

Intraoperative Vaginoscopy and Transrectal Ultrasound to Facilitate Genital Reconstruction in a Female Patient with Clitoromegaly due to Congenital Adrenal Hyperplasia: A Case Report

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Abstract

We present the case of a 29-year-old female who had atypical genitalia due to congenital adrenal hyperplasia secondary to 21-hydroxylase deficiency who underwent reconstructive surgery. We performed a vaginoscopy and intraoperative ultrasound to facilitate the procedure as we consider that this approach could reduce the risk of complications.

Keywords: Vaginoscopy; Reconstructive Surgery; Congenital Adrenal Hyperplasia; Transrectal Ultrasound; Clitoromegaly

Introduction

Classic congenital adrenal hyperplasia (CAH) is the most common disorder of sex development (DSD), presenting with atypical genital appearance and a 46 XX karyotype in the newborn period [1].

More than 90% of all cases are secondary to 21-hydroxylase deficiency [2]. This group of patients present in the neonatal period without palpable gonads and atypical genital due to endogenous virilization.

Prevalence varies according to geographic area and race, affecting approximately 1 in 15,000 livebirths [3].

Patients with 46 XX CAH and virilization will have a common urogenital sinus (UGS) and different degrees of clitoral hypertrophy, with normal vagina, cervix, uterus, and ovaries. The aim of the surgical treatment is not only to restore anatomy but to achieve a functional and cosmetic outcome [4].

The optimal timing of surgery is challenging and requires shared decision-making with the family [5].

We report the case of a female with clitoromegaly and low confluence UGS due to CAH who underwent genital reconstruction.

Case Report

A 29-year-old patient was referred to our department from her local hospital to consider the possibility of genital reconstructive surgery.

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She was diagnosed with CAH after birth. Karyotype analysis was 46 XX. She underwent clitoroplasty and labiaplasty at the age of 2 and she had menarche at the age of 13.

On examination there was a small pseudopenis of 3 cm with a narrow orifice just below it (Image 1). There was no palpable gonad. The remaining female secondary sexual features were normal. Additional examination of the patient did not find any other gynaecological or systemic disorders.



Image 1: Clinical appearance before surgery.

Gynaecological ultrasound showed a cystic lesion bellow the normal uterus, compatible with a hydrocolpos. The ovaries were normal. The evaluation of the internal genital organs showed the common channel 1.5 cm in length, as a distal fusion of the vagina and urethra.

Patient was discussed in our multidisciplinary meeting, and the decision was to perform feminizing genitoplasty with clitoroplasty and vaginoplasty as the procedures of choice.

The procedure started with cysto-vaginoscopy to confirm the anatomy seen preoperatively on MRI and USS, verify the length of the vagina, the connection with the cervix and the location of the bladder neck.

After visualising the anatomy, a bladder catheter was installed in the bladder.

The upper part of her vagina was not connected to the introitus. The rectal ultrasound was used to facilitate the visualization of the hydrocolpos (Image 2). Therefore, the upper vaginal space was opened under ultrasound guidance and clear liquor drained from it. Methylene blue was used to rule out a bladder connection.

Vaginoscopy was then performed, and the cervix was visualised. The next step was creating the lower part of the vagina and the detachment of the vagina from the confluence of the common UGS. A vertical incision was created on the perineum and the separated vagina was then brought out as a separate opening bellow the urethra. Both were then separated and connected to the introitus (Image 3a).

Clitoral reduction was then performed. The skin of the shaft was mobilised down to the base of the corpora cavernosa by a coronal incision. The next step was the dissection of the neurovascular (NV) bundle from the corpora cavernosa finding an avascular plane with

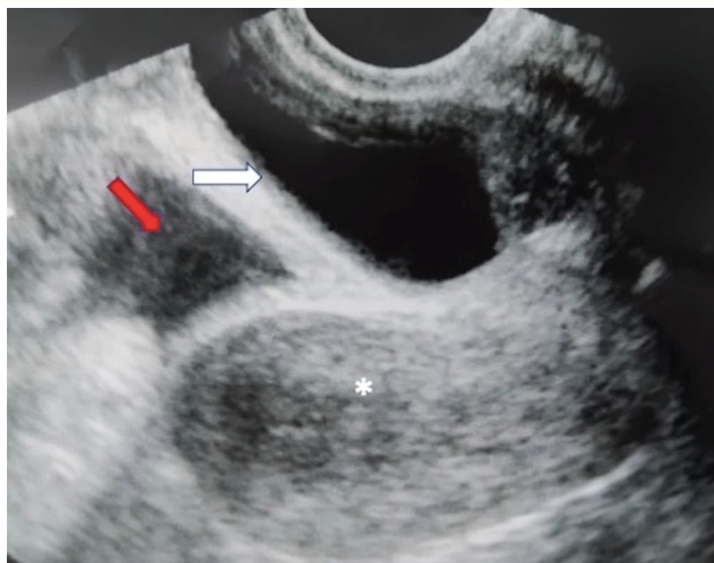


Image 2: Intraoperative USS showing hydrocolpos (white arrow), bladder (red arrow) and uterus (white asterisk).

a longitudinal incision at 9 and 3 o'clock. The pedicle was isolated proximally from the tunica albuginea and a loop was placed through the space created to facilitate the rest of the dissection of the pedicle from distal to proximal (Image 3b). Once the corpora were dissected from the dorsal NV pedicle, until their bifurcation under the symphysis pubis, the corpora cavernosa were removed (Image 3c). Then we had a dorsal strip containing dorsal arteries, nerves, and deep dorsal vein of clitoris under Buck's fascia supplying the glans. Afterwards, one third of the lateral aspect of the clitoroideal skin was removed to reduce the size of the gland. Finally, the clitoris was reattached to the corporeal stumps near the lower edge of the pelvis with absorbable sutures (Image 3d).

Discussion

In 46, XX CAH, the vagina opens into the dorsal wall of the urethra at a variable distance from the bladder neck but lower than where the verumontanum is usually located in the male urethra. This junction is also at variable space from the perineum based on the development of the urethra and the increased thickness of the pelvic floor muscles [6]. Prader classification suggested the height of the urethro-vaginal confluence was associated to the level of external virilization, but we now know this is incorrect. The sagittal fusion of the genital folds could range from an almost normal vulva to a complete scrotal-like aspect. Always, the gonads are not present in the genital folds [7].

Centralization of care is extremely important as genitoplasty procedures are rare. The Chicago Consensus Statement state that only those surgeons experienced in the surgery of DSD should perform these procedures [8].

Optimal timing for surgery is controversial, as it is the surgical technical of choice. Some authors claim the psychological benefits of early surgery, whereas other colleagues prefer surgery at a later stage due to the need for further reconstructive surgery and the inability to obtain signed consent in childhood [8,9].

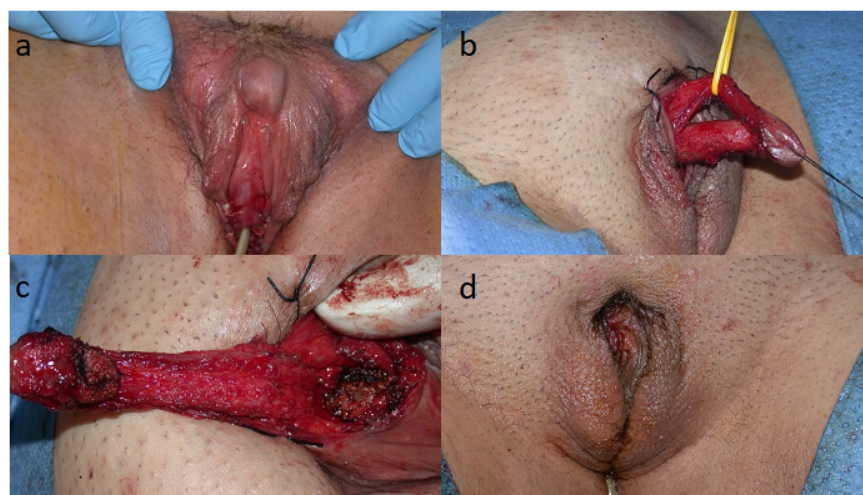


Image 3: a) Clinical appearance after vaginoplasty. b) Traction on the loop separating the neurovascular pedicle from the corpora cavernosa. c) Removal of the corpora cavernosa. d) Postoperative appearance.

Sturm and colleagues, in 2015 [1], evaluate the surgical treatment in patients with CAH and found that approximately 90% of feminizing genitoplasty procedures include a vaginoplasty.

Leslie and colleagues [4] state every reconstructive procedure should always begin with a cystovaginoscopy to measure the location of the confluence relative to the introitus and the bladder neck, verify the presence of a sole cervix and confirm the anatomy visualized with genitography.

In 2016, Lindert, *et al.* [10], compared the results of the ultrasound with the findings of endoscopy which was performed before surgery. The length of the UGS, the length of the bladder neck and the distance to the vaginal opening into the UGS was measured. In more than 90% of the patients the entry of the vagina into the UGS could be recognized. The ultrasound also was an effective tool to precisely measure the length of the UGS and the length of the bladder neck.

They concluded that the use of this technique could potentially reduce the morbidity and the costs associated with endoscopy and genitography.

Blanco and colleagues [11] performed a transrectal ultrasound in three patients with CAH and concluded it is ideal to visualize the vagina and the pelvic floor.

In our patient an intraoperative transrectal ultrasound and a cysto-vaginoscopy was performed to confirm the anatomy and to reduce the risk of complications during the reconstructive procedure.

In this particular case the hydrocolpos was drained under USS guidance and the vagina was then safely connected to the introitus.

Clitoroplasty was finally performed, based on the technique described by Kumar, *et al.* [12] who emphasized the importance to preserve the NV pedicle leading to the clitoris while removing the erectile tissue. This procedure preserves the physiological function of the clitoris while removing a variable segment of the corpora cavernosa.

Conclusion

Reconstructive treatment of CAH is complex and should be performed by experienced surgeons. The objective of surgery is to isolate the urethra from the vagina and to reposition the proximal aspect of the vagina to its usual place in the perineum. Intraoperative vaginoscopy and transrectal ultrasound could facilitate this step allowing us to verify the anatomy beforehand, reducing the risks of complications.

Conflict of Interest

The authors certify that there is no conflict of interest with any financial organization regarding the material discussed in the manuscript.

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Authors Contribution

All authors contributed equally to the manuscript and read and approved the final version of the manuscript.

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