

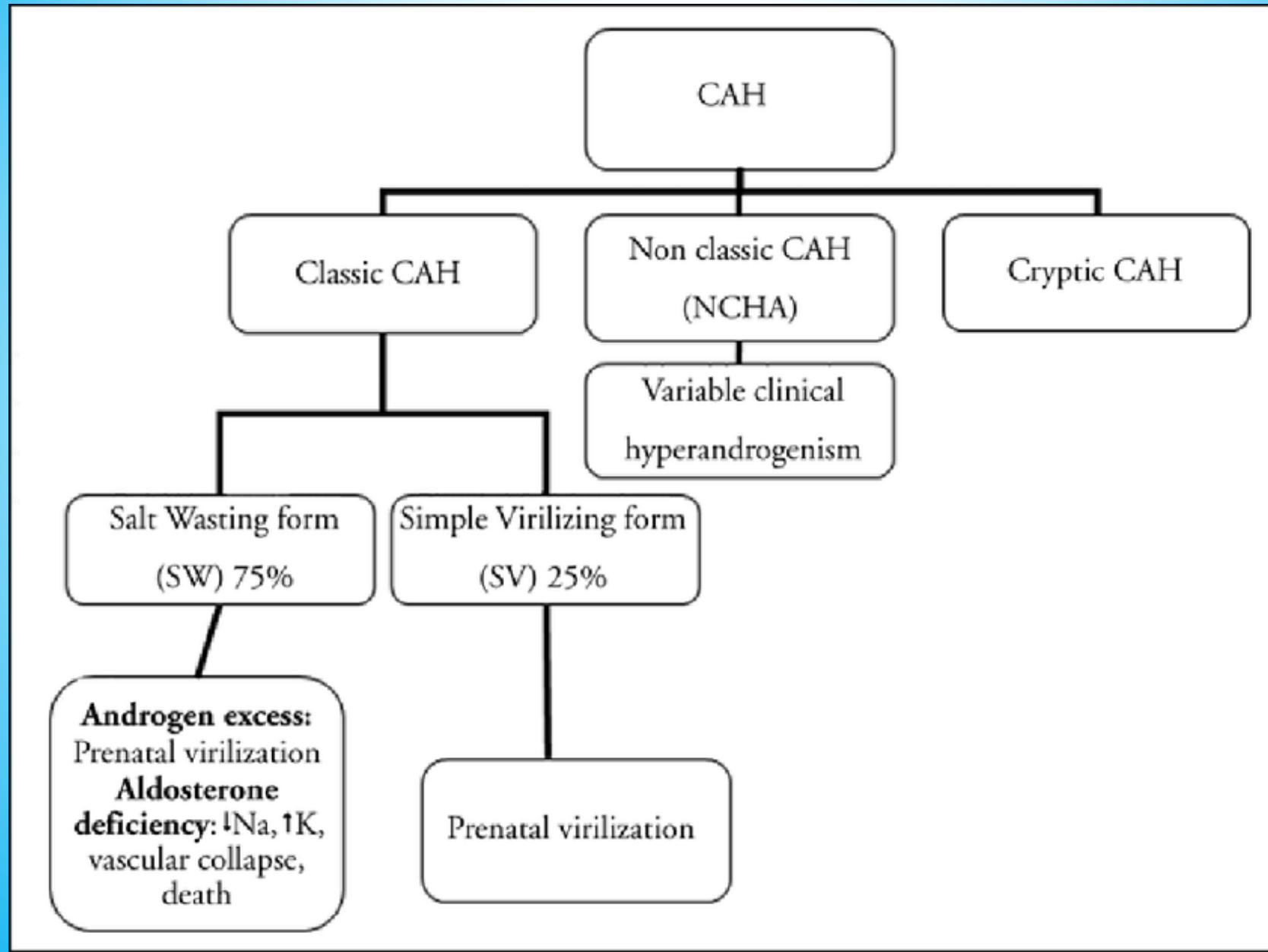
CONGENITAL ADRENAL HYPERPLASIA: REVIEW FROM A SURGEON'S PERSPECTIVE

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INTRODUCTION

- ▶ Condition is characterized by impaired cortisol synthesis
- ▶ Most commonly due to 21-hydroxylase deficiency-95%
- ▶ It is the most common type of disorder of sex development(DSD) for XX karyotype
- ▶ Treatment with cortisol replacement became available in the 1950s
- ▶ Enzyme deficiency leads to a block in cortisol synthesis followed by a build-up of cortisol precursors which, in turn, are diverted to androgen synthesis
- ▶ The increased androgen concentration triggers a variable degree of virilisation in female newborns

CLASSIFICATION OF CAH



CLASSIFICATION- PRADER SCALE



No virilization



Stage 1



Stage 2



Stage 3



Stage 4



Stage 5



MANAGEMENT

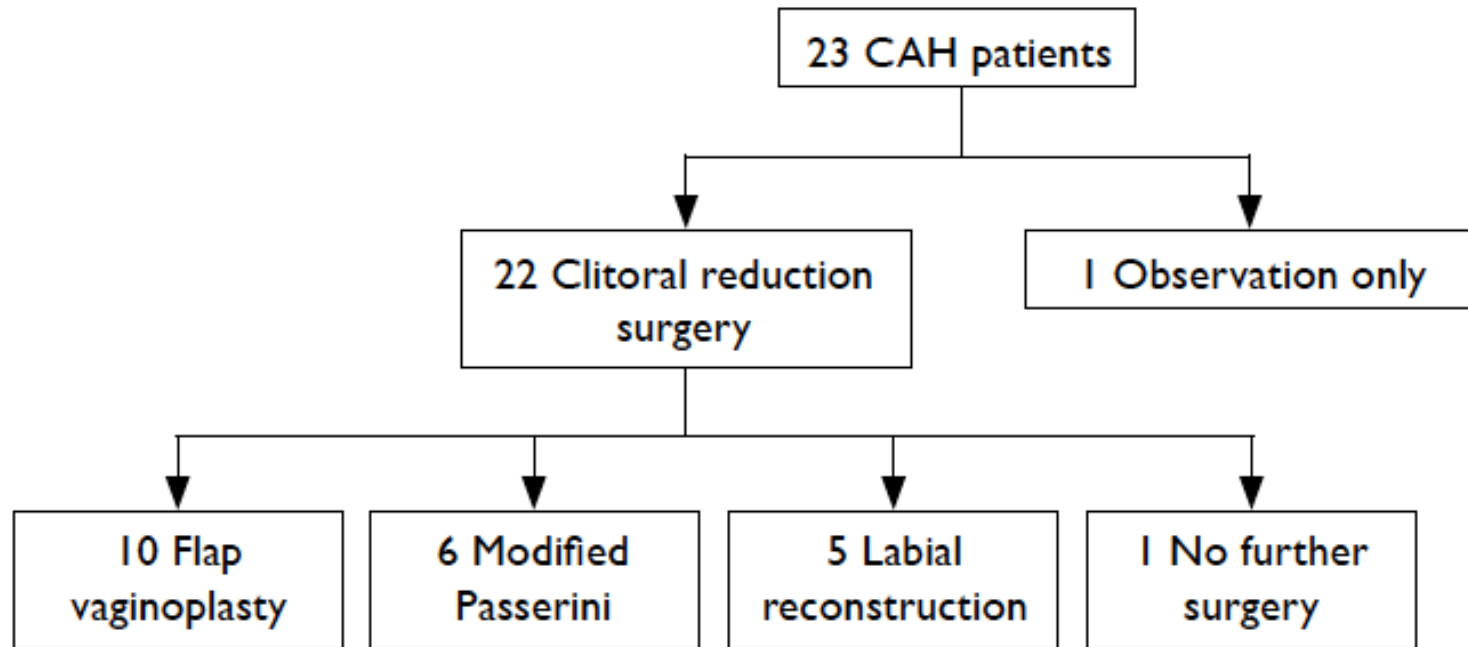


FIG. Surgical management of all patients with congenital adrenal hyperplasia (CAH)

SURGERY

CLITOROPLASTY

(Prader scale 3-5)

- Clitoridectomy (amputation)
- Corporal sparing techniques (clitoral recession)
- Clitoral reduction (clitoroplasty)

VAGINOPLASTY

(Prader 3-5 and UGS)

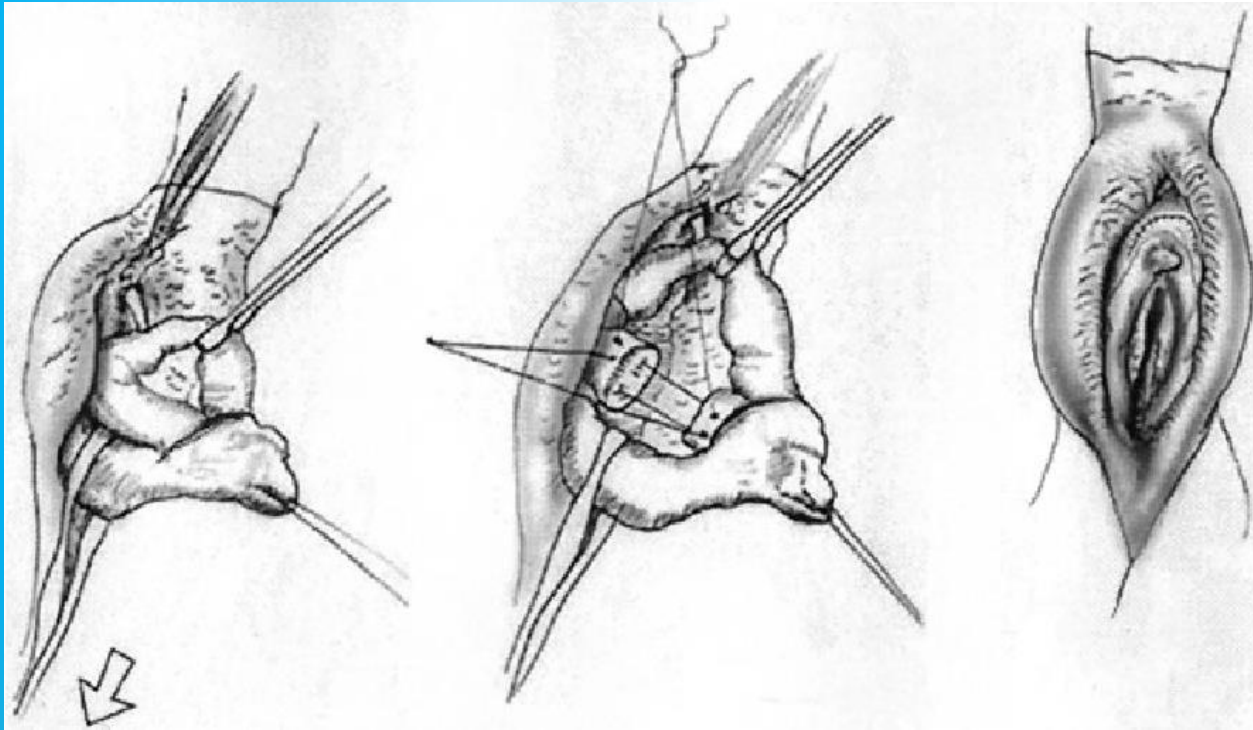
- Flap vaginoplasty (U/ Fortunoff And Passerine-glazel)
- Urogenital sinus mobilization (PUM,TUM)
- Vaginal pull through (ASTRA)

LABIAPLASTY/ PERINIOPLASTY

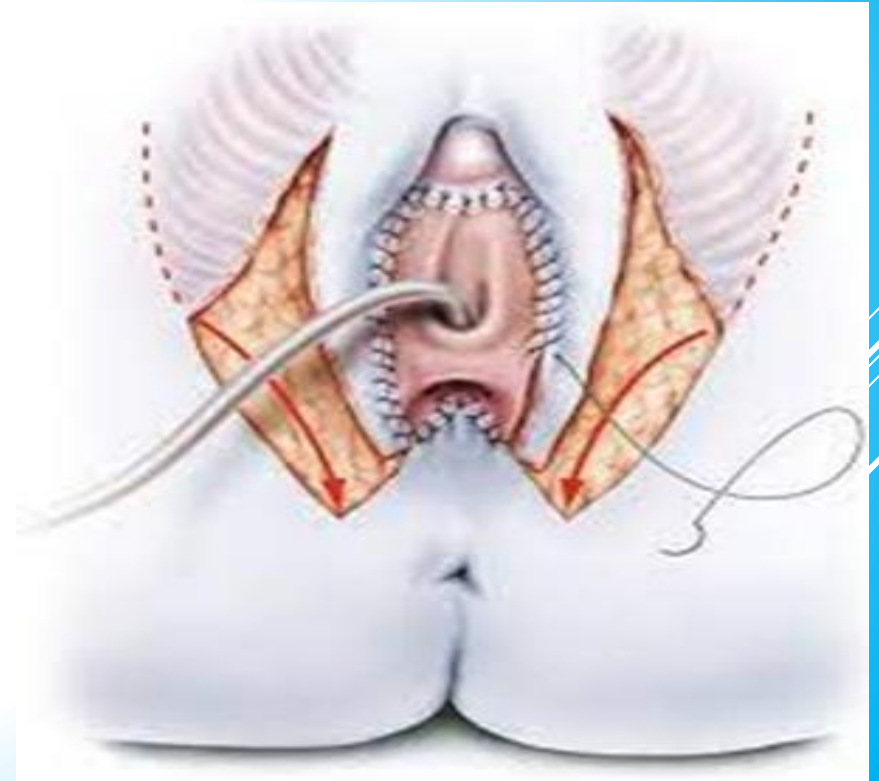
- Skin from genital tubercle-labia minora
- Rugated labioscrotal fold-trimmed and mobilized-labia majora

SURGICAL DETAILS

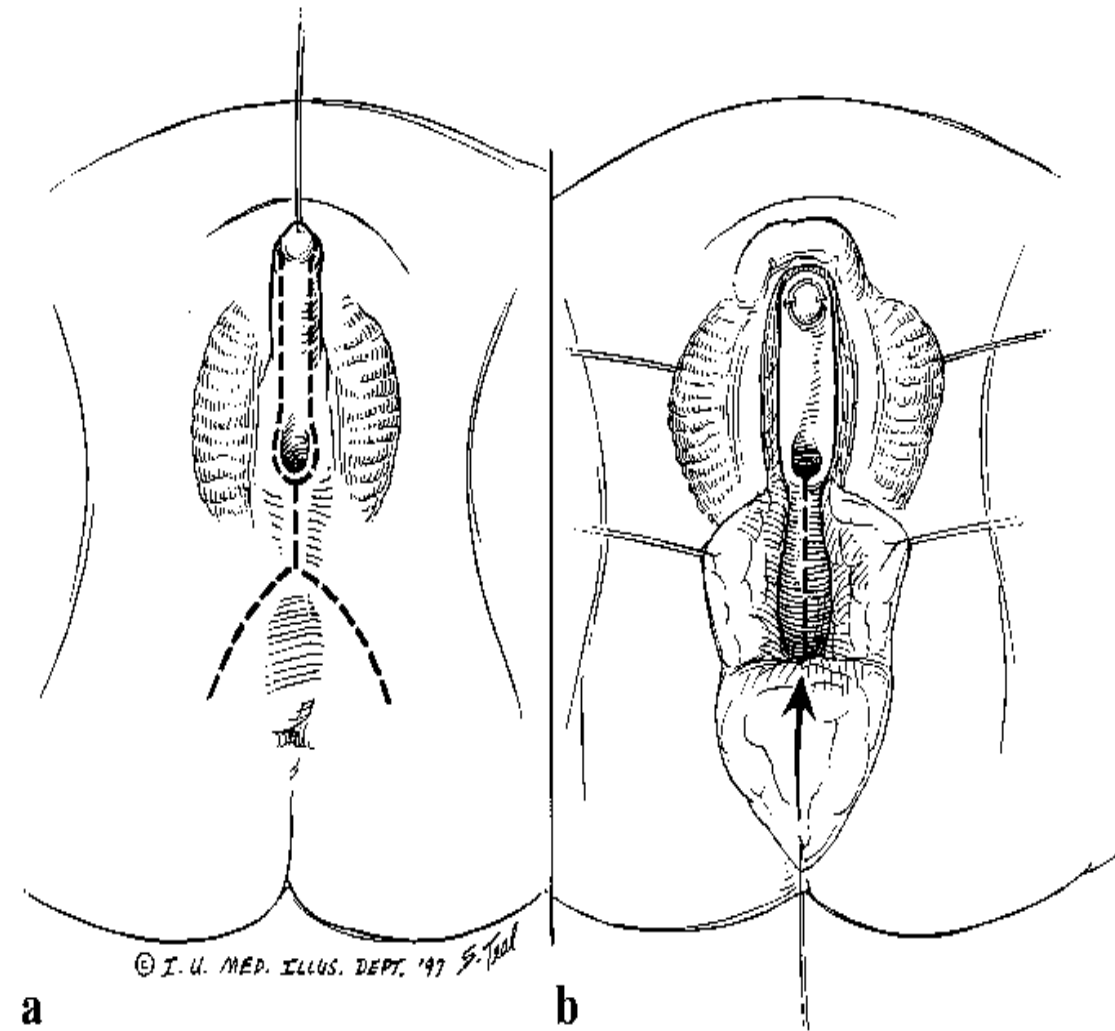
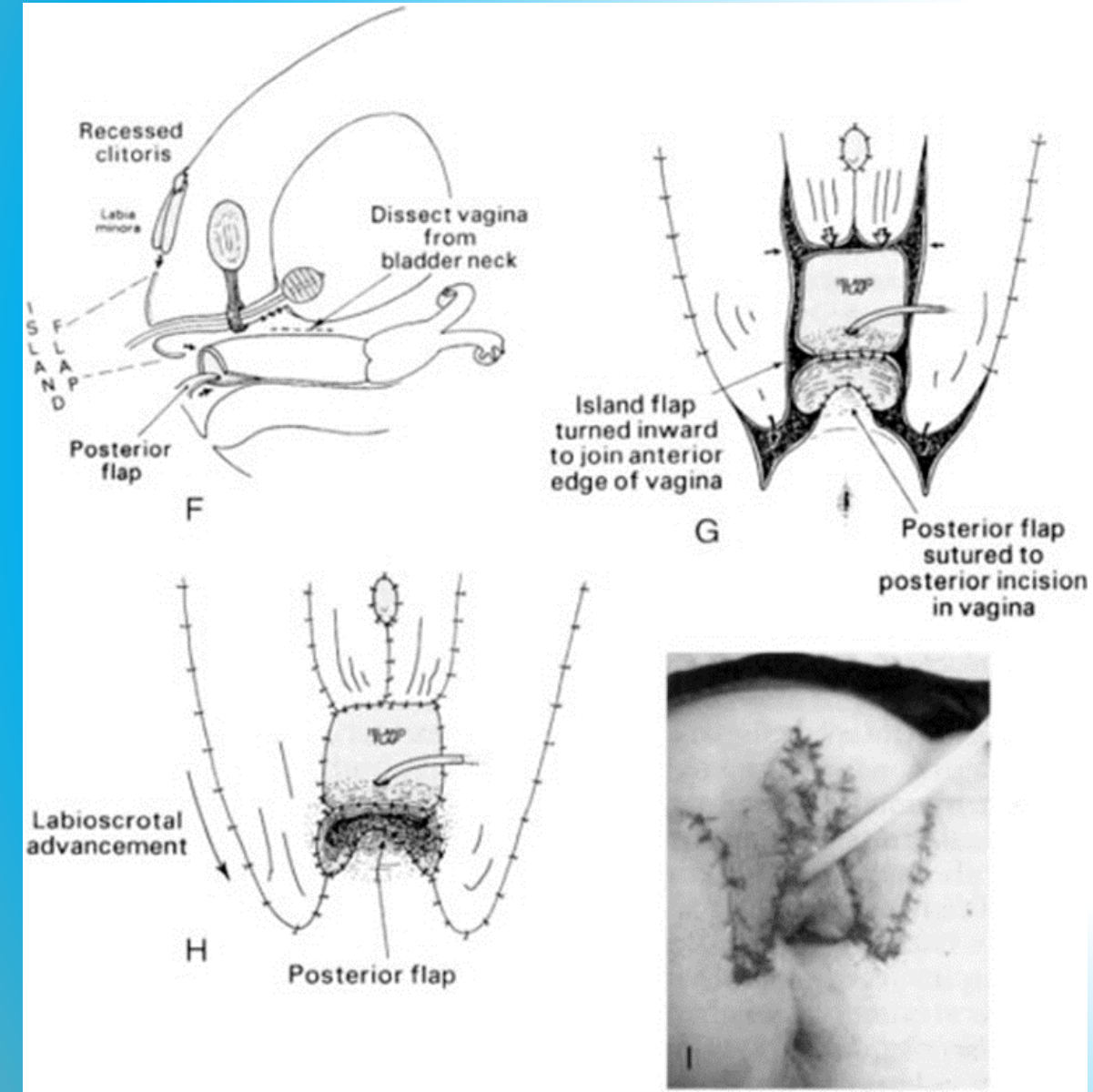
CLITOROPLASTY



LABIOPLASTY



VAGINOPLASTY



OUR CASES

NO.	AGE	PRADER	TYPE	SURGERY	OUTCOME	F/U
1	1.5yrs	III	Salt wasting	Single stage	Satisfactory	On meds
2	3yrs	IV	Salt wasting	Single stage	Satisfactory	On meds
3	2.5yrs	IV	Salt wasting	Single stage	Satisfactory	On meds
4	4yrs	III	Simple virilising	Clitoroplasty +labiaplasty	Satisfactory	-
5	11mths	V	Salt wasting	Single stage	Satisfactory	On meds
6	3.5yrs	IV	Non classic	Single stage	Satisfactory	-
7	2.5yrs	V	Salt wasting	Single stage	On vaginal dilatation	On meds



PREOP



POSTOP

POST OP OUTCOME



CURRENT SURGICAL RECOMMENDATIONS

The attachment of the clitoris to the pubic arch should not be disrupted

Dorsal neurovascular bundles should be preserved. There may be significant lateral branches along the corporal tissue

The neurovascular bundles branch distally in a fan fashion toward the glands clitoris

Reduction of the clitoral erectile tissue is better achieved with a ventral incision along the urethral plate

The urethra and vagina should not be separated

Dissection of the bladder from the uterus below the bladder neck should be avoided

Using labial and clitoral skin to cover the new introitus is more physiological than rotational skin flaps from the buttocks and perineum

OUTCOME

- ▶ There is controversy on functional outcome of clitoral surgery despite using modern techniques
- ▶ Genital sensation have been altered by surgery, but sexual outcome, and patient satisfaction may not be impacted
- ▶ Flap vaginoplasty performed at young age will need future minor or major surgical revision.
- ▶ Passerini-Glazel Genitoplasty or labioscrotal flaps developed frequent stenosis
- ▶ En-block urogenital mobilization with the use of urogenital or skin flaps are promising, with excellent short term and medium results
- ▶ Two stage feminizing genitoplasty, delaying vaginal reconstruction until or after puberty seems to have excellent functional outcome

BRAIN IMPRINTING

The brain organization theory :

- ▶ The fetal brain develops in traditional male direction through a direct action of androgens on the developing nerve cells
- ▶ In the traditional female direction in the absence of androgens
- ▶ This indicates that sexual development, including sexual orientation, are programmed into our brain structures prenatally
- ▶ Androgen levels affecting the masculinization of the brain may also vary, leading to individual differences

SOCIAL AND PSYCHOLOGICAL GENDER

- ▶ Prenatal elevated androgen levels have shown to influence children's sex-typical play behaviors
- ▶ Females with CAH have shown to demonstrate more male-typical rough and “tomboyish” behavior
- ▶ Biological and social factors seem responsible for the development of gender identity disorder
- ▶ Peer pressure and expectations from parents affect the psychology of the individuals markedly

IS NO SURGERY AN OPTION?

- ▶ One stage genitoplasty at an early age will render in a normal looking and innervated genitalia
- ▶ It will function normally in the future **if hormonal substitution and continued medical attention is carried out through-life**
- ▶ When counseling parents of these patients we should clearly state that we think **rearing kids with sex-congruent looking external genitalia is more reasonable than leaving them untouched until puberty to avoid physiological trauma**

CONCLUSION

- ▶ Controversies exist regarding optimal timing, indications and techniques for feminizing genitoplasty procedures
- ▶ Female gender assignment is the only option to maintain fertility in individual
- ▶ 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 as well as cosmetic and functional outcomes.
- ▶ To promote shared decision making (SDM)

THANK YOU