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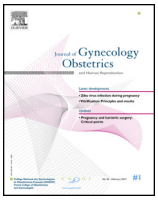

Letter to the Editor

Uterus transplantation in patients with complete androgen insensitivity syndrome (CAIS): Why CAIS cannot be considered as an indication in France

In their recommendations for indications of uterus transplantation (UTx), the French College of Gynaecologist and Obstetricians (CNGOF) French Uterus Transplantation Committee stated that "UTx is currently therefore contraindicated in CAIS for two reasons: absence of an ovary and a XY karyotype [1].

We would like to object this statement and to explain why the existence of a XY karyotype could not be considered as the reason of a non-indication of UTx.

Complete Androgen Insensitivity Syndrome (CAIS) is a rare genetic disorder, characterized by a complete tissue insensitivity to testosterone leading to a female phenotype (without any external genital atypia in the complete form) in an affected individual with a 46, XY karyotype. Peripheral androgen insensitivity is related to a lack of function of the androgen receptor. The diagnosis is generally raised in a girl with an ovarian hernia or a primary amenorrhea with normal breast development in an adolescent girl, associated with the absence of uterus. The hormonal profile of CAIS is characterized by high plasma testosterone (normal or above adult male levels) and high LH levels.

The diagnosis is confirmed by a 46, XY karyotype and the genetic analysis of the androgen receptor gene. Clinically, individuals with CAIS present with female external genitalia. Pubic hair is poorly developed. The clitoris is normal and there is at least a vaginal cup. The gonads have differentiated to testes and the typical location of testes is intraabdominal, in the inguinal canals or in the labia. Due to normal production of anti-Mullerian hormone by the gonads, the Mullerian structures regress (the uterus and the upper part of the vagina). Given the risk of gonadal malignancy, early prophylactic gonadectomy was usually performed. But optimal timing of the gonadectomy is still controversial. Immature germ cells are likely to be present mostly at a young age [2]. Cryopreservation of gonadal tissue in CAIS patients as maturation of immature germ remains experimental [3]. Data on sexuality in female with CAIS are limited but have shown that they have a female-behavior and sexuality [4]. Vaginal hypoplasia can be handled by self-dilatation of the vagina which provide satisfactory introitus in most cases. Surgery may be indicated in case of failure of self-dilatation but should be exceptional. Vaginal dilatations and vaginal surgery were performed at the earliest in adolescence, on the patient 's request. Infertility is a major issue for women with CAIS. Individuals can adopt or might to use choose IVF with donor oocytes, partner's sperm and a surrogate mother to achieve a pregnancy but the last procedure is not currently authorized in France. The possibility of a uterine transplantation would allow a pregnancy with oocyte donation. However, we acknowledge that uterine transplantation is indicated to women who can obtain a spontaneous pregnancy. Overall, we agree that uterine transplantation is not indicated in CAIS women but not because of their karyotype.

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