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# Congenital uterovaginal abnormalities, it's embryogenesis, surgical management and clinical implications

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Objective

Congenital Mullerian duct malformations are a challenging group of conditions for surgeons and need surgical experience and skill. Accordingly, the aim of this study is to present the diagnosis, surgical management, and clinical implications of congenital uterovaginal abnormalities.

Methods

Between 1980 and 2015, 8 patients with congenital uterovaginal abnormalities were diagnosed. In one patient a unique case of an unusual horseshoe shaped double uterus communicating via a transverse canal along with agenesis of the cervix and vagina was noted, and utero-vaginal agenesis was diagnosed in 6 patients. Complete androgen insensitivity syndrome with its female phenotype associated with bilateral testicular tissue in the inguinal canal with an accompanying absence of the ovaries, uterus, uterine tubes, vagina, and an imperforate hymen, was diagnosed in one patient. Clinical examination of all the patients revealed well-developed secondary sexual characteristics. A modified McIndoe vaginoplasty procedure was the surgical treatment common to all patients to treat vaginal agenesis. The surgery was performed by a consultant (Dr. K.G. Paul) using the standardized surgical technique.

Results

An unusual Mullerian duct anomaly, uterus bicornisacollis, was successfully corrected by uteroplasty and a new cervix was constructed. Complete vaginal agenesis was corrected by a modified McIndoe vaginoplasty technique. None of the patients had any significant post-operative complications.

Conclusion

Knowledge of congenital uterovaginal abnormalities diagnosed in this study is essential for surgeons, clinical anatomists, radiologists, and morphologists who may increase the success of their diagnostic evaluations and surgical approaches to the region.

Keywords: Androgen insensitivity syndrome; Infertility; Vaginal agenesis; Wolffian duct

## Introduction

The reproductive system or genital system is a system of sex organs within an organism involved in producing offspring. In men, the genital system includes the prostate, testes, and penis. In women, it includes the ovaries, fallopian tubes, uterus, and vagina. A basic understanding of the embryology of the reproductive tract is essential to restore reproductive and sexual functioning and normalize genital anatomy whenever possible.

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tion (Supplementary Fig. 4). The surgery was performed by consultant Dr. K.G. Paul using the standardized surgical technique. In this operation, the labia are retracted with Allis clamps, a transverse incision is made in the epithelium, and blunt finger dissection is then carried out to create a new vaginal space or canal (optimum vaginal length of 10–12 cm) between the rectum and bladder. Two proposed paddle-shaped full-thickness skin grafts approximately 10 cm in length and 6 cm in width over the inguinal ligaments from the anterior superior iliac spine to the pubic tubercles were harvested from the patient (Supplementary Fig. 5). After harvesting the full-thickness skin graft, to close the wound, the skin is stretched and sutured using a synthetic absorbable ture (Supplementary Fig. 5). These full-thickness skin grafts allow for sufficient penetration of the transudation nutrients from the bed of the graft that are necessary for nutrition during the first 72 hours until microcapillary growth has been completed, which reduces postoperative contraction. An artificial mould was created using a dental impression material and a full-thickness skin graft. A mould was shaped using 3M Putty Material and covered with a layer of soft sponge and a condom. Subsequently, the condom was applied, tied on to its open end. The size of the mould corresponding to the neovagina was selected (Supplementary Fig. 4). The full-thickness skin graft was then sewn over this mould with the deep surface of the graft facing outward so as to be in contact with the newly created vaginal wall. The margins were sutured together with synthetic absorbable suture and introduced into the neo-vaginal canal or space (Supplementary g. 4). To hold the skin graft covered mould in place for 12 days, the labia were sutured at the midline with interrupted 0-nylon sutures without tension. A mould covered with a layer of soft sponge and a condom was removed on the 12th postoperative day. The new vagina was thoroughly inspected and irrigated with normal saline solution.

## Results

An unusual Mullerian duct anomaly, uterus bicornis (double uterus) acollis (absence of cervix), was successfully corrected by uteroplasty and a new cervix was constructed. Complete vaginal agenesis in 7 patients was corrected by a modified McIndoe vaginoplasty technique. Clinical examination performed postoperatively revealed a neovagina of adequate length and caliber. None of the 7 patients had any significant post-operative complications.

## Discussion

The genital system is concerned with the maintenance and propagation of the species. In women, the internal genitalia include ovaries, fallopian tubes, the uterus, and the vagina; the external genitalia include mons pubis, labia majora and minora, the clitoris, the vestibule, and the perineum. Gonads initially develop in females (ovaries) and males (testes) from the undifferentiated genital ridge at approximately the 5th week of development, and gonadal differentiation becomes apparent at approximately the 7th week of embryonic life. In embryos of both sexes, the primitive sex ducts are indifferent and consist of 2 paired ducts—mesonephric (Wolffian) and paramesonephric (Mullerian). With the development of the testis from the genital ridge, the mesonephric duct is retained in males as the duct system of the testis and the paramesonephric duct mostly degenerates. In females, however, the paramesonephric duct plays an important role in the development of reproductive organs and the mesonephric duct and its tubules mostly regress.

At first, these paired bilateral Mullerian ducts pass caudally (through the cranial vertical part), lateral to the mesonephric duct. In the pelvis, they cross (through the intermediate horizontal part) ventral to the mesonephric duct and grow medially. During the 8th week, they reach the caudal end of the mesonephric duct and to contact and fuse with their counterparts (caudal vertical part) to form a Y shaped uterovaginal bulb or tubercle that bulges into the dorsal of the urogenital sinus. Caudal vertical parts of both Mullerian ducts fuse in the caudo-cranial direction, which normally occurs between the 6th and 11th weeks of gestation. The partition between them completely disappears and forms a single duct utero-vaginal canal by the end of 3rd month. The cranial part of the utero-vaginal canal forms the entire uterus, and points of fusion of the 2 ducts represent the site of the future fundus. The cranial vertical parts and most of the intermediate horizontal parts of each Mullerian duct form the respective fallopian tubes. Any disruption of Mullerian duct development and fusion during embryogenesis can result in a broad and complex spectrum of congenital abnormalities, termed Mullerian duct anomalies. A uterus is absent in 2% to 7% of pa-

tial for surgeons, clinical anatomists, radiologists, and morphologists to increase the success of reproductive diagnostic evaluation and surgical approaches to the region.

## Conflict of interest

No potential conflict of interest relevant to this article was reported.

## Ethical approval

This study was reviewed and approved by the appropriate institutional human research ethics committee (reference number: HREC10AUG19).

## Patient consent

Each patient's informed consent for the purpose of this study (i.e. publication without disclosure of personal identity) was obtained.

## Supplementary materials

Supplementary Figures associated with this article can be found online at https://doi.org/10.5468/ogs.2020.20046.

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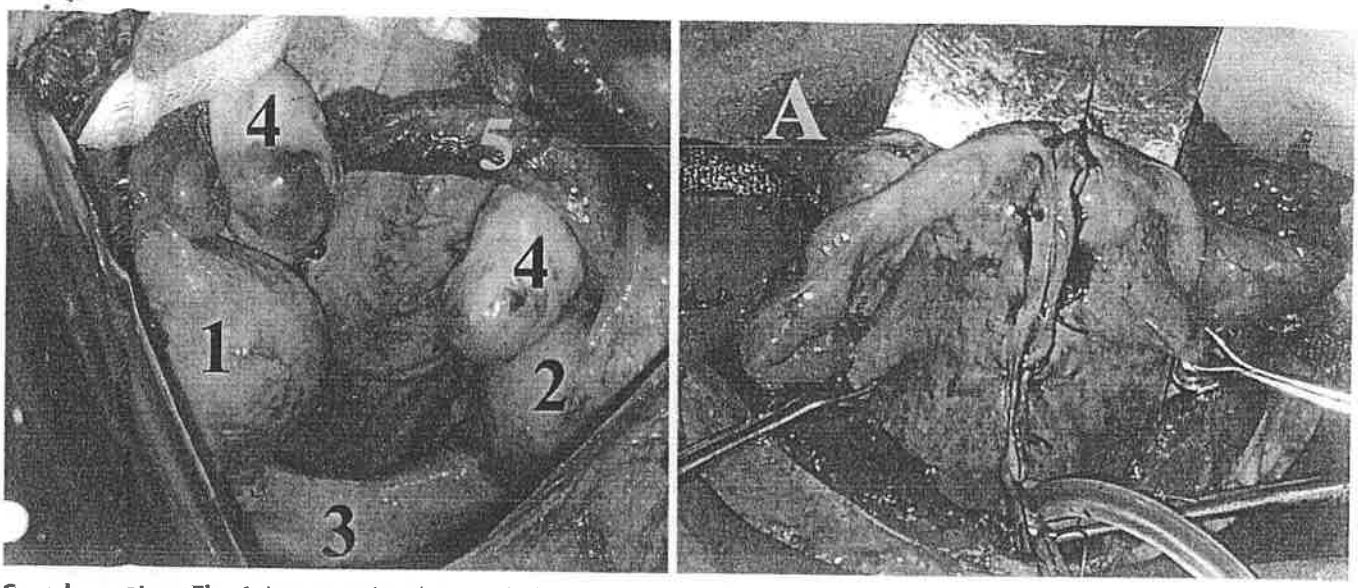
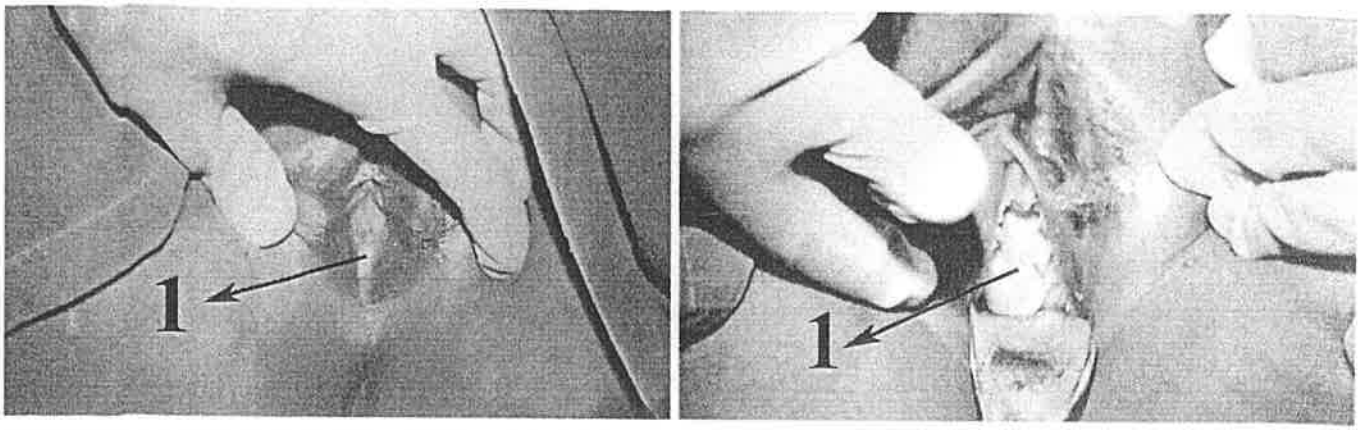
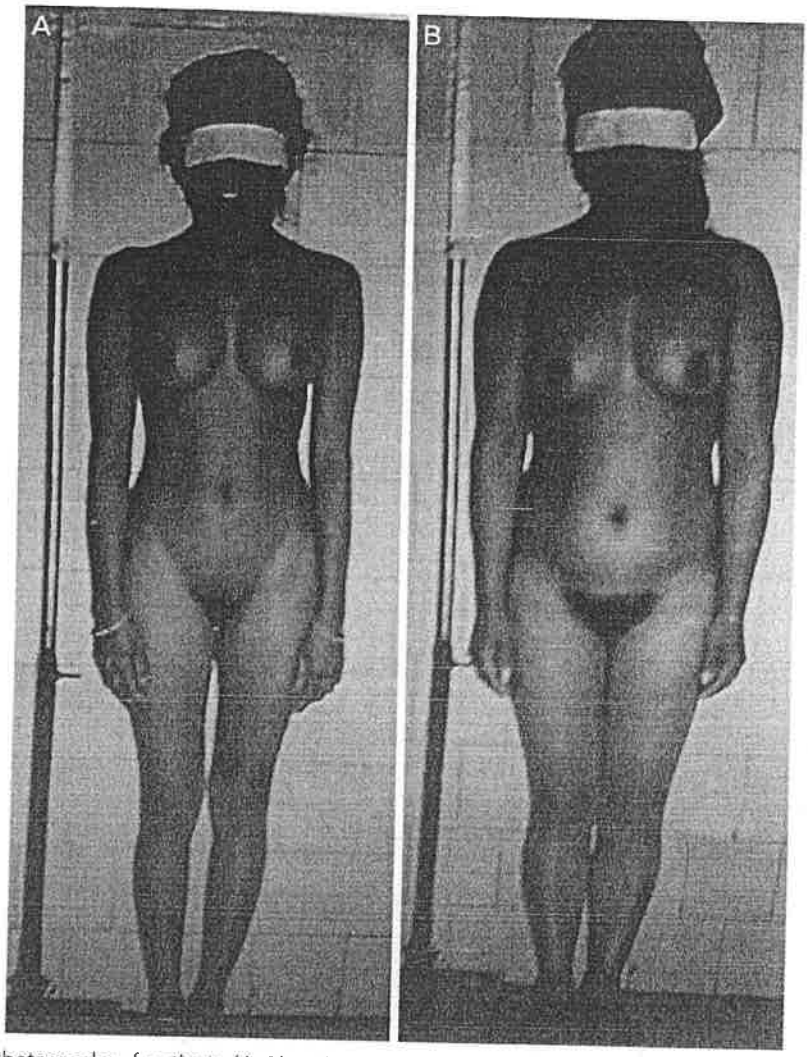


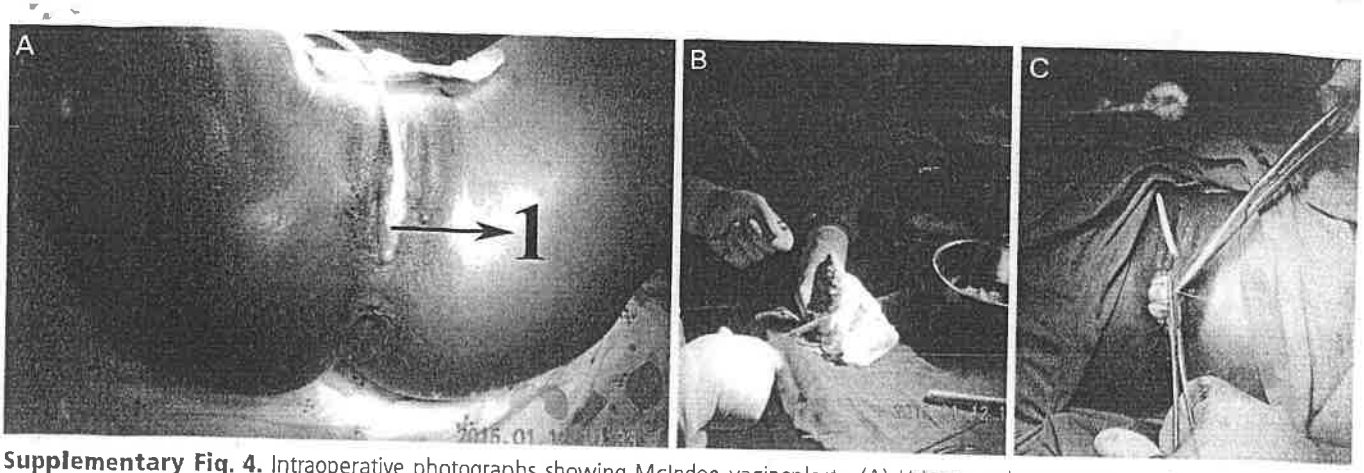

**Supplementary Fig. 1.** Intraoperative photograph showing uterine malformation and uteroplasty; 1 and 2, double uterus; 3, transverse canal; 4, ovaries; 5, fallopian tubes; A, uteroplasty.




**Supplementary Fig. 2.** Photographs showing congenital absence of the vagina; 1, vagina.




**Supplementary Fig. 3.** Photographs of patients (A, B) with complete androgen insensitivity syndrome with well-developed secondary sexual characteristics. The patient has a 46, XY karyotype and bilateral testicular tissue in the inguinal canal with an absence of ovaries, erus, uterine tubes, vagina, and an imperforate hymen.




**Supplementary Fig. 4.** Intraoperative photographs showing McIndoe vaginoplasty. (A) Urinary catheterization under anaesthesia; 1, congenital absence of the vagina. (B) Mould lined by the full-thickness skin graft. (C) Mould lined by the full-thickness skin graft inserted to the newly constructed vaginal space.