Feminizing Genitoplasty For CAH: At Which Age??

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Introduction

- Reconstructive surgery for girls with CAH is usually performed for the following reasons:

1. To improve the cosmetic appearance of the genitals
2. To allow for vaginal-penile intercourse
3. To achieve an unobstructed, sex-typical manner for urination
Debate:??

➢ the age at which surgery is best conducted,?
➢ which type of procedure offers the best outcome? And
➢ whether surgery should be performed in all patients

EARLY???

Advocates of surgery in infancy, maintain that:

☑ the procedures are easier to perform
☑ the results are better in the young child
☑ the stigmatization of living with genital ambiguity is reduced for the patient and family.
LATER???

Those favoring later reconstructive surgery note that:

- the decision more appropriately rests with the patient rather than the family
- the surgical options for genital reconstruction will be preserved in the event that the initial sex assignment was incorrect and a sex change is desired, or the patient does not desire genitoplasty

Remember:

- Many adult patients who have undergone reconstructive surgery are dissatisfied with the outcome because of reduced sexual satisfaction and function
- There is a lack of data to provide adequate guidance as to the best timing and surgical approach.
Also,

- No evidence indicating that either early or late surgery better preserves sexual function.

- Choice of surgical procedure is dependent on the specific anatomy, which differs substantially between patients with CAH.

Anatomy:
Feminizing genitoplasty

Feminizing genital surgery comprises:

1. Clitoroplasty
2. Vaginoplasty
3. Labioplasty

Clitoral surgery; techniques

- Subtunical removal of erectile (corporal) tissue ventrally,
- Sparing the glans with its neurovascular supply
- In an attempt to avoid ablative surgery, a corporal-sparing clitoroplasty with the goal of preserving all sensation-bearing structures
- Glans reduction can be done either by wedge resection or by de-epithelization; the latter may cause less disruption to sensation
Another technique
Clitoral surgery; timing

- surgery reduces innervation of the clitoris?.... Debate
- Adult women who have undergone clitoral surgery in infancy report:
  ✓ reduced sexual sensation?
  ✓ poorer sexual function when compared to normal controls and also to women with clitoromegaly who had not undergone surgery

Clitoral surgery; timing

- Sexual pleasure is the only role for the clitoris
- Surgery may have effects on sexual response

Because of these concerns the Chicago consensus and subsequently the Endocrine Society guidelines on the management of CAH both recommend that:
- clitoral surgery should not be performed in girls with mild degrees of virilization
- However in the case of moderate and severe virilization, both guidelines currently recommend that feminizing surgery including clitoral reduction
Clitoral surgery; timing

• Supporters of surgery in early childhood cite normalization of the appearance of the external genitalia in accordance to the assigned gender.
• No studies have linked early genital surgery to successful gender outcome.
• Girls with virilized genitalia left intact may suffer unwarranted social interactions with their peers (e.g. the locker room time at school), leading to embarrassment and social withdrawal.

Clitoral surgery; timing

• Furthermore, the idea of “doing nothing” sometimes represents a stressful concept for some parents who have difficulties coping with the appearance of their child’s genitalia.
• Lastly, despite the limited experience, feminizing genitoplasty in postpubertal girls is perceived by some surgeons as being technically more demanding.
Vaginoplasty: timing

❖ Since the vagina has no function in early childhood, vaginoplasty could be postponed until after puberty.
❖ Advantages of such approach would be the inclusion of the patient in the consent process as well as the ability to comply with post-operative vaginal dilation.
• In addition, manipulation of the vagina may be easier in the setting of higher estrogen levels.
• *On the other hand*, not performing vaginoplasty at the time of clitoroplasty precludes the use of the mucosa from the urogenital sinus and excessive prepuce that would be discarded and therefore no longer available for reconstruction of the distal vagina.
• This may be of importance in cases of high urogenital sinus (UGS) or a very small vagina.
**ETHICAL ISSUES: CONSENT, \**

**TIMING OF SURGERY**

- A newborn infant with ambiguous genitalia cannot contribute to decisions relating to gender assignment, nor provide informed consent for any genital surgery undertaken in infancy or childhood.
- Responsibility for these difficult decisions lay with the parents, in the light of advice and information received from the doctors caring for their child.
- Debate?? Now ??
• However, some patient groups and ethicists assert that responsibility for this decision belongs, as a right, to the affected individual.

• So, surgery should be deferred until such an age when the individual can make informed decisions.

• However, it is extremely difficult to contemplate rearing a child against a background of such uncertainty.
• Likewise the prospect of leaving a severely virilized genital phenotype uncorrected in an otherwise ‘normal’ female ‘i.e. a little girl with a penis’ would be unacceptable to most parents

• An impact on psychological development and psychosocial adjustment of rearing girls with an uncorrected male genital phenotype.
The website of the Adrenal Hyperplasia Network lists as:

- An abuse of human rights ‘**genital surgery performed without informed consent of patient done as a child for more sociological acceptance than real medical need**’

And

- condemns ‘cosmetic surgery performed on infants with no evidence that the surgery is necessary and evidence that it could be damaging
• Also, ‘clitoral surgery should only be done with informed consent of the patient, not at wishes of parents or medics for social reasons as it might cause major damage to sensate function’.

• Putting aside the ethical question of the rights of the child vs the parents there is a growing body of evidence that the website statement relating to the outcome of clitoral surgery is correct.

Conclusion

• Regardless of sex of rearing, many girls with CAH, at some point, require genital reconstructive surgery.
• Should only be performed by surgeons with specific expertise working within the context of a specialist multidisciplinary team, there is
• ongoing debate as to both
  ➢ the timing of surgery and
  ➢ which procedure should be chosen.
Conclusion

• We should advise patients and families.

• Non-surgical clinicians would benefit from an understanding of the indications for surgery as well the general principles and outcomes of the most commonly used procedures.

• Many factors will determine the procedure chosen but the final decision as to technique will lie with the operating surgeon alone.

Conclusion

• need for long-term functional and psychosexual outcome studies to formulate a more evidence-based approach.

• Aggressive and unnecessary early surgery is unjustifiable.

• leaving some girls with an obviously uncorrected male genital phenotype, is likely to prove unacceptable to most parents.
Our data in Alexandria University:

### Age Distribution Among Studied Children

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### Distribution of Complications Among Studied Age Groups

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Finally:

- When to do?
  Young or adult

Thanks