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Predicting sexual problems in young adults with an anorectal malformation or Hirschsprung disease

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ABSTRACT

Aim: The aim of this study was to examine the prevalence of sexual dysfunction and distress and to assess whether sexual functioning could be predicted by psychosocial factors in childhood and adolescence in patients with an anorectal malformation or Hirschsprung disease.
Material and methods: In 1998 patients completed a psychosocial questionnaire: The Self-Perception profile. To assess the prevalence of sexual distress and sexual functioning in adulthood (2015) the Female Sexual Function Index (FSFI), The Female Sexual Distress Scale (FSDS) and the International Index of Erectile Functioning (IIEF) were used.
Results: In total 74 patients returned the questionnaires (26.2%). 36.8% of women reported sexual dysfunction and 45% experienced sexual distress. In our male sample 8.8% reported mild to moderate erectile dysfunction. In females perceived self-competence in adolescence was associated with sexual distress ($p < .01$). In male adolescents associations were found between perceived self-competence in romantic relations ($p < .01$) and in close friendships ($p < .05$) and sexual desire in adulthood.
Conclusion: Pediatric surgeons should be more aware of sexual problems patients may face at older age as a result of their congenital disease and treatment. More standardized care and follow-up are needed.
Prognosis study–Level II.

Each year approximately 80 children with an anorectal malformation (ARM) or Hirschsprung disease (HD) are born in the Netherlands [1,2]. ARM comprises a wide spectrum of anomalies of the distal anus and rectum, as well as the urogenital tract. The malformation can be as little as an anal membrane, but it can also present with a rectourethral, rectovesical or rectovestibular fistula and even with a cloacal malformation. The severity of the malformation, choice of operative procedure and additional anomalies, such as severe sacral dysplasia, can be of influence on the functional prognosis of these patients [3]. Mild ARMs are known to carry a much better prognosis than severe ones (e.g. rectourethral, rectovestibular and cloacal malformation) [4].

In HD there is a variable length of aganglionic, nonperistaltic colon, which is surgically resected soon after birth. The disease can be limited to the distal rectum and sigmoid colon, but can also involve the entire colon and even include the small intestine. We know from literature that in this patient population functional problems of varying degrees may be encountered, with some suggestion of improvement in symptoms over time [5–9].

The outcome of patients with these congenital anomalies has improved significantly in the last decades. This is the result of improved neonatal care, and improved and safer surgical techniques, which are based on anatomical reconstruction without blind dissections. This is again the result of a better understanding of the pathological anatomy and physiology of these defects [10]. Despite this improvement, functional problems (fecal or urinary incontinence, soiling and constipation) can still be a concern for some patients reaching adulthood. However it is suggested that many patients with mild ARMs and patients with HD, treated with a TEPT, can have a largely normal functional outcome by adulthood [8,11,12].

Functional problems can have an impact on (heath-related) QoL of patients [10,13–15]. But patients with bowel symptoms can also report a normal QoL.

As patients are followed more often into adulthood, other problems are faced. Sexual function has been described to be impaired in patients with ARM and HD. This may be caused by associated urogenital anomalies (26% of males, 30%–35% of females) in patients with ARM or by the extensive pelvic surgery in childhood [16–21]. In patients with HD, urogenital anomalies are generally absent. However older patients with HD often had extensive pelvic surgery, resulting in iatrogenic

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injuries which caused urological and sexual problems. Recently a study was published that stated that the TEPT did not result in more urological and sexual problems than healthy matched controls [22].

During the last decades there has been an increased interest in researching psychosocial factors that can lead to problems in sexual functioning. A study from the early eighties has shown us that the ability to "date" during adolescence is positively correlated with normal sexual development [23]. It has been shown that higher levels of self-competence and self-esteem contribute positively in developing normal romantic and (sexual) relationships [24]. We know that children and adolescents with ARM or HD can have lower self-competence [25]. This could be the cause of problems in starting (sexual) relationships and friendships, in adulthood. These specific relationships and factors between psychosocial functioning and sexual functioning in ARM and HD patients have hardly been studied. To date research into predicting and preventing problems in sexual functioning owing to these psychosocial factors is nonexistent.

The aim of the present study was dual: to examine the prevalence of sexual dysfunction and distress in ARM and HD patients and to examine whether disease type and perceived self-competence measured in childhood or adolescence were predictive for sexual functioning in adulthood.

1. Material and methods

1.1. Patients

This is a follow-up study of the NAHO study, A study on psychosocial functioning and quality of life (QoL) in patients with ARM and HD performed in 1998. Exclusion criteria were lack of understanding the Dutch language, mental retardation or Down syndrome. In addition, patients who were lost to follow-up or deceased were excluded. In that study 316 patients (164 ARM and 152 HD), between 8 and 16 years old, from all pediatric surgical centers in The Netherlands (6 centers) returned the questionnaires and were included in the study [25–28].

In 2015 we started a follow-up study concerning sexual functioning of patients included in the NAHO study in 1998 and to examine whether self-competence, in childhood or adolescence, was a predictor for sexual functioning in adulthood. All patients who had been included in the NAHO study and could be traced in 2015 were sent questionnaires about sexual functioning and an informed consent letter. Data were only used when both questionnaires and a signed informed consent form were returned. This was a multicenter study. Approval for this study was given by the local medical ethical committee, and was valid for all participating hospitals.

Almost all patients were not in regular follow-up at the time of this study. During childhood and adolescence they did receive systematic surgical follow-up by a pediatric surgeon.

1.2. Perceived self-competence in childhood and adolescence (NAHO 1998)

Perceived self-competence was assessed by the Self-Perception profile. Two versions were used: The Self-Perception Profile for Children (SPC, age 8–13) and the Self-Perception Profile for adolescents (SPA, age 14–19). The SPC consists of 36 items and the SPA of 40 items. These items are divided in subscales, covering social acceptance, behavioral conduct, scholastic competence, athletic competence, physical appearance, global self-worth, romantic relations (only SPA) and close friendships (only SPA). Each item is scored with a range of 1–4, with higher scores indicating higher perceived self-competence. The SPC and SPA have good psychometric qualities including good internal consistency, test–retest stability and a theoretically meaningful correlation with child-, parent- and teacher-reports of psychopathology and personality [29].

1.3. Sexual functioning in adulthood (follow-up study 2015)

1.3.1. Female sexual functioning

The Female Sexual Function Index (FSFI) assesses female sexual functioning over the past month. The FSFI consist of nineteen items divided into six domains, covering sexual desire, arousal, lubrication, orgasm, satisfaction and pain. Individual items are scored on a 5-point Likert scale, with a sixth option when there was no sexual activity over the past month. The score range of the FSFI is 2–36 with a maximum score of 6 per domain. A higher score indicates better sexual functioning. A score 26 or less is defined as sexual dysfunction [30]. The FSFI is a valid and reliable instrument with good internal consistency on the total score scale ($\alpha = 0.79$) [31].

The Female Sexual Distress Scale (FSDS) is used to measure female sexual distress. The FSDS consists of twelve items from which a total score is calculated. The score range is 0–48, with higher scores indicating higher distress. Scores of 15 or higher are defined as having sexual distress [32]. The internal consistency and stability has proven to be very good [31].

1.3.2. Male sexual functioning

The International Index of Erectile Function (IIEF) assesses male sexual functioning in the past month. The IIEF consists of fifteen items divided into five domains, covering erectile function, orgasmic function, sexual desire, intercourse satisfaction and overall satisfaction. Individual items were scored on a 5-point Likert scale with a sixth option when there was no sexual activity in the past month. The score range on the IIEF is 5–75. For the separate domains the score ranges are 1–30 for erectile function, 0–10 for orgasmic function, 2–10 for sexual desire, 0–15 for intercourse satisfaction and 2–10 for overall satisfaction. A higher score indicates a better sexual functioning. A total score on the erectile function scale of 5–7 points is defined as severe erectile dysfunction, 8–11 as moderate, 12–16 as mild to moderate and 17–21 points as mild erectile dysfunction. The IIEF has a good internal consistency and adequate construct validity [33].

1.4. Disease classification and performed operation

Anorectal malformations were classified according to the Krickenberg International Classification system [34]. Surgical techniques that were used were: posterior sagittal anorectoplasty (PSARP), anoplasty, anterior sagittal anorectoplasty (ASARP), cut-back, posterior sagittal anorectovaginourethroplasty (PSARVUP) or other. HD was classified according to the length of the aganglionic segment of the bowel. When the aganglionosis was maximum up to the splenic flexure it was defined as short segment. If the aganglionosis was found beyond the splenic flexure it was defined as long segment. Surgical techniques that were used were: Duhamel, Rehbein or Lester–Martin procedure.

1.5. Statistical analysis

To examine the role of disease type on the development of sexual dysfunction, differences in sexual functioning between HD and ARM patients were examined by conducting Mann–Whitney U tests. Cohen's d effect sizes were calculated for all Mann–Whitney U tests and then categorized.

To examine whether self-competence in childhood or adolescence was predictive for sexual functioning in adulthood Spearman's/Pearson rank order correlation coefficients were calculated. Dependent variables were female sexual functioning and distress and male sexual functioning (overall scores and separate items). A p-value of <0.05 was considered significant.

All analyses were performed with the Statistical Package for Social Sciences (SPSS version 23).

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