



Gender Reassignment Surgery in a Case of Androgen Insensitivity Syndrome

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Abstract

Gender reassignment surgery plays an important role in relieving the physical, emotional and psychological discomfort of individuals with gender dysphoria. The collaborative effort between the surgeon, behavioural scientist, and a medical physician, endocrinologist is recommended. The surgery can be performed with good outcomes and a low rate of complications. We report a case of gender reassignment surgery in a 32 year old karyotypic male with androgen insensitivity syndrome and brought up as a female since birth.

Keywords: Androgen insensitivity syndrome; Amenorrhea; Gonadectomy; Testes

Introduction

Gender Reassignment Surgery (GRS) is an effective treatment for patients with gender dysphoria after appropriate diagnosis and psychotherapy [1]. Techniques for male to female GRS are well documented in the literature, many of which have been proven to produce good functional and aesthetic outcomes [2]. Modern surgical techniques include creation of a vagina allowing for coitus and the creation of a sensible neo-clitoris for sexual orgasm [3-5].

Male to female GRS can be performed with a low rate of major complications. Sigurjonsson et al. [2] retrospectively studied 205 male to female GRS performed during the period 2000 to 2013. The median age of the patients was 33 (range 18 to 76) years. Major complications such as rectovaginal fistula, deep vein thrombosis and deep infections were rare. The most common minor complications were bleeding and infection. Hess et al. [6] evaluated the effect of male to female GRS on sexuality in 254 patients. In total, 119 patients completed the questionnaires after a median of 5.05 years since surgery. Of the study participants, 33.7% reported a heterosexual, 37.6% a lesbian, and 22.8% a bisexual orientation related to the self-perceived gender. Of those who had sexual intercourse, 55.8% rated their orgasms to be more intensive than before, with 20.8% who felt no difference. Most patients were satisfied with the sensitivity of the neoclitoris (73.9%) and with the depth of the neovaginal canal (67.1%). We report a case of gender reassignment surgery in a 32 year old karyotypic male with androgen insensitivity syndrome and brought up as a female since birth.

Case Presentation

A 28 year old phenotypic female patient approached the urological services of the hospital with complaints of primary amenorrhea. She was examined in detail. On examination it was observed that the patient had bilateral hemi-scrotum with a palpable gonad on either side. The external genitalia appeared like a severely hypospadiac phallus (Figure 1). The patient had no facial hair and the body appearance was feminine. The patient's breasts appeared like that of a male adult. Abdominal ultrasonography and laparoscopy revealed no female reproductive organs in the abdomen. Serum free testosterone levels were 7.5 pg/ml and total testosterone was 821.1 pg/ml. Serum LH and FSH levels were within normal range. Blood karyotyping revealed 46XY pattern. The patient was diagnosed to be a case of severe hypospadias with androgen insensitivity.

The patients' relatives and the patient were counselled. The patient wanted to remain a female and more over she had affection and attraction towards male characters. Following a 2 year period

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Received Date: 04 Jan 2019

Accepted Date: 25 Feb 2019

Published Date: 27 Feb 2019

Citation:

Nerli RB, Powar R, Musale A, Jamadar
RK, Deole S, Hiremath MB. Gender
Reassignment Surgery in a Case of
Androgen Insensitivity Syndrome. *Ann
Plast Reconstr Surg.* 2019; 3(1): 1022.

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Figure 1: External genitalia appears like a severely hypospadiac phallus.



Figure 2: a) Space was created for the neo vagina between the urethra and bladder on one side and the rectum on the other side. b) The penile skin cylinder is inverted and closed at its perpetual end, which then is place into the space created for the neo vagina.

of counseling and follow-up, the patient was advised bilateral orchiectomy as the first step. Bilateral orchiectomy was performed and the patient started on female hormones. Following a follow-up period of another year, patient was advised gender reassignment surgery. These surgeries were done in stages due to financial constraints from the patients' side. Gender assignment surgery was performed with help from the Department of Plastic surgery.

Surgery

The patient was placed in lithotomy position. A vertical perineal incision was made from mid-scrotum to a point 1 cm short of the anal margin [5]. The penis was then degloved. The glans penis with neurovascular bundle was separated. Both the corporal bodies were resected completely and also the corpus spongiosum. The central tendon was divided and space was created for the neovagina between the urethra and bladder on one side and the rectum on the other side (Figure 2a). The penile skin cylinder was inverted and closed at its

preputial end which then was placed into the space created for the neovagina (Figure 2b). The position for the urethra and clitoris were marked and button holes were created for the same. The quarter of the glans penis was left uncovered to form the clitoris and rest of it was de-epithelialized and buried below the skin. The urethra was shortened and anastomosed to the skin opening with a catheter in it. The labia majora were created from the scrotum folds. A soft mould was placed within the vaginal tube. Postoperative dressing was changed on alternate days with subsequent vaginal dilatation. The patient was closely followed up and was taught self vaginal dilatation (Figure 3a-3c). At three months the patient was comfortable and was happy with the outcome.

Discussion

The development of a fetus into male occurs only when an XY zygote directs the bi-potential gonad to become a testis (sex determination), which in turn secretes sufficient amounts of active androgen to produce the male phenotype (sex differentiation) [7]. Resistance to the action of androgens results in development of a female phenotype i.e. a hormone- resistance syndrome. Androgen insensitivity syndrome is defined as a disorder resulting from complete or partial resistance to the biological actions of androgens in an XY man or boy with normal testis determination and production of age-appropriate androgen concentrations. The pathophysiology of this syndrome is based on the mechanism of action of androgens [8].

Typically the patients with complete androgen insensitivity syndrome present with either primary amenorrhoea in adolescence or inguinal swellings during infancy. An adolescent with the disorder has breast development and a pubertal growth spurt at the appropriate age, but no menses. Development of oestrogen-dependent secondary sexual characteristics occurs as the result of excess aromatization of androgens. Pubic and axillary hair is usually absent or can be present in sparse amounts. The prevalence of complete androgen insensitivity syndrome ranges from one in 20,400 to one in 99,100 genetic males on the basis of a proven molecular diagnosis [9].

Management of androgen insensitivity syndrome should address functional, sexual, and psychological issues such as disclosure, gonadectomy and subsequent hormone replacement, creation of a functional vagina, and provision of genetic advice [9]. Adults with complete androgen insensitivity syndrome rarely report sexual problems once a reasonable vaginal length has been achieved. If surgery is needed, it should be delayed until consent is given and the woman is able to manage dilator therapy herself after surgery.

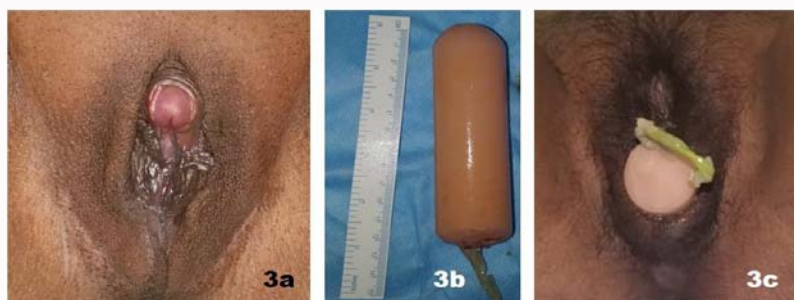


Figure 3: a) Post-operative figure showing reduced clitoris and created labia majora. b) Soft mold used for vaginal dilatation. c) Vaginal dilators used for dilating neo vagina.

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