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Laparoscopic-assisted vaginal pull-through: A new approach for congenital adrenal hyperplasia patients with high urogenital sinus

Jacques Birraux, Faustin Tambo Mouafo[1], Sophie Dahoun[2], Veronique Tardy[3], Yves Morel[3], Pierre Mouriquand[4], Claude Le Coultre, Pierre-Yves Mure[4]

ABSTRACT

Background: To open vaginal cavity to the pelvic floor is part of surgical treatment for urogenital sinus (UGS) in girls with congenital adrenal hyperplasia (CAH). For high UGS, this operative procedure can be challenging and may jeopardise urinary continence. Combined perineal and laparoscopic approaches could be useful to minimise perineal dissection and to facilitate the vaginal lowering. **Patients and Methods:** We report the procedure of a laparoscopic-assisted vaginal pull-through for supra-sphincteric UGS in a 5-year-old girl with CAH. Laparoscopic dissection of the vagina from the posterior wall of the bladder and urethra, division of the confluence and vaginal pull-through to the perineum are described. **Discussion:** The technique is derived from laparoscopic-assisted treatment for high ano-rectal malformations. Compared with current procedures for treatment for high UGS, laparoscopic-assisted approach allows mobilising vagina with minimal dissection of perineum and complete preservation of urethra. Another major advantage is to provide a direct vision for dissection of the space between rectum and urethra prior to vaginal pull-through. **Conclusion:** Laparoscopic-assisted vaginal pull-through appears to be an interesting approach for high UGS in CAH patients, reducing dissection and risk of urinary incontinence. This new approach needs to be strengthened by other cases.

Key words: Congenital adrenal hyperplasia, laparoscopy, urogenital sinus, vaginal pull-through

INTRODUCTION

Congenital adrenal hyperplasia (CAH) is a rare disease, which induce high levels of androgens, consecutive to an enzymatic deficiency in the biosynthetic pathway from cholesterol to the different corticosteroids. In female foetuses, this results in varying degrees of virilisation of external genitalia and incomplete separation of urethra and vagina with confluence of both structures in a common channel called urogenital sinus (UGS). The severity of virilisation was described by Prader.[1] It is known that these two characteristics are not always linked, meaning that a low degree of external virilisation may cohabit with high UGS, and *vice versa*.[2] Interestingly, this is also not linked to the severity of the enzymatic activity deficiency.[3] Rarely, confluence may be very high, supra-sphincteric, with a vaginal confluence having a *veru montanum*-like appearance.

Paediatric Surgery, Department of Pediatrics Children's Hospital, University Hospital of Geneva, Rue Willy Donzé 6, 1211 Genève 14, Switzerland, [1]Department of Surgery and Specialties, Faculty of Medicine and Biomedical Science, Pediatric Surgery Unit, Yaounde Gyneco-Obstetric and Pediatric Hospital of Yaounde, University of Yaounde I, Yaounde, Cameroon, [2]Medical Genetic, Department of Genetics University Hospital of Geneva, Rue Michel Servet 1, 1211 Genève 14, Switzerland, [3]Department of Molecular Endocrinology and Rare Diseases, Molecular Endocrinology and Rare Diseases, University Claude Bernard Lyon 1, Hôpital Femme Mère Enfant, [4]Department of Paediatric Surgery, University Claude Bernard Lyon 1, Hôpital Femme Mère Enfant, 69677 Bron Cedex, France

Address for correspondence:
Prof. Pierre-Yves Mure,
GHE-HFME, 59, Boulevard Pinel, 69500 Bron, France.
E-mail: pierre-yves.mure@chu-lyon.fr

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Corrective surgery in CAH girls is a controversial issue in paediatric urology, especially regarding clitoral reduction. In this report, we will focus on vaginoplasty that is less debated. The guiding principles of this surgery were proposed by Hendren and Crawford in their description of the vaginal pull-through procedure.[4] This manoeuvre can be quite straightforward in patients with a low UGS, the confluence being almost at perineum. For supra-sphincteric confluence, the procedure may appear challenging, potentially leading to severe complications especially regarding urinary continence. The Hendren's principles are still followed, but numerous modifications have been proposed. Some of those modifications use an extensive dissection of the perineum, hence also a better mobilisation of the vagina, such as the total urogenital mobilisation (TUM).[5] Different approaches have also been proposed, for example, transtrigonal approach.[6] More recently, anterior sagittal transrectal approach (ASTRA) has been described.[7] Nevertheless, these techniques remain complex, with limited exposure and extensive perineal dissection.

Keeping in mind Georgeson's description of laparoscopic transsection of recto-urethral fistulae for ano-rectal malformations,[8] we postulated that laparoscopic approach of high UGS and separation of the vagina from the urethra could be possible. We were backed up by the experience of Fuchs *et al*.[9] reporting laparoscopic approach for a non-hormonal UGS, not linked to CAH.

We report the laparoscopic approach of a high UGS in a CAH girl, done at the same time as feminising genitoplasty.

PATIENTS AND METHODS

A 3 -year-old girl with genital ambiguity was referred to our institution without previous medical assessment. She presented a severe virilisation, Prader V stage, with the clitoral organ measuring 5 cm in length and 2.5 cm broad, a sub-coronal urethral meatus, and complete fusion of the genital fold with scrotal aspect [Figure 1].

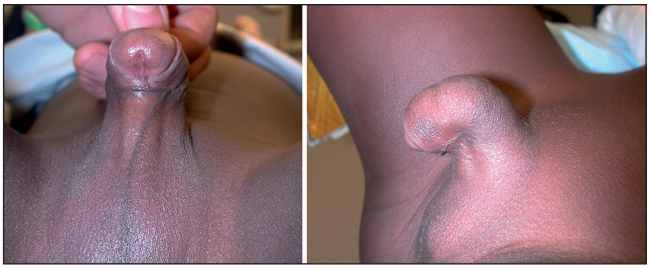

Figure 1: Masculinised external genitalia

At birth, she was declared male. Familial history revealed a boy deceased soon after birth, 2 years before the birth of our patient. Initial work-up revealed a 46,XX caryotype. Genetic analysis shows compound heterozygote for two severe mutations of CYP21 gene. Ultrasound and magnetic resonance imaging of the pelvis revealed normal female internal organs and showed high UGS. Voiding cystourethrography showed the confluence of the vagina and urethra about 15 mm below the bladder neck. Due to the severity of the case, we decided to make a diagnostic cystoscopy, which revealed a male-like urethra with confluence between urethra and vagina 15 mm below the bladder neck, having an aspect of *veru montanum*. Vaginoscopy reveals a normal-appearing vagina of 5 cm depth. Cervix uterus was normal looking. For 2 years, the child and her parents were regularly seen in a multidisciplinary team, including endocrinologist, geneticists, psychologist and paediatric surgeons. During this follow-up period, parents had a good understanding of the situation of their child, and common decision was made to change the sex of rearing.

Surgery began with a new urethro-cystoscopy. A 2 Fr Fogarty catheter was placed into the vagina and a 10 Fr Foley catheter in the bladder. We then continued with a standard laparoscopic approach. Revision of the pelvis showed two normal ovaries and annexes and a normal uterus. The patient was positioned in Tredelenburg, and procedure was begun with opening of the peritoneal fold just in front of uterus wall. Dissection was conducted in this almost avascular plan. The two ureters were visualised but not dissected. The Fogarty balloon was used to localise the proximal part of the vagina. The anterior vaginal wall was dissected by blunt dissection up to the urethro-vaginal confluence, which was divided [Figure 2] after complete dissection of the vaginal end

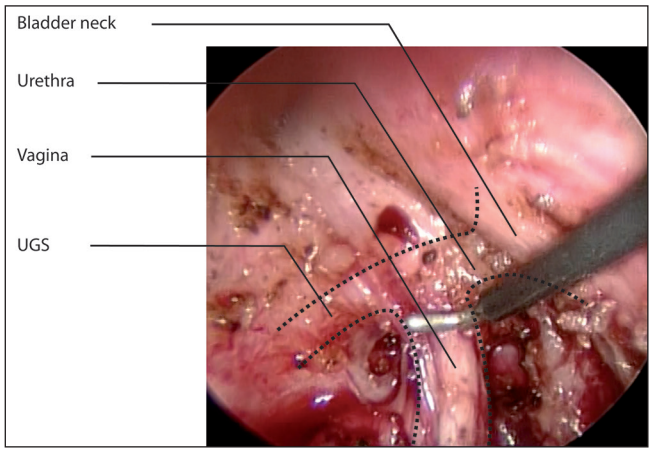

Figure 2: Division of vaginal fistula

with a McGill clamp. The defect on the urethra was not sutured. Lateral aspects of the vagina were freed from surrounding tissues. Vascularisation of the distal part of the vagina was clearly maintained. Genitoplasty was pursued with a median opening of the labial fold fusion and UGS up to its bend towards bladder. A Fortunoff's flap was achieved. A space was created between the rectum and urethra up to the abdominal cavity, using a Kelly clamp. Laparoscopic control allowed to secure the direction of this dissection. The opening was then progressively enlarged with Hegar dilatators, till size 17 [Figure 3]. We then introduced a 10 mm unarmed trocar, through which the distal part of the vagina was seized with a non-traumatic clamp. Taking care not to twist it, the vagina was then pulled downwards without tension to its definitive position and fixed to the Fortunoff's flap posteriorly and to the urethra anteriorly. Feminising genitoplasty was completed, with clitoral neuro-vascular bundle preservation[5] [Figure 4]. Bleeding was minimised using bupivacaini hydrochloridum anhydricum 0.25% Adrenalin (Adrenalin 1:200,000) (AstraZeneca®) in local injection. Following procedure, a perineal hematoma results in partial dehiscence of the Fortunoff's flap. This could necessitate local revision later on. Urinary catheter was left in place for 10 days, and cystography at this time showed a well-healed urethra, with no urine leaks. Two months later, a clinical work-up with a cystovaginoscopy was organised. The child was doing well, had no pain or urine leaks. Cystoscopy showed a normal looking urethra, without stenosis, measuring 32 mm from the bladder neck to the vulva. The gain of the urethral length between the previous confluence and the actual position was 17 mm. Vaginoscopy demonstrated a well-vascularised, normal looking vagina, including the distal part, measuring 60 mm from cervix to vulva. The glans clitoris was well vascularised.

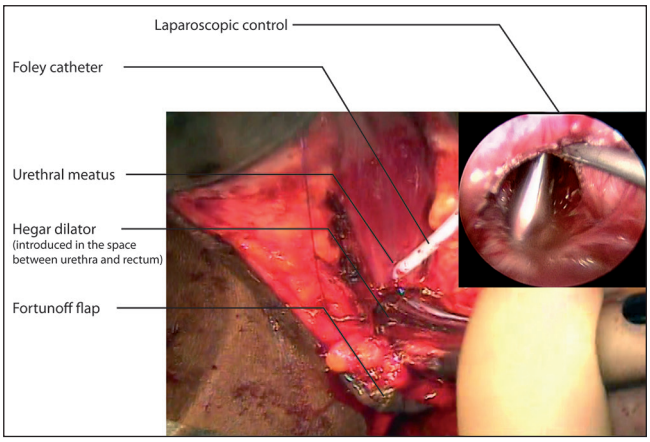

Figure 3: Hegar dilatation

DISCUSSION

Bringing the vagina to the perineum in patients presenting with a high UGS is the most delicate part of the procedure and can lead to per- or post-operative complications.[10] Hendren and Crawford first described the vaginal pull-through in 1969,[4] using a perineal approach in seven such patients, allowing a significant step forward in the options provided for these patients. Despite this progress, there remained concerns regarding continence following this procedure.[11] In 2005, a study showed that two-thirds of adult women who underwent genital feminisation surgery in infancy presented with lower urinary tract symptoms.[12] These results and the difficult exposure during this surgery have led to technical modifications such as TUM[5] and more recently ASTRA.[7] A recent review has shown satisfactory results in regards to continence with these modified techniques.[13] Unfortunately, these studies only included children, and the mean follow-up was shorter than 4 years. As long-term results are lacking, we favour any technical improvement reducing the perineal dissection and thus preserving the surrounding structures.

As proposed by Rink *et al.*,[14] we believe that the decision making in regards to vaginoplasty should rely on the distance from the confluence to the bladder neck rather than the length of UGS. The short urethra measured between the confluence and the bladder neck (15 mm) in our case meant that an exclusively perineal approach did not seem appropriate.

The laparoscopic-assisted posterior sagittal anorectoplasty to treat high anorectal malformations was first described by Georgeson *et al.* in 2000[8] is well codified[15] and the results are known.[16] Taking into account this acquired experience and the report

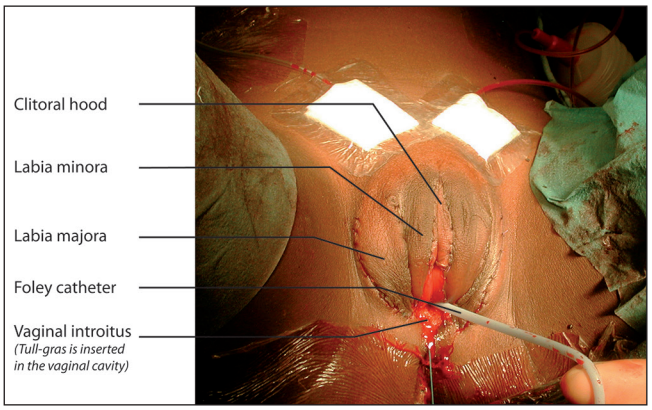

Figure 4: Final aspect of genitoplasty

of the team of Netherland in a different condition,[9] the technique appeared suitable for high UGS in CAH patients to mobilise the vagina without extensive perineal dissection and tension. Moreover, it allows for the creation of a normal length urethra using the distal part of the UGS as a substitute of urethra down to the perineum. Last but not least, the laparoscopic approach offers a safer dissection when creating the space between urethra and rectum which is done under visual control.

CONCLUSION

Laparoscopic-assisted vaginal pull-through appears to be useful in order to minimise perineal dissection and to facilitate vaginal lowering in CAH patients with high UGS. Even as a preliminary report, the technique appears to be considered when surgical correction of this very rare condition is intended. Only long-term follow-up and reports from other surgical teams will allow to validate this approach.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Prader A. Genital findings in the female pseudo-hermaphroditism of the congenital adrenogenital syndrome; morphology, frequency, development and heredity of the different genital forms. Helv Paediatr Acta 1954;9:231-48.
2. Lee PA, Houk CP. Key discussions from the working party on Disorders of Sex Development (DSD) evaluation, Foundation Merieux, Annecy, France, March 14-17, 2012. Int J Pediatr Endocrinol 2013;2013:12.
3. Sugiyama Y, Mizuno H, Hayashi Y, Imamine H, Ito T, Kato I, *et al*. Severity of virilization of external genitalia in Japanese patients with salt-wasting 21-hydroxylase deficiency. Tohoku J Exp Med 2008;215:341-8.
4. Hendren WH, Crawford JD. Adrenogenital syndrome: The anatomy of the anomaly and its repair. Some new concepts. J Pediatr Surg 1969;4:49-58.
5. Rink RC, Metcalfe PD, Cain MP, Meldrum KK, Kaefer MA, Casale AJ. Use of the mobilized sinus with total urogenital mobilization. J Urol 2006;176:2205-11.
6. Cavailher H, Braillon G, Spay G. Treatment of vesico-vaginal fistula by trans-trigonal approach. Lyon Chir 1970;66:65-6.
7. Salle JL, Lorenzo AJ, Jesus LE, Leslie B, AlSaid A, Macedo FN, *et al*. Surgical treatment of high urogenital sinuses using the anterior sagittal transrectal approach: A useful strategy to optimize exposure and outcomes. J Urol 2012;187:1024-31.
8. Georgeson KE, Inge TH, Albanese CT. Laparoscopically assisted anorectal pull-through for high imperforate anus — A new technique. J Pediatr Surg 2000;35:927-30.
9. Fuchs J, Warmann SW, Seitz G, Schäfer J, Schröder M, Obermayr F. Laparoscopically assisted vaginal pull-through for high urogenital sinus: A new surgical technique. Urology 2012;79:1180-3.
10. Rink RC, Pope JC, Kropp BP, Smith ER Jr, Keating MA, Adams MC. Reconstruction of the high urogenital sinus: Early perineal prone approach without division of the rectum. J Urol 1997;158:1293-7.
11. Peña A. Total urogenital mobilization — An easier way to repair cloacas. J Pediatr Surg 1997;32:263-7.
12. Davies MC, Crouch NS, Woodhouse CR, Creighton SM. Congenital adrenal hyperplasia and lower urinary tract symptoms. BJU Int 2005;95:1263-6.
13. Ludwikowski BM, González R. The surgical correction of urogenital sinus in patients with DSD: 15 years after description of total urogenital mobilization in children. Front Pediatr 2013;1:41.
14. Rink RC, Adams MC, Misseri R. A new classification for genital ambiguity and urogenital sinus anomalies. BJU Int 2005;95:638-42.
15. Podevin G, Petit T, Mure PY, Gelas T, Demarche M, Allal H, *et al*. Minimally invasive surgery for anorectal malformation in boys: A multicenter study. J Laparoendosc Adv Surg Tech A 2009;19 Suppl 1:S233-5.
16. Bischoff A, Peña A, Levitt MA. Laparoscopic-assisted PSARP — The advantages of combining both techniques for the treatment of anorectal malformations with recto-bladderneck or high prostatic fistulas. J Pediatr Surg 2013;48:367-71.