



# SURGICAL HYSTEROPROSTATECTOMY?

## A 59-YEAR-OLD MALE PATIENT WITH PROSTATE CANCER FOUND TO HAVE PERSISTENT MÜLLERIAN DUCT SYNDROME WITH UTERUS AND FALLOPIAN TUBES.

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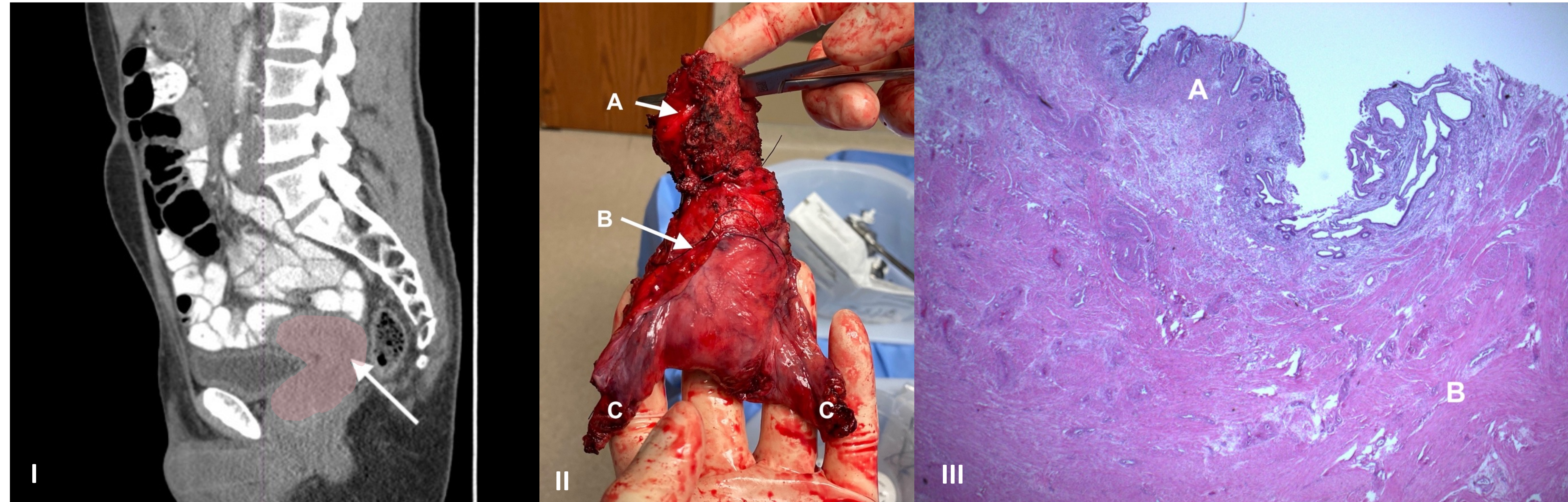


### Background

- Persistent Mullerian duct syndrome (PMDS) is a rare condition where the Mullerian structures fail to regress completely, resulting in male patients having both male and female internal organs.
- Most PMDS patients are diagnosed in infancy during laparoscopic orchidopexy, but in rare cases, adults may present later in life with complaints of infertility and a past history of undescended testicles
- Here, we present a case of an adult PMDS patient with prostate cancer and a history of bilateral undescended testicles.

### Case

- 59 y.o. African American patient
- PMH: IVDU, alcohol induced CHF, cardiac pacemaker, hepatitis C, infertility, and bilateral cryptorchidism
- Labs: Presented with an **elevated prostate specific antigen (PSA)**
- PSH: failed orchidopexy at 11 years old
- Physical exam: penile **chordee**, a shrunken scrotum without testes, and a **tender right inguinal mass (undescended testicle)**
- Pathology: Prostate biopsy showed **unfavorable intermediate risk prostate cancer**
- Imaging: CT scan for prostate cancer also revealed suspected uterine tissue (Figure 1 I)



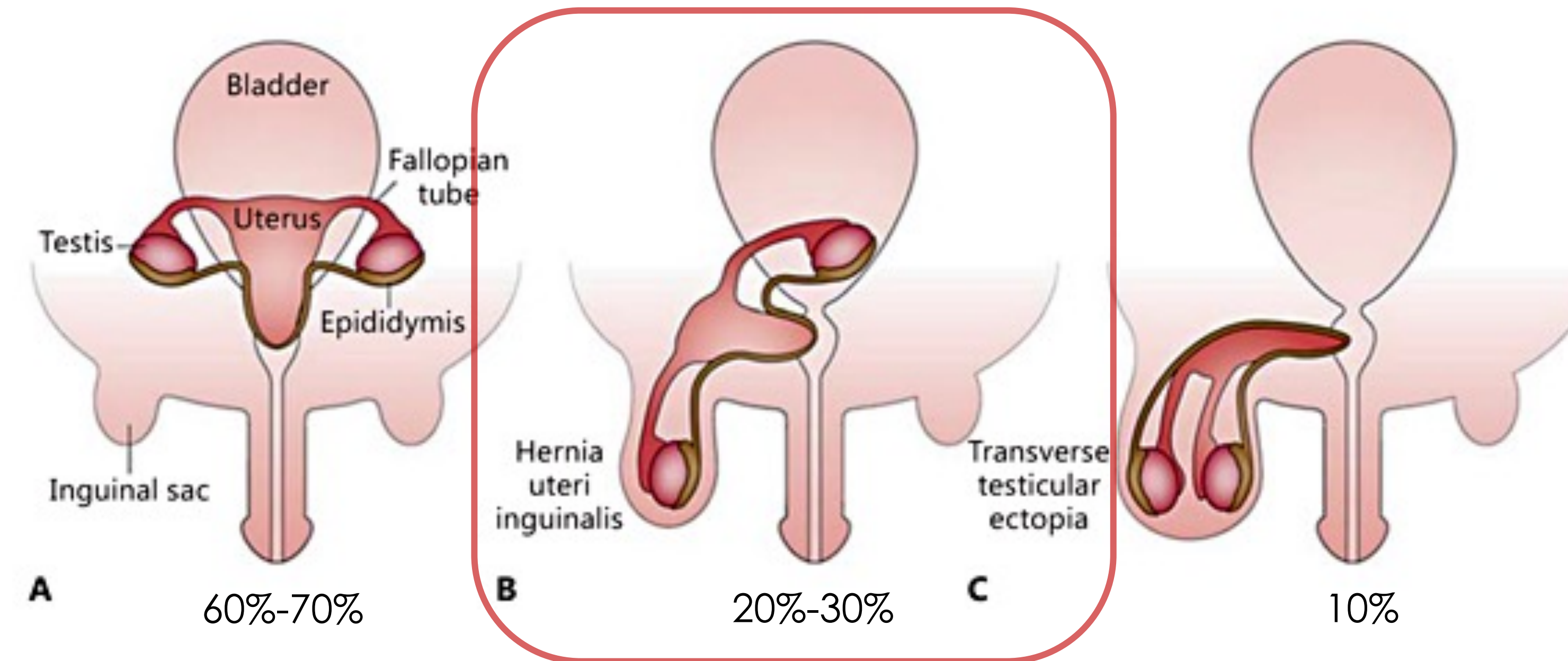
**Figure 1.** I: CT scan with abnormal uterine tissue present. II: uterine tissue removed from patient with cervix and prostate (A), uterus (B), and Fallopian tubes (C). III: Pathology slide of uterine tissue with poorly differentiated endometrium (A) and surrounding myometrial stroma (B).

### Surgery and Follow-up

#### Prostatectomy with staging + Hysterectomy

##### Figure 1 (II)

- **LEFT testicular vessels** and vas deferens were found to have **blind endings** consistent with vanishing testicle
- **RIGHT** showed **atrophic vessels entering** the internal inguinal **ring**.
- Surgical pathology: revealed **positive lymph nodes**,
- Follow-up: patient desired to treat with inguinal orchiectomy over medical androgen suppressive therapy.
- Surgery revealed atrophic right testicular nubbin that was excised, and the patient's total free testosterone dropped from 293 ng/dL to 10 ng/dL
- To date he remains asymptomatic with a PSA <0.1.



**Figure 2.** I: Illustrated abnormal development and migration of testes in patients with PMDS. A: bilateral intra-abdominal testes (female form); B: Hernia uteri inguinalis (male form); C: Transverse testicular ectopia (male form).

### Conclusion

- In PMDS **orchidopexy** is **recommended**, but **excision of Mullerian tissue is not required**.
- This case also illustrates the **importance** of a **thorough history and physical** in all patients, especially **marginalized populations** with barriers to care, such as a longstanding history of IV drug and alcohol use.

### Discussion

- PMDS testicles can be present in male or female forms (Figure 2). Our patient presented with hernia uteri inguinalis as one testicle was present in the scrotum.
- Over **80% of PMDS** patients are homozygous for either **abnormal anti-Mullerian hormone (AMH)** or the **AMH receptor (AMHR2)** genes
- AMH or AMHR2 normally cause involution of the uterus, fallopian tubes and upper third of vagina.
- **Infertility** is thought to as female structures develop in **close proximity to ejaculatory ducts, obstructing them**
  - Another thought is **azoospermia** due to the **hostile environment** for Sertoli cells when testicles fail to descend and remain in the abdomen or inguinal canal
- In PMDS patients, it is **suggested to NOT remove Mullerian structures** if no risk factors are identified to avoid further damage to testicles and fertility
- Concern with our patient: This patient never obtained a urology referral or further work-up until elevated PSA despite physical exam findings, health co-morbidities, and frequent physician visits
- This is particularly concerning as normal repaired undescended testicles become cancerous in 5%-18% and PMDS carries a 33% risk
- This delay could be due to limited access to quality care experienced by illicit substance users and the poorer health outcomes experienced by African Americans when compared to Caucasians.