International Journal of Medicine Research
ISSN: 2455-7404; Impact Factor: RJIF 5.42
Received: 19-12-2018; Accepted: 21-01-2019
www.medicinesjournal.com
Volume 4; Issue 1; January 2019; Page No. 69-73

Current management of the feminizing genitoplasty in congenital adrenal hyperplasia with low vaginal confluence

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Abstract
Purpose: Highlighting the importance of evolution of techniques of feminizing genitoplasty in congenital adrenal hyperplasia(CAH) with low vaginal confluence.
introduction: Surgical aims for virilized girls with CAH are to create cosmetic and functional external genitalia from a sexual, reproductive and urinary standpoint.
observation: Patient aged 1 year and 7 months, without specific antecedents, followed for CAH, with karyotype 46, XX, and signs of virilization stage III of Prader. the patient benefited from the posterior based omega-shaped flap vaginoplasty and the clitoroplasty technique of subtunical excision of erectile tissue with preservation of the neurovascular bundle.
Conclusion : Surgical technique for virilized genitalia in CAH has evolved from excisional surgery of the clitoris, to very refined surgery with precise anatomical land- marks to preserve the majority of nerve terminals in reductions clitoroplasty. Several aspects of vaginoplasty and introitoplasty had been developed to improve cosmetic appearance.

Keywords: congenital adrenal hyperplasia, vaginoplasty, clitoroplasty

1. Introduction
Congenital adrenal hyperplasia (CAH) leads to phenotypic changes due to excessive endogenous androgen production during fetal development in genetic females [1].
It is the most common diagnosis in virilized infants with 46, XX DSD.1 Even so it is a rare diagnosis, occurring in approximately 1/5,000 to 20,000 individuals [2].
Treatment of children with classic CAH consists of steroid administration with subsequent androgen suppression, and surgical repair of ambiguous genitalia [3].
Surgical aims for virilized girls with CAH are to create cosmetic and functional external genitalia from a sexual, reproductive and urinary standpoint.Concerns remain as to the potential need for additional procedures, long-term effects on psychosocial and sexual function, and cosmetic and functional outcomes [2].

2. Observation
Patient aged 1 year and 7 months, without specific antecedents, followed for CAH, diagnosed in post-natal period on endocrinological evaluation. She ‘s under Hydrocortisone and fludrocortisone.
The patient has a karyotype 46, XX, the examination of the external genitalia finds signs of virilization stage III of Prader. (Figure 1)
Pelvic ultrasound found female type internal genitalia with bilateral adrenal hypertrophy.
A genitography allowed opacification of the uro-genital sinus which is located low, with evidence of a posterior oblong cavity corresponding to the vaginal cavity and opacification of the bladder.
An endoscopy showed low vaginal confluence (figure 2)

Fig 1: Girl virilized stage III of Prader

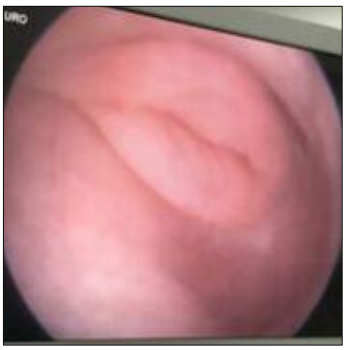

Fig 2 : Endoscopy confirmed the low vaginal confluence

After the informed consent of the parents by explaining the genital feminizing technique and the functional and aesthetic consequences, the patient benefited from the posterior-based omega-shaped flap vaginoplasty figure (3, 4,5,6).

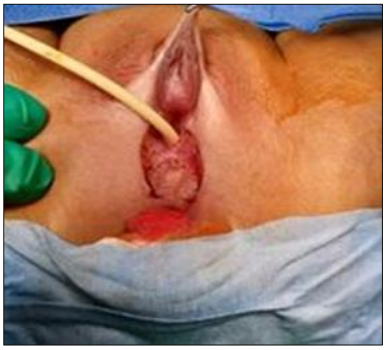

Fig 3: Construction of a posterior-based omega-shaped flap

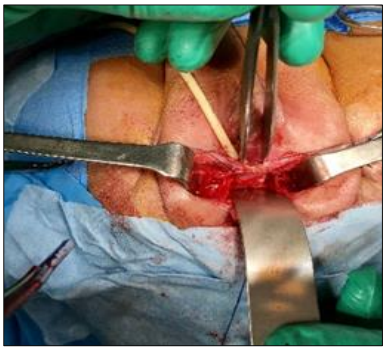

Fig 4: opening of the urogenital sinus and identification of the vagina

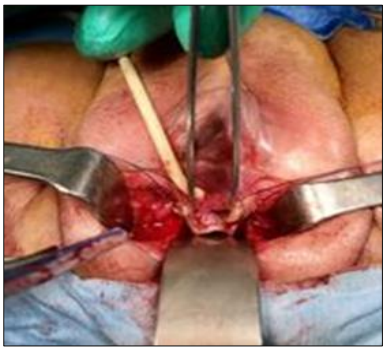

Fig 5: opening the vagina

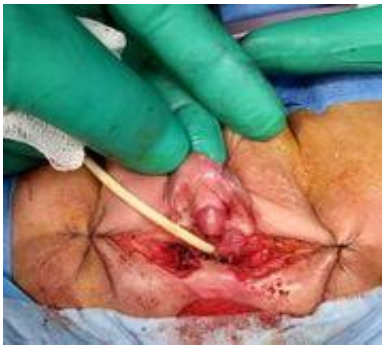

Fig 6: anastomosis with posterior-based omega-shaped flap

And the clitoroplasty technique of subtunical excision (figure 7,8) of erectile tissue with preservation of the neurovascular bundle. (The surgical technique is shown in Figs)

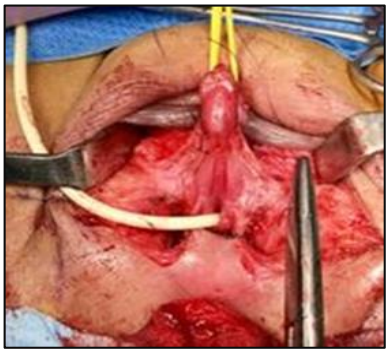

Fig 7: Two ventral incisions on the Buck ' s fascia at 5 and 7o ' clock., the ventral skin strip is well preserved

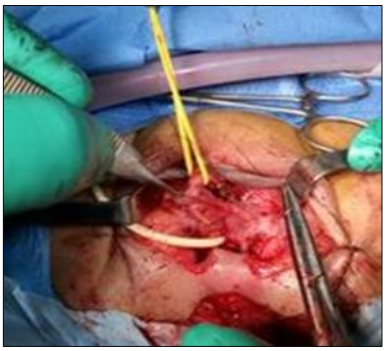

Fig 8: subtunical excision of erectile tissue with neurovascular preservation of the clitoris

The patient is kept hospitalized for 10 days keeping the foley catheter. The aesthetic consequences were satisfactory (figure 9)

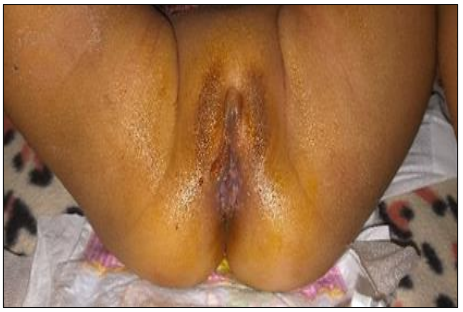

Fig 9: The appearance after feminizing genitoplasty

3. Discussion

The Congenital adrenal hyperplasia (CAH) diagnosis should be established as soon as possible to permit early gender reassignment and steroid replacement therapy as well as a discussion on the surgical correction of ambiguous genitalia. In children, medical treatment involves the administration of hydrocortisone, 10 – 18 mg / m2, divided into 3 daily doses. Fludrocortisone must be given to patients with the salt-wasting form as well as to those with the simple virilizing form in whom mineralocorticoids are slightly deficient [3].

The primary goals of feminizing genital reconstruction are to create a normal-looking, sensate clitoris, provide an adequately sized and appropriately situated vagina, and create normal-appearing female external genitalia [4].

Due to the complex nature of this condition, the best surgical management and timing of intervention remain the subject of controversy [5].

The Current practice includes corrective genital surgery to separate the labia, reduce the size of the clitoris and separate the vagina and urethra. The surgery is usually a 'one-stage' procedure in infancy, although many patients require further surgery in adolescence to facilitate menstrual flow and allow penetrative sexual intercourse. There are concerns with various aspects of this management, including the necessity of the procedure, timing of surgery, and long-term outcomes for sexual function and cosmesis [6].

Feminizing genital reconstruction in CAH with low vaginal confluence

3.1 Vaginoplasty

Patients with low vaginal confluence should have their "complete repair including vaginoplasty, perineal reconstruction, and clitoroplasty be done simultaneously at an early age" [7].

In the low form the virilization is corrected by clitoroplasty associated with a'cutback' vaginoplasty [8].

In the 'cutback' vaginoplasty the flap-vaginoplasty described by Fortunoff *et al.* is usually used, where an inverted U-flap is sutured onto the urogenital sinus previously opened in its ventral aspect. Although such a flap is conveniently vascularized, its wide base allows no imbrication of the lateral flaps that are to form the labia majora, leading to rather inadequate cosmesis [8].

The Fortunoff flap does not reproduce normal female anatomy, but shortens the perineum, conferring an abnormal appearance, brings hair baring skin into the vagina and precludes mucosal lining of the posterior vaginal fourchette.

When used primarily by itself in cases of low urogenital sinus confluence this approach allows perineal access to the vagina, and the flap successfully widens a narrow vaginal introitus [9].

In the posterior-based omega-shaped flap vaginoplasty the labia majora may be overlapped at the base of the omega (Fig. 9), thus consistently producing better cosmesis of the external genitalia. To maintain flap vitality it is important that a broad base of subcutaneous tissue be spared.

This allows for the construction of longer and larger flaps, thus avoiding the common stenosis found at the anastomosis site between the vagina and the flap [8].

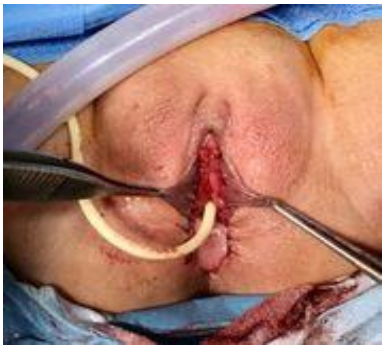

Fig 9: The labia majora may be overlapped at the base of the omega

3.2 Clitoroplasty

Surgical technique for virilized genitalia in CAH has evolved from excisional surgery of the clitoris, the most innervated organ in the human body, to very refined surgery with precise anatomical land- marks to preserve the majority of nerve terminals in reductions clitoroplasty [10].

Accordingly, surgery that either ablates the glans altogether or does not preserve its nerve supply is not acceptable.

When the phallus is small, either clitoral resection or plication techniques may produce a satisfactory result. However, any procedure that does not remove the erectile tissue leaves the possibility of later enlargement with the problems of an unacceptable cosmetic appearance and possible painful engorgement during sexual stimulation [11].

Resection of the erectile tissue with careful preservation of the nerve supply to the glans appears to be the most reasonable procedure. To spare the parents the offending presence of large phallic organ in a child who they have been assured is a little girl, we advocate performing this clitoral surgery as soon as the child's medical condition has stabilized. We have found no difficulty when the operation was done in the first few months of life [11].

Young performed the first clitoroplasty for correction of ambiguous genitalia in the late 1930s. Up until the 1970 s, clitoral amputation was considered as the procedure of choice for clitoral hypertrophy, based on the belief that the clitoris was not necessary for normal sexual function [3].

Over time, with a better understanding of the fundamental role of the clitoris for female sexual function, clitoral amputation was abandoned and replaced by more conservative procedures such as clitoral plication, concealment, recession and reduction. Clitoral recession or plication consists of burying the clitoris under the skin by fixating it to the pubic bone with nonabsorbable sutures, without damaging the neurovascular bundle. Despite initial satisfactory cosmetic results, longterm follow-up studies have shown that those patients experienced painful erections upon sexual arousal due to engorgement of the clitoral corporal bodies. To overcome such complications, Schmid was the first to report the excision of corporal tissue while preserving the glans and neurovascular bundle [3].

Since then, all clitoroplasty techniques involving resection of corporal tissue have been based on Schmid 's preservation of the neurovascular bundle. Glans devascularization was one of the most severe complications related to these techniques. In 1983, Kogan *et al*. reported on a subtunical excision of erectile tissue by incising laterally through Buck 's fascia. Preservation of the neurovascular bundle with maintenance of a good blood supply to the glans was obtained on a consistent basis by keeping most neurovascular fibers attached to the tunica, decreasing the risk of damage.In 1995, Gearhart *et al*. showed that neural conduction was preserved in the dorsal neurovascular bundle, but a correlation between their fi ndings and a normal adult sexual life could not yet be established. Baskin *et al*. further clarified the anatomy of the clitoris, stating that a ventral incision on the clitoral corporal body would not only preserve the neurovascular bundle but also neural branches that fan out laterally [3].

The ventral skin strip also is well preserved and contributes to the blood supply to the glans clitoris [11].

More recently, Poppas *et al*. modified the clitoroplasty technique of subtunical excision of erectile tissue by performing 2 ventral incisions on the Buck ' s fascia at 5 and 7o ' clock.

These authors believe that the great majority of neural fibers are maintained, resulting in better preservation of erectile and sensitive function, it is essential to perform the dissection in the exact plane (below the second layer of the Buck ' s fascia, just above the tunica albuginea), in order to avoid injury to

the neurovascular bundle [3].

If the correct plane is entered, dissection is rather easy and safe. they have not had any case of glans ischemia in their feminizing genitoplasty series, probably due to the proper dissection technique mentioned above. Although important progress has been made in the field of clitoroplasty [3].

With that in mind, Pippi Salle *et al*. described an alternative technique to standard clitoroplasty, which spares all clitoral components. The neurovascular bundle and glans are dissected off the corpora cavernosum following the technical principles described above. The 2 corporal bodies are then completely separated in the midline, rotated downward, and hidden in pouches constructed in the labia majora. Preliminary results were encouraging, making this technique an attractive option due to the possibility of preserving genital sensitivity as well as the reversibility of the procedure in cases of gender dysphoria [3].

Although clitoroplasty with excision of part of the erectile tissue is still the most commonly used surgical technique to reconstruct the clitoris to date, it is important to bear in mind that this operation has its own risks, such as glans necrosis, impairment of clitoral sensitivity as well as harm to sexual function. Therefore, discussion of these issues with parents when contemplating surgical correction of genital ambiguity is imperative [3].

3.3 Timing of Surgery

Controversies exist on the optimal timing, indications and techniques for feminizing genitoplasty procedures. Opponents of early surgery cite concerns regarding the frequent need for further reconstructive procedures, the lack of multiinstitutional long-term data on function and cosmesis in adulthood, and the inability to obtain informed consent of the patient in infancy. Advocates of early surgery argue that there may be psychosocial benefits for patient and parent when there is consistency of anatomy with gender of rearing, potentially better quality of genital tissue with exposure to maternal estrogens in infancy and the relatively minor procedures anticipated later in life compared to initial reconstruction [2].

Knowledge of practice patterns remains limited to physician surveys and retrospective series from high volume centers. At the Fourth World Congress of the International Society of Hypospadias and Disorders of Sex Development 48 of the 61 surgeons (78%) surveyed preferred to perform feminizing genitoplasty before age 2 years.5 A study from the largest DSD referral center in the United Kingdom between 2001 and 2012 showed a minimal decrease in clitoroplasty or overall surgical reconstruction rates in infancy or early childhood.6 This study was encouraging since it demonstrated improved cosmetic outcomes and a decreased need for secondary procedures [2].

The age at which surgery is most appropriate is still not proven. There are, however, a considerable number of experts who support the following statements:

- The availability and quality of tissue is much better in the first 6 months of life;
- Pediatric surgeons are well-prepared for surgery on infants;
- Early surgery creates less uncertainty for parents, a more secure parent-child relationship and more stable developmental options for the child [12].

3.4 Sexual Function and Genital Sensitivity Following feminizing Genitoplasty for Congenital Adrenal Hyperplasia

Many controversies persist regarding clitoral sensitivity and viability following commonly used techniques. Sexual function has not been fully evaluated in adult CAH women who had clitoral surgery early in life. Expectations are high and rely on the fact that new techniques may improve the results obtained with outdated procedures [3].

Informed consent should be based not just on the technical aspects of surgery and risks, but on a developed understanding and appreciation of potential implications for future sexual lives [13].

The family must be informed that clitoroplasty carries potential risks of impaired sensitivity and procedures for a narrow vagina are often required before intercourse [14].

It is sensitive to avoid patient stigmatization, be clear with surgical expectations, avoid repeated pelvic examinations when not crucially needed, especially throughout toddler and child development, offer patients and family attention with a multidisciplinary team with special emphasis in psychological support and encourage participation in self-help groups [10].

4. Conclusion

Surgical technique for virilized genitalia in CAH has evolved from excisional surgery of the clitoris, the most innervated organ in the human body, to very refined surgery with precise anatomical land- marks to preserve the majority of nerve terminals in reductions clitoroplasty. Several aspects of vaginoplasty and introitoplasty had been developed to improve cosmetic appearance and avoid vaginal and introital stenosis.

We have little information on the results at long term of this surgery.

A regular follow-up to assess for long-term complications is necessary and it is vital to inform families about the different management options with all the risk and benefits of the genital feminizing reconstruction.

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