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Case Report

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Genital Ambiguity and Challenges in Gender Assignment

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Abstract

This is a case presentation involving two siblings of four and six years of age with female gender identity from a neighboring country with 46,XY DSD and a 20 years old youngster from Northwest Ethiopia with an alleged male sex but with a 46,XX DSD; but definitely all the three with obvious ambiguous external genitalia. The 46XX, DSD with Congenital Adrenal Hyperplasia (CAH) generated more challenges as opposed to the siblings who were diagnosed to have Complete Androgen Insensitivity Syndrome (CAIS). The case presentations are unique as they were referred for sex identification by physicians unlike the usual late complaints of patients with primary amenorrhea or absence of menarche, failure to attain penetrative vaginal sexual intercourse or primary infertility. It was evident that the role of the parents, grandparents, relatives and the healthcare professionals is very much impacting in the sex assignment of the clients, although the final preferential gender choice is made by the individual himself. The atypical external genitalia, contrary to the sex of rearing so far, are the hallmark of the abnormality and source of confusion of the clients under investigations. An early and thorough neonatal physical examination and random checkups of pediatric age groups underscores the complex issues of the subsequent multidisciplinary approach towards the sex assignment. The situations glare the psychosexual and physical development of those involved and the undesirable consequences in their respective families, relatives and their respective societies where beliefs, religion and culture play significant role in the social upbringing of the individual.

The main objective of the presentation is to increase the degree of awareness among healthcare professionals of the existence of such congenital anomalies in the communities and is also intended to contribute to the national and world literature.

Keywords: Disorder of Sexual Development (DSD), Ambiguous Genitalia, Complete Androgen Insensitivity Syndrome (CAIS), Congenital Adrenal Hyperplasia (CAH), Gender Assignment, Siblings, Addis Ababa, Ethiopia

Introduction

Ambiguous genitalia are a disorder of sexual development or differentiation (DSD). DSD is a spectral congenital abnormality, and not a disease phenomenon that influences the chromatin/ chromosomal sex, gonadal sex, sex of rearing/assigned sex, external anatomical/ physical appearance and culture-influenced preferential gender-based naming and addressing despite maintaining the individual gender. Notably, those with genital ambiguity lack the distinctive features of normally attributed descriptions of either a male or female gender. Hence, the external genitalia incompletely display characteristic of both sexes and the external and internal organs do not match; and are thus quite atypical [1,2].

The incidence of all types of DSD is between 1 in 2500 to 1 in 4500 although Congenital Adrenal Hyperplasia (CAH) tends to be specific and stands at 1 in 15,000. The figure could be much higher than often cited where proper registration is not the usual norm and services are not widely available especially in low resource settings. Moreover, accurate family history of such disorders is hushed and could not be obtained. In advanced high resource setups and western countries, genital ambiguity/DSD can be occasionally determined prenatally in the presence of family history of DSD. Hence, the incidence is highly variable as illustrated by the fact that for instance ambiguous genitalia in the Kingdom of Saudi Arabia (KSA) is 1 in 2500, in Egypt 1 in 3000, in Germany 2 per 10,000 live births and in the United Kingdom 1 in 18,000 [3-5].

The Prader classification dealing with virilization of the external genitalia [6,7]. Tanner staging of the external secondary sex characteristics (size of the breasts and genitals, testicular volume,

pubic hair) [8,9]. Quigley scale of seven classes from fully masculinized to fully feminized genitalia and the PVE (Phallic length, Vaginal location and External genital appearance) system after Rink among many others have been introduced to classify the condition and apply in the evaluation and management of patients especially with DSD [10-12]. In lieu of the management plan, this is quite indicative for the need to stage each of the cases on their merits and highly invidualize the clinical approaches.

The amalgam of the mental, family and personal history, at times invading the private life of the individuals, and thorough physical examinations are very instrumental in the understanding and documentation of such complex clinical issues. The imaging, tissue biopsy, hormonal and genetic investigations demand resource, financial capabilities, time, patience and energy [13-16]. The complicating factor of parenthood, social complexities, clients' psychosocial and psychosexual orientations, the tedious, time consuming, costly advanced multidisciplinary medical investigations and consultations, multistage surgical management, psychological and psychiatric rehabilitative treatment courses among many other issues are resource demanding and at times nerve wrecking and academically challenging [17].

The holistic management approach comprises of unthinkable array of multidisciplinary work force. Hence, an organizational setup with a tertiary facility, human and financial resource capability of handling such complex scenarios that are legally and as well medically mandated to handle such cases should be in place as a center of excellence [18].

The previous publications mainly dealt with aspects of primary amenorrhea, primary infertility, and delay in the development of pubertal milestones and sexual dysfunction [19-22]. The current presentation specifically addresses the issues pertaining to ambiguous genitalia and is intended to add to the world and national literature from our perspective and share the costly and intricate diagnostic workup and management difficulties of patients with DSD.

Case Presentation

Case 1

Two siblings of 4 and 6 years of age dressed in female customary clothes were referred to us for an alleged high index of genital ambiguity. They were brought to the institution accompanied by the parents and a language interpreter. They were assigned female sexes, named and raised so far as girl children and dressed like girls even though upon deep interrogation, it was found out that they had their reservations and preferred to be treated like males. They are born to a Para VIII mother and gender distribution wise, seven of them are females, two of whom had undergone female genital cutting (FGM) and the only son as well is circumcised. With the exception of the two referent cases, the remaining five daughters have not so far been screened and parents are warned of not attempting to circumcise them in any circumstance as the occurrence of the disorder may involve similar and multiple siblings.

Figure A1: The cute and curious children of 6 and 4 years of age, respectively, a focus of attention, as seen from their pictures and dressed in traditional female clothes cannot be differentiated from the general population of similar gender and age

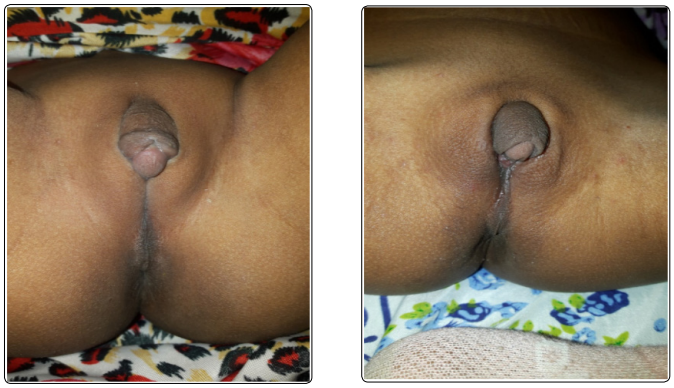

Figure A2: The impressive genital findings of patients 6 and 4, with their phallus and scrotal folds, respectively. They both have easily palpable inguinal masses.

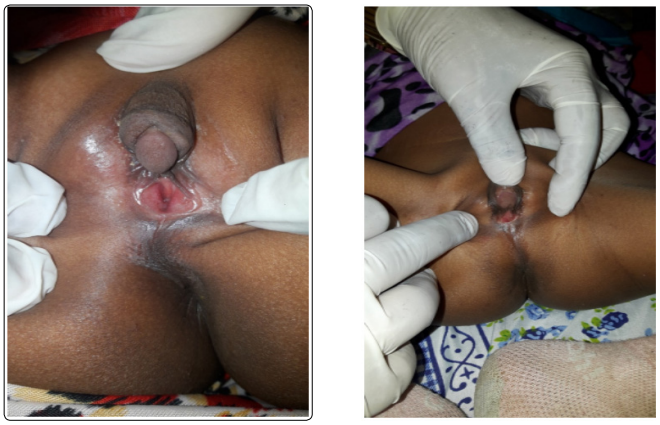

Figure A3: Note the phallus, hypospadias, bilateral scrotal folds and a blind vaginal pouch as attested by a probe of patient 6 and 4 years of age, respectively.

Ultrasonography

An ultrasound examination of both children is performed with a full bladder and portrayed that the kidneys are of normal size, shape and in normal position. The uterus and ovaries are not visualized and hence absent; but there are bilateral, each of which measured 2.5x2.7 cm, non-tender mobile inguinal masses suggestive of cryptorchidism.

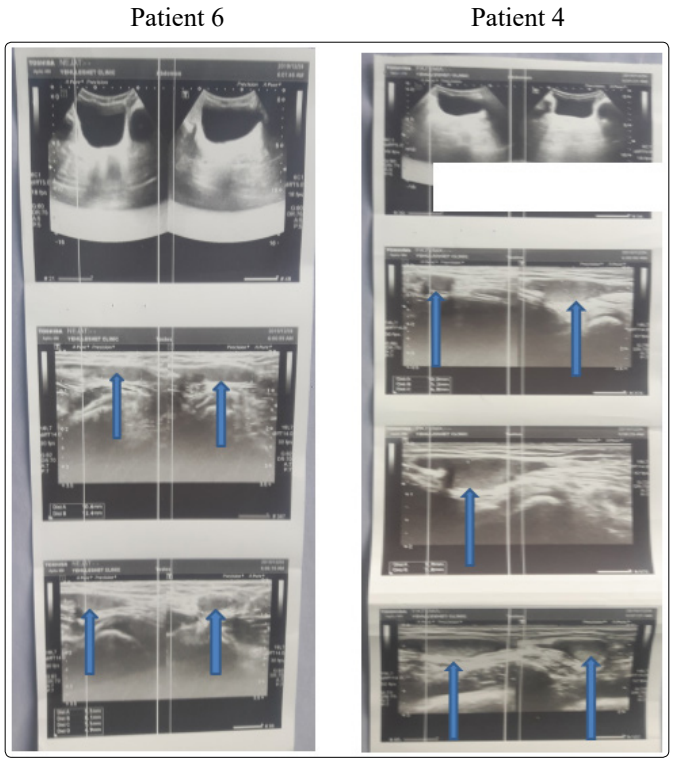

Figure A4: The kids had bilateral, freely mobile, non-tender ovoid structures with no demonstrable inguinal hernias as clearly illustrated above

Genetics

Test Results

The DNA which was obtained from the patients' blood was screened for anomalies on sex chromosomes and androgen insensitivity related gene (AR) regions on Xq12. Several probes that are targeting different regions of the autosomal and sex chromosomes were used to determine the karyotype. Four genes: (DMRTI on 9p24, CYP17A1 on 10q24, SRD5A2 on2p23 and HSD17B3 on 9q22) which are all known to have an effect on gonadogenesis; a few genes linked with sex reversal and disorders of sexual dysfunction: CXSORF21 and ROB1 (DAX) located on Xp21.2, NR5A1 n 9q33, and SOX 9on 17q24) have also been included. The CYP21A2, AR, CAH, 5α-reductase and sex reversal as gonadogenesis related genes did not show significant copy number changes.

Conclusion/Interpretation

The subtelomeric screening as well as the sex chromosome investigations of the siblings shows that the karyotype appears to be 46 XY and assertively the Y chromosome is present in both of them.

Table A1 Laboratory investigations

Relevant Laboratory Investigations and Results				
Category	Investigations (Hormones)	Identification		Normal Ranges (Lab given for males)
Fertility Panel		Patient 4 years	Patient 6 years	
	FSH*	1.90	1.56	0.68-6.7mIU/ml
	LH**	0.16	0.19	<0.1-6.0 mIU/ml
	Estradiol	<5.00	<5.00	6.0-270 pg/ml
	Progesterone	0.36	0.29	Below ranges in ng/ml
	Testosterone	<7.00	<7.00	Pre-pubertal in ng/ml
	Prolactin	5.01	6.40	2.8-29.2ng/ml

Adrenal Gland	ACTH*** (plasma)	9.02	4.79	7.2-63.3 pg/ml
	Cortisol	12.10	13.39	4.30-22.40 ug/dl
AMH	AMH****	19.88	23.00	<3.27 ng/ml

*Follicle Stimulating Hormone **Luteinizing Hormone ***Adrenocorticotrophic hormone ****Anti Mŭllerian Hormone.
NB: High levels of serum AMH implies the presence of testes in CAIS distinguishing it from complete gonadal dysgenesis.

Conclusion and Recommendation

The two siblings were diagnosed to have Complete Androgen Insensitivity Syndrome (CAIS). They were referred to a pediatric surgeon for correction of the inguinal-placed testes for orchidopexy and repair of the hypospadias. The parents were advised to raise them up as males and also subject the rest of the female children for further scrutinization.

Case 2

The second case history relates to a 20 years old diploma graduate referred for the purpose of clarification of the sexual ambiguity. He presented with a complaint of unduly enlarging bilateral breast tissue since the age of 14 despite having a phallus of appreciable size. He was assigned a male sex and named accordingly. Thus, he was raised, dressed, addressed and moreover got enrolled at schools as a male. He has three, one elder and two younger sisters and all are of Christian faith and ardent followers. No family history of infant deaths extracted or shock syndrome was explained by the mother and as well by the client. He is almost always accompanied by the mother who at times shows breakdown with tears.

He always shied in exposing his genitalia even as a child and opted for loneliness throughout his upbringing; and shared his problem with an only female college mate in whom he confided. Having had an impasse during security checkups with security guards at the bank, the friend would withdraw money for him on his behalf and at times reciprocally rewards her also financially when in need. It was a traumatizing process recollecting the disclosure. He claims to have had occasional sexual urge but never masturbated nor was engaged in any fetish practices or made a pass on anybody and for that matter never suffered of discrimination in religious practices or places or sexual orientation and never experienced sexual violence, abuse, intimidation and/or coercion.

Figure B1: Very normal, well developed breasts with normal anatomical size differences and with more of absolute female features

Figure B2: Note the compensatory posture with nuchal flexion, kyphosis and lordotic belly in trying not to display the breasts as feminine features!

He found it very difficult to seek a job as a male teacher in the presence of such noticeable big breasts. He sought for a solution for this specific manifestation, got admitted to a surgical ward for mastectomy and kept in a hospital for seventeen days wherefrom he was discharged for psychiatric consultation and referred to us for further investigations and help in the establishment of a clear diagnosis with regard to the sex identification.

Upon presentation, he is described as a short statured (150 cm), lean with a weight of 47 kg, shy but communicative and dressed in jeans with a male jacket and exaggerated abdominal curvature with forward leaning shoulders. He had a common short cut male hairstyle with his scalp hairline distribution not receding. He has smooth shiny facial texture without a blemish and with no hirsutism (beard and moustache) and signs of androgenization/virilization like hoarseness of voice and acne. He has no webbing of the neck, pigeon/shield chest, pectus cavum, widely spaced nipples and showed no increased carrying angle and cardiac anomalies. The client revealed no skeletal deformities and hence was not subjected for radiological examinations.

Figure B3: The most striking features include an enlarged clitoris resembling a fair sized penis with labial folds looking like scrotum. Looks more like a male than female!!!

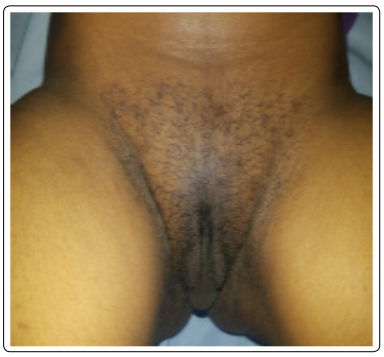

Figure B4: Note the female type typical pubic hair distribution, labial/scrotal empty folds, the hanging macro clitoris

Figure B5: Demonstration of the phallus/clitoris with closed meatus, hypospadias with fused labia/closed vagina in addition to the lateral folds

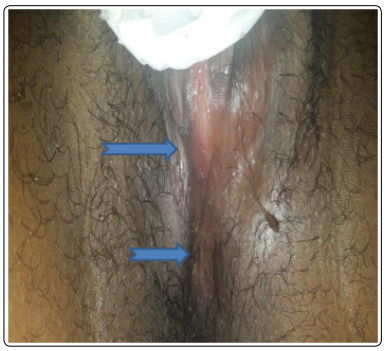

Figure B6: The hypospadias with fusion of both sides of the labia in the middle, with no opening or bulging suggestive of imperforate hymen with cryptomenorrhoea

Table 1: Prader Classification

Type	Degree of virilization of external genitalia
Type 0 (P0)	Normal female external genitalia
Type 1 (P1)	Clitoral hypertrophy
Type 2 (P2)	Clitoral hypertrophy, urethral and vaginal orifices present, but very near
Type 3 (P3)	Clitoral hypertrophy, single urogenital orifice, posterior fusion of the labia majora
Type 4 (P4)	Penile clitoris, perineoscrotal hypospadias, complete fusion of labia majora/minora
Type 5 (P5)	Complete masculinization (normal looking male genitalia) but no palpable testes
Type 6 (P6)	Normal male presentation, no hypospadias, normal testes

Genetics

Test Result

The DNA which was obtained from the patient's blood was screened for anomalies on sex chromosomes and androgen insensitivity related gene (AR) regions on Xq12. Several probes that are targeting different regions of the autosomal and sex chromosomes were used to determine the karyotype. Four genes: (DMRTI on 9p24, CYP17A1 on 10q24, SRD5A2 on2p23 and HSD17B3 on 9q22) which are all known to have an effect on gonadogenesis; a few genes linked with sex reversal and disorders of sexual dysfunction: CXSORF21 and ROB1 (DAX) located on Xp21.2, NR5A1 n 9q33, and SOX 9 on 17q24) have also been included. Particular attention was given to the copy number of the sex chromosomes. The peripheral chromosomal analysis as well as sex targeting probes shows the karyotype to be 46XX. The probes that specifically target Y chromosome did not produce signals indicating its absence. The AR, and CYP21A2 genes did not show significant copy number changes.

Conclusion/Interpretation

The Karyotype appears to be 46 XX and Y chromosome is absent and no indication of X monosomy. The NROBI or DAX shows significant copy number change. The AR, CAH related CYP21A2 and the gonadogenesis related genes did not show significant copy number changes. However, subtle changes such as point mutations and balanced translocation and other genetic changes that cannot be detected by the method employed and yet having clinical significance cannot be ruled out.

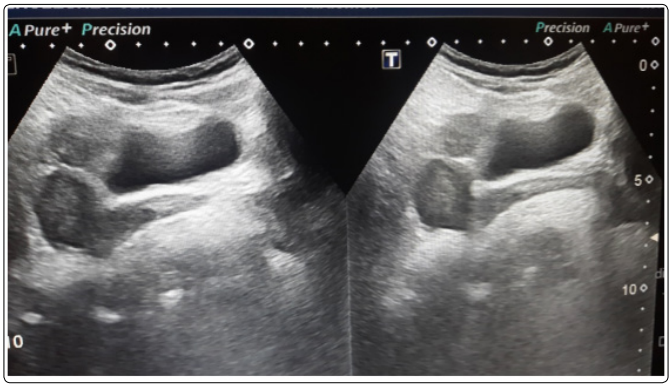

Figure B7: A full bladder, closed introitus, vaginal canal, anteflected and anteverted uterus with an ovary

Ultrasonography

The evaluation clearly demonstrated a vaginal canal (25.3x17.7x20.1mm), well developed uterus (39.4x27.0x28.3 mm), and ovaries (17.5x12.4mm) with ovarian follicles as opposed to Mayer-Rokitansky-Kuster-Hauser Syndrome. There was no obvious haematocolpos, haematometra, haematosalpinx and/or fluid in the pouch of Douglas suggestive of cryptomenorrhoea. The report excluded urinary tract anomalies.

MRI

Uterus has normal size, mildly anteverted position and homogenous myometrial signal and normal endometrial thickness and signal intensity. The cervix has normal caliber and signal intensity. The vaginal canal appears to have normal course and signal intensity. The ovaries appear to have normal size, shape and signal intensity, and few small sized intraovarian follicular cysts noted in the right ovary. The final impression is summed up as normal, well developed female internal organs with uterus, cervix, vaginal canal and ovaries as shown below.

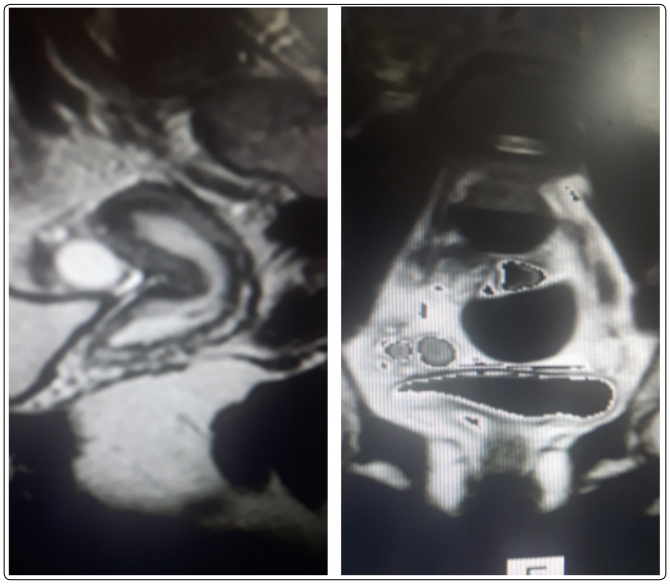

Figure B8: An MRI picture with a full bladder, closed introitus, vaginal canal, anteflected and anteverted uterus with the right ovary and its intraovarian cysts

Colpography and Urethrogram

General procedure

The client was thoroughly informed of the details of the procedure, possible adverse outcomes like anaphylactic shock and finally written and signed consent was obtained. Accordingly, an IV cannula was inserted through the fused labia/closed vagina about the possible site of the vaginal opening. Iodine containing contrast material (Iopamiro 370mg l/ml) was introduced through the placed cannula. The vaginal canal and vault are well demonstrated and appear to have normal course and caliber with accumulation of the dye at its apex. Further introduction of contrast material showed normal outline of the cervix and uterus. There was no intravasation of contrast material. It is to be underlined that special caution was exercised not to, in anyway, open the vaginal opening non-consensually and prematurely and undesirably embarrassing the gender identity of the individual.

Through the urethral opening, a paediatric catheter was placed, the Foley catheter balloon inflated with air and dye introduced into the bladder. It was observed that there was normal course and caliber of the urethra and bladder has normal position and outline.

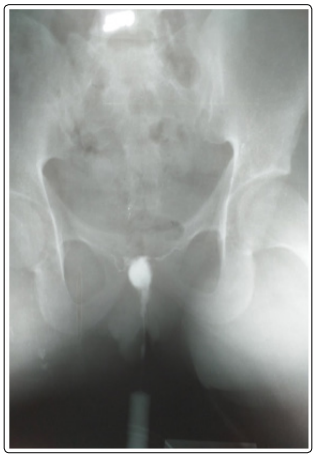

Figure B9: The loaded syringe with contrast and the IV cannula that was inserted into the vaginal canal through the fused labia is depicted in this picture. Note the accumulation of the dye at the vaginal apex with impressive resistance

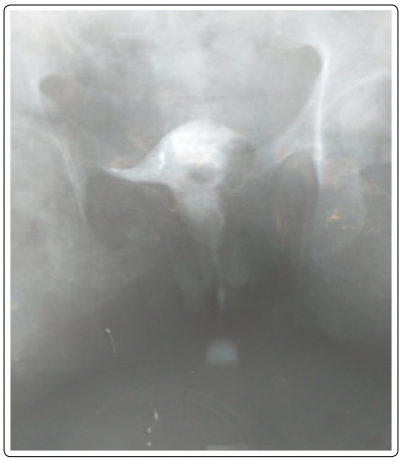

Figure B10: Note the introduction of the dye into the uterine cavity and the cornua, the vaginal canal and relieve of the initial accumulation of the contrast at the apex.

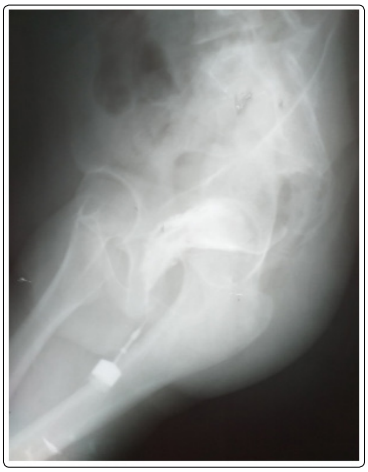

Figure B11: Lateral view of the pelvic X-ray with loaded syringe-in-situ, the vaginal canal and uterine silhouette

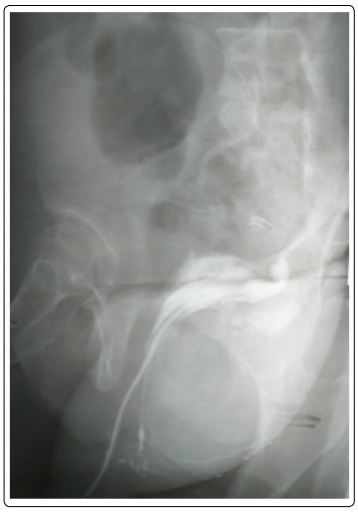

Figure B12: A pediatric catheter was introduced and its balloon inflated with air and contrast. Note the IV cannula and the vaginal canal outlining and the catheter separately which are demonstrative of the fact that there is no single urethral/vaginal opening.

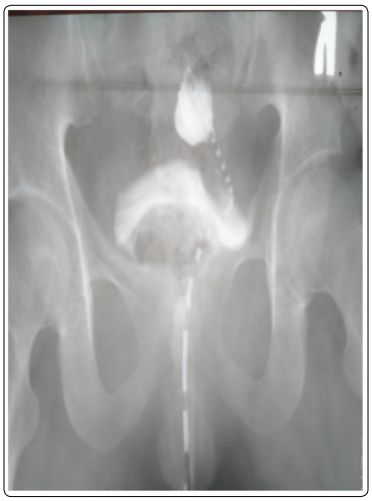

Figure B13: Partial pneumocystography with the broken lines illustrating the urethral length, the bladder and the uterine silhouette.

Table B1: Laboratory investigations

Relevant Laboratory Investigations and Results			
Category	**Investigations (Hormones)**	**Results**	**Normal Ranges (Lab given for females)**
Fertility Panel	FSH	3.80	2.5-10.2 mIU/ml
	LH	6.95	1.2-12.5 mIU/ml
	Estradiol	49.6	15-112 pg/ml
	Progesterone	1.56	0.36-1.21 ng/ml
	Testosterone	162.48	280-1080 ng/ml
	Prolactin	13.5	3.1-23.03 ng/ml
Adrenal Gland	ACTH(plasma)	21	10-48 pg/ml
AMH	Cortisol (morning)	16.85	5.72-19.42 ug/dl
	AMH	18.05	1.5-9.95 ng/ml
Electrolytes	Sodium	142.8	135-145mmol/l
	Chloride	92.2	98-108mmol/l
	Potassium	3.42	3.5-5.5mmol/l
	Calcium (total)	2.43	2.2-2.9mmol/l
	Calcium (iodized)	1.25	1.1-1.4mmol/l

NB

With the exception of AMH which may be explained on the basis of the intraovarian cysts as in polycystic ovary syndrome (PCOS), all the tested hormones and electrolytes are within normal limits

Conclusion and Recommendation:

All the investigations weigh-in heftily asserting the fact that the individual is absolutely a female with 46XX, DSD. It is feasible that feminizing reconstructive genitoplasty including corporal body operations, repair of the hypospadias, creating adequate vaginal opening along with the labia majora and minora formation to retain his inherent internal organ structures and external appearance, supplemented with hormonal therapy, may ultimately improvise on his fertility chances. His preferred sex identity, however, is a male and requires rehabilitative stepwise psychiatric therapeutic sessions with the pros and cons analyzed and delivered to the client, parents and relatives to finalize the intended desire in respect of the individual's reproductive right and choice. It is instrumental to underline the fact that he can spontaneously menstruate or experience his menarche at old age or hormonally induced and eventually have children!

Discussion

Ambiguous genitalia could be quite obvious at birth, shortly after birth and at times more so following the pubertal period due to the influence of the sex hormones or the developing secondary sex

characteristics and the phenotypic appearance of the individual. Consequently, genital ambiguity has in the course of time generated historical interest and analysis, modified our sociocultural perceptions and interactions, invigorated our medical practices, impacted our faith/religious teachings, challenged the strict interpretation of the legal systems, and assumed political and socially vibrant issues vis-à-vis the concept of sexual and reproductive health rights and services (23,24).

The decision of sex assignment and societal gender based allocations of functions become complex and the long term sequelae includes the embedment of scarred self and embarrassment to the entire family and relatives. The associated social, cultural, psychological stigma, family honor being at stake, personal sexual conflicts and taboos are immensely distressful situations to all. The brunt of parental feelings of guilt, disgrace, disgust, shame and stigmatization hampering the future chances in marital bondage of their progeny with others tends to be a stumbling block in our socio-cultural structure and practices (17,25).

The bipotential gonads under the influence of determinant factors encoded by the Y chromosome (sex determination) and in the presence of male hormones induce the gonads to differentiate into testes (sex differentiation). The somatic sexual differentiation results in a typical female phenotype which in a sense is a neutral sex. The etiologic factors incriminated in the pathogenetic mechanism of DSD and processes are many. The most common causes in females are the androgens from the adrenal glands (CAH, placental aromatase deficiency), insufficient secretion of androgens and their metabolites, maternal ingestion of substances with male hormone activity during gestation especially in the first trimester with labial fusion and also after 12 weeks with clitoromegaly but without labial fusion, and male hormones generating tumor in the female fetus. Of interest, severe clinical manifestations of CAH and the hallmarks of pigmentation, salt wasting disease, hyponatremia and shock have not been experienced by our sole patient with the condition who, for sure, is not a candidate for a late onset CAH. The possible causes in males genome are Leydig cell aplasia, 5-alpha-reductase deficiency, maternal ingestion of substances with female hormone activity during pregnancy and androgen insensitivity syndrome as in the two sisters (4,14,17). Hence, our cases can be analyzed through the bidimensional causal perspectives.

In addition to the hormonal etiologies, ambiguous genitalia have genetic basis of sex disorder with alteration of localization of specific genes specifically with Androgen Insensitivity Syndrome which is X-linked autosomal recessive inheritance though sometimes X-linked dominant gene with incomplete penetrance is a possibility. The uterus, ovaries and fallopian tubes are absent because of the fact that the testes secrete normal amounts of Müllerian Inhibing Factor (MIF). The rest of the five sisters of the siblings deserve to be given the chance of being screened for similar disorders in as much as in the second case with mild CAH, falling under Prader IV with macro clitoris, and his three sisters at home who have not so far brought to our attention. In the course of history taking and physical examination, the parents were strictly advised to refrain from performing routine circumcisions in the light of a probability of causing irreversible phallic trauma bearing in mind that it is more difficult to reconstruct a functional penis with all its characteristics rather than create a vagina (26,27,28,29).

The psychosocial, religious, cultural and psychosexual complexities are typically exemplified by the study from Pakistan whereby the rejection of intersex people, deprivation of education and basic sexual and reproductive health services and rights and subsequent isolation leading to internal conflicts, helplessness, hopelessness and increased suicidal tendencies. The coyness, secretive and feeling of being in isolation with failure to establish intimate interpersonal relationship with indeterminate gender identity of the patient with CAH has been worrisome as opposed to the two kids who already had individually accepted that they are different and more like males than females unlike the parents' impositions. Extreme caution was exercised in extracting maternal history of drug ingestion or virilizing gynecological tumors in order to avoid displaced anger or premature disclosure of the clinically determined sex identity before appropriate psychiatric consultation (30,31).

Consanguinity is a common cultural and traditional practice among the different Muslim and Somali societies and deserves attention as the occurrence of hereditary anomalies tends to be more common where relatives intermarry. Hence, it is essential that premarital and pre-conception counseling should be provided in such a situation in order to rule in or out the presence of family history of X-linked recessive autosomal diseases whether and if they exist in the family pedigree (33). Two Ethiopian siblings in Saudi Arabia with CAH were also included in the study by Al Jurayyan (32). The sex of a fetus is now increasingly known long before the birth of the child through the determination of the karyotypes, analyzing the chorionic villi, amniotic fluid samples, or maternal circulating free fetal DNA coupled with the use of three-dimensional ultrasonography. Moreover, we strongly recommend that meticulous prenatal workup with strong existence of such background history can be thought of and furthermore neonatal, paediatric, pubertal, premarital and thorough preconception counseling ought to be in place (34,35).
In ambiguous genitalia, the AMH is highly variable with low, normal or higher levels. Though it tends to be constant throughout the childhood period in both sexes, boys have 35 times higher levels than girls. In all the three cases under discussion, their AMH values are raised and are meaningful in the presence of testicular tissue in the CAIS siblings and a polycystic ovary in the CAH case. Hence, it remains to be a useful clinical parameter applicable in the workup of patients with DSD. Moreover, it is worth noting en passé that it is a useful instrument employable in the workup of patients with premature ovarian failure, gonadal dysgenesis (XO), granulosa cell tumors with precocious puberty, hypogonadotropic hypogonadism state and in aftermath of cancer treatment with alkylating gonadotoxic agents (36,37,38).

A complete history and physical examination coupled with an array of investigations establish the cornerstones of the clinical diagnosis. The final diagnosis and management of such patients also demand a hoard of specialties that may involve and include a geneticist, radiologist, pediatric endocrinologist and urologist, plastic surgeon, psychologist, psychiatrist, gynecologist, social worker or counselor and a lawyer. The multidisciplinary management options must ensure long term successful outcome with healthy puberty and adulthood with pleasurable and safe sex life in the fulfillment of individual sexual and reproductive health rights and services. The patients, parents or guardians must be well informed with the DSD under the principle of informed choice leading them to informed consent for the ultimate intervention. It is to be appreciated the timing of reconstructive surgeries should be ideally in infancy but

in all our cases their presentation is relatively late, but still doable. Generally, the managing of ambiguous genitalia poses complex challenges and entails expenses difficult to bear in the absence of insurance scheme or other means of covering the medical expenses. We have appreciated these loopholes in carrying out the relevant specifics during the workup of the patients and considered them as serious limitations has it not been for the generous subsidy by colleagues (14,28,34,35).

The Ethiopian Constitution guarantees the right to equality and protects people's rights and upholds Universal Declarations of Human Rights, International Convention on Civil and Political Rights (39). The country is a signatory of the many Conventions, Declarations and Resolutions pertaining to sexual and reproductive health rights and services. Historically, it is a home of a variety of religions and deeply entrenched in traditional conservative values. There exist no provisions with regard to gender transformation or sex re-assignment and gender identity altering surgical operations based solely on pure volition and on own choice outside the classical orthodox male and female categorization even in the Renewed Family Code of 2000 (40). We believe per se that the management of the underlying DSD is not affected unlike transformative management alternatives. For the individual with 46 XX, DSD who claimed to be a male with a virilized mentum and was brought up accordingly as a male and desires to be transformed into a male within an Ethiopian setup would be an uphill battle difficult to fulfill the individual's own aspirations! We are committed to provide him with the required long term stepwise psychiatric consultations and finally facilitate the surgical, hormonal and rehabilitative treatment modalities. We are cognizant of the international experience, for instance that of Christiane Vôlling and the Malta law/declaration, that non-consensual medical and surgical interventions can entail intricate legal repercussions and it might as well can be operational in our setup be it on the ground of incomplete or lack of comprehensive counseling or irreversible damage per se of the treatment sequale (41,42).

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