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Gender assignment in disorders of sex: An Islamic perspective from Saudi Arabia

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Abstract

Genital ambiguity, is defined as a group of congenital conditions in which development of chromosomal, gonadal, or anatomic sex is atypical. It constitutes a major social emergency and the decision making in relation to sex assignment has been perceived as an extremely disturbing and difficult to both families and health care professionals. It can also be a medical emergency as it may lead to life-threatening adrenal crisis. Each patient should be evaluated individually by a well-coordinated experienced multidisciplinary team of specialists in a higher center capable of managing such disorders. The team should include a geneticist, neonatologist, endocrinologist, pediatric surgeon, urologist, psychiatrist and psychologist, a social worker, and other specialists such as a nurse, and gynecologist to be consulted whenever needed.

The estimated incidence was found to be approximately 0.1% to 0.2% worldwide i.e, 1:4500-5000 live birth. The 46XX is the most common cause, while the majority of these patients congenital adrenal hyperplasia (CAH) is the most underlying aetiology. The 46XY DSD is a heterogeneous disorders that often result from a disruption in the production or response to testosterone, dihydrotestosterone, or Mullerian inhibitory substances. Chromosomal DSD includes condition resulting from abnormal meiosis, including Klinefelter syndrome (47XXY) and Turner syndrome.

The evaluation of patients with sex disorders requires a through history, physical examination, karyotype, serum electrolytes, and specific hormones, such as 17-OH-progesterone.The pediatric radiologist has a major role in diagnosis.

In Saudi Arabia, a general guidelines were suggested by the council of senior ulama to manage patients with DSD in accordance with the islamic laws. This should be the practice in other muslim countries. Health professionals need to be aware of the existence of such recommendation for a better care of patients.

Keywords: Genital; Ambiguity; Islamic perceptive; Gender; Assignment; Multidisciplinary team

1. Introduction

Genital ambiguity (uncertain or atypical genitalia) represents a wide range of congenital conditions in which the chromosomal, gonadal or anatomic sex is atypical. It constituted a major complex social and medical emergency that requires prompt response by a well-coordinated multidisciplinary team of specialists. Several conditions produce significant salt loss which, if unrecognized, and early treated may lead to shock and even death. The team should include a geneticist, neonatologist, endocrinologist, pediatric surgeon, urologist, psychiatrist and psychologist, a social worker, and other specialists such as a nurse, and gynecologist to be consulted whenever needed (1-11).

Limited data are available on the exact incidence of DSD with genital ambiguity at birth, which has been estimated to be approximately 0.1% to 0.2% world-wide, i.e, 1 in 4500-5000 live births. Thyen and associate (12) reported from

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Germany that ambiguous genitalia incidence was four-fold higher in infants of non-German origin compared with the general population. This was attributed to an increase in autosomal recessive forms of disorders of sex due to higher rates of consanguinity in migrant population. In support to this the higher incidence of genital ambiguity in Saudi Arabia were the consanguinity rate is high (8,13), the incidence of ambiguous genitalia was reported as 1 in 2500 live-births (14-16). Likewise that of 1 in 3000 live births in Egypt (17). Rate of disorders of sex is even higher in some other countries like, Turkey. Where Aydin and associates (18) have reported higher incidence of 1.3 in 1000 live births.

In 2006, the Lawson Wilkins Pediatric Endocrine Society (LWPES) and the European society for Pediatric Endocrinology (ESPE) published a consensus statement of intersex disorders and proposed the umbrella term disorders of sex development "DSD" instead of terms like "intersex", pseudo hermaphroditism (PH), "hermaphroditism" " sex reversal ", which are often perceived as pejorative by patients and can be confusing to both health professionals and parents. This was modified later (19-21). Congenital adrenal hyperplasia (CAH) is the most common cause for DSD in the newborn world-wide. Other notable causes include partial androgen insensitivity syndrome (PIAS). Unfortunately, a great number of cases do not have an identifiable causes, particularly among 46XY DSD, which highlight the great importance of an experienced team with multidisciplinary focus. Congenital malformations can also cause disorders of sex development (DSD), many of which have a primary genetic cause, and others which have a more structural or developmental causes and do not have any hormonal causes. These include VATER/VACTER associations or cloacal exstrophy.

The main objective of this article is to outline the clinical approach to the management of patients with disorders of sex. Gender related issues from the perspective of the islamic jurisprudence are also highlighted and discussed.

2. The practical approach of diagnosis of disorders of sex development "DSD"

The management of patients with genital ambiguity and disorders of sex should be managed in a highly specialized medical center, where a multi-disciplinary team of specialists capable of managing such disorder is present. A thorough history and physical examination were essential. Special hormonal investigations and serum electrolytes can identify the etiology. Establishing the genetic sex (karyotype) should be the first step in the management (25-27)

The pediatric Radiologist has a major role in determining the internal anatomical structures. Abdominal ultrasonography remains the first choice. (Figure 1), this could include genitography, (figure 2), and magnetic resonance imaging (MRI), (figure 3). (28-32).

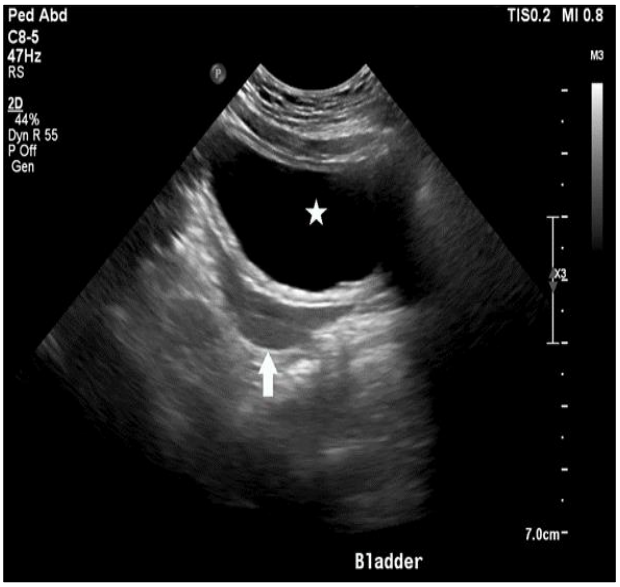

Figure 1 Ultrasound pelvis demonstrates presence of the uterus (white arrow). The urinary bladder (white star)

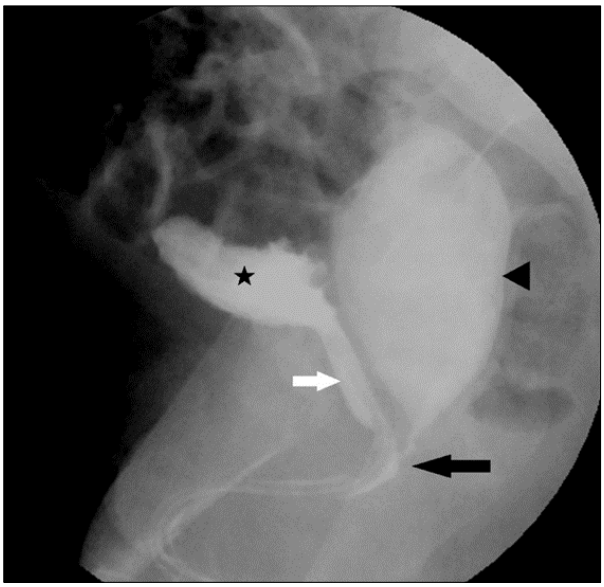

Figure 2 Fluoroscopy genitogram demonstrates normal appearing urinary bladder (black star) and urethra (white arrow) with dilated upper vagina (black arrowhead) with communication with lower urethra (lack arrow)

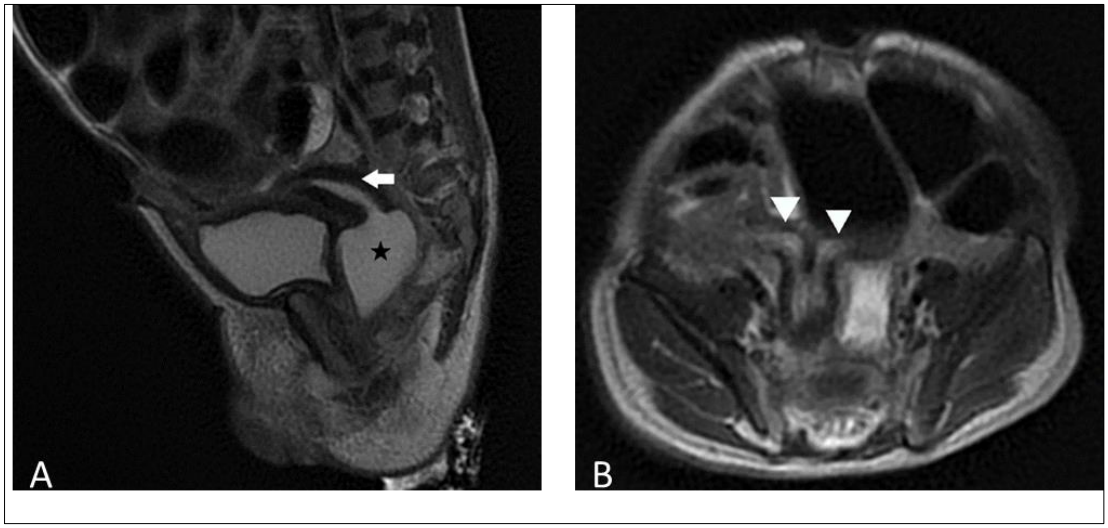

Figure 3 (A and B) MRI pelvis T2 weighted image sagittal (A) and axial (B) demonstrates presence of a dilated upper vagina (black star) and the uterus (white arrow) with two separate cavities (white arrow heads) (bicornute uterus). No ovaries or testes were visualized

The following represents the diagnostic algorithm for patients in which the gonads are either palpable or not (figure 4). However, decision making algorithms based on the karyotype, and other hormonal investigations were available (27, 33). Special procedures like laparoscopy or Laparotomy with gonadal biopsies might be needed at times. However, a biopsy as well as laparoscopy is not required when the diagnosis is clearly established biochemically or by gene studies. (34)

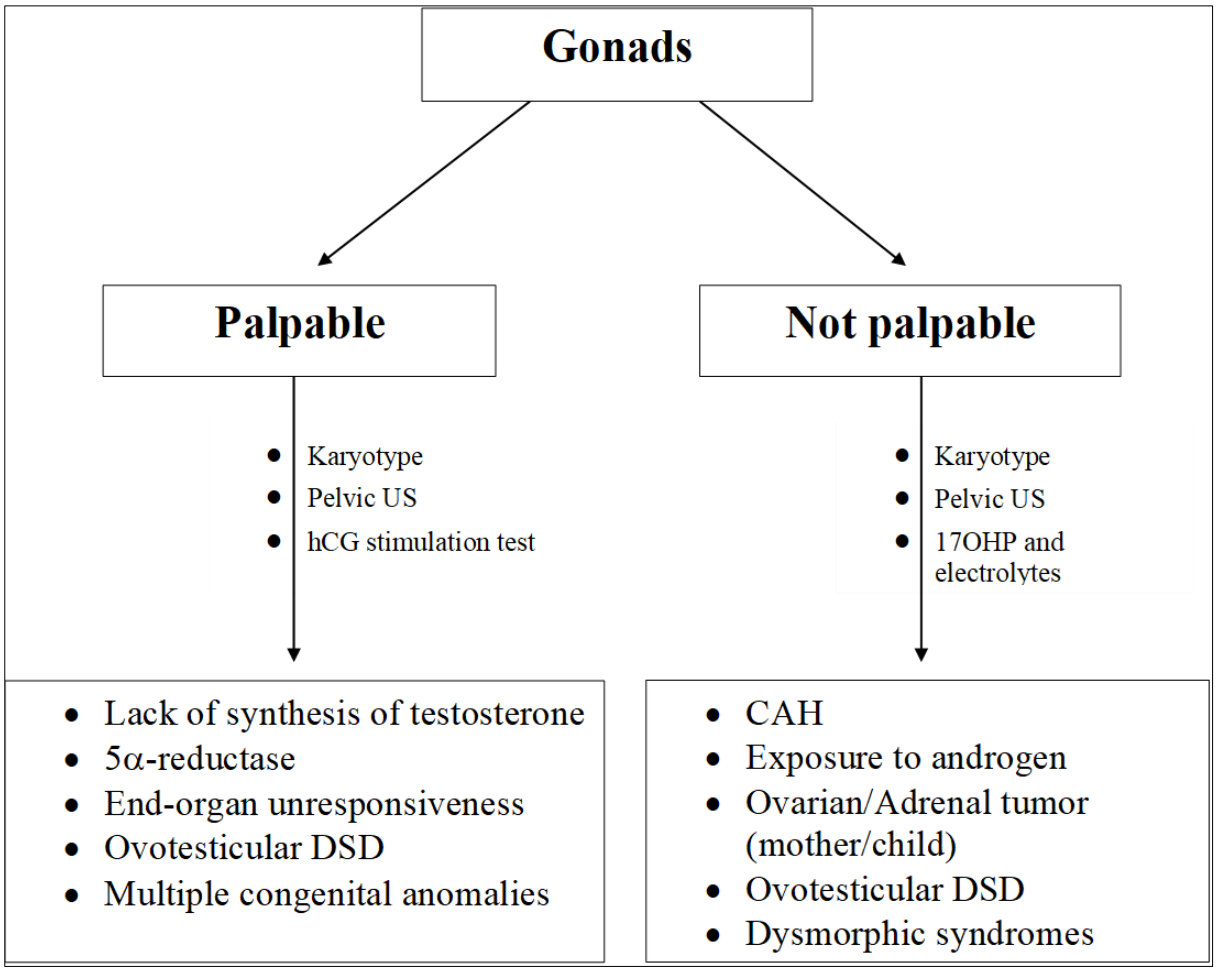

Figure 4 Diagnostic Algorithm for patients with ambiguous genitalia, based on gonadal palpation

3. Discussion on issues related to ambiguity of genitalia

Management of patient with DSD is undoubtedly complex and challenging. Biomedical, psychological, cultural and religious factors, among others, seems to be influential (14,35-38). Kuhnle and Krahl (39) demonstrates earlier the impact of culture on sex assignment and gender development in patients with disorders of sex. This was in accordance with Meyer-Bahlburg (40). Although several studies from different Muslim countries like Saudi Arabia, Egypt and Turkey which indicated preference of male sex despite of the gonadal makeup and karyotype (41-43). Also Warne and Raza have reported that In India and Pakistan children with disorders of sex are more likely to be raised as males.(44) Nevertheless, not all countries have preferences for male sex as stated by Zainuddin and Mahdy from Malaysia (45).

Great need for a better psychological support, to encourage self-acceptance, and counseling for individuals with DSD and their parents. It is always considered as a difficult task that requires a skillful personals. It should be clear, frank and honest. (51-54) The need for encounter and exchange ideas with other parents with similar experience with DSD are found to be helpful. Furthermore, there is a need for a continuous physiological support and counseling. Efforts should be made to improve strategies and quality of counseling. Incorporating a religious scholar with good understanding of genital ambiguity into the multidisciplinary team will ease the discussion with patients and their family. Finally, in each country establishing a central body called '' Gender assignment council '' to ensure a better control. Their contact telephone number and E-mail should be available to facilitate communication.

Furthermore, patient and or family should be fully informed of the condition and the possible outcome including surgical procedures (55-61). This will improve decision making and acceptance.

In Saudi Arabia the council of senior Ulama, the highest religious authority in the country, suggested a set of guidelines to manage patients with DSD. (62-64)

There is an impression fostered by the local clinical experience as published earlier (14,47) That these guidelines are practical and easy to apply Zabidi (65) has reviewed the issues related to genital ambiguity from both the Islamic and medical perspectives. Management of patients with DSD and genital ambiguity continues to be challenging and complex. To find a solution for all the issues from one Islamic council seems to be insufficient. Certainly, there is a need to bridge the gap between the medical professionals and Islamic scholars around the world. Guidelines for gender assignment and issue related to the decision-making process require in-depth research.

4. Conclusion

The management of patients with genital ambiguity is challenging and complex. There is an urgent need to establish unified general guidelines to manage patients with genital ambiguity. A combined efforts between the council of senior Ulama of Saudi Arabia and the International Islamic Fiqh Academy of the muslim world league should be encouraged. Furthermore, counseling strategies and psychological support need to be improved in quality. Exchange with other patients, or parents with DSD with similar experience has been of importance. In addition, further studies on the subject should be executed. Finally, to ensure a better care, health professionals need to be aware of the existing guidelines.

Compliance with ethical standards

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