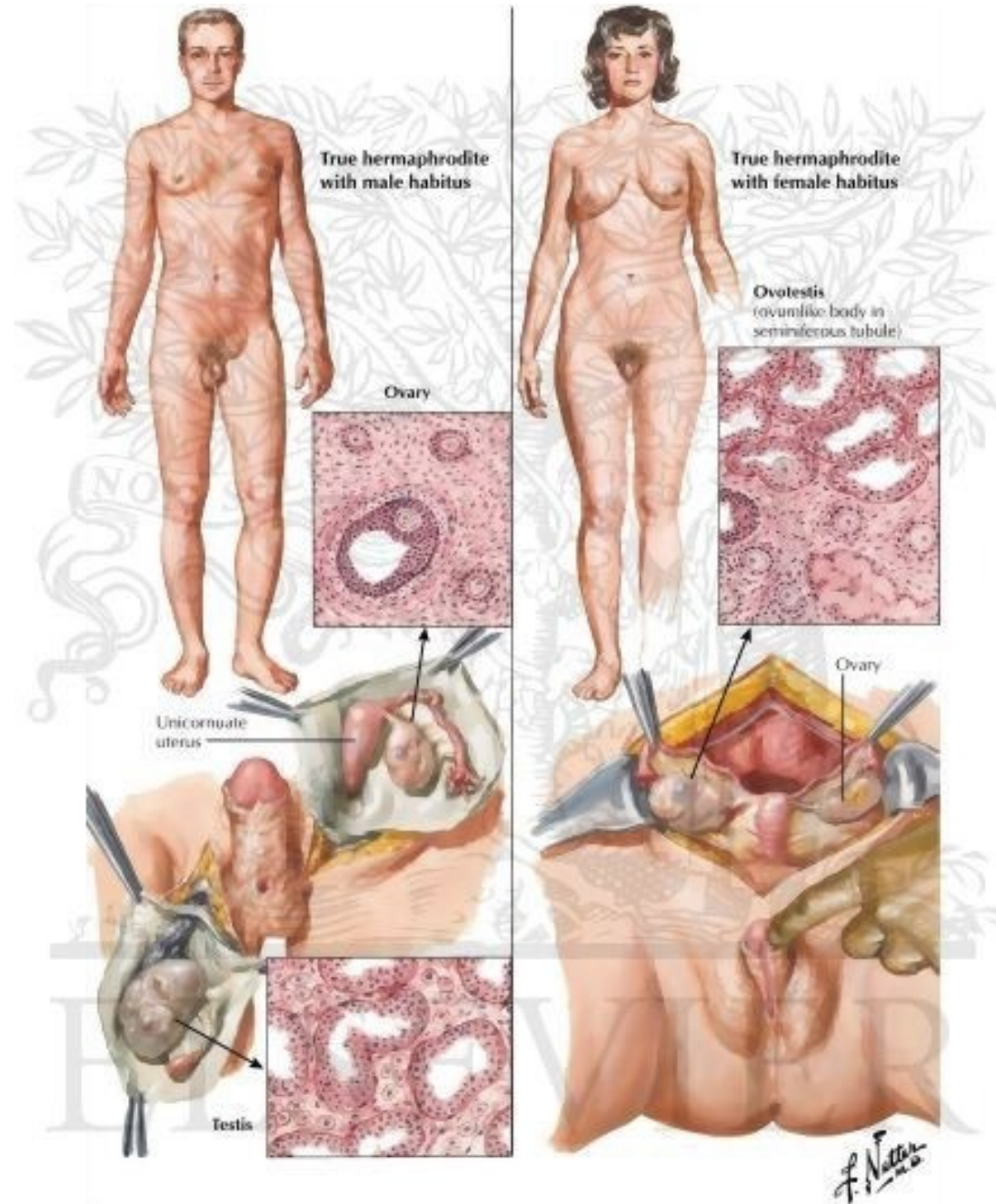
**45 X – Turnerův syndrom**



# Intersexuální malformace

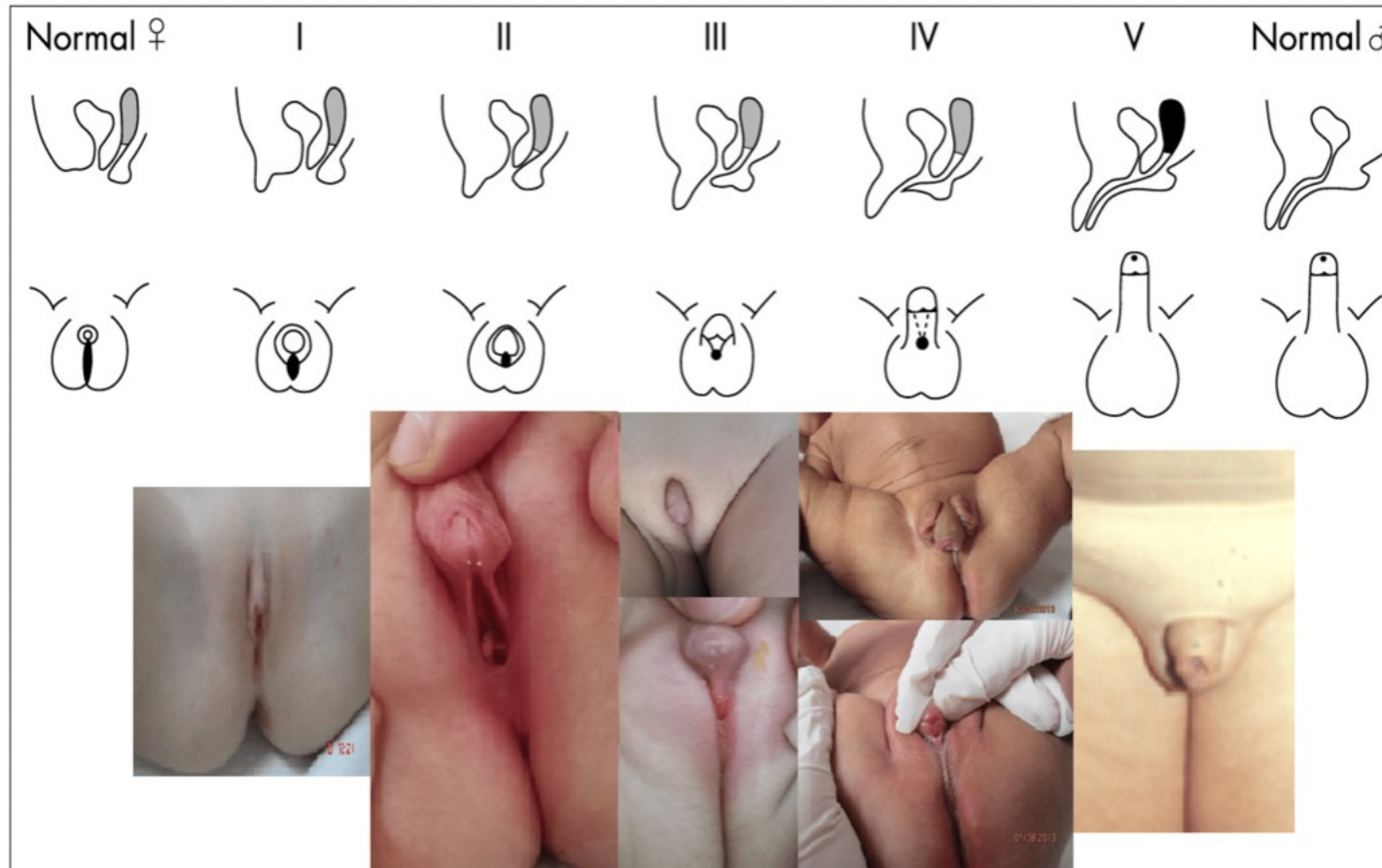
## *Hermaphroditismus verus*

- Ovaria i testes, ev. ovotestis
- Velmi vzácný



## *Pseudohermaphroditismus femininus u CAH*

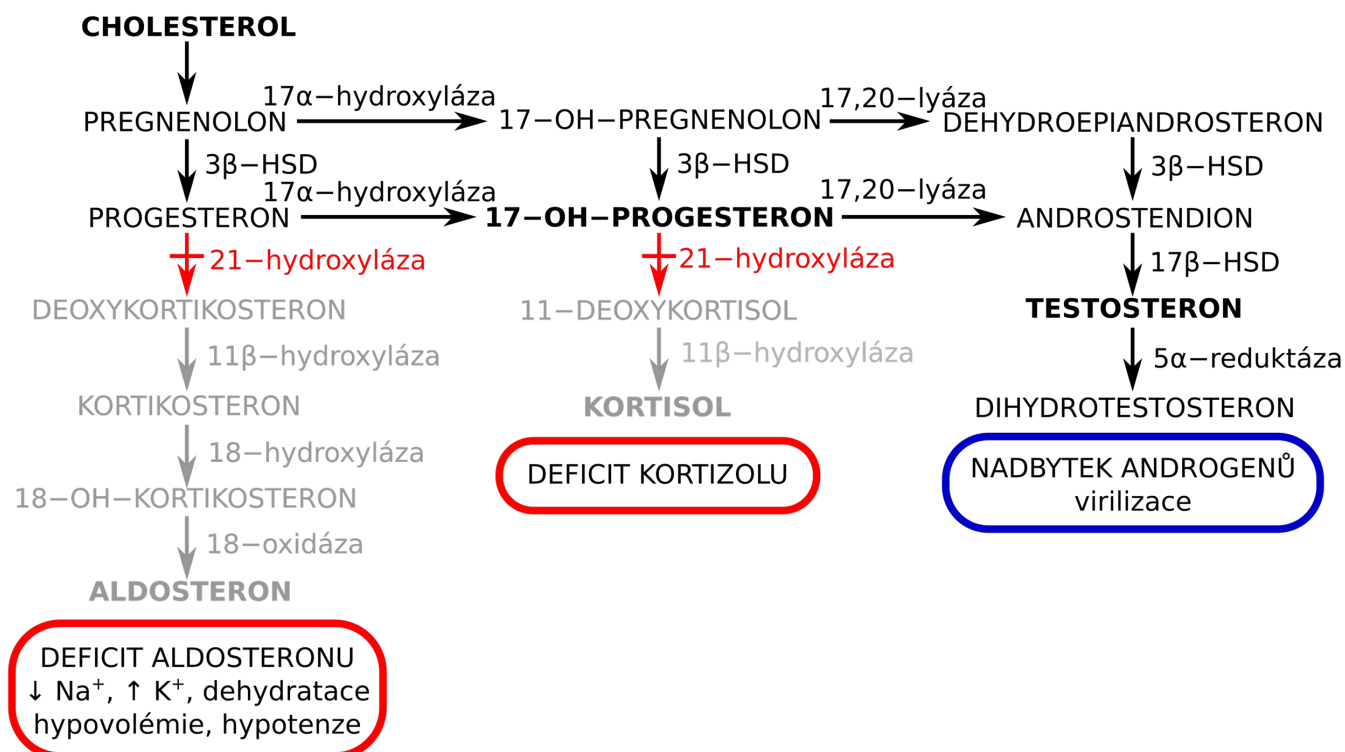
- 46 XX
- Distální úsek pochvy zužený, vnitřní pohl. orgány normální
- Nejčastěji – peniformní klitoris, skrotiformní labia





# Kongenitální adrenální hyperplázie

- AR
- Defekt enzymů → porucha syntézy hormonů (nejč. 21-hydroxyláza)



## **Klasická forma**

### *Se solnou poruchou*

- Dívky – virilizace, solná krize, smrt

### *Prostá virilizující*

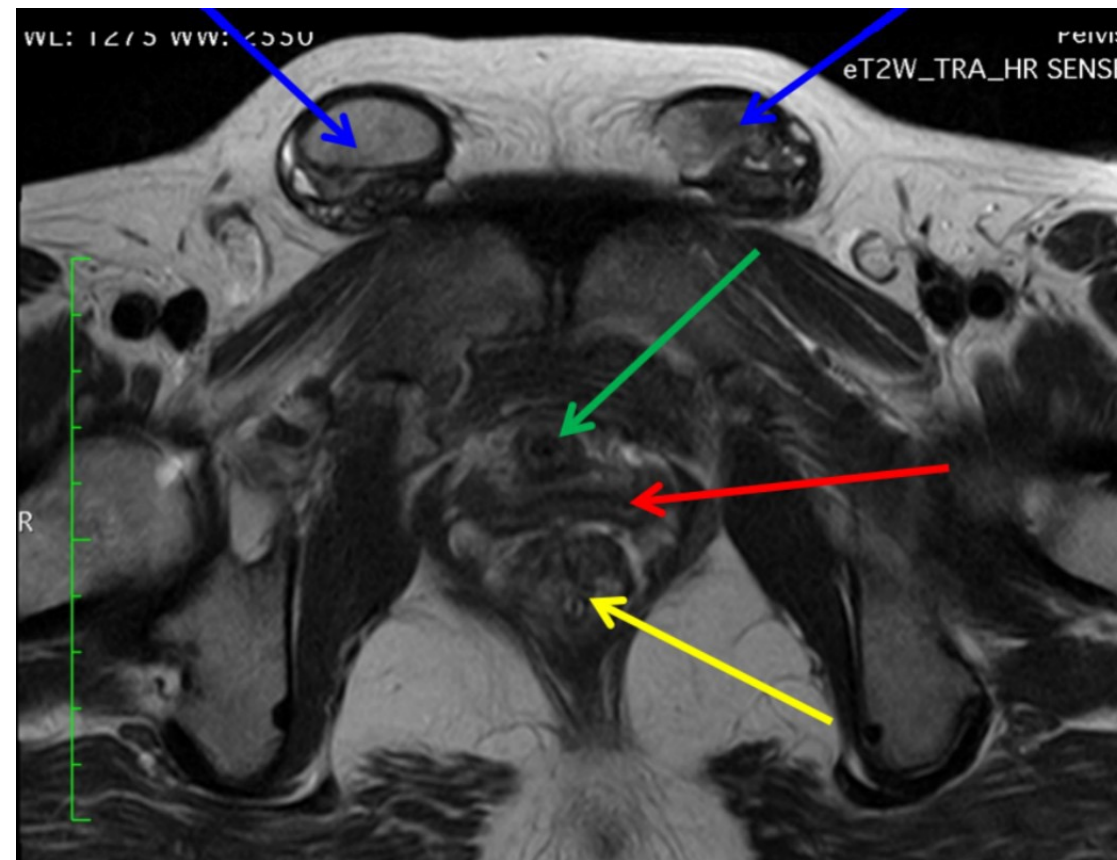
- 1% aktivity
- Dívky – virilizace, pseudopubertas praecox heterosexualis, nízká finální výška

## **Late-onset forma**

- 20-50% aktivity
- Virilizace dívek, poruchy cyklu

## *Pseudohermaphroditismus masculinus – syndrom testikulární feminizace*

- 46, XY
- Ženská zevní rodidla
- Produkce MIF → nejsou vnitřní rodidla



# Poruchy Müllerových vývodů

## Poruchy průchodnosti - GYNATRÉZIE

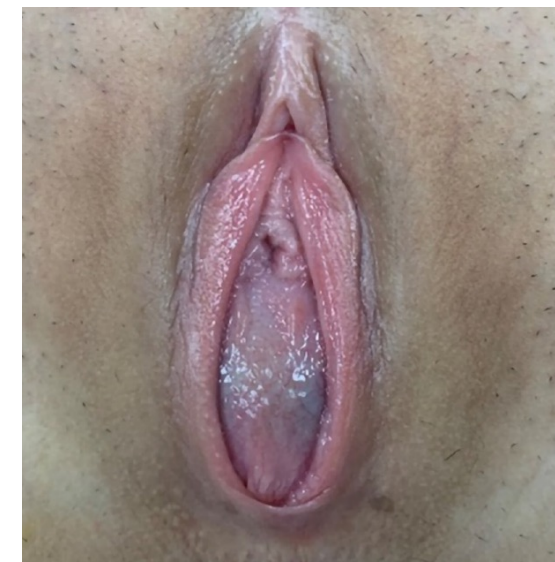
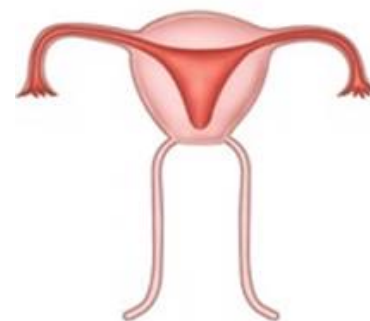
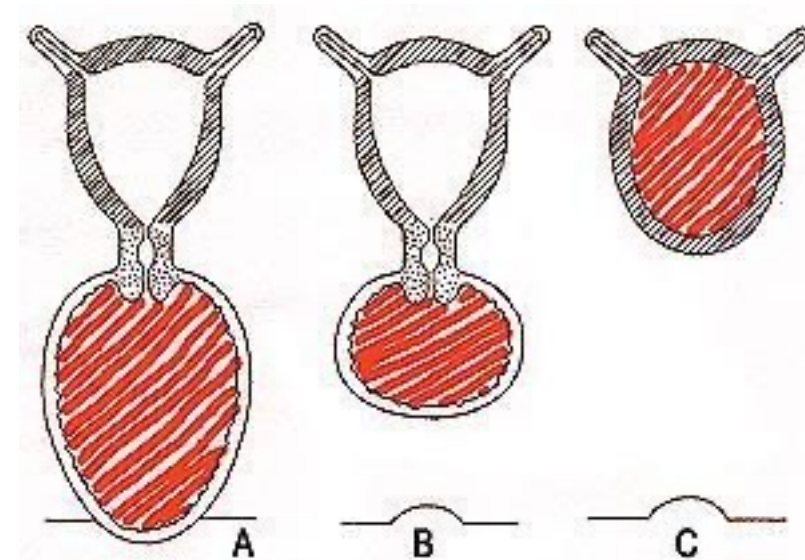
### Atresia hymenalis

- Mucocolpos
- Hematocolpos až hemoperitoneum

### Aplasia partis distalis vaginae

### Atresia cervicis uteri

### Aplasia vaginae

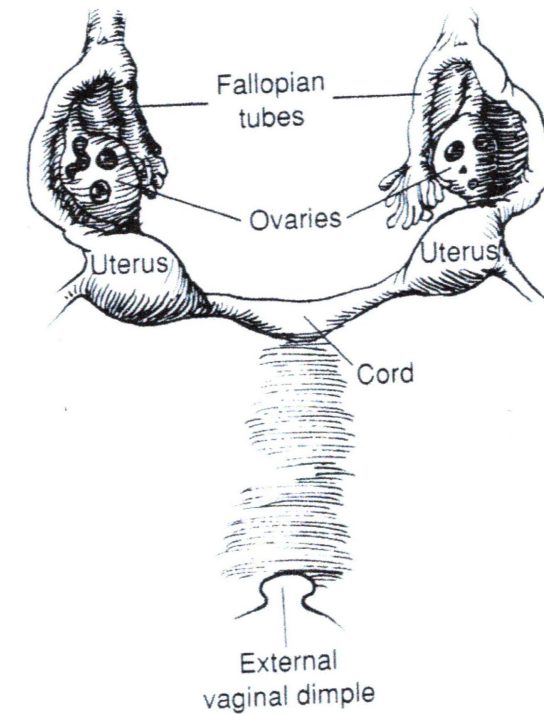
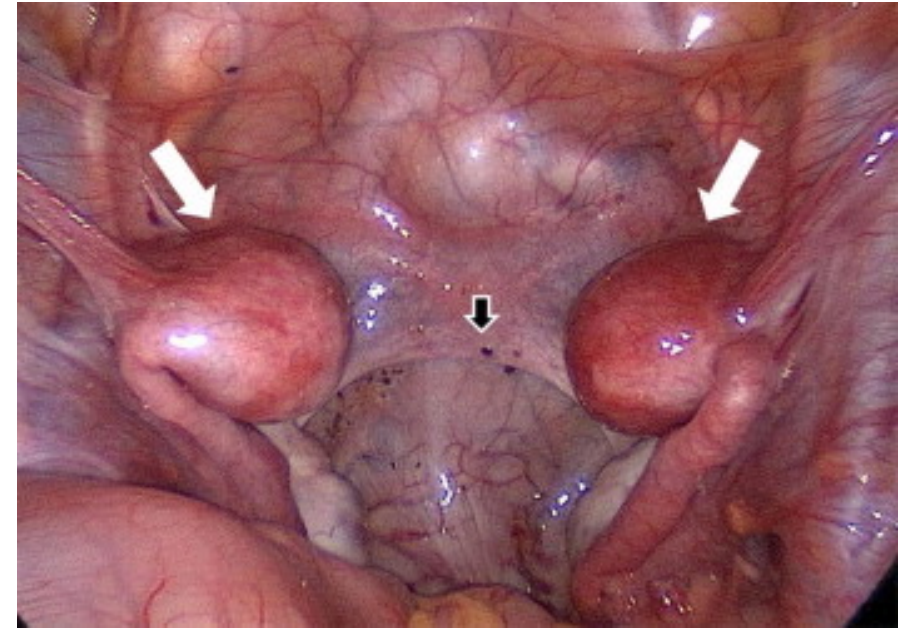


*Atrézie = neprůchodnost*  
*Aplázie = nevyvinutí*

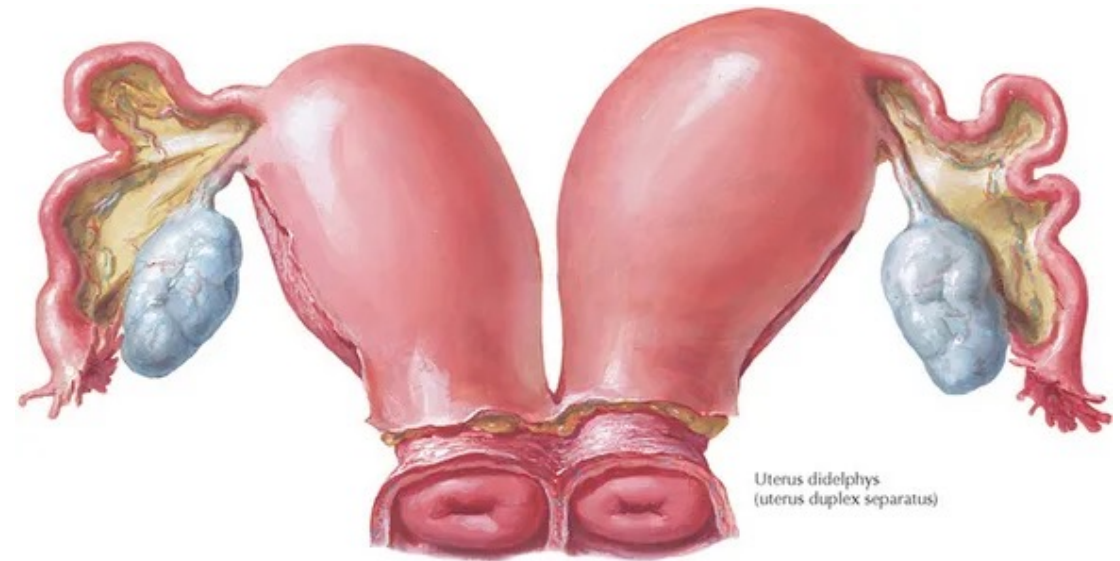
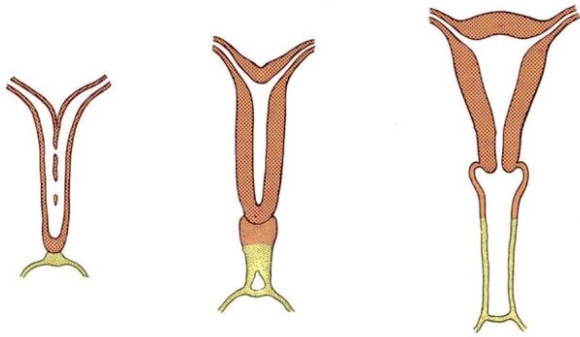
## Syndrom Rokitanského-Küsterův

- Úplná aplázie dělohy a pochvy

operace



# Poruchy splývání

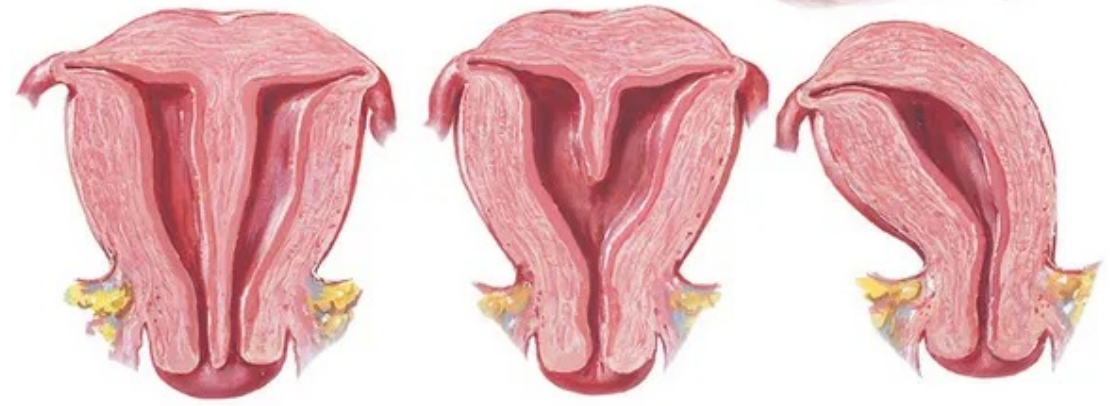
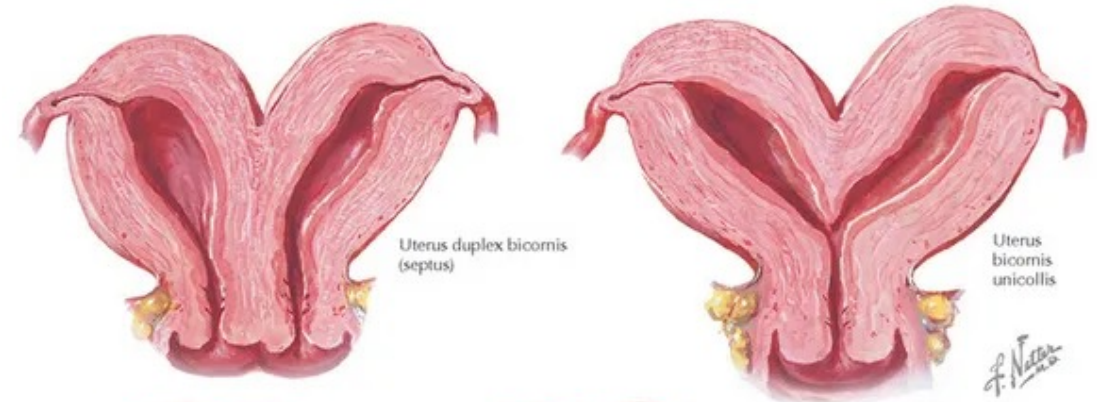


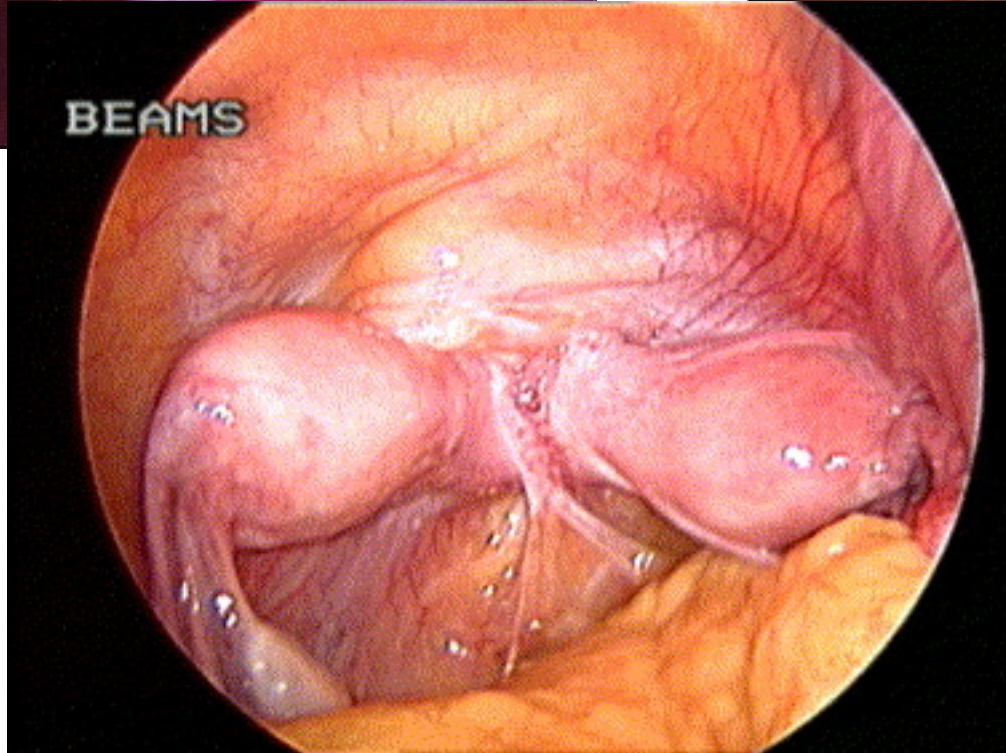
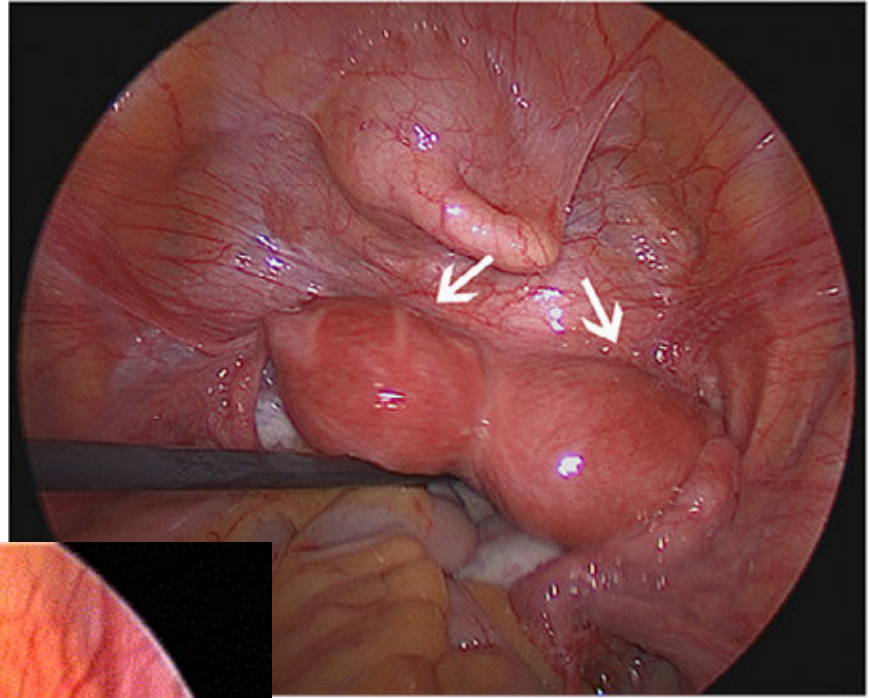
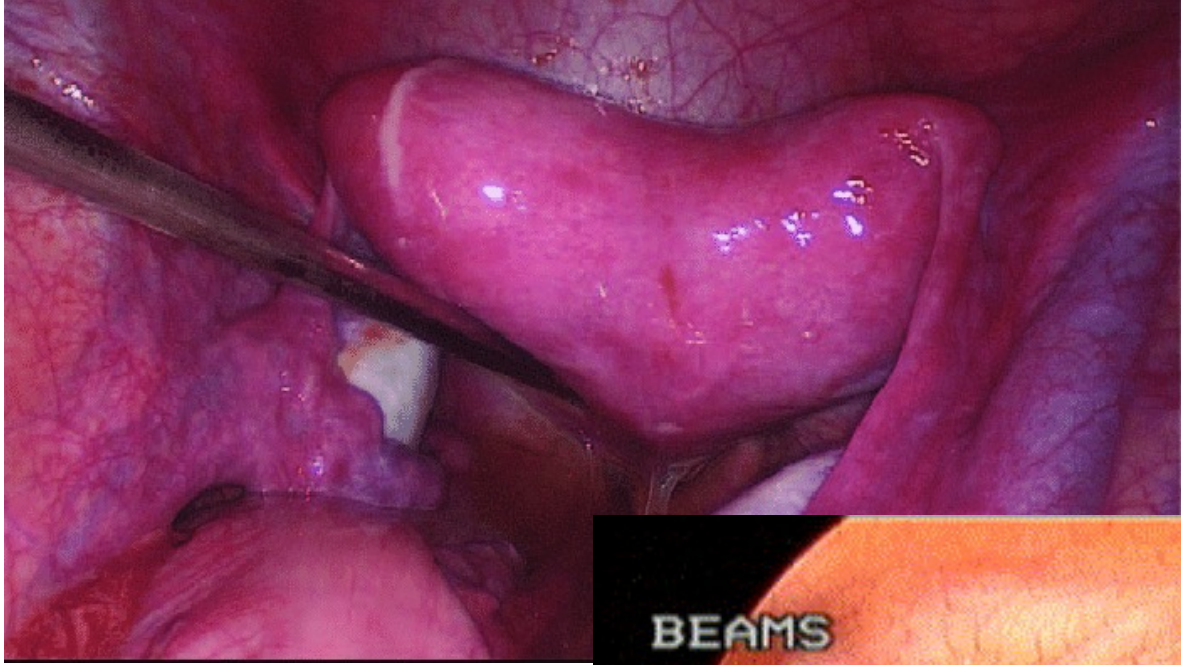
## Jednoplášťové

- Zvenku jednotná, abnormalita uvnitř

## Dvouplášťové

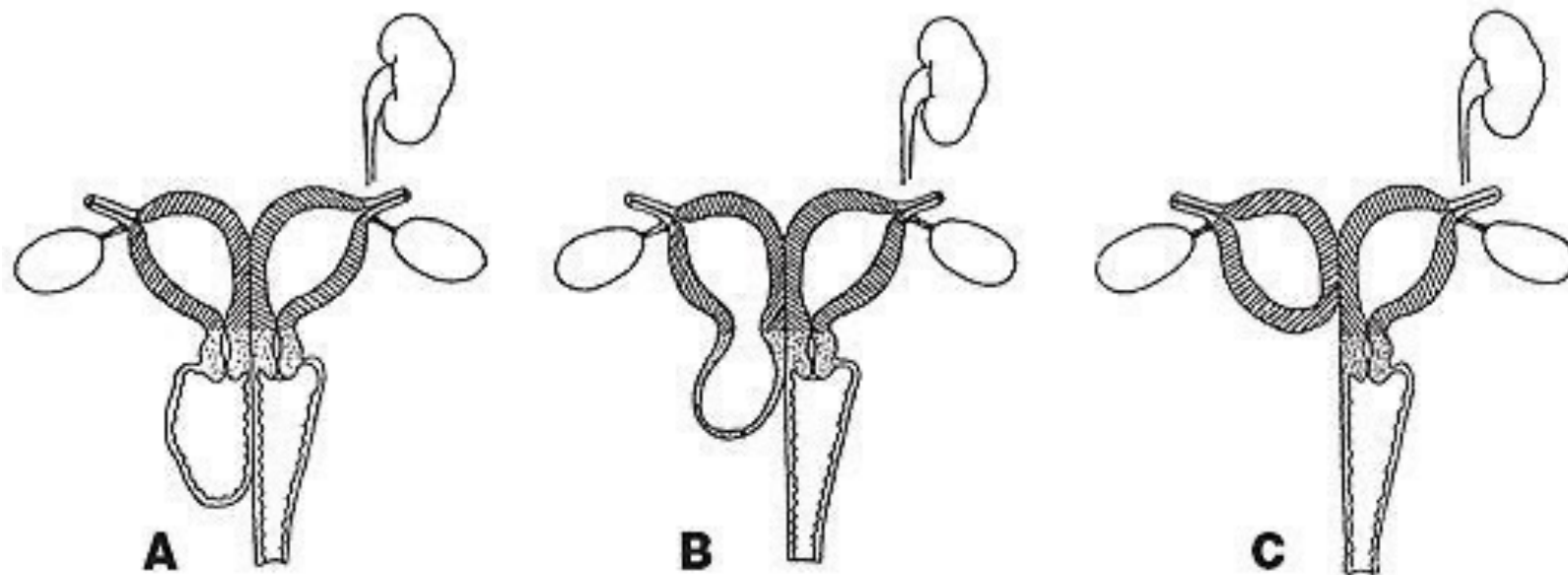
- Abnormalita patrna i zevně





## Kombinované poruchy – syndrom inkompletního zdvojení

- Nesplynutí Müllerových vývodů a nespojení s kloakální výchlípkou
- Vždy aplázie ledviny a ureteru ipsilaterálně





Class U0/Normal Uterus



Class U1/Dysmorphic Uterus

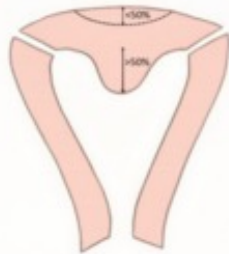


a. T-shaped



b. Infantilis

Class U2/Septate Uterus

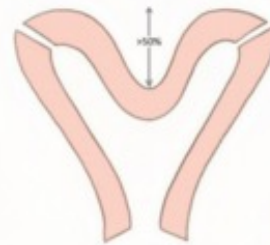


a. Partial



b. Complete

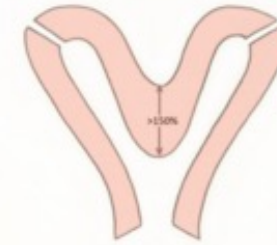
Class U3/Bicorporeal Uterus



a. Partial



b. Complete

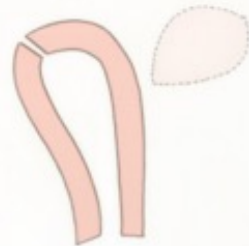


c. Bicorporeal Septate

Class U4/Hemi Uterus



a. With rudimentary cavity



b. Without rudimentary cavity

Class U5/Aplastic Uterus



a. With rudimentary cavity

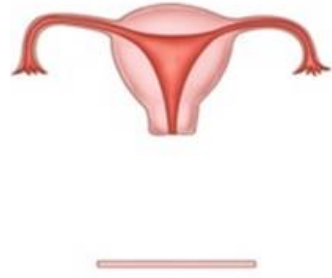


b. Without rudimentary cavity

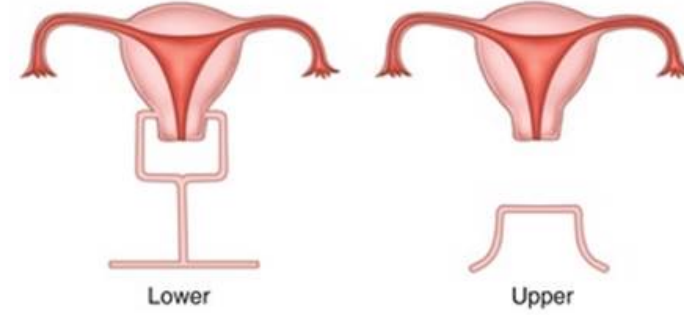
Class U6/Unclassified Cases

**a**

1. Vaginal aplasia : Complete



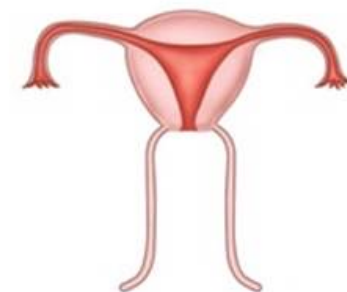
2. Vaginal aplasia : Partial



3. Cervical aplasia



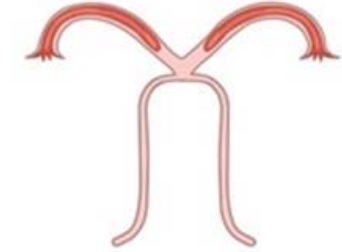
4. Cervical hypoplasia



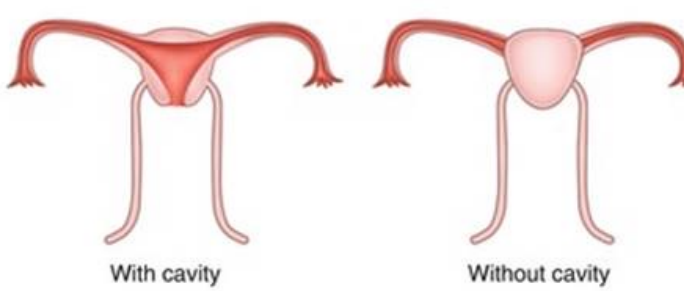
5. Tubal aplasia



6. Uterine aplasia



7. Uterine hypoplasia



8. Combined: commonest findings  
Cervico-vaginal agenesis



Cervico-utero-vaginal agenesis  
(Rokitansky syndrome)



# Sdružené vývojové vady

## **U pseudohermaphroditismu:**

Hypospadie

Epispadie

