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Children With Anorectal Malformations, Hirschsprung Disease, and Their Siblings: Proxy Reports and Self-Reports

[†]*Esther E. Hartman,* [‡]*Frans J. Oort,* [||]*Daniel C. Aronson,* [||]*Alida F.W. van der Steeg,* [||]*Hugo A. Heij,* [#]*Ernest van Heurn,* [††]*Gerard C. Madern,* [‡‡]*David C. van der Zee,* [¶]*Ivo de Blaauw,* [§§]*A. van Dijk, and* [§]*Mirjam A.G. Sprangers*

ABSTRACT

Objectives: The aim of the present study was to compare parent proxy reports with that of self-reports of children with anorectal malformations (ARMs) or Hirschsprung disease (HD) and healthy siblings and thereafter was examine whether these comparisons differed between patients and their siblings.

Methods: Parents (n = 98) of either children with ARM (n = 44) or HD (n = 54) and a healthy sibling (n = 98) recruited from the 6 Dutch pediatric surgical centers and from the ARM and HD patient societies were included in this cross-sectional multilevel study. Agreement between child self-reports and parent proxy reports was compared through mean differences and through (intraclass) correlations. We conducted multilevel analyses to take dependencies between assessments within families into account.

Results: All of the children (children with ARM or HD and their siblings) reported more pain and symptoms than their parents reported. We also found that only children with ARM or HD reported less positive emotions than their parents. Furthermore, higher correlations were found between parent proxy reports and patient-self reports than between parent proxy reports and sibling self-reports on cognitive functioning and social interaction.

Conclusions: Parents tend to overestimate the physical functioning of both their ill and healthy children, and overestimate the emotional functioning of only their children with ARM or HD. Furthermore, children with ARM or HD and parents agree more on health-related quality of life domains than healthy children and parents.

What Is Known

- In health-related quality of life assessments, parent reports cannot substitute child reports.
- Hence, a combination of self-reports and proxy reports is preferred.
- The ability of parents to rate their child's health-related quality of life is dependent on the health of the child.

What Is New

- Until now, no studies compared self- and proxy reports of children with anorectal malformation or Hirschsprung disease.
- Moreover, far most studies examining the level of agreement between parent reports and self-reports of ill children versus healthy children compared 2 separate dyads of parents and children.
- The present study included a triad of parents and children with anorectal malformation or Hirschsprung disease and a healthy sibling.

Key Words: anorectal malformations, Hirschsprung disease, parent proxy reports, quality of life, self-reports, siblings

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Anorectal malformations (ARMs) and Hirschsprung disease (HD) are congenital anomalies, often leading to chronic bowel dysfunction. Despite neonatal surgery, children and adolescents with ARM or HD may never develop adequate bowel control and suffer from long-lasting functional problems, varying from persistent, severe constipation to soiling or complete fecal incontinence (1,2). Consequently, these patients have to cope with substantial functional problems, which affect their health-related quality of life (HRQOL) (3).

Although definitions of HRQOL vary widely, there is consensus about 2 central aspects. First, HRQOL should be regarded as a multidimensional construct incorporating ≥3 broad domains,

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From the [†]Department of Clinical and Medical Psychology, Center of Research on Psychology in Somatic Diseases, Tilburg University, Tilburg, the [‡]Department of Education, Faculty of Social and Behavioural Sciences, the [§]Division of Medical Psychology, University of Amsterdam, the [||]Department of Paediatric Surgery, University Hospital, VU University Medical Centre/Emma Children's Hospital AMC, Amsterdam, the [¶]Department of Paediatric Surgery, University Hospital, Radboud Hospital, Nijmegen, the [#]Department of Paediatric Surgery, University Hospital, Maastricht, the [††]Department of Paediatric Surgery, University Hospital, Utrecht, The Netherlands, the [‡‡]Division of Paediatric Surgery, Queen Elisabeth Central Hospital, College of Medicine, University of Malawi, Blantyre, Malawi, and the [§§]Department of Paediatric Surgery, University Hospital, Groningen, The Netherlands.

Address correspondence and reprint requests to Esther E. Hartman, Developmental Psychology/CoRPS, Tilburg University, Room T502, PO Box 90153, 5000 LE Tilburg, The Netherlands (e-mail: e.e.hartman@uvt.nl).

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including physical, mental, and social functioning (4,5). Second, considering the subjective character, HRQOL should be assessed from the patient's perspective whenever possible (6), following that self-reports would be most appropriate. Large-scale studies showed that young American children from 5 years up were able to provide reliable and valid self-reports, for both healthy and chronically ill children (7–10), but these results were not replicated in Dutch samples (11,12). So, from which age on self-reports could be used is still not clear, but it is generally accepted that children are able to reliably provide self-reports from 8 years up (6). Nevertheless, in some situations patients are not able to complete HRQOL instruments, for instance when they are too ill or too young, and parent reports are needed in such cases. Research consistently showed that parent reports, however, cannot substitute child reports (13) because self-reports and proxy reports do not perfectly agree (14–20). Parents and children seem to have different views on how the child feels and functions (21). Hence, to obtain multiple perspectives, it is recommended that in HRQOL assessments, a combination of self-reports and proxy reports is preferred (22).

An important factor that affects differences between HRQOL ratings of parents-as-proxies and children themselves is the health of the children. Few studies examined the effects of health status on the level of agreement between child self-reports and parent reports, but all of the results indicated more agreement between parents and children with a chronic disease compared with parents with healthy children (23,24). Moreover, parents tend to rate the HRQOL of healthy children as higher (21,25) than children rate their own HRQOL, whereas parents of children with a chronic health condition appear to rate the HRQOL of their children as lower than the children rate their HRQOL themselves (14). These studies examining the level of agreement between parent proxy reports and self-reports, however, compared 2 separate dyads of parents and children: parents and healthy children versus parents and ill children. A recent review by Limbers and Skipper (26) showed that 3 studies examined concurrently the HRQOL of children with a chronic disease and that of their siblings including both self-reports and proxy reports (27–29). Results showed that the HRQOL of siblings was better than that of children with a physical chronic illness but that findings varied by respondent (child self-report vs parent proxy report) in correspondence with the results with the separate dyads (eg, 23). Parents tended to rate healthy sibling HRQOL better than healthy siblings reported their own HRQOL. Further results of comparisons between self-reports and proxy reports, however, of these few studies were inconclusive. Two studies found that parent proxy reports overrated the HRQOL of their ill children and underrated the HRQOL of the healthy siblings, whereas in the other study parents overrated both the HRQOL of their ill and healthy children. Moreover, only one of these studies also examined the level of agreement between self- and proxy reports showing low agreement between proxy reports and self-reports of both ill and healthy siblings (27). In sum, only very few studies included child self-reports and proxy reports of children with a chronic illness and of healthy siblings simultaneously. Moreover, these studies showed inconsistent results, and only one of them simultaneously examined self-reports and proxy reports of both children and siblings by comparing the mean differences and correlations. Examining both mean differences and correlations adds additional information to the level of agreement between self-reports and proxy reports (25). For example, it is possible for scores to be correlated (ie, linearly related) but also show statistically significant differences in mean scores (25,14). Hence, how the HRQOL of siblings and chronically ill children is exactly related, taking into account child-reports and parent proxy reports, is still unknown.

Parents of children with ARM or HD have large responsibilities in the treatment of their children, including the performance of anal dilatations and/or washouts (30,31). Furthermore, care of these patients involves frequent hospital visits and visits to a general practitioner, and sometimes paramedical or psychosocial care is also needed (32–36). Hence, parents are intensively involved in the care of their children with chronic diseases, probably resulting in better insight into the HRQOL of these children leading to the hypothesis that parents will agree more with their children with ARM or HD than with their healthy children. Moreover, from the literature, it is expected that parents will overrate the HRQOL of their healthy child while underrating the health of their child with ARM or HD.

The aim of the study was to examine the extent to which parents were better proxies for their ill children (ARM or HD) than for their healthy children. First, parent proxy reports were compared with self-reports of children with ARM or HD and healthy siblings and thereafter was examined whether these comparisons differed between patients and their siblings. In the present study, parents who had a child with ARM or HD and a healthy sibling were included.

METHODS

Participants and Procedure

All of the parents of children with ARM or HD (8–16 years) were recruited from the 6 Dutch pediatric surgical centers and from the ARM and HD patient societies. Not included were parents who lacked basic proficiency in Dutch and children with ARM or HD who were mentally retarded and/or had Down syndrome, had a cloaca, were untraceable, or had died. Thereafter, only families with both a child with ARM or HD and a healthy child in the same age range (8–16 years) were selected. In case of multiple eligible siblings, only the one nearest in age of the child with ARM or HD was included. A total of 123 parents completed an informed consent to allow both of their children to participate in this study; 98 (80%) assessments included patient self-reports, sibling self-reports, and parent proxy reports for both ill children (ARM or HD) and healthy children (sibling). Parent proxy reports were completed by either the father or the mother. The medical ethics committees of all of the 6 pediatric surgical centers approved the study.

Measures

Quality of Life

HRQOL was measured with the TNO-AZL Child Quality of Life (TACQOL) questionnaire, containing a child self-report form and a parent proxy report form. In the present study, the child self-report was also used for the siblings. The child self-report and the parent proxy report include identical items only differing in the first person or third person. The TACQOL has been validated in a Dutch population of children ages 8 to 16 years and their parents (37). Within the child self-report, 2 different versions are available, a child form (8–11 years) (38,39) and an adolescent form (12–16 years) (40). Both versions consist of 56 identical items, but the adolescents' form contains 31 additional items that are not applicable to younger children, such as questions about homework, body image, and worries about the future. In the present study, we used only the 56 identical items to enable analyses of children and adolescents combined. These items were aggregated to form 5 health-related functioning scales, ''pain and symptoms,'' ''basic motor functioning,'' ''autonomy,'' ''cognitive functioning,'' ''interaction with parents and peers,'' and 2 emotion scales: ''experience of positive emotions'' and ''experience of negative emotions.'' We omitted the ''autonomy'' scale (α ranged from 0.24 to 0.71) because of poor reliability, as is also advised in the manual (38). The ''basic motor functioning'' scale (α ranged 0.45 to 0.62)

appeared to be insufficiently reliable for self-reports and proxy reports too. With each item of the health-related functioning scale, the respondent could indicate whether or not a specific problem occurred in the past few weeks, with 3 response-options ''never,'' ''occasionally,'' and ''often.'' If a problem occurred, the respondent was then asked how he/she felt about this problem: ''fine,'' ''not so good,'' ''quite bad,'' and ''bad.'' For each item, the 2 responses were combined into 1 single item score ranging from 0 to 4 (''never'' 4 and ''occasionally'' or ''often'' combined with ''fine'' 3, with ''not so good'' 2, with ''quite bad'' 1, and with ''bad'' 0). With the emotion scales, respondents indicated on a Likert scale whether the presented feeling occurred recently in the past few weeks (never, occasionally, and often). With all of the 7 scales, higher scores correspond to better HRQOL.

Background Characteristics

Clinical variables were extracted from medical records and included ''disease-severity'' (mild vs severe), ''presence of additional congenital anomalies'' (yes vs no), and ''presence of a permanent stoma'' (yes vs no). For ARM, ''mild'' versus ''severe'' referred to low defects (bucket handle, covered anus, anterior displaced anus, and perineal or vestibular fistula) versus more complex defects (urethral, vesical or vaginal fistula, or no fistula). For HD, ''mild disease'' was defined as aganglionosis of a common (or usual) segment (rectum or sigmoid), and ''severe disease'' referred to aganglionosis of a long segment (colon descendens, colon transversum, colon ascendens, or ileum). Note that the classification of the severity of ARM and HD is arbitrary. We choose to classify the severity of ARM by the complexity of the malformation because more complex forms are more frequently associated with more symptoms (41,42). The classification of the severity of HD was based on the length of the affected part of the bowel because longer affected segments in HD are usually associated with worst physical outcome/more symptoms than short (or usual) segments of the bowel (42,43). In addition, with ARM, we frequently observed the VACTERL-association, which includes vertebral-, anorectal, cardiac-, tracheoesophageal-, renal-, and limb defects (44). Demographic characteristics included ''sex'' (male vs female) and ''age.''

Statistical Analyses

Sample Characteristics

We tested for differences in age and sex between children with ARM or HD and their healthy siblings. The χ^2 tests were used for sex and *t* tests for age. The level of significance was set at 5%.

Comparing Child Self-Reports and Parent Proxy Reports of Child HRQOL

To compare the mean HRQOL of child self-reports with parent proxy reports and to examine whether these comparisons differ between the children with ARM or HD and their healthy siblings, multilevel analyses were conducted (taking into account dependencies between assessments within families). The advantage of multilevel analysis is that it utilizes all of the available information (also including information from incomplete cases) to obtain maximum likelihood estimates of regression effects (representing mean differences between reports) and (intraclass) correlations (between reports) (45). The TACQOL subscales Pain and Symptoms, Cognitive Functioning, Social Interactions, and Positive and Negative Emotions served as dependent factors. Type of report (child self-report, sibling self-report, and parent proxy report) and health status of the child (healthy or with ARM/HD) were used as independent variables. Child age and sex were added to the model as covariates. The multilevel models were parameterized in such a way that the intercept represents the mean self-report of children with ARM or HD and regression coefficients represent deviations of sibling self-report and parent report from self-report of children with ARM or HD. As all of the dependent variables are rescaled to an overall mean of 0 and a standard deviation of 1, these regression coefficients can be interpreted as effect sizes *d*. According to Cohen (46), rule of thumb effect sizes *d* of 0.2, 0.5, and 0.8 can be considered small, medium, and large; *d* also applies to a child's sex. The regression coefficients of the continuous variable child's age, however, can be interpreted as effect size *r*. According to Cohen, *r* of 0.1, 0.3, and 0.5 can be considered as small, medium, and large, respectively (46).

To further examine the relative agreement between parent proxy reports and self-reports, the multilevel analyses were repeated for the children with ARM or HD and the siblings sample separately, and we examined the (intraclass) correlations between proxy reports and self-reports. As a rule of thumb, we considered differences between 2 (intraclass) correlations significant when the point estimate of 1 (intraclass) correlation falls outside the confidence interval (CI) of the other correlation (CI criterion). For all of the analyses, SPSS version 19 (IBM SPSS Statistics, Armonk, NY) was used.

RESULTS

Sample Characteristics

Patient and sibling characteristics for disease groups (ARM/HD) are given in Table 1. The total sample consisted of 98 patients (44 children with ARM and 54 children with HD), 98 siblings and 98 parents. There were a larger proportion of boys with HD than healthy boys ($P = 0.00$), which was expected because of the male-female ratio in HD (47). Moreover, children with ARM or HD were significantly younger than their siblings ($P = 0.00$ and $P = 0.03$, respectively). Therefore, sex and age were included as covariates in the multilevel analyses.

HRQOL Compares Self-Reports of Children (ARM, HD, and Siblings) Versus Parent Reports

Table 2 shows the main effect of ''parent report'' on ''Pain and Symptoms,'' indicating that children (children with ARM or HD and their siblings) reported more pain and symptoms than their parents reported ($\beta = 0.286$, $P = 0.002$; reversely scored). This effect is the same for children with ARM or HD and healthy siblings, according to the insignificant interaction effect ($\beta = 0.157$, not significant). We also found main effects of ''parent report'' ($\beta = 0.352$, $P = 0.000$) and ''report of healthy children'' ($\beta = 0.307$, $P = 0.033$) on ''Positive Emotions,'' but the latter effect is cancelled by a significant interaction effect ($\beta = -0.283$, $P = 0.027$), indicating that only children with ARM or HD reported less positive emotions than their parents. In addition, analyses of the covariates showed that higher age of patients with ARM or HD was related to better social functioning ($\beta = 0.151$, $P = 0.032$) and less negative emotions ($\beta = 0.227$, $P = 0.003$), and that girl siblings reported less positive emotions than boy siblings ($\beta = -0.316$, $P = 0.009$).

Overall, Table 3 shows higher (intraclass) correlations between parent proxy reports and patient reports than with sibling reports on all of the HRQOL domains, but only the difference between the (intraclass) correlations for cognitive functioning ($r = 0.75$, CI 0.64–0.82 vs $r = 0.51$, CI 0.35–0.64) and social

TABLE 1. Characteristics of children and adolescents with ARMs or HD, and of their healthy siblings

	ARM (n = 44)	Siblings of patients with ARM (n = 44)	HD (n = 54)	Siblings of patients with HD ARM (n = 54)
Sex				
Male (%)	27 (61)	26 (59)	47 (87)*	27 (50)*
Female (%)	17 (39)	18 (41)	7 (13)*	27 (50)*
Age				
Mean age, y (range)	10.9 (8–16)*	12.4 (8–16)*	11.5 (8–17)*	12.5 (8–16)*
Missing n (%)	—	7 (7.1)	—	—
Disease severity				
Mild (%)	15 (34)	NA	36 (68)	NA
Severe (%)	24 (55)	NA	17 (32)	NA
Missing (%)	5 (11)	NA	1 (2)	NA
Additional congenital anomalies				
Yes (%)	19 (43)	NA	6 (11)	NA
No (%)	20 (46)	NA	47 (87)	NA
Missing (%)	5 (11)	NA	1 (2)	NA
Stoma				
Yes (%)	1 (2)	NA	4 (7)	NA
No (%)	43 (98)	NA	50 (93)	NA

ARM = anorectal malformation; HD = Hirschsprung disease.
* $P < 0.05$.

interaction ($r = 0.77$, CI 0.55–0.77 vs $r = 0.22$, CI 0.02–0.40) for children with ARM or HD met the CI criterion.

DISCUSSION

When comparing mean differences, results showed that parents reported better physical functioning than both their ill (ARM or HD) and healthy children reported themselves. Furthermore, parents reported more positive emotions for their children with ARM or HD, than these children reported themselves. In contrast, the parents did not differ in their ratings of positive emotions (or any other domains) from those of their healthy children. Hence, taking self-reports as the criterion standard, parents seem to overestimate their child's HRQOL on physical functioning (which applies to both ARM/HD and healthy children) and on the experience of positive emotions (which only applies for ARM/HD).

TABLE 2. Significance tests of differences in quality-of-life domains between self-report of children with ARMs or HD versus self-report of healthy siblings versus parent reports

	Pain and symptoms β (SE)	Cognitive functioning β (SE)	Social interactions β (SE)	Positive emotions β (SE)	Negative emotions β (SE)
Intercept (mean self-report ARM/HD)	−0.187 (0.155)	−0.167 (0.157)	0.012 (0.146)	−0.032 (0.152)	−0.034 (0.154)
	$P = 0.229$	$P = 0.289$	$P = 0.933$	$P = 0.833$	$P = 0.825$
Parent report (mean deviation from self-report)	0.286 (0.088)	0.062 (0.078)	−0.092 (0.080)	0.352 (0.096)	−0.038 (0.091)
	$P = 0.002$	$P = 0.434$	$P = 0.253$	$P = 0.000$	$P = 0.675$
Report of healthy children (mean deviation from report of ARM/HD)	0.094 (0.140)	0.181 (0.145)	−0.152 (0.128)	0.307 (0.142)	0.128 (0.123)
	$P = 0.503$	$P = 0.215$	$P = 0.238$	$P = 0.033$	$P = 0.302$
Parent report of healthy children (mean deviation parent report of ARM/HD)	0.157 (0.134)	−0.019 (0.127)	−0.159 (0.136)	−0.283 (0.126)	−0.074 (0.129)
	$P = 0.242$	$P = 0.884$	$P = 0.247$	$P = 0.027$	$P = 0.567$
Age patients (deviation from mean age in standard deviations)	−0.045 (0.072)	0.064 (0.067)	0.151 (0.070)	−0.012 (0.061)	0.227 (0.074)
	$P = 0.528$	$P = 0.342$	$P = 0.032$	$P = 0.842$	$P = 0.003$
Age siblings (deviation from mean age in standard deviations)	−0.087 (0.072)	−0.011 (0.067)	−0.085 (0.071)	−0.056 (0.061)	−0.071 (0.074)
	$P = 0.227$	$P = 0.87$	$P = 0.226$	$P = 0.358$	$P = 0.336$
Girl reports (mean deviations from boy reports)	−0.036 (0.171)	0.123 (0.159)	0.180 (0.165)	−0.091 (0.144)	0.039 (0.175)
	$P = 0.832$	$P = 0.443$	$P = 0.277$	$P = 0.534$	$P = 0.825$
Girl sibling reports (mean deviation from boy sibling reports)	−0.201 (0.139)	−0.125 (0.130)	−0.245 (0.135)	−0.316 (0.118)	−0.113 (0.143)
	$P = 0.151$	$P = 0.339$	$P = 0.072$	$P = 0.009$	$P = 0.431$
HD reports of patients and parents (mean deviation from ARM reports)	0.096 (0.144)	0.126 (0.135)	0.116 (0.140)	−0,108 (0.122)	−0.022 (0.149)
	$P = 0.505$	$P = 0.355$	$P = 0.411$	$P = 0.831$	$P = 0.881$

All of the continuous variables are standardized to z scores (a higher score means better functioning) and all other variables are binary coded. The intercept can be interpreted as the mean self-report of a child with ARM or HD, and the regression coefficients can be interpreted as the mean difference with other reports. Regression coefficients can be interpreted as Cohen effect size d: ''small'' (0.20), ''medium'' (0.50), and ''large'' (0.80), except for the regression coefficients of the continuous variable child's age, which can be interpreted as effect size r, with 0.1, 0.3, and 0.5 indicating small, medium, and large effects (1). ARM = anorectal malformation; HD = Hirschsprung disease.

TABLE 3. Agreement between parents and children for ARM or HD children and healthy siblings separately

	Patient-parent $r^{\dagger}$ (CI)	Sibling-parent $r^{\dagger}$ (CI)
Generic HRQOL		
Pain and symptoms	0.62 (0.48–0.73)	0.50 (0.33–0.64)
Cognitive functioning	0.75 (0.64–0.82)*	0.51 (0.35–0.64)*
Social interactions	0.68 (0.55–0.77)*	0.22 (0.02–0.40)*
Positive emotions	0.60 (0.46–0.71)	0.49 (0.32–0.63)
Negative emotions	0.59 (0.44–0.71)	0.50 (0.33–0.63)

ARM = anorectal malformation; CI = confidence interval; HRQOL = health-related quality of life.

* $P < 0.05$.

† According to Cohen, r of 0.1, 0.3, and 0.5 can be considered as small, medium, and large effects, respectively.

Next to comparing mean differences between child and parent reports to examine ''absolute'' agreement, the ''relative'' agreement between child and proxy reports was examined with correlations (48,49) because self- and proxy reports may show no differences in mean scores but low correlations, or vice versa (25,14). Hence, analyzing the data in several ways provides additional information about the agreement between self- and proxy ratings.

From the analyses at the relative level, results showed that parent reports were more strongly correlated with self-reports of their children with ARM or HD than with those of their healthy children, specifically on the domains of cognitive functioning and social interactions.

An examination of literature indicates that parents tend to overestimate the HRQOL of their healthy children but that they underestimate the HRQOL of their children with a health condition and that parents agree more with their children with a chronic disease as compared with their healthy children (23,50). Hence, in contrast to the extant literature, parents tend to overestimate the emotional functioning of their children with ARM or HD and the physical functioning of both their ill and healthy children at an absolute level, but in concordance with the literature, parents consistently agree more on a relative level with their ill children than with their healthy children.

Strength of the study was that we included a triad sample of parents of both a child with ARM or HD and a healthy child, which allowed us to examine whether the same parents rate the HRQOL of children with ARM or HD differently than that of their healthy children. This strength, however, also implied that the parent and child dyads in the present study were not independent observations. Thus, how a parent rates 1 child may have impacted how they rate their other child, maybe causing an overestimation of the HRQOL of their child with ARM or HD. Parents completed the proxy versions of the questionnaires of their 2 children in succession, maybe causing them to—though erroneously—think that their child with ARM or HD is actually not doing worse as compared with their healthy child. An explanation for the higher associations of the HRQOL reports between ill children and parents may be that parents are more involved in the care of, and feel more responsible for, children with a chronic disease (51,52), which may result in better insight into the well-being of the ill child. In line with this explanation, some studies found higher parent-child agreement in the child's HRQOL when children were more severely ill (53,54).

One of the limitations of the study was that the number of fathers and mothers who completed the proxy version of the questionnaires was unknown. The present study probably relied on mothers' proxy reports, similar as in earlier work that mentioned sex of the proxy respondents (21). Another study limitation was that we were not able to determine to what extent selection bias may have played a role because we were not able to assess the characteristics of the patients who were not willing to participate, as a result of confidentiality considerations.

To prevent the effects of the larger proportion of boys in the HD group and the effects of age, we controlled for sex of the child (of the children with ARM, HD, and the siblings) and age in the multilevel analyses. Additional advantage of using multilevel analyses is the possibility of the use of all of the available data, also data from dropouts. Furthermore, by using multilevel correlations, the ratio between subject variability and total variability was reflected, correcting for any systematic difference providing a more reliable reflection of true concordance between self- and proxy reports.

CONCLUSIONS

The results revealed that parents overestimated the physical functioning of both their children (ARM/HD and healthy), and the level of positive emotions of their children with ARM or HD. In the context of health care delivery, health care professionals should ask the children themselves about possible (hidden) emotional problems. At a relative level, parents agreed more with their child with ARM or HD than with their healthy child on cognitive functioning and social interactions, probably because of more involvement in their children with ARM or HD as a result of the large responsibility in self-care. As parents and children each offer unique information, we agree with previous recommendations that HRQOL assessments should be completed by both children and parents as proxies (21).

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