ORIGINAL RESEARCH

Psychosexual Well-Being after Childhood Surgery for Anorectal Malformation or Hirschsprung's Disease

Desiree van den Hondel, MD,* Cornelius E.J. Sloots, MD, PhD,* Johanna M. Bolt, MD,†
Rene M.H. Wijnen, MD, PhD,* Ivo de Blaauw, MD, PhD,‡ and Hanneke IJsselstijn, MD, PhD*

*Department of Pediatric Surgery, Erasmus MC—Sophia Children's Hospital, Rotterdam, The Netherlands; †Department of Reproduction and Development, Erasmus MC, Rotterdam, The Netherlands; ‡Department of Pediatric Surgery, Amalia Children's Hospital, Radboudumc, Nijmegen, The Netherlands

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ABSTRACT

Introduction. Anorectal malformations (ARMs) and Hirschsprung's disease (HD) are congenital malformations requiring pelvic floor surgery in early childhood, with possible sequelae for psychosexual development.
Aims. To assess psychosexual well-being in adult ARM and HD patients related to health-related quality of life.
Methods. Eligible for this cross-sectional two-center study were all patients aged ≥18 years who had been operated for ARM or HD. Exclusion criteria were intellectual disability, comorbidity affecting sexual functioning, and cloacal malformation.
Main Outcome Measures. Participants completed the International Index of Erectile Functioning, Female Sexual Functioning Index, Female Sexual Distress Scale, Hirschsprung and Anorectal Malformation Quality of Life Questionnaire, and sexual education questionnaire.
Results. Response rates were 32% and 37% for ARM and HD patients, respectively. We studied 70 participating ARM and 36 HD patients (median age 26 years). We excluded 10 patients with sexual inactivity in the past 4 weeks. Six of 37 men with ARM (16%) reported moderate to severe erectile dysfunction, vs. two of 18 men with HD (11%). Thirteen and 10 of 26 women with ARM (50% and 38%) reported sexual dysfunction or sexual distress, respectively, vs. eight and three of 15 women with HD (53% and 20%). Quality of life and type of malformation or operation were not associated with self-reported psychosexual problems. Addressing sexuality with special interest to the congenital anomaly during medical care was reported to be insufficient by 42 ARM (60%) and 22 HD patients (61%).
Conclusion. Approximately 13% of male ARM and HD patients reported erectile dysfunction, while 50% female ARM and HD patients reported sexual dysfunction not related to quality of life or type of malformation. Both ARM and HD patients felt a need for better addressing sexual concerns during medical care. Further research is needed to optimize form and timing of this education. **van den Hondel D, Sloots CEJ, Bolt JM, Wijnen RMH, de Blaauw I, and IJsselstijn H. Psychosexual well-being after childhood surgery for anorectal malformation or Hirschsprung's disease. J Sex Med 2015;12:1616–1625.**

Key Words. Anorectal Malformation; Hirschsprung's Disease; Psychosexual Functioning; Sexuality

Introduction

Anorectal malformations (ARMs) and Hirschsprung's disease (HD) are rare congenital colorectal anomalies. Almost all children with these anomalies require pelvic floor surgery in early childhood. In ARM patients, the pelvic floor is hypoplastic [1], and often a fistula is present from the rectum to perineum or to the urogenital tract. Associated anomalies are frequently of urogenital origin, such as hypospadias—present in 26 to 40% of the male ARM patients [2–4]—or of the internal reproductive system in females [5]. In HD, the distal colon is functionally obstructed due to

aganglionosis of the last part of the colon, and associated congenital anomalies are less prevalent [6].

Children operated upon for congenital anomalies are now usually being followed into adolescence. In ARM and HD patients, this follow-up generally focuses on continence because constipation and soiling are still everyday problems (7–40% and 5–45%, respectively) [7,8]. However, ARM and HD patients might also be at risk for experiencing psychosexual problems as well, considering the pelvic floor surgeries in both diseases and considering the hypoplastic pelvic floor and urogenital comorbidity in ARM patients. Three studies indeed indicate some form of impaired sexual functioning (e.g., erectile dysfunction, vaginal stenosis or dyspareunia) both in ARM and HD patients [9–11]. These studies were however conducted in small patient groups, and results were mostly obtained through self-developed questionnaires, so these results cannot be unequivocally extrapolated. It is important to obtain more detailed information on psychosexual well-being so that we can anticipate in current treatment of psychosexual problems, and so, we can improve education and counseling of future ARM and HD patients and their parents.

Aims

The aims of this study were threefold: (i) to investigate whether ARM and HD patients experience sexual dysfunction or sexual distress in adulthood; (ii) to investigate whether these problems are related to the type of malformation or to the quality of life; and (iii) to determine whether ARM and HD patients in their follow-up period missed that their caregiver addressed sexuality and sexual concerns with special interest to the congenital anomaly during medical care.

Methods

Study Sample

All patients treated for ARM or HD in one of the two participating university pediatric surgery centers (Erasmus MC—Sophia Children's Hospital, Rotterdam and Amalia Children's Hospital, Radboudumc, Nijmegen, the Netherlands) who were at least 18 years of age at time of the study (i.e., born in 1995 or before) were eligible for inclusion. Excluded were patients with a cloacal malformation, intellectual disability (as documented in the medical records), or severe comorbidity affecting sexual functioning, for example, malignancies that required chemotherapy. Patients were identified by a systematic search in the electronic patient files and in databases of patients with congenital anomalies kept by the pediatric surgeons. Patient characteristics were obtained from the medical records. This study was approved by the Erasmus MC Medical Ethical Review Board and the Radboudumc Medical Ethical Review Board. Patients with ARM and HD were analyzed separately because of the great difference in etiology and associated anomalies.

Classifications and Procedures

ARMs were classified according to the Krickenbeck classification [12]. Surgical techniques were described as mentioned in the medical records: anoplasty, posterior sagittal anorectoplasty (PSARP), abdominosacroperineal pull through operation (i.e., with laparotomy), and other/unknown operations.

In HD, the length of the aganglionic segment was retrieved from the surgical records and pathology reports and was classified as short (rectosigmoid), long (descending, transverse, and ascending colon), or total colonic aganglionosis. The surgical techniques were described as mentioned in the records and included Rehbein operation, Duhamel operation, and other operations.

Chronic urogenital comorbidity or complications (both further referred to as chronic urogenital comorbidity) that could have a negative influence on sexuality were documented. These included hypospadias, recurrent urinary tract infections, neurogenic bladder requiring intermittent catheterization, and other congenital anomalies of the genital tract (excluding nondescended testes and phimosis).

Questionnaires

A letter was sent to the patient's last known address with general information on the purpose of this study, and the patients were invited to return a reply card. Those who returned the reply card were sent more detailed study information, the patient consent form, and questionnaires. The latter two could be returned in a prestamped envelope. The following questionnaires were used:

International Index of Erectile Functioning (IIEF-15)

The IIEF-15 contains 15 questions covering five domains: erectile function (EF), orgasm, desire,