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K. B. Ashok

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Correspondence concerning this article should be addressed to Dr. KB Ashok; Midnapore Medical College and Hospital, India / Email: krsnasok@yahoo.co.in

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"Sex" is gift of nature. What if she betrays?

KB Ashok *

Midnapore Medical College and Hospital, India

* *Corresponding author*

ABSTRACT

Introduction: Intersex is a condition of abnormal sexual maturation resulting in abnormal karyotype or primary or secondary sexual characteristics which can be diagnosed by ambiguous genitalia or abnormal characteristics in adolescent. Prevalence of this rare condition is too low (1 in 20,000 live births). It is very confusing for a doctor to determine the treatment chapter which includes proper diagnostic technique, sufficient time for interpretation and intervention and informing the patient. But the steps are too confusing and controversial. Prescribed case made us highly confused by its characteristics which was none of 5-alpha reductage deficiency syndrome, Reifensteine Syndrome, Pseudo-harmaphodism and also having a balance between a male and female which is very rare and unique. As the diagnosis was difficult to define, initiatives were taken to reveal the proper diagnosis for better treatment so that this kind of cases in future could be managed properly.

Case Presentation/Description: 15 years old unmarried girl came to Gynaecology outpatient department complaining of primary amenorrhoea, gradual swelling of the external genitalia since 12 years of age and hoarseness of voice since last month.
Physically she was tall, with male pattern pubic hair, tanner stage 0 sized breast, having cliteromegaly rather micropenis, perineal hypospadius, palpable mass in both labia major and a blunt space resembling vagina in between.
Laboratory investigations revealed very high Testosterone (246ng/dl) and Thyroid Stimulating Hormone (8.09 mcIU/L) but low level of Follicle Stimulating Hormone (13.83IU/L) and Oestrogen (17pg/dl).
Cytogenic study showed 46XY/46XX mosaic chromosomal pattern.
In laparoscopy there was no internal female genital organ.
Biopsy of the labial mass was done after operation which depicted seminiferous tubules lined by sertoli cells with no sperm production and interstitial fibrosis.
Case was very much mysterious as it was in a balance to support both sexes. After informing the family members first, the society had become an obstruction for the doctors to treat with freedom. Then according to the will of the family members and for the sake of the society diagnosis was hidden from patient and managed by Gonadectomy, Clitorectomy, and Vaginoplasty in same sitting with hormonal therapy for life long to make her a "girl" again.
After 2 months of the treatment patient came with a good respond towards the hormone therapy. Our case proved that 'Intersex' is a mystery of nature and do not always follow the known clinical features.

Discussion: Sometimes we treat the patient for sake of society, family etc. and never think about the patient's will. This case was not a different in this aspect. Doctors were bound to follow the rules made by society without protest. Patient of INTERSEX is one of them who have nothing to do against the Nature's betray. More they need physical rehabilitation, ramification, adaptation and a helping hand to come out of gasping agony inside.

Keywords: Intersex, Pseudo-harmaphodism, sex disorder

Introduction

Intersex is a condition of abnormal sexual maturation resulting in abnormal karyotype or primary or secondary sexual characteristics which can be diagnosed by ambiguous genitalia or abnormal characteristics during adolescent period. Prevalence of this rare condition is too low (1 in 2,000 live births) and that is only on visibility of ambiguous genitalia.[1] If the victims who are revealing their identity in adolescence would be counted then the prevalence becomes higher. It is very confusing for a physician to determine the treatment chapters for this particular disorder which includes proper diagnostic technique, sufficient time for interpretation and intervention. The most difficult job for a doctor comes when the time says to inform the patient and their family about the actual identity of the person. On the other hand these steps are too confusing and controversial worldwide. Prescribed case made us highly confused as it nowhere was matched with the already diagnosed cases available in literatures and publications and the controversial recommendation of the different physicians and surgeons on the management procedure. As the diagnosis was difficult to define, initiatives were taken to reveal the proper diagnosis for better treatment so that this kind of cases in future could be managed properly.

Case Presentation/Description

15 years old unmarried girl came to Gynaecology outpatient department complaining of primary amenorrhoea, gradual swelling of the external genitalia since 12 years of age and hoarseness of voice since last month. There was a history of treatment taken for pharyngeal infection from local quack.

Physically she was tall, with male pattern pubic hair, tanner stage 0 sized breast, having cliteromegaly rather micro-penis of 1.8 cm. length, perineal hypospadius, palpable mass in both labia major and a blunt space resembling vagina in between.

Laboratory investigations revealed very high Testosterone (246ng/dl) [normal in female 20 – 80ng/dl] and Thyroid Stimulating Hormone (8.09 mcIU/L) [normal 0.35-5.50 mcIU/L] but low level of Follicle Stimulating Hormone (13.83IU/L) [normal 2.52-10.2 IU/L] and Oestrogen (17pg/dl) [normal 2.8-29.2ng/dl].

Cytogenic study showed 46XY/46XX mosaic chromosomal pattern.

In laparoscopy there was no internal female genital organs i.e. complete absence of mulerian duct.

Biopsy of the labial mass was done after operation which depicted seminiferous tubules lined by sertoli cells with no sperm production and interstitial fibrosis.

Case was very much mysterious by its characteristics which were none of 5-alpha reductage deficiency syndrome, Reifensteine Syndrome, Pseudo-harmaphodism and also having a balance between a male and female which is very rare and unique.

After informing the family members first, the society had become an obstruction for the doctors to treat with freedom. Then according to the will of the family members and for the sake of the society diagnosis was hidden from patient and managed by Gonadectomy, Clitorectomy, and Vaginoplasty in same sitting with hormonal therapy for life long to make her a “girl” again.

After 2 months of the treatment patient came with a good respond towards the hormone therapy. Our case proved that ‘Intersex’ is a

mystery of nature and do not always follow the known clinical features.

Discussion

All the investigations & examinations confuse each other in a controversial manner. Though most of the confusion occurred by:

1. 46XX/46XY mosaic chromosomal pattern.
2. Histology of biopsy shows a turn to the Androgen Insensitivity Syndrome (AIS).

Cytologically confusion aired while we matched the case with available theory (See Table 1).

Again with literature confusion came in role as paper published my Catherine et al.[2] reported that:

1. 47% of the 47XY female patients had an accurate diagnosis
2. 32% of them were inaccurate
3. 15% were changed
4. 6% had unknown etiology

As the confusion and controversy came in for diagnosis, altogether the management had become more confusing.

This case could be treated as BOY for:

1. Breast was male variety
2. External genitalia was male type with micropenis & testicles
3. Mulerian ducts were completely absent
4. external appearance and voice was male type

This case could be treated as a GIRL too for:

1. She was a girl in past life
2. Spermatogenesis absent
3. Minimal chance of Seminoma in the future
4. Social demand
5. Surgical ease

Conclusion

Sometimes we treat the patient for sake of society, family etc. and never think about the patient's will. This case was not a different in this aspect. Doctors were forced to follow the rules made by society without protest. Victim of INTERSEX is one of them who have nothing to do against the Nature's betray. More they need physical rehabilitation, ramification, adaptation and a helping hand to come out of gasping agony inside.

References

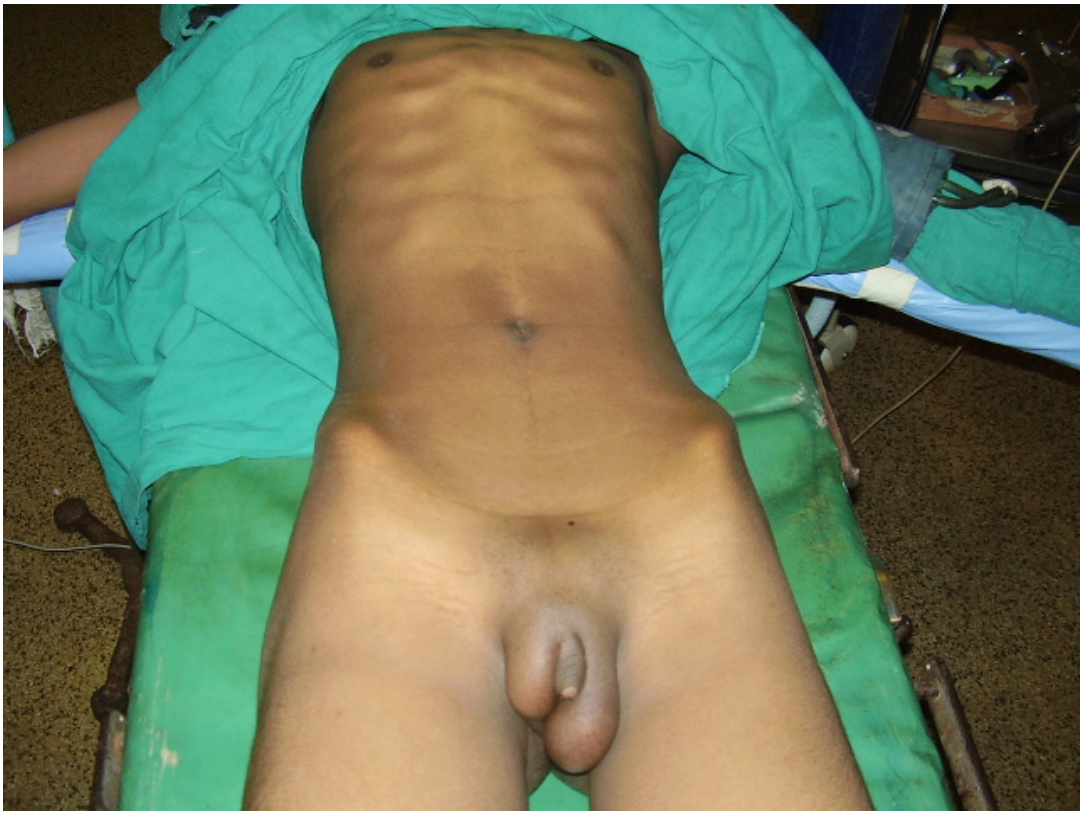
[1] http://www.menstuff.org/issues/byissue/intersex.html accessed on 29.01.2011.

[2] Catherine L. Minto, Naomi S. Crouch, Gerard S. Conway, Sarah M. Creighto. XY females: revisiting the diagnosis. BJOG: an International Journal of Obstetrics and Gynaecolog October 2005, Vol. 112, pp. 1407–141

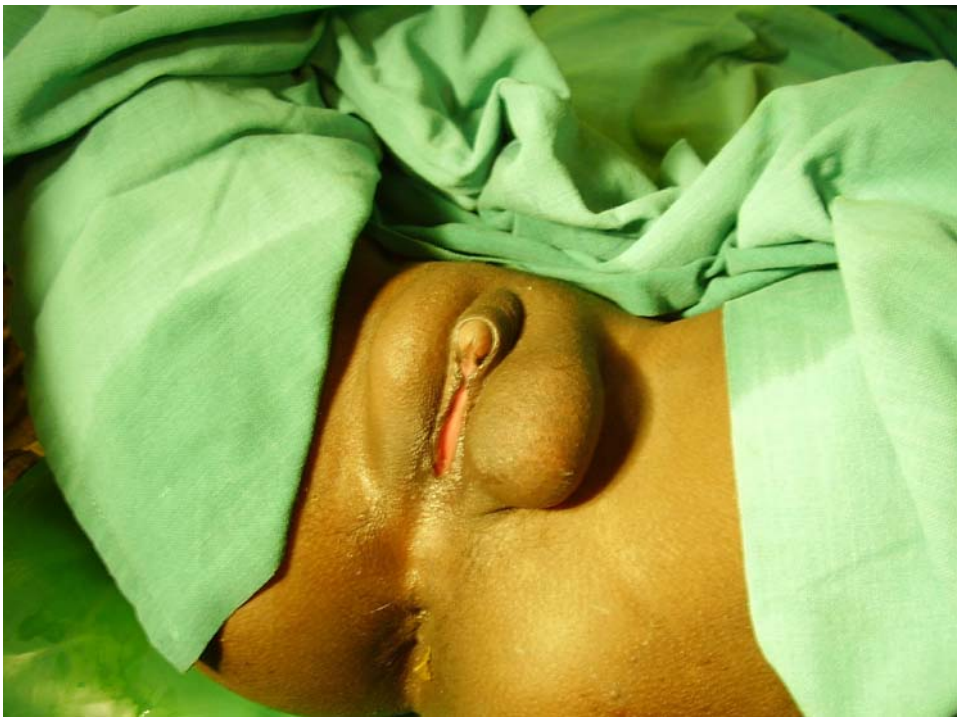
Table 1

Component	5-alpha Reductage deficiency	Reifensteine syndrome	Pseudo-hermaphroditism	Our case
Karyotype	46XY	46XY	46XX / 46XY	46XX/46XY
Spermatogenesis	Decreased	Absent	Present	Absent
Mulerrian	Absent	Absent	Present	Absent
Wolffian	Male	Male	Male	Male
External genitalia	Female	Male hypospadius	Both sexual characters	Male perineal type hypospadius
Breast	Male	Gynaecomastia	Female	Male

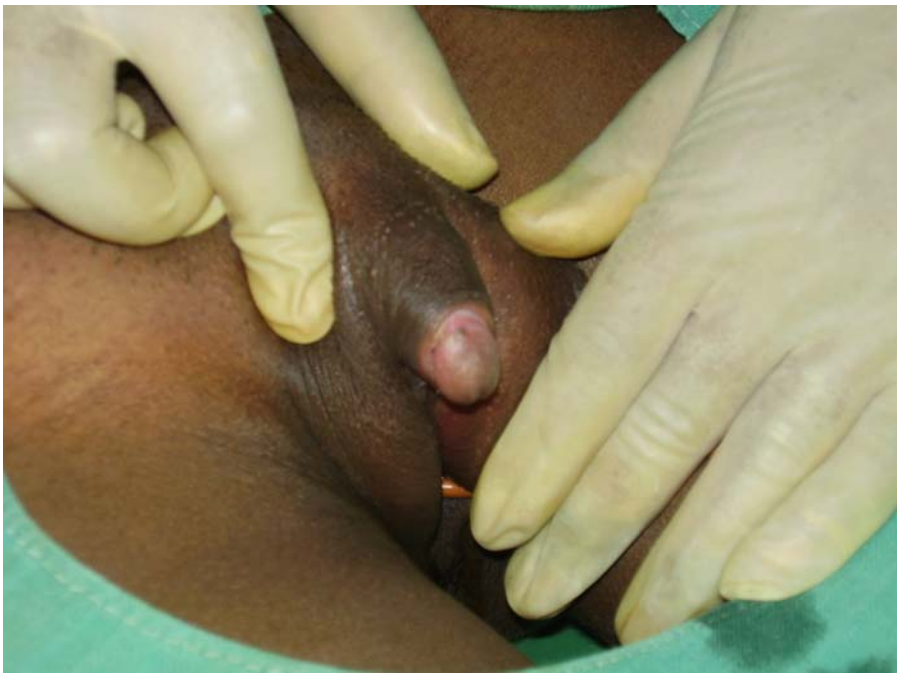
PICTURES (ANNEX)



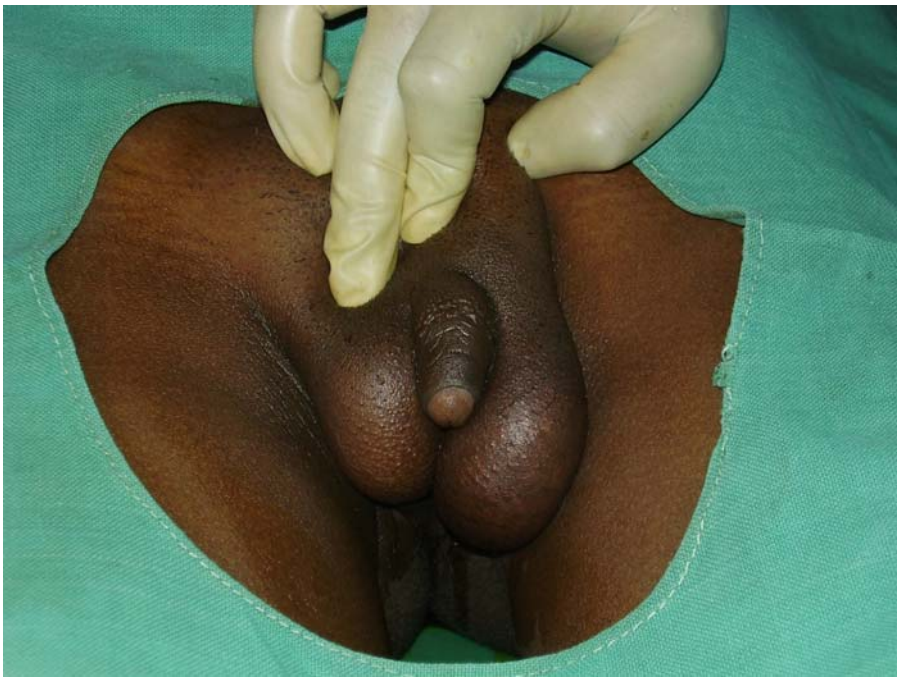
"Physical examination of the patient"



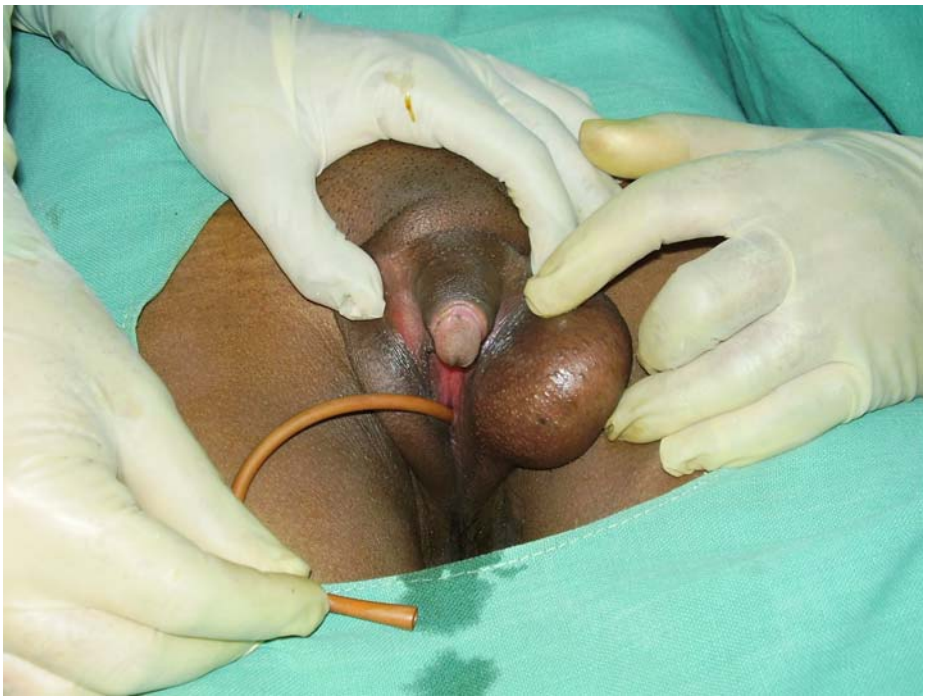
"A blunt vagina in between the swelled labia"



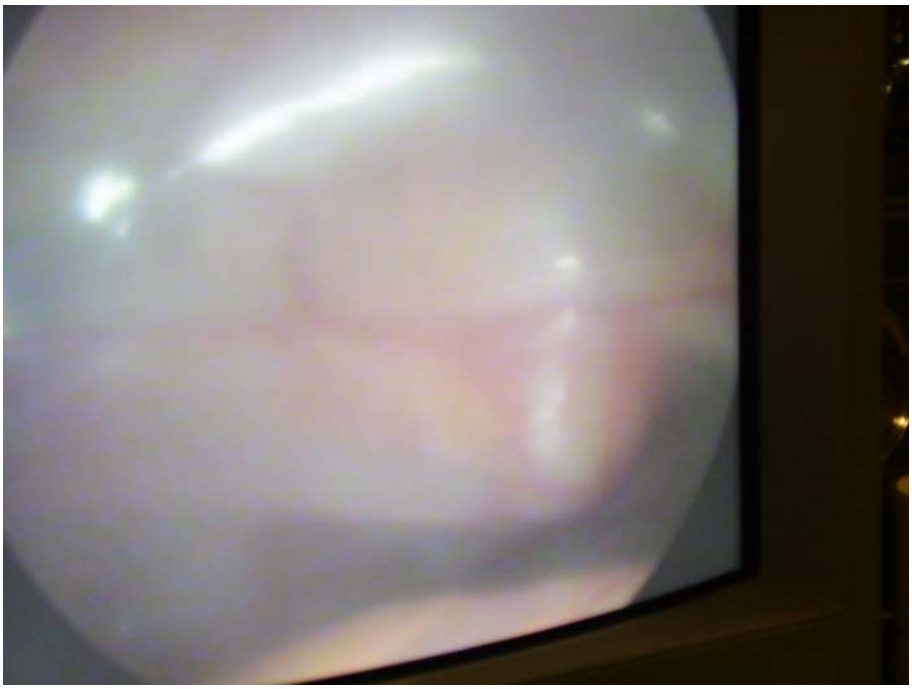
"Micro-penis"



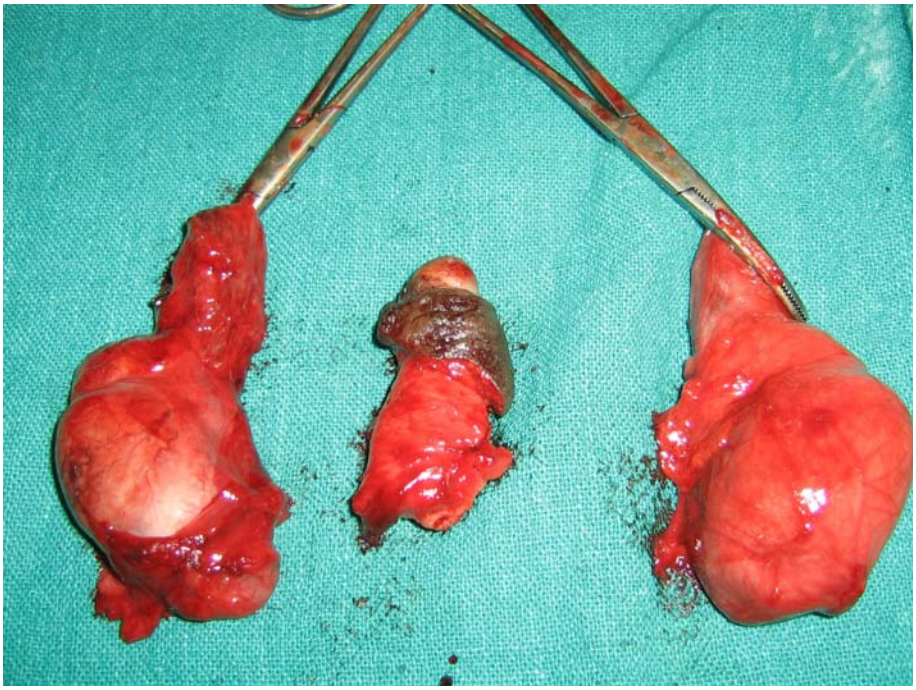
"Palpable lump in both labia major"



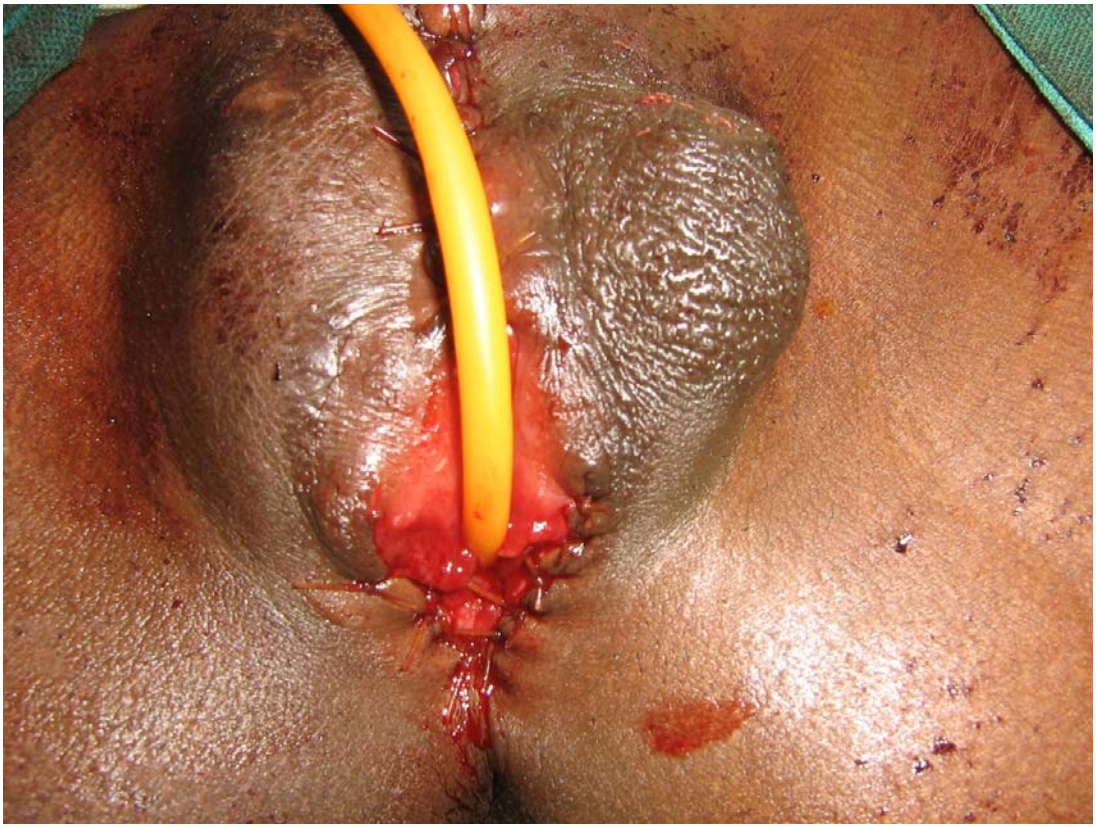
"Perineal type of hypospadius"



"On laparoscopy, no internal female genitalia present"



"Post gonadectomy and clitorectomy picture of excised gonads on both side on micro-penis"



"Post operative picture after vaginoplasty"