

## **AN IGNORED MULLERIAN REMNANT: CASE REPORT**

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**Introduction:** The authors describe a case of disgenetic male pseudohermaphroditism assigned to female sex at birth. The patient underwent skin flaps vaginoplasty at 6 years with complete failure. A complete re-assessment of the patient anatomy revealed a Mullerian remnant. Its use for a neovaginoplasty provided optimal result in long term follow-up.

**Materials and methods:** a 9 year-old patient with disgenetic male pseudohermaphroditism (XY cariotype) was referred to our Center. At birth was assigned to female sex because of micropenis, ambiguous external genitalia with single perineal opening and no response to hCG test. At 6 months a bilateral gonadectomy plus reductive clitoroplasty was performed with subsequent complete clitoris atrophy. At 6 year old a perineal skin flap neovaginoplasty failed resulting in a complete retraction of the neovagina.

When the patient was referred to us showed female genitalia aspect with clitoris and vaginal absence and a one centimetre deep introitus. A genitography showed a low confluence urogenital sinus with a Mullerian remnant 5 cm long, ignored in the past patient history. Through a perineal approach it was divided from the urethra and brought to the perineum where labia minora were rearranged. Postoperatively, self dilatations plus oestrogen substitution therapy and psychological support was started. The 3 years follow up shows a 9 cm long neovagina with optimal introitus (hegar n° 24) and good adaptation to female role.

**Discussion:** management of intersex disorders is a challenge because of their wide anatomical variability. Neonatal sex assignment of disgenetic male pseudohermaphroditism should be decided by a multidisciplinary team considering anatomy and future sexual function. A thoroughly anatomical assessment should be performed to ensure the right surgical strategy for each case. The case presented shows how well a narrow and apparently rudimentary mullerian remnant may well develop a fully functional vagina even in a XY pseudohermaphroditism.

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