SEXOLOGICAL AND PSYCHOLOGICAL PROBLEMS AFFECTING WOMEN WITH CONGENITAL MALFORMATIONS OF GENITAL ORGANS – CLINIC OBSERVATION

SEKSUOLOGICZNE I PSYCHOLOGICZNE PROBLEMY U PACJENTEK Z WRODZONYMI WADAMI ROZWOJOWYMI NARZĄDÓW PŁCIOWYCH – OBSERWACJE KLINICZNE

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Abstract

Introduction. The data on psychological status of patients with congenital defects of urogenital tract are very scarce, yet it was confirmed that reactions to the information about diagnosed anomaly and then patients' attitudes toward treatment vary with a kind of malformation, patient's age and personality.
Aim of the study. The aim of the study was to analyze the correlation between the severity of malformation of urogenital organs and psychological status of the patient.
Material and methods. 102 female patients with congenital anomalies of the genital tract, hospitalized in Gynecogical Clinic in the years 1983–2000, were analyzed. All presented patients needed multidirectional diagnostics and treatment. Diagnosis was made based on clinical examination, including psycho-sexological consultation. Malformations diagnosed in the patients were divided into 5 groups.
Results. Malformations diagnosed in the patients were divided into the following groups: Group I: Congenital atresias of the female genital tract (gynatresias) – imperforate hymen; 34 patients; all treated effectively by incision of hymen (hymenectomy). Group II: Vaginal and uterine septa; 11 patients; these malformations develop in the fetal period as a consequence of inappropriate degeneration of paramesonephric ducts' apposed walls. Group III: Duplications of the female genital organs – double uterus and vagina. 13 patients, these malformations arise from complete failure of unification of the paramesonephric ducts in parts that normally join to form uterus and vagina. Group IV: Rudimentary genital tract and genital tract hypoplasia; 15 patients, these malformations follow an incomplete development of paramesonephric Müllerian ducts. Group V: Congenital absence of the vagina and uterus (aplasia) – the Rokitansky syndrome; 29 patients. Awareness of severe malformations, especially those resulting with inability to give birth, had strong impact on patient's psyche.
Conclusions. It is essential to point, that clinical management of patients with malformations of the genital tract, particularly in Rokitansky syndrome, must include psychological counseling. Medical professionals should be aware of psychosocial effects of these disorders.

Keywords: malformation, genitalia, sexuology, psychology, gynecology.

Streszczenie

Wstęp. Dane dotyczące stanu psychicznego pacjentów z wrodzonymi wadami układu moczowo-płciowego są bardzo rzadkie, ale stwierdzono, że reakcje na informację o nieprawidłowości i diagnozowane postawy pacjentów wobec leczenia zależą od rodzaju deformacji, wieku i osobowości pacjenta.
Cel pracy. Obserwacje kliniczne dotyczące związku między rodzajem wad rozwojowych żeńskich narządów płciowych a aspektami psychologicznymi.
Materiał i metody. Analizowano przypadki 102 pacjentek Kliniki Ginekologii (z lat 1983–2000) z wrodzonymi wadami rozwojowymi narządów płciowych, u których przeprowadzano wielokierunkową diagnostykę i leczenie uwzględniając aspekty psychoseksuologiczne. Zależnie o rodzaju wad wyodrębniono 5 grup pacjentek.
Wyniki. Według wyodrębnionych grup: I. Gynatrezje (tzw. „zarośnięcia") 34 pacjentki z brakiem otworu w błonie dziewiczej (hymenektomia zastosowana we wszystkich przypadkach dała dobry rezultat). II. 11 pacjentek z przegrodami w obrębie pochwy i macicy. III.13 pacjentek ze zdwojeniem macicy i pochwy. IV. 15 pacjentek z szczątkowymi narządami płciowymi pochodzącymi z przewodów Miilera. V. 29 pacjentek z agenezją macicy/pochwy, tzw. zespół Rokitanky'ego. We wszystkich przypadkach rozpoznanie wady rozwojowej wiązało się ze stresem i problemami psychoseksualnymi, jednak najmocniej wyraża się to u pacjentek z wadami uniemożliwiającymi odbywanie stosunków płciowych i posiadanie potomstwa – (grupa V).
Wnioski. Pacjentki z wrodzonymi wadami rozwojowymi narządów płciowych wymagają interdyscyplinarnej opieki medycznej I psychologicznej, w sposób szczególny dotyczy to pacjentek z zespołem Rokitansky'ego.

Słowa kluczowe: malformacje, genitalia, seksuologia, psychologia, ginekologia.

Introduction

The Müllerian system, also known as the paramesonephric system, the mesonephric or Wolffian system. It starts cephalad, extends caudad. As it comes down, it is in close approximation to the developing renal system and urinary collection system. This has strong implications in the practices of gynecology because if major anomalies in one of these systems are detected, we will find corresponding anomalies in the other. Teratogenic factors acting about 8–10 weeks of embryonic life are the general cause of these malformations. Extent and intensification of abnormalities depend on kind of evoking factor and time of its activity (gestational age, duration) [1, 2].

Aim of the study

The aim of the study was to analyze the correlation between the severity of malformation of urogenital organs and psychological status of the patient.

Material and methods

102 female patients with congenital anomalies of the genital tract, hospitalized in Gynecogical Clinic in the years 1983–2000, were analyzed. All presented patients needed multidirectional diagnostics and treatment. Diagnosis was made based on clinical examination, including sexological consultation.

Results

Malformations diagnosed in the patients were divided into the following groups:

Group I Congenital atresias of the female genital tract (gynatresias) – imperforate hymen; 34 patients; all treated effectively by incision of hymen (hymenectomy).

Group II Vaginal and uterine septa; 11 patients; these malformations develop in the fetal period as a consequence of inappropriate degeneration of paramesonephric ducts' apposed walls.

Group III Duplications of the female genital organs – double uterus and vagina; 13 patients; these malformations arise from complete failure of unification of the paramesonephric ducts in parts that normally join to form uterus and vagina.

Group IV Rudimentary genital tract and genital tract hypoplasia; 15 patients; these malformations follow an incomplete development of paramesonephric Müllerian ducts.

Group V Congenital absence of the vagina and uterus (aplasia) – the Rokitansky syndrome; 29 patients.

It was proved that reactions to the information about diagnosed anomaly and then patients' attitudes toward treatment are influenced by the type of malformation, patient's age and personality. Awareness of severe malformations, especially those resulting with inability to give birth, had strong impact on patient's psyche.

Discussion

1. Vaginal anomalies

Embryology

After the 17th week of gestation, the female reproductive tract arises from the paired mullerian ducts, which eventually fuse, canalize, and join the urogenital sinus. Further differentiation of these structures results in the formation of fallopian tubes and uterus. The upper vagina is formed from the mullerian duct, whereas the lower one develops from the urogenital sinus. The external female genitalia differentiate after 12 weeks' gestation. The genital tubercle becomes the clitoris, the genital swellings form the posterior fourchette and labia major, and the labia minor arise from the genital folds [1, 2, 3].

2. Absent vagina

Congenital vaginal agenesis, also known as the Mayer-Rokitansky syndrome, develops when the vaginal plate fails to canalize. Patients with this syndrome are normal genetic female patients (46, XX) and present at puberty with primary amenorrhea and normal external female genitalia. Many of these patients have renal anomalies also, including renal agenesis; renal ectopia; or fusion anomalies, including horseshoe kidneys or crossed fused renal ectopia. Other abnormalities include skeletal anomalies. These findings, once seen together, are described as the MURCS (mullerian duct aplasia, renal aplasia, and cervicothoracic somite) association [1, 3, 4].

3. Vaginal obstruction

Obstruction of the vagina most likely results from an incomplete formation of the vaginal canal during gestation. Hydrocolpos is defined as vaginal obstruction only, whereas hydrometrocolpos refers to blockage and distension of the vagina and uterus. Imperforate hymen, vaginal septum, or vaginal atresia may be among the more common anatomic problems. In newborn infants, an abdominal or introital mass may be noted on physical examination. In some cases, urinary tract obstruction may result from extrinsic compression of the bladder or ureters. At puberty, amenorrhea may be the initial presentation. Obstruction of menstrual flow with distension of the vagina or vagina and uterus is termed hematocolpos or hematometrocolpos, respectively. Other abnormalities of vaginal canalization can result in a transverse vaginal septum, with or without obstruction [1, 5].

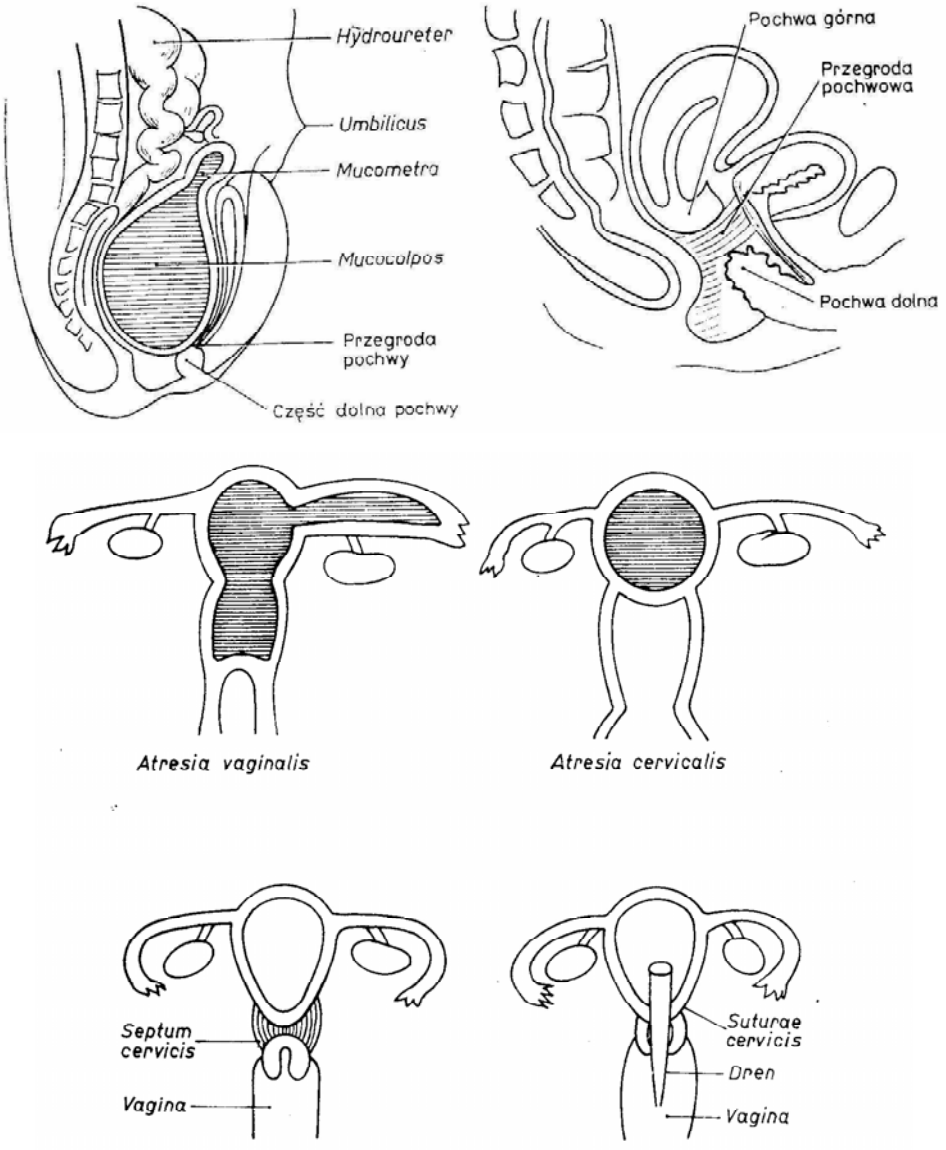

Figure 1. Type of obstruction of the vagina [8]

4. Vaginal or uterine duplication

Disruption of the fusion process of the mullerian structures can result in a variety of duplication anomalies. A result of partial fusion can lead to a bicornuate uterus with a single vagina or complete duplication with uterus didelphys and vaginal duplication. As discussed earlier, various portions of these duplicated segments may be obstructed. Many of these patients have unilateral renal agenesis [1, 6].

Evaluation and Management

Detection of vaginal anomalies depends on the time of presentation, varying from an abdominal mass to bulging introitus in newborn infants to cyclical abdominal pain and amenorrhea at puberty. Obvious findings of an absent or blind-ending vagina lead to the diagnosis of vaginal agenesis. Rectal examination with bimanual palpation in older patients may help to detect masses or asymmetry of the internal genital structures [1, 3, 4]. As noted earlier, sonography, computed tomography, and MR imaging may be helpful in determining the anatomic relationships of the internal structures and, when present, the area of obstruction. Surgical treatment is tailored to specific abnormalities. In cases of vaginal segment obstruction by a transverse web, simple incision may be the only procedure required [1, 5, 6]. In cases of vaginal agenesis, extensive reconstruction, including the use of isolated bowel segments and split thickness skin grafts, may be required. These procedures may require consultation from practicioners of various specialties, including pediatric urology, pediatric surgery, plastic surgery, and gynecology. As in the case of ambiguous genitalia, vaginal anomalies may present complex diagnostic and therapeutic challenges to practitioners [1, 7, 8].

Psychosocial problems

Heller-Boersma et al. [9] who reviewed literature on psychological impact of Rokitansky syndrome suggested that most patients learn about the diagnosis in adolescence. They believed that trauma from distressing diagnostic procedures and psychological adjustment to the new situation (i.e. the prospect of childlessness) may pose major psychological difficulty for the patient. Coming to terms and learning to live with the impairment could be a long process often accompanied by feelings of inadequacy and confusion over one-'s psychosexual identity. Heller-Boersma and her colleagues (9) also studied psychological characteristics of vaginal agenesis patients and controls. They showed that even many years after diagnosis young adults with Rokitansky syndrome were characterized by high levels of depression and phobic anxiety. Additionally, these women had lower self-esteem, considered themselves ineffective and had substantially more feelings of interpersonal alienation and distrust than controls. Their research also confirmed that women with congenital vaginal agenesis were prone to eating disorders, especially to bulimia. Moreover, investigators argued that because of its intimate character surgical procedures for congenital vaginal agenesis such as dilator treatment could add up to feelings of shame and disturbance. In their study the amount of time (i.e. years since diagnosis) was found to have no influence on patient's psychological status. On the other hand, a great deal of women with Rokitansky syndrome who were subjects in Heller-Boersma investigation were able to sustain satisfying relationships with their partners. Furthermore, authors found the future for women with this gynecological problem very optimistic because with the development of in vitro fertilization and surrogacy they would have a good chance of overcoming infertility, having biological children of their own and living a full life.

Conclusions

It is essential to point, that clinical management of patients with malformations of the genital tract, particularly in Rokitansky syndrome, must include psychological counseling. Medical professionals should be aware of psychosocial effects of these disorders.

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