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Anorectal malformation & Hirschsprung's disease: A cross-sectional comparison of quality of life and bowel function to healthy controls☆,☆☆,★

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ABSTRACT

Background: Patients with anorectal malformation (ARM) and Hirschsprung's disease (HD) face long-term disturbance in bowel function even after definitive surgery. This study evaluates the quality of life (QOL) of patients with ARM and HD, and compares them to healthy controls using self-report questionnaires.
Methodology: A prospective study was performed recruiting patients with ARM or HD from September 2013 to December 2014 who had primary surgery done in our institution at least 2 years prior to participation. Age-matched and gender-matched controls were enrolled from our patients with minor outpatient complaints. All participants completed the following PedsQL™ scales (maximum score 100): 4.0 Generic Core Scales, 3.0 General Well-Being (GWB) Scale and 2.0 Family Impact (FI) Module. All were also scored on bowel function (BFS), with a maximum score 20. Appropriate statistical analysis was performed, with significance level <0.05.
Results: There were 193 participants: 87 controls, 62 ARM, 44 HD. When comparing Core, GWB and FI scores, there were no significant differences between groups although controls had best scores indicating best QOL and general wellbeing, with least impact of the child's health on the family. BFS was significantly different with controls having best and ARM worst scores. There were no significant differences in scores between parent and child indicating intradyad consistency. There was significant positive correlation between BFS and Core ($p < 0.0001$), and between BFS and GWB scores ($p < 0.005$); and significant negative correlation between BFS and FI scores ($p < 0.0001$).
Conclusions: Bowel function impacts quality of life. Those with ARM and HD can achieve good quality of life comparable to controls, based on patient and caregiver self-reported outcomes.
Type of study: Prospective comparative study
Level of evidence: Level II.

Anorectal malformation (ARM) and Hirschsprung's Disease (HD) are the 2 most common congenital abnormalities of the large bowel presenting in infancy that require surgical correction, with a similar incidence of approximately 1 in 5000 live-births respectively, although there are population variations in incidence.

In both these conditions, despite corrective surgery done in infancy, many children continue to experience disturbances in bowel function up to adulthood, with an adverse effect on quality of life [1]. Jarvi et al reported at least occasional fecal soiling in 50% of adults with HD. In another study of 83 children with low type ARM, Rintala et al found that only 15% had completely normal bowel function [2,3].

While both ARM and HD are conditions seen across all ethnicities, most published studies on quality of life in patients with ARM or HD describe Caucasian populations [2–12] with a paucity of literature from other parts of the world [13–16].

The aim of our study was to evaluate quality of life (QOL) and bowel functional outcome in children with ARM or HD compared to age and gender matched controls using previously described QOL and bowel function scores. We hypothesized that patients with ARM or HD would have significantly worse scores compared to controls.

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Table 1
Distribution of types of fistula in ARM patients according to the Krickenbeck classification.

Type of fistula	No (%)
Perineal	21 (38)
Vestibular	8 (14)
Urethral	7 (13)
Prostatic	8 (14)
Vesical	5 (9)
No fistula	3 (5)
Cloaca	4 (7)
TOTAL	56 (100)

Records confirming site of fistula were unavailable in 6 patients.

Table 2
Length of aganglionosis in HD patients.

Length	No (%)
Rectosigmoid	29 (73)
Long segment	7 (18)
Total colonic	4 (10)
TOTAL	**40 (100)**

Records confirming length of aganglionosis were unavailable in 4 patients.

1. Methods

This was a prospective study with recruitment occurring between September 2013 and December 2014. Institutional ethical approval was obtained (CIRB 2012/703/D).

1.1. Participants

1.1.1. Patients with ARM or HD

All patients with ARM or HD who had primary surgery done in our institution at least 2 years prior to participation were eligible.

We excluded the following:

a. Those who had primary corrective surgery for ARM or HD done elsewhere.
b. Age less than 2 years.
c. Patients who had corrective surgery less than 12 months prior to the time of intended recruitment.

1.1.2. Controls

Controls were age-matched and gender-matched to ARM and HD participants, and enrolled from our population of patients with minor outpatient complaints, for example, those with skin or subcutaneous lumps amenable to excision as day surgery cases.

We excluded the following from our controls:

a. Those with recent history of acute gastrointestinal illness.
b. Those with chronic medical disorders requiring active treatment or sustained long-term follow-up.

1.2. Recruitment, interviews and study setting

Eligible patients who were scheduled for outpatient follow-up were identified and approached at the time of their clinic appointment in a sequential manner. Consent for participation was taken by a doctor from the study team. However, questionnaires were administered by an independent researcher to minimize the risk of bias in the participant's responses. Participants were asked to complete the questionnaires and return them during the visit. Only those fluent in reading and writing in English were included. On average, each participant took between 10 and 20 min to complete the full set of questionnaires.

We did not recruit inpatients as we felt that those requiring admission for an acute medical event would potentially report nonrepresentative lower QOL scores and thus provide unfair comparison to healthy controls.

1.3. Questionnaires and assessments

1.3.1. Pediatric Quality of Life Inventory™ (PedsQL™) [17]

The following PedsQL™ questionnaires were used. When both child self-reports and parent proxy reports were available within the design of the questionnaire, we invited both patient and caregiver to complete the reports.

a. Generic Core Scales 4.0 (Core)—This is a widely used validated modular approach to measuring health-related QOL in healthy children and adolescents and those with acute and chronic health conditions. This scale is designed to measure the core dimensions of health as delineated by the World Health Organization (physical, emotional, social), as well as role (school) functioning. Both child self-reports (available for children aged 5 years onwards) and parent proxy reports were used, with maximum score of 100 indicating best quality of life.
b. General Well-Being Scale 3.0—This scale measures general health for children 8 years old and above, with both child self-reports and parent proxy reports available. In contrast to Core scales which require respondents to relate answers to their health, the GWB scale assesses the respondents' overall view of their life.
c. Family Impact Module 2.0—This module measures the impact of pediatric acute and chronic health conditions on parents and the family, and is available for all ages, with only parent proxy reports required for scoring.

1.3.2. Bowel function score

This was a quantitative score first developed by Holschneider and modified by Lindahl et al assessing various aspects of bowel function, with a maximum score of 20 indicating best bowel function [7].

Questionnaires were given to respondents in the following order: Generic Core Scale, General Well-Being Scale, Family Impact Module, Bowel Function Score. In this way, we minimized possible bias from poor BFS on reported QOL scores.

1.4. Other associated anomalies and type of ARM/HD

We also collected data on the presence of associated congenital malformations that could additionally impact QOL. We defined 'major' anomalies as those that required medical intervention. For example, a large ventricular septal defect requiring long term antifailure medication was classified as 'major' while a patent ductus arteriosus that closed spontaneously was not. We also recorded the type of ARM according to the Krickenbeck anatomical classification, and the length of aganglionosis in HD.

1.5. Statistical analysis

We used chi-squared tests for categorical data and Kruskal–Wallis tests for nonparametric continuous variables. Spearman correlation tests were used to identify the relationship between bowel function scores and QOL scores. Where both child self-reports and parent proxy reports were completed, we used the Wilcoxon signed-rank test for nonparametric comparison of paired data. A significance level of $p < 0.05$ was taken.

2. Results

Based on eligibility criteria, there were 451 potential patients identified from our operative database, of whom 172 were no longer on active follow-up, leaving 279 patients. Our study protocol allowed only for recruitment at an outpatient visit, leaving us with our final group of 193 participants: 87 controls, 62 ARM and 44 HD patients. Types of ARM and length of aganglionosis in HD participants are listed in Tables 1 and 2.

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