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Mental Health, Psychosocial Functioning, and Quality of Life in Adolescents With Hirschsprung Disease

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ABSTRACT

Background: Studies of mental health in adolescents with Hirschsprung disease (HD) are scarce. This cross-sectional study investigates mental health, psychosocial functioning and quality of life in HD adolescents.
Methods: Adolescents (12–18 years) treated at the Department of pediatric surgery at Oslo University Hospital were invited for participation. Mental health was assessed by interview; Child Assessment Schedule (CAS) and questionnaires; parental Child Behavior Checklist (CBCL) and adolescent Youth Self-Report (YSR). Psychosocial functioning was rated by Child Global Assessment Scale (cGAS). Adolescent Quality of Life was assessed by Pediatric Quality of Life inventory (PedsQL) and chronic family difficulties (CFD) by interview. Medical records were reviewed for somatic history.
Results: Thirty-seven adolescents, 28 males, median age 14.3 years, participated. By CAS interview, 8 of 37 (44% of females and 14% of males) fulfilled criteria for psychiatric diagnosis all within emotional and related disorders. Twenty-seven percent had CBCL internalizing scores and 16% had YSR internalizing scores in clinical range indicating emotional problems. By interviewer rated cGAS, 27% were scored in clinical range. By PedsQL 16% reported reduced psychosocial health score. Increased CFD, lower psychosocial functioning and reduced QoL as well as less paternal education were significantly associated with psychiatric diagnosis. Twice as many (4/8) adolescents who either had a stoma or bowel management had a psychiatric diagnosis compared to those who had neither stoma nor bowel management (7/28).
Conclusion: Nearly one in four adolescents with HD fulfilled criteria for psychiatric diagnosis. Mental health problems were associated with reduced psychosocial function and reduced QoL.
Level of evidence: III.

1. Introduction

Hirschsprung disease (HD) is a congenital disorder characterized by lack of ganglion cells most commonly in the distal bowel, but can be variable in the length of involvement affecting approximately one in 5000 live births [1–4]. Surgical management of HD usually involves removal of aganglionic bowel and restoration of bowel continuity early in life. It is well known that many HD children experience impaired bowel function and reduced quality of life (QoL) [5]. Even though many HD children experience improved bowel function and QoL as they get older, impairment may persist into adolescence [6–10].

Generic QoL covers subjective experiences such as satisfaction, joy, mastery and meaning, as well as the level of psychological distress and negative emotions. Mental health assessment provides a more detailed description of possible psychosocial, cognitive, behavioral and emotional problems and psychiatric diagnoses. A psychosocial functioning score is based on mental health assessment. This assessment of mental health and psychosocial functioning is time demanding and should involve an experienced child- and adolescent

Abbreviations: ASEBA, Achenbach System of Empirically Based Assessment; CAS, Child Assessment Schedule; CBCL, Child Behavior Checklist; CFD, Chronic family difficulties; cGAS, Child Global Assessment Scale; HD, Hirschsprung disease; PedsQL, Pediatric quality of life inventory; QoL, Quality of life; YSR, Youth Self-Report.

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psychiatrist/psychologist. Validated interviews are the gold standard when examining mental health and psychosocial functioning. Thus, mental health assessment and diagnostics provide an in depth evaluation of problems patients may have and which QoL assessment does not evaluate.

Several studies have explored both generic and disease specific QoL in HD patients, and the majority find that QoL in most HD patients is negatively affected if bowel function is impaired [1–4]. While QoL in HD patients is often studied, assessment of mental health and psychosocial functioning is limited[7,11–16]. A Norwegian study performed 26 years ago reported interview-based mental health assessment and psychosocial functioning in 19 HD adolescents and found no difference between HD adolescents and a general population sample [12]. A British study from 2002 compared emotional and behavioral problems in 15 children/adolescents with total colonic aganglionosis (TCA) and 15 with rectosigmoid aganglionosis (RSA) [14]. Based on questionnaire reports Ludman et al. found that the TCA patients reported more emotional and behavioral problems than those with RSA [14].

Due to the lack of comprehensive data on mental health and psychosocial functioning in young people with HD, we aimed to assess mental health, psychosocial functioning, and generic QoL in HD adolescents. Secondly, factors which could impair mental health and generic QoL were explored.

2. Patients and methods

2.1. Study design, patient population, surgery, and follow-up routines

This is a single-center cross-sectional study. Guidelines for Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) were applied [17].

Non-intellectually impaired adolescents, aged 12–18 years, having undergone surgery for HD and/or follow-up at department of pediatric surgery at Oslo University Hospital were eligible for inclusion. The department is a tertiary referral center and member of the European Reference Network for Rare and Inherited Congenital Anomalies (ERNICA) [18]. Adolescents were identified from surgical logbooks and the hospital's electronic database. They were invited to participate by mail or during outpatient clinics. In 2020, 12–16 years adolescents were invited to participate, and in 2021 HD adolescents up to 18 years were invited to increase sample size.

Patients were operated with various surgical techniques depending on the length of the aganglionic segment and the surgeons' preference (Table 1) [19–22].

The anastomosis in patients operated with a transanal endorectal pull-through, was calibrated after two to three weeks and then according to surgeons' preference, but at least a few times during the first two months. If needed, the parents performed anal dilatations/calibrations at home. Patients having undergone a Duhamel procedure had the anastomosis examined around one month postoperatively. Postoperative follow-up included frequent outpatient visits the first year and thereafter depending on symptoms and availability for follow-up at the patients' local pediatric department. A multidisciplinary team, as also recommended by Vilanova-Sanchez et al. [23], was available for the patients during follow-up in the present study.

Table 1
Clinical characteristics of adolescents with Hirschsprung disease[a].

N	37
Males	28 (76%)
Age at interview, (years)	14.3 (12.3–18)
Age at diagnosis, (months)	
Males	1 (0–73)
Females	2 (0–58)
Length of aganglionosis	
Rectosigmoid	30 (81%)
Long segment and total colon aganglionosis	4 (11%)
Near total intestinal aganglionosis	3 (8%)
Surgery	
Transanal pull-through (±laparoscopy)	27 (73%)
Duhamel	6 (16%)
Initial stoma	3 (8%)
Rehbein	1 (3%)
Hospital admissions >2 weeks, before 5 years	22 (59%)
Longest hospitalization before 5 years, (days)	17 (0–413)
Anal dilatations/calibrations by parents	7 (19%)
Duration, (months)	2 (0.75–7.75)
Age at cessation, (months)	5 (4–12)
Bowel management or stoma	12 (32%)
Fecal incontinence	8 (22%)

[a] Continuous variables; median (min–max), dichotomous variables numbers (percent).

2.2. Collection of somatic data

Patient demographics, including length of aganglionosis and operative technique were retrospectively retrieved from medical records. In addition, disease related factors that might impact mental health and psychosocial functioning were recorded. These included age at diagnosis, number of hospitalizations before 5 years of age, length of hospitalizations, anal dilatations/calibrations performed by parents, presence of a stoma, fecal incontinence, and use of bowel management. A pediatric surgeon (ATH) who had not operated or been involved in follow-up of the adolescents, recorded current clinical status in the outpatient clinic. TCA was defined as aganglionosis extending to maximum 50 cm of ileum, whereas ultra-long aganglionosis described aganglionosis proximal to the last 50 cm of ileum. Fecal incontinence was scored dichotomously by the adolescents as either having or not having fecal incontinence limiting daily life. Anal dilation/calibration by parents was defined as a daily or weekly procedure using a Hegar dilator or a finger at home. In retrospect, it was impossible to decide whether the procedure was performed as a prophylactic calibration to avoid stricture or dilatation of a postoperative stricture. Bowel management denotes regular use of either antegrade or retrograde enemas.

2.3. Evaluation of mental health and psychosocial functioning

Adolescent mental health was assessed by a child- and adolescent psychiatrists (RH or HG) during a semi-structured interview, the Child Assessment Schedule (CAS). The parents were interviewed separately by child- and adolescent psychiatrist (HG) about family functioning, psychosocial issues and adolescent health in general.

The interviews took place between January 2020 and December 2021. Due to the Covid-19 pandemic in 2020/2021 and travel restrictions, some CAS interviews were conducted by video consultation. The interview procedure was similar for digital and physically attended interviews. The digital interviews were performed using a secure, double encrypted video link.

2.4. Interview assessment

2.4.1. Child Assessment Schedule (CAS)

The semi-structured interview CAS has been shown reliable and valid [24]. First, the adolescent answers questions about school, activities, friends, hobbies, worries, family, fears, mood, self-image, expression of anger, somatic concerns, and psychotic symptoms.

The second part of the interview is an in-depth interview with questions related to duration and severity of mental health related problems identified during the first part of the interview. A psychiatric diagnosis was generated based on the CAS interview if the adolescent fulfilled diagnostic criteria according to the International Diagnostic and statistical Manual of Mental health Disorders 4th edition (DSM IV).

2.4.2. Child Global Assessment scale (cGAS)

Based on the CAS response, the child- and adolescent psychiatrist scored the adolescent's overall psychosocial functioning according to cGAS. The scale is a continuous measure from one to 100 with scores ≤70 indicating significant clinical problems [25]. The cGAS measure is widely applied in child and adolescent mental health services [26].

2.4.3. Chronic family difficulties (CFD)

During interviews with the parents, the family situation was scored with a sum score of chronic family difficulties (CFD) based on family economy, parental employment situation, housing conditions, psychosocial support, family health, parental relation, and parental collaboration on child upbringing. Each aspect is scored from nil to six, where nil means no psychosocial strain, and six means huge strain. Forty-two is the highest possible total difficulty score. A high CFD score has been reported to have a significant negative effect on psychosocial functioning in adolescents [27].

2.4.4. Questionnaire assessment

2.4.4.1. Achenbach system of empirically based assessment (ASEBA). ASEBA consists of comprehensive questionnaires screening for internalizing (emotional) and externalizing (behavioral) problems in children and adolescents. The parental report, Child Behavior Checklist for ages 6–18 (CBCL 6–18), and Youth Self-Report (YSR) assess behavioral problems and adaptive functioning, strengths, and competence [28,29]. Scoring ASEBA yields standardized total scores (T-scores) based on American norms as there are no specific Norwegian norms for the ASEBA. For the internalizing (emotional) and externalizing (behavioral) grouping of problems T-scores <60 are considered to be in the normal range. T-scores between 60 and 63 denote borderline range, meaning possible mental health problems, whereas T-scores >63 are in the clinical range. Clinical range indicates mental health problems [28]. In the present study, patients with borderline T-scores are analyzed together with patients having T-scores in the clinical range as T-scores >60 may be indicative of mental health problems and need for further diagnostic assessment [28].

2.4.4.2. Pediatric quality of life inventory (PedsQL). The psychometric properties of the Norwegian version of PedsQL 4.0 have been evaluated in the general population and found satisfactory [29]. The generic self-report PedsQL is applied in the present study. The questionnaire consists of 23 items on health status with self- and parental report forms. PedsQL assesses four domains of health (emotional-, social-, physical- and school-functioning) which is summarized in a total score. The psychosocial score of the PedsQL questionnaire includes 15 questions from the three domains emotional-, social-, and school-functioning. A score of 100 points indicates optimal QoL, while scores ≤70 indicates reduced QoL based on previous studies in Norwegian adolescents [29].

2.5. Statistics

To investigate the relationship between psychiatric diagnosis after CAS interview, mental health assessed by questionnaire reports (CBCL, YSR), and PedsQL against possible explanatory factors, cross table and bivariate analyses were performed with IBM SPSS software for windows version 28. Due to non-normality, nonparametric tests were applied in bivariate comparisons. A p-value <0.05 was considered statistically significant. Continuous variables are presented as median (min–max) and analyzed with Mann Whitney U test and categorical variables with Pearson chi square test as appropriate.

The agreement between the interviewers (reliability) were analyzed by intra class correlation (ICC) for CAS continuous scores, and with Cohens Kappa for CAS diagnostic agreement. Both measures range 0–1.00, the closer to 1.00 the higher agreement between raters.

2.6. Ethics

The study was approved by the Regional Ethics Committee for Medical and Health Research Ethics Norway (2018/2009). Written informed consent was obtained from all adolescents and parents. The hospital's Data Protection Officer also approved (18/19101).

3. Results

3.1. Demographics

Of 50 identified HD adolescents, five had intellectual impairment, two were lost to follow up, and six declined. Consequently, 37/43 (86%) of eligible adolescents participated.

The majority were males with RSA (Table 1). There were 5/9 (56%) females and 2/28 (7%) males with aganglionosis extending past the rectosigmoid colon. None of the patients had undergone bowel transplantation. One patient received parenteral nutrition for less than 5 nights per week at the time of follow-up.

The families in this study were characterized by highly educated parents, 62% of mothers and 50% of fathers had a bachelor degree or higher as compared to 40% and 35% in the general population. In line with this, 97% of HD parents had a full time job in comparison to 70% in the general population. There was no difference between HD adolescents and general population regarding living with both parents (78% vs 77%) [30].

3.2. Interviews

Twenty-seven of 37 (73%) were interviewed at the hospital and 10/37 (27%) by video link. The CAS-interviews lasted from one to 3 h, independent on whether the interview took place at the hospital or on the digital platform (results not shown). The majority (34/37) of the interviews were performed during the Covid-19 restrictions.

3.3. Mental health and psychosocial functioning

Of the 37 adolescents, 8 (22%) fulfilled DSM IV criteria for a psychiatric diagnosis. The diagnoses were within emotional problems such as anxiety, depression, phobia, and functional/conversion (Table 2). These findings corresponded well with the self- and parental-reports from the ASEBA questionnaire who showed increased problem scores for internalizing (emotional) problems (Table 2).

Ten of 37 (27%) adolescents had cGAS ≤70 indicating clinically significant problems in psychosocial functioning (Table 3).

With the present DSM IV revision the ICC rater 1 and 2 was 0.986 for CAS symptom score. For psychiatric diagnosis by CAS interviews Kappa $\kappa = 1$ indicated complete agreement on diagnosis between the two raters. The ICC for cGAS in the present study was 0.94 indicated a strong agreement between the raters.

Table 2
Mental health, psychosocial functioning and quality of life in adolescents with Hirschsprung disease[a].

Mental health	
CAS score[b] (n = 37)	
Total score, problems	15 (1–78)
Total score, symptoms	11 (1–63)
DSM IV diagnosis[c] (n = 37)	8 (22%)
Generalized anxiety disorder	2 (5%)
Social anxiety disorder	2 (5%)
Unspecified anxiety disorder	1 (3%)
Recurrent depression (moderate)	1 (3%)
Specific isolated phobia	1 (3%)
Other specified dissociative disorder	1 (3%)
ASEBA - CBCL score[d] (n = 33)	
Internalizing score	10 (27%)
Externalizing score	1 (3%)
ASEBA - YSR score[e] (n = 32)	
Internalizing score	6 (16%)
Externalizing score	1 (3%)
Psychosocial functioning	
cGAS[f] (n = 37)	82 (41–100)
Quality of life[g]	
Self-report (n = 31)	
Total score	90.9 (61.4–100)
Psychosocial score	88.3 (61.7–100)

[a] Continuous variables; median (min–max), dichotomous variables numbers (percent).
[b] Child Assessment Schedule.
[c] Diagnosed by statistical Manual of Mental health Disorders, 4th edition after CAS interview.
[d] Child Behavior Checklist, parental reports and adolescents with T-score in borderline/clinical range ≥60.
[e] Youth Self- Report, adolescents with T-score in borderline/clinical range ≥60.
[f] Children's Global Assessment scale.
[g] The PedsQoL questionnaire (self-report).

3.4. Quality of life

Six of 31 (19%) adolescents reported reduced QoL with PedsQL total score ≤70, and 5/31 (16%) had psychosocial score ≤70 (Table 2).

3.5. Factors associated with impaired mental health and reduced QoL

Median CFD score 5 (0–32), indicated low psychosocial strain for many families and adolescents.

Low paternal education, increased CFD score, cGAS ≤70, and self-reported PedsQL total and psychosocial score ≤70 were all significantly associated with fulfilling criteria for psychiatric diagnosis (Table 3). The association between diagnosis and lower education level for mothers did not reach significance (Table 3). There was a trend towards higher frequency of having a psychiatric diagnosis in females (44%) compared to males (14%) (Table 3). Disease and treatment related factors like anal dilatation/calibration performed by parents, bowel management/permanent stoma, fecal incontinence, age at HD diagnosis, length of aganglionosis, number of hospitalizations ≤ 5 years of age, and duration of longest hospitalization during the first five years were not significantly associated with adolescent mental health problems. Nevertheless, relatively more adolescents with aganglionosis proximal to rectosigmoid, or experiencing bowel management or stoma had a psychiatric diagnosis (Table 3).

Analysis with dichotomized PedsQL psychosocial score against factors associated with psychiatric diagnosis revealed no significant association with CFD. The association with paternal education in years persisted. Relocation last year and female sex were significantly associated with reduced psychosocial QoL score (Table 4).

Table 3
Comparison of possible risk factors affecting mental health in adolescents with or without a psychiatric diagnosis[a].

	N	Psychiatric diagnosis	No psychiatric diagnosis	p
Demographics				
Sex (Male)	37	4/8 (50%)	24/29 (83%)	0.056
Lives with both parents[a]	36	5/8 (63%)	24/28 (86%)	0.218
Relocation last year[a]	37	0/8 (0%)	5/29 (17%)	0.198
Age at interview, (years)[b]	37	14.8 (12.3–18)	14.3 (12.8–17.4)	0.580
Maternal education, (years)[b]	36	13.5 (10–16)	15.5 (9–20)	0.080
Paternal education, (years)[b]	35	12 (10–15)	15 (1–20)	0.010*
Chronic family difficulties	37	12 (0–32)	4 (0–15)	0.031*
Medical history				
Anal dilatation/calibration by parents[a]	36	1/8 (13%)	6/28 (21%)	0.574
Bowel management or stoma[a]	36	4/8 (50%)	7/28 (25%)	0.176
Fecal incontinence[a]	37	2/8 (25%)	6/29 (21%)	0.368
Age by HD diagnosis, (months)[b]	37	0 (0–58)	1 (0–73)	0.583
Length of aganglionosis[a]	37			0.130
Rectosigmoid		5/8 (63%)	25/29 (86%)	
Proximal to rectosigmoid		3/8 (38%)	4/29 (14%)	
Number of hospitalizations ≤ 5 years[b]	37	3.5 (2–9)	5 (0–13)	0.490
Longest hospitalization ≤ 5 years[b]	37	14 (7–87)	17 (0–413)	0.618
Child Global Assessment Scale (cGAS) ≤70[a]	37	6/8 (75%)	4/29 (14%)	<0.001*
Quality of life[a]				
Self-reported total score ≤70	31	3/6 (50%)	3/25 (12%)	0.003*
Self-reported psychosocial score ≤70	31	3/6 (50%)	2/25 (8%)	0.012*
Miscellaneous factors[a]				
Other chronic diseases	37	0/8 (0%)	6/29 (21%)	0.160
Impaired vision and/or hearing	37	4/8 (50%)	6/29 (21%)	0.098
School problems	37	4/8 (50%)	7/29 (24%)	0.157
Ever bullied	36	1/8 (13%)	9/28 (32%)	0.274

*p < 0.05.
[a] n (percent), Pearson chi square test.
[b] Median (min–max), Mann Whitney U test.

Table 4
Possible risk factors affecting psychosocial Quality of life in adolescents.

Self-report	N	Psychosoc QoL≤70	Psychosoc QoL >70	P
Demographics				
Sex (male)[a]	31	2/5 (40%)	22/26 (85%)	0.029*
Lives with both parents[a]	30	5/5 (100%)	19/25 (76%)	0.221
Relocation last year[a]	31	2/5 (40%)	2/26 (8%)	0.048*
Age at interview, (years)[a]	31	14.5 (13.3–16.6)	14.3 (12.3–17.9)	0.979
Maternal education, (years)[b]	30	12 (9–16)	16 (10–20)	0.057
Paternal education, (years)[b]	29	10 (7–17)	15 (1–19)	0.003*
Chronic family difficulties (0–42)[b]	31	2 (0–10)	5 (0–32)	0.548
Medical history				
Anal dilatation/calibration by parents[a]	31	0/5 (0%)	3/25 (12%)	0.414
Bowel management or stoma[a]	30	2/4 (50%)	8/26 (31%)	0.448
Fecal incontinence[a]	31	1/5 (20%)	5/26 (19%)	0.397
Age by HD diagnosis, (months)[b]	31	0 (0–14)	1.5 (0–73)	0.331
Length of aganglionosis[a]	30	3/5 (60%)	22/25 (88%)	0.125
Rectosigmoid		2/5 (40%)	3/25 (12%)	
Proximal to rectosigmoid				
Number of hospitalizations ≤ 5 years[b]	31	7 (2–13)	5 (0–11)	0.514
Longest hospitalization ≤ 5 years (days)[b]	31	23 (7–413)	13.5 (0–204)	0.257
Miscellaneous factors[a]				
Other chronic diseases	31	1/5 (20%)	3/26 (12%)	0.605
Impaired vision and/or hearing	31	3/5 (60%)	7/26 (27%)	0.147
School problems	31	1/5 (20%)	7/26 (27%)	0.746
Ever bullied	30	2/5 (40%)	5/25 (20%)	0.334

*p ≤ 0.05.
[a] n (percent), Pearson chi square test.
[b] Median (min–max), Mann Whitney U test.

4. Discussion

The most important and novel finding in this study exploring mental health and psychosocial functioning in HD adolescents, is that 22% fulfilled criteria for a psychiatric diagnosis. Furthermore, having a psychiatric diagnosis was associated with reduced psychosocial function and QoL.

4.1. Mental health

Nearly one in four of the HD adolescents fulfilled the criteria for a psychiatric diagnosis. This is more common than reported in the general adolescent population [31–35]. Population studies show that approximately 15% of adolescents have some kind of mental illness before the age of 18 [32]. The incidence of mental illness in Norwegian adolescent males was 6% in 2016, while the female incidence was 20% [30]. Thus, the incidence of mental illness in this HD population with mainly males, equals that of adolescent females in the general population. The main problems among HD adolescents were emotional which is equal to that found in females in the general female population. In line with this, conduct and behavioral problems which is common in the general male population, were not found in the HD population [33,36]. Taken together the increased incidence of mental illness and the uncommon pattern of male psychiatric diagnoses, our findings indicate that mental health problems are more common in HD adolescents than in the general population.

Our finding that emotional problems were most common is in line with the study performed 26 years ago in Norwegian HD adolescents [12]. A recent Swedish questionnaire-based cohort study reported higher frequency of depression among both male and female HD patients compared to a control group [16]. Thus, the pattern of predominating emotional problems appear consistent [31].

4.2. Psychosocial functioning

Psychosocial functioning was significantly associated with being diagnosed with a psychiatric diagnosis. Scores ≤70 indicate probable and ≤60 likely need for treatment according to established norms [37]. More adolescents were scored with problems in psychosocial functioning ≤70, cGAS, than those who fulfilled psychiatric diagnostic criteria. A few adolescents with lower cGAS score had some daily challenges related for instance to school and peer relations, but otherwise functioned well and did therefore not fulfill the criteria for a psychiatric diagnosis. In a review article by Diseth et al. the HD-studies reported no over-representation of psychosocial problems in children and adolescents compared to general population controls [13]. No control group was available leaving this unanswered for the present study.

4.3. Pediatric quality of life

Even though the PedsQL questionnaire provides no information about the kind of mental health problem the adolescent may have, the PedsQL scores were significantly associated with having a psychiatric diagnosis in the present study. In the study by Diseth, no QoL assessment was reported thus we have found no other studies reporting how QoL-measures relate to interview based mental health assessment for HD adolescents [12].

We could not demonstrate any disease-related factors significantly associated with reduced PedsQL scores. Some studies on generic QoL in HD patients report impaired bowel function negatively affecting QoL [2,4], while others do not [38]. The different findings may be explained by use of different QoL questionnaires, different grading of bowel function and different age of the patients. Furthermore, type II errors may also explain the conflicting results as some studies have small patient population.

4.4. Mental health and QoL: possible stressors and vulnerabilities

4.4.1. Bowel function and treatment

We could not identify any particular treatment modality to be associated with mental health problems. Many patients had undergone anal calibrations/dilatations and rectal wash outs. This could be stressful and influence mental health as reported by Diseth [39]. These procedures were performed during early age in this study population and performed under general anesthesia if the

child seemed troubled. This may explain why anal procedures did not influence mental health. Furthermore, we did not find that fecal incontinence, having a stoma and following a bowel management program were significantly associated with having a psychiatric diagnosis, although bowel management/having a stoma was relatively more common in the group with a psychiatric diagnosis and call for further and larger studies. A review article from 2017 did not demonstrate that fecal incontinence negatively influenced psychosocial functioning and QoL [13]. Ludman et al. reported more mental health problems in TCA patients [14]. Though not significant with the present small sample, we also found more adolescents with a psychiatric diagnosis in those with more extensive disease with aganglionosis proximal to rectosigmoid.

Several factors may represent mental health stressors when it comes to coping with a chronic disease. These include precautions, procedures and medication. Adolescents typically do not want to different from their peers. Time consuming procedures such as bowel management may be a stressor in that particular developmental phase. In adolescence immediate needs like participation in peer activities may as often override the long term perspectives and benefits related to precautions and procedures [40]. The intensive use of social media emphasizing success when it comes to appearance as well as achievements, may also represent a particular stressor for the HD adolescents [41,42].

4.4.2. Parental factors

Parents' education and socioeconomic status (SES) may affect children's mental health [43,44]. This corresponds well with our data showing that less parental education was associated with mental health problems in their children [45]. Thus, it is important to focus on parental understanding and coping in order to reduce mental health problems in adolescents [45]. Why the parents in this study were more educated than in the general population is most likely a coincidence or it could be that highly educated parents are more likely to encourage their teenagers to participate in a study like this. Given the general findings of less mental health problems in offspring of families with longer parental education and higher SES, this could imply a bias towards less mental health problems in the present study [43,44].

The findings indicate that generic PedsQL reflects aspects of mental health issues in adolescents with HD, but with some differences when it comes to significant background factors. CFD was not significantly associated with PedsQL scores and appeared to affect mental health more than QoL. Dealing with necessary daily routines and procedures regarding bowel habits, food intake and related issues may be more stressful for families with other vulnerabilities [45]. Similar findings of influential family factors are reported in the ARM study by Ludman 1994 [46], and emphasizes the need for multi-disciplinary approach [23].

5. Strength and limitations

The main strength of this study is the thorough examination of mental health and psychosocial function using validated and well recognized methods. There are few drop-outs as 86% of eligible adolescents were included. The relative low number of patients make sub-analyses difficult, for instance identifying predictors for mental health problems. Furthermore, a control group would have strengthened the study as we have relied on register data and recent epidemiological studies. The study was performed during the Covid-19 pandemic. This was a potential stressor for all children and adolescents, and we do not know to what extent this has influenced our data. In spite of that, we think the findings from this study bring important and new information about HD adolescent mental health and psychosocial functioning.

6. Conclusions

HD adolescents seem to have an increased occurrence of psychiatric diagnoses of which emotional problems were dominating. Furthermore, reduced QoL was associated with mental health problems. The results emphasize the need for a multi-disciplinary team in the follow-up of HD patients. This team must include health professionals that can identify and help HD adolescents with mental health and psychosocial challenges.

Previous communication

Preliminary results were presented at BAPS 2022.

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Conflicts of interest

The authors have no conflicts of interest to declare.

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