Journal of Pediatric Urology (2018) **14**, 198—199

Video Bank

Genitoplasty in newborn females with adrenogenital syndrome: Focus on the reconstruction technique and its outcomes

Marjan Waterloos, Tom Claeys, Maxime Sempels, Erik Van Laecke, Piet Hoebeke, Anne-Françoise Spinoit

University Hospital Ghent, Department of Pediatric Urology, De Pintelaan 185, 9000 Gent, Belgium

Correspondence to: M. Waterloos, University Hospital Ghent, Urology, De Pintelaan 185, Gent, Belgium

marjan.waterloos@hotmail.com (M. Waterloos)

Keywords
Congenital adrenal hyperplasia; Genitoplasty; Adrenogenital syndrome

Received 15 June 2017
Accepted 8 February 2018
Available online 8 March 2018

Summary
The adrenogenital syndrome is an autosomal recessive disorder in which an enzyme defect in the steroid pathway leads to excessive prenatal exposure of androgens. In the female fetus, masculinization of the external genitalia is observed. Surgery aims for functional and aesthetical reconstruction. Many techniques have been described. A video of our modified pull-through reconstruction technique is hereby presented. A retrospective descriptive database was created with patients who underwent genitoplasty for a CAH-associated genital condition. A video demonstrating the reconstructive technique was recorded while operating on a 9-month-old girl. Prior to surgery a cystoscopy is performed to evaluate the length of the urogenital sinus. Surgery starts with creating a reversed U-flap, after which the urogenital sinus is mobilized. The corpora cavernosa are released and the neurovascular bundle is isolated. To create vaginal space the urogenital sinus is subsequently separated. The vaginal introitus is anchored to the perineal skin flap. Labia minora are created by splitting the preputial skin. Finally excessive skin tissue is resected. Twenty-two female patients underwent reconstructive surgery for the adrenogenital syndrome in a tertiary referral centre over 16 years. Median age at surgery was 3 months (0—190). Median follow-up was 36 months (0—108) after surgery. A good functional and aesthetical outcome was observed. The modified pull-through technique, illustrated by this video, provided satisfactory results with a low complication rate. Follow-up until adulthood is needed to evaluate long-term outcomes.

Introduction

The adrenogenital syndrome is a condition caused by congenital adrenal hyperplasia (CAH). CAH is caused by an autosomal recessive disorder in which normal müllerian development takes place but an enzyme defect in the steroid pathway leads to a hormonal deficiency. This deficiency is a 21-hydroxylase deficiency in 90% of the cases, but is variable, thereby explaining the wide spectrum of clinical presentation [1].

It is the most frequent clinical presentation of disorder of sexual development, and concerns 85% of all children presenting with ambiguous genitalia. It is characterized in the female newborn by masculinization of the external genitalia resulting from excessive prenatal exposure to androgens.

Surgery aims for functional and aesthetical reconstruction of the female external genitalia. Several techniques have been reported. Most frequently described vaginoplasty techniques are the inverted U-flap vaginoplasty, pull-through technique, total and partial urogenital mobilization. Clitorectomy is no longer a preferred technique, with nerve-sparing clitoroplasty being the golden standard [2,3].

A video of our modified pull-through reconstruction technique, focusing on anatomic restoration, is hereby presented.

Supplementary video related to this article can be found at https://doi.org/10.1016/j.jpurol.2018.02.011.

Materials and methods

A retrospective descriptive database was created of patients who underwent genitoplasty for a CAH-associated genital condition between 1999 and 2016. Descriptive statistics were applied.

A video demonstrating the reconstructive technique was recorded while operating on a 9-month-old girl.

https://doi.org/10.1016/j.jpurol.2018.02.011

Prior to reconstructive surgery a cystoscopy was performed for evaluation of the urogenital sinus (low or high confluence) and for further reconstruction technique assessment. The modified pull-through technique was applied, with maximal preservation of all genital sensitive tissue.

This technique starts with creating a reversed U-flap to define the future introitus. Subsequently the urethral plate is sacrificed to allow mobilization of the urogenital sinus. To release the corpora cavernosa, clitoris deglovement needs to be performed. The neurovascular bundle is subsequently isolated and the corpora cavernosa are resected or plicated. The urogenital sinus is separated, thereby creating the vaginal space. After further developing the vaginal introitus, it is anchored to the perineal skin flap. The clitoris is fixated in anatomic position. The preputial skin is split to create the labia minora, after which clitoroplasty is further realized. To finalize the procedure the excessive tissue of the labia majora is resected.

The overall result is a female-like presentation of the external genitalia with preservation of all sensitive tissues.

Outcome and complications were evaluated during follow-up consultations by questioning the parents.

An ethics committee approved this study.

Discussion

Surgery was performed in 22 female patients. Median age at surgery was 3 months (0—190). Median follow-up was 36 months (0—108) after surgery.

The modified surgical technique for performing a genitoplasty was described stepwise. A good functional and aesthetical outcome was observed: re-intervention was required in two patients for minor aesthetical procedure (V-Yplasty of mons pubis).

As a result of some controversy regarding use of such techniques, the most recent procedures were performed with a corporal-sparing technique, allowing for a reversible procedure later in life in case of gender dysphoria [4].

All parents of the operated girls in our series reported a satisfactory outcome.

Conclusion

Reconstructive surgery for adrenogenital syndrome was performed in 22 patients in a tertiary referral centre over 16 years. Our technique, illustrated by a video, provided satisfactory results with a low complication rate. However, long-term follow-up, using standardized questionnaires, until adult life, is necessary to evaluate cosmetic and functional outcome in adulthood.

Conflict of interest

None.

Funding

None.

References

[1] Gearhart J, Rink R, Mouriquand P. Pediatric urology. Saunders; 2010.

[2] Wang LC, Poppas DP. Surgical outcomes and complications of reconstructive surgery in the female congenital adrenal hyperplasia patient: what every endocrinologist should know. J Steroid Biochem Mol Biol 2017;165:137—44.

[3] Yankovic F, Cherian A, Steven L, Mathur A, Cuckow P. Current practice in feminizing surgery for congenital adrenal hyperplasia; a specialist survey. J Pediatr Urol 2013;9(6 Part B): 1103—7.

[4] Pippi Salle JL, Braga LP, Macedo N, Rosito N, Bagli D. Corporeal sparing dismembered clitoroplasty: an alternative technique for feminizing genitoplasty. J Urol 2007;178(4 Pt 2):1796—801.