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Abstract
Purpose:
This pilot study was designed to assess bowel function and quality of life (QoL) in children and adolescents with congenital colorectal malformations (CCM) during the first UK COVID lockdown period.
Methods:
Changes in health were assessed through semi-structured interviews, gastrointestinal functional outcomes using Krickbeck scoring and QoL by the modified disease-specific HAQL (Hirschsprung's disease anorectal malformation quality of life questionnaire). The State-Trait Anxiety Inventory (STAI)TM for adults was used to assess parental anxiety.
Results:
Thirty-two families were interviewed; 19 (59%) reported no change in their child's health during the lockdown, 5 (16%) a deterioration and 8 (25%) an improvement. Neither the severity of the CCM, nor the degree of bowel dysfunction, correlated with any deterioration. The HAQL score was not correlated to a change in health. Anxiety scores ranged from no anxiety to clinical concerns. Telemedicine was well accepted by 28/32 parents (88%); however, in-person appointments were preferred if there were clinical concerns.
Conclusion:
In the follow-up of children and adolescents with CCM during the first UK lockdown using telemedicine we found that over half had stable health conditions. Patients needing additional care could not be predicted by the severity of their disease or their bowel function alone.

Keywords (separated by '-') Anorectal malformation - Hirschsprung's disease - Quality of life - Sars-CoV-2 - COVID-19 - Telemedicine

Footnote Information **Supplementary Information** The online version contains supplementary material available at <https://doi.org/10.1007/s00383-021-04971-6>.

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2 Children with congenital colorectal malformations during the UK 3 Sars-CoV-2 pandemic lockdown: an assessment of telemedicine 4 and impact on health

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8 Abstract

9 **Purpose** This pilot study was designed to assess bowel function and quality of life (QoL) in children and adolescents with
10 congenital colorectal malformations (CCM) during the first UK COVID lockdown period.

11 **Methods** Changes in health were assessed through semi-structured interviews, gastrointestinal functional outcomes using
12 Krickenbeck scoring and QoL by the modified disease-specific HAQL (Hirschsprung's disease anorectal malformation
13 quality of life questionnaire). The State-Trait Anxiety Inventory (STAI)TM for adults was used to assess parental anxiety.

14 **Results** Thirty-two families were interviewed; 19 (59%) reported no change in their child's health during the lockdown, 5
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16 correlated with any deterioration. The HAQL score was not correlated to a change in health. Anxiety scores ranged from
17 no anxiety to clinical concerns. Telemedicine was well accepted by 28/32 parents (88%); however, in-person appointments
18 were preferred if there were clinical concerns.

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20 found that over half had stable health conditions. Patients needing additional care could not be predicted by the severity of
21 their disease or their bowel function alone.

22 **Keywords** Anorectal malformation · Hirschsprung's disease · Quality of life · Sars-CoV-2 · COVID-19 · Telemedicine

23 Introduction

24 The arrival of Sars-CoV-2, the coronavirus responsible for
25 the current pandemic, forced hospitals and health care sys-
26 tems to reorganize services for many patients. Non-urgent
27 appointments were rescheduled, and telemedicine was rap-
28 idly implemented.

29 The national lockdown imposed by the UK government
30 on March 23rd, 2020 meant that schools were closed for all
31 children, apart from those of key workers. Children's lives
32 were disrupted in an unprecedented way.

33 Children born with a congenital colorectal malformation
34 (CCM), such as Hirschsprung's disease (HSCR) or anorectal
35 malformations (ARM), are followed at our institution by a

dedicated team of surgeons and nurses. Our children's colo- 36
rectal clinic became virtual in line with our hospital's pan- 37
demic response, offering telephone or video consultations. 38

Constipation and fecal incontinence are known long-term 39
complications in both mild and severe forms of congenital 40
colorectal malformations [1, 2]. The congenital colorectal 41
clinic aims to monitor and alleviate abnormal bowel func- 42
tion, using a bowel management program [3]. 43

We hypothesized that lockdown and modified access to 44
health care might alter the bowel function and the quality of 45
life of children and adolescents with CCM. Therefore, the 46
study's main aim was to assess bowel function and quality 47
of life of children and adolescents with CCM during the 48
lockdown. Two secondary objectives were to (1) assess 49
the parental anxiety and (2) collect the opinions of parents 50
regarding telemedicine. 51

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52	Patients and methods		
53	Participants		
54	Forty patients aged 5–16 years with CCM operated on and	physical symptoms (8 items), emotional functioning (9	93
55	followed at the congenital colorectal clinic at Birmingham	items), use of laxative diet (2 items), urinary continence (4	94
56	Women's and Children's Hospital, who had an appoint-	items), and social functioning (3 items). For patients with	95
57	ment scheduled during the lockdown period (April–May	a stoma, extra items (8) related to the stoma are included	96
58	2020), were asked to participate by their surgeon (IJ, OG,	and items related to bowel motions omitted. Responses are	97
59	AL) or the colorectal nurses (GF, EH). During the first UK	scored (0–3) depending on the frequency (never, sometimes,	98
60	lockdown period, regular outpatients' appointments were	often, very often) of the specific problem during the past	99
61	replaced by telephone clinics at our institution and no patient	7 days. The values are transformed into a score for each	100
62	was seen face to face except in emergency situation. Lack of	dimension (0–100) which are summed for all seven dimen-	101
63	English proficiency was an exclusion criterion.	sions (0–700).	102
64	Severity of the malformation	There are validated and culturally adapted French, Dutch,	103
65	The severity of an ARM was classed as mild (perineal and	Swedish and Italian versions; an English translation exists	104
66	vestibular fistula in girls, perineal fistula in boys) and severe	but has not been validated.	105
67	(cloaca and recto-vaginal fistula in girls, recto-urethral fis-	Questions were administered over the phone by two	106
68	tula in boys). In HSCR, severity depended on the agangli-	authors (ES, CS) not involved in patient care. The question-	107
69	onic segment length: mild was rectosigmoid aganglionosis,	naire has not been validated for those with developmental	108
70	and severe was long-segment or total colonic aganglionosis.	delay or a cloaca. Such patients were included in our study,	109
71	Semi-structured interviews	and their results are reported separately.	110
72	Semi-structured interviews were conducted over the phone	Assessment of parental anxiety	111
73	by two authors (ES, CS), who were not involved in patient	Anxiety in a parent or primary carer was assessed using	112
74	care. Two dimensions were explored: parental perception	the State-Trait Anxiety Inventory (STAI) TM for adults. The	113
75	of their child's health related to the colorectal disease and	STAI consists of two questionnaires of 20 items each: the	114
76	access to health care. The questions are summarized in	first questionnaire (state anxiety) measures how the subject	115
77	Online Appendix 1.	feels at a particular moment in time, the second (trait anx-	116
78	Assessment of digestive functional outcome	ety) how they feel generally. Examples of items are "I feel	117
79	Functional outcome was assessed following the Krickenbeck	at ease", "I feel upset", "I am a steady person". The psycho-	118
80	scoring system (see Online Appendix 2). Participants man-	metric properties of both questionnaires are well-established	119
81	aged with retrograde colonic enemas or antegrade enemas	[6]. After gaining consent, the questionnaires were sent elec-	120
82	were excluded from this assessment.	tronically or by mail.	121
83	Quality of life assessment	Statistical analysis	122
84	Quality of life was assessed using a disease-specific ques-	Demographical data are presented descriptively. Average	123
85	tionnaire, the modified HAQL (Hirschsprung's disease	HAQL scores are reported as mean \pm standard deviation.	124
86	anorectal malformation quality of life questionnaire) [4,	Student's <i>t</i> test was used to look for difference in HAQL	125
87	5]. There are three age-specific versions of the question-	scores according to severity of malformation, presence of	126
88	naires (6–11, 12–16, > 17 years); proxy questionnaires exist	soiling or constipation or a deterioration in health. Statisti-	127
89	for two younger age groups. Seven dimensions that cover	cal analyses were performed using GraphPad Prism 9.0.0	128
90	physical, emotional and social functioning, as well as dis-	(GraphPad Software, San Diego, CA, United States).	129
91	ease-related symptoms, are explored by multiple items: the	Results	130
92	presence of diarrhea (4 items), fecal continence (6 items),	Population characteristics	131
		Out of the 40 patients identified, 32 [21 boys (67%), 11 girls	132
		(33%)] participated, 5 (13%) accepted to participate but did	133
		not answer our subsequent calls and 3 withdrew (8%). Eleven	134
		children had Hirschsprung's disease, of whom 4 (36%) had	135
		long segment/total colonic aganglionosis. Twenty-one chil-	136
		dren had an anorectal malformation, of whom 10 (48%) had	137

138 severe malformations including 2 patients with a cloaca.
139 Therefore, according to the predetermined classification
140 18/32 (56%) of children had congenital colorectal conditions
141 of mild severity and 14/32 (44%) were classified as severe.

142 Fourteen (44%) patients had other co-morbidities (see
143 Table 1). Ten (31.5%) children were managed with mechanical
144 irrigation via ACE stomas or transrectally. A further four
145 (12.5%) children had stomas (3 colostomies, 1 ileostomy).
146 Four (12.5%) children had developmental delay.

147 Telemedicine

148 During the first UK lockdown period, all 32 patients were
149 followed by telephone clinics as scheduled. No patient pre-
150 sented to an Emergency department.

151 Fourteen out of 32 (44%) parents preferred virtual clinics,
152 4 (12%) parents stated they disliked them, and 14 (44%) felt
153 they were as good as face-to-face clinics. All parents who
154 disliked telephone clinics have a child with a severe ARM.
155 Among the 28 parents who were satisfied with virtual clinics,
156 16 (57%) spontaneously commented that they would
157 like their child to be seen face to face if there was a problem.

158 Health during lockdown

159 In our group of patients, 19 out of 32 (59%) reported no
160 change in their child's health during lockdown while 8 out
161 of 32 (25%) reported an improvement and 5 out of 32 (16%)
162 commented that their child's health had deteriorated. There
163 was no correlation between the severity of the congenital
164 colorectal malformation, the degree of bowel dysfunction
165 and the deterioration in the child's health (Table 2). There
166 was no correlation with the overall HAQL score and the
167 report of a deterioration in the child's health (Table 3).

168 Parents repeatedly stated that the reason for an improve-
169 ment in their child was (i) that they had better control of
170 their child's diet, (ii) better control of their child's sched-
171 ular (including the timing of stooling) and (iii) the absence
172 of peers' judgment. Less physical exercise and less social

173 contact were the reasons given to explain health deteriora-
174 tion during the lockdown.

Functional assessment

175 Seventeen out of eighteen (94%) children who did not have
176 either an ACE or a stoma were assessed with the Krick-
177 beck assessment tool. In our sample of patients, 9 (53%)
178 had no soiling, and 8 (47%) had no constipation (Table 2).
179

Quality of life

180 Out of 32 patients, 25 (78%) patients' quality of life was
181 assessed with the HAQL; four patients did not complete
182 the questionnaire, one child had such severe developmental
183 delay that questions in the social dimension were not appli-
184 cable and the parents of two children who were 6 and 7 years
185 old respectively did not feel that they were able to answer
186 questions in the social dimension due to their young age.
187 All but one of the questionnaires was answered by a parent.
188

189 The mean (\pm SD) score was 594 (\pm 92) from a maximum
190 of 700. In our group, quality of life appeared independent
191 of the malformation severity (Table 3). An ACE or stoma
192 were the only factors that significantly negatively affected a
193 child's quality of life, $p=0.035$. Constipation did not appear
194 to affect the quality of life. There was a trend towards a
195 worse quality of life in children who soiled, although this
196 did not reach statistical significance, $p=0.057$.

197 The HAQL score was not related to a reported deteriora-
198 tion in a child's health.

Parental anxiety

199 Eight parents (24%) returned the STAI questionnaire. The
200 range of anxiety scores was from no anxiety to clinical con-
201 cerns. Parents of children with a severe congenital colorectal
202 malformation had higher trait anxiety. There was no differ-
203 ence based on the type of malformation, the presence of
204 colostomy or ACE (Table 4).
205

Table 1 Demographics

Demographics	Mild HSCR ($n=7$)	Severe HSCR ($n=4$)	Mild ARM ($n=11$)	Severe ARM ($n=10$)	Total ($n=32$)
Male:female	6:1	2:2	5:6	8:2	21:11
Isolated condition	5 (71%)	1 (25%)	7 (64%)	5 (50%)	18 (56%)
Co-morbidities	2 ADHD	1 Short gut 1 Chromosomal (T21) 1 Deaf	2 Chromosomal (T21, Di George) 1 Urological 1 ADHD	1 VACTERL 1 VACTERL + chromosomal 2 Urological 1 Haem	14 (44%)
Developmental delay	0	1 (25%)	2 (18%)	1 (10%)	4 (12.5%)
Enema/ACE	1 (14%)	0	4 (36%)	5 (50%)	10 (31%)
Stoma	2 (29%)	2 (50%)	0	0	4 (12.5%)

Table 2 Survey results according to severity and type of colorectal condition

	Mild HSCR (<i>n</i> =7)	Severe HSCR (<i>n</i> =4)	Mild ARM (<i>n</i> =11)	Severe ARM (<i>n</i> =10)	Total (<i>n</i> =32)
Change in Health					
Improved	1	1	4	2	8 (25%)
Deteriorated	2	0	1	2	5 (16%)
No change	4	3	6	6	19 (59%)
Telemedicine					
Preferred	4	2	5	3	14 (44%)
Disliked	0	0	0	4	4 (12%)
Neutral	3	2	6	3	14 (44%)
Krickenbeck Scoring	<i>n</i> =4	<i>n</i> =2	<i>n</i> =6	<i>n</i> =5	<i>n</i> =17
Soiling					
Grade 0	3	1	3	2	9 (53%)
Grade 1	0	0	2	2	4 (24.5%)
Grade 2	1	1	1	1	4 (24.5%)
Grade 3	0	0	0	0	0
Constipation					
Grade 0	2	1	3	2	8 (47%)
Grade 1	0	0	2	0	1 (6%)
Grade 2	2	1	1	2	7 (41%)
Grade 3	0	0	0	1	1 (6%)
HAQL (mean, sd)	<i>n</i> =7 539 (± 112)	<i>n</i> =4 611 (± 41) Without DD <i>n</i> =3 609 (± 50)	<i>n</i> =8 617 (± 106) Without DD <i>n</i> =6 616 (± 124)	<i>n</i> =6 617 (± 58) Without cloaca <i>n</i> =4 620 (± 58)	<i>n</i> =25 594 (± 92) Without DD <i>n</i> =22 591 (± 98) Without DD OR cloaca <i>n</i> =20 588 (± 101)

Table 3 HAQL scores according to the severity of the malformation, presence of ACE/stoma, presence of soiling/constipation and deterioration in health

	Yes	No	<i>p</i> value
Severe malformation	614 (50)	581 (112)	0.388
Presence of ACE/stoma	551 (114)	628 (55)	0.035*
Soiling	599 (50.3)	657 (48)	0.057
Constipation	601 (40)	647 (62)	0.147
Deterioration in health	593 (61)	591 (99)	0.97

Mean (standard deviation)

Discussion

Our study of health changes in children with CCMs during the UK Sars-CoV-2 lockdown showed no change in 19 out of 32 (59%) according to parental reporting; 5 out of 32 (16%) reported a deterioration and 8 out of 32 (25%) an improvement. There was no correlation between the severity of the colorectal malformation or the quality of life assessment and the report of a change in the child's health condition. Parents linked improvement with a better diet, a more regular stooling timing, and an absence of peers' judgment;

Table 4 STAI state and trait assessment in our group (*n*=8)

	Mild (<i>n</i> =3)	Severe (<i>n</i> =5)	ARM (<i>n</i> =4)	HSCR (<i>n</i> =4)	With colostomy (<i>n</i> =2)	No colostomy (<i>n</i> =6)	With ACE (<i>n</i> =2)	No ACE (<i>n</i> =6)
STAI-State	40 (14)	31 (17)	39 (15)	30 (17)	22 (3)	38 (16)	29 (0)	36 (18)
STAI-Trait	41 (4)	27 (4)	34 (6)	30 (10)	27 (1)	34 (8)	37 (5)	31 (8)

Mean (standard deviation)

deterioration was attributed to decreased physical exercise and social contacts. Anxiety among carers ranged from no anxiety to pathological levels suggesting that they would benefit from targeted medical management.

The absence of a clear link between bowel function and health perception in our study mirrors the fact that the effect of fecal incontinence or constipation on the quality of life of children with congenital colorectal malformation remains unclear. In the mild form of ARM, Grano et al. found that fecal incontinence negatively affected various dimensions of adolescent quality of life [7]. However, studying a comparable population of children with mild ARM, Wigander et al. did not show any correlation between functional problems and quality of life [8]. Similarly, follow-up studies of quality of life in HSCR patients showed equivocal results. Variable degrees of functional impairment were found in pediatric and adult patients and were not always associated with altered overall quality of life when compared to healthy controls [9, 10]. However, studies of HSCR patients with total colonic aganglionosis have shown a negative impact on quality of life compared with healthy peers [11].

The absence of a relationship between disease severity, bowel function, and quality of life makes it difficult for clinicians to predict the patients and parental needs. In a standard setting, Hartman et al. suggested that patients' perception of the disease and their life coping strategies were as important as disease severity and additional congenital anomalies, in terms of their influence on quality of life [12]. Our findings illustrate that identifying patients at risk for health problems cannot be identified by relying solely on disease severity, bowel function or quality of life and inquiry into psychosocial resources should be sought.

Social relations constitute one of the domains of the quality of life as defined by the World Health Organization. Bowel dysfunction may influence the health-related quality of life of patients with CCMs. In a series of 33 adolescents with ARMs, Diseth and Emblem found that incontinence to flatus or stool was associated with psychiatric or psychosocial impairment [13]. However, Ojmyr-Joelsson et al. described a series of 25 school-aged children with severe ARMs who reported either constipation or fecal incontinence, and did not show lower emotional health or self-esteem and reported good relationships with their friends [14]. During the time of our study, social relationships were turned upside down due to the lockdown. Some parents perceived this as a benefit to their child's health, for example, if their child experienced bullying at school; however, for other children a lack of social interaction was a hindrance to their well-being.

Our study has several notable limitations. The sample size is small and not representative of the actual distribution of CCMs. Patients with a severe form of ARM or HSCR constitute a larger proportion of our cohort when compared to the

expected distribution [15, 16]. Likewise, patients using an ACE for either fecal incontinence or constipation are over-represented in our cohort. This selection bias may stem from the fact that participants were selected during a short period through medical outpatient appointments and nurse-led clinics—where there is a focus on maintaining adequate washout and stoma bag supplies for patients. However, this selection bias allowed us to have a qualitative assessment of a broad spectrum of children with congenital colorectal malformations and associated anomalies, ranging from patients doing well with minimal intervention to those in a structured bowel management program.

Secondly, the outcomes are, for the most part, reported by the parents and may not reflect the child's or the adolescent's point of view. Although the adolescent or the older child was allowed to express themselves, the vast majority declined.

The third important limitation is the lack of functional and quality of life scores gathered during the period preceding the lockdown. Therefore, no comparison can be drawn in absence of a reference score.

Telemedicine in pediatric surgery was not considered the usual standard of care [17]. During the Sars-CoV-2 pandemic, most of our patients with chronic health conditions have been managed by telemedicine. However, this delivery of pediatric surgical services by telemedicine has not been formally assessed. In a survey of parental perception of telemedicine in a pediatric population, Abdulhai et al. showed that it was supported by 70% of the respondents regarding postoperative visits and 68% for subspecialty evaluations [18]. In our study it was only possible to perform telephone clinics. Despite this, 88% of the parents were happy and satisfied to have a telemedicine follow-up. Fifty-seven percent wanted to see their physician if they experienced a problem. It can be assumed that this high percentage might improve by use of video clinics. Unfortunately, this technology did not yet exist at the beginning of the first lockdown in our hospital in 2020.

Despite its limitations, this study allowed us to have a broad look at our patients' health during the lockdown and investigate telemedicine's efficacy. A more extensive cohort study would be required to confirm these preliminary results and inform a change in practice to focus on the patient and parent-reported quality of life as part of routine care. The evidence of psychosocial concerns influencing quality of life suggests that many of these patients would benefit from more structured psychological support.

Conclusion

Severity of the CCM or degree of bowel dysfunction are not linked to deterioration of health during the Sars-CoV-2 pandemic induced lockdown period. Follow-up of these children

319 and adolescents by telemedicine is possible but the assess-
320 ment should contain both disease-specific and psychosocial
321 questions to allow detection of patients at risk for quality of
322 life problems.

323 **Supplementary Information** The online version contains supplement-
324 ary material available at <https://doi.org/10.1007/s00383-021-04971-6>.

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328 Declarations

329 **Conflict of interest** There are no disclosures or conflicts of interest.

330 **Ethical approval** The study was registered and approved as service
331 evaluation following assessment using the UK NHS research govern-
332 ance assessment tool (<https://www.hra-decisiontools.org.uk/research/>)
333 by the local Research Governance department at our institution (Bir-
334 mingham Women's and Children's NHS Foundation Trust) and deemed
335 not to require ethical approval (CARMS-30563).

336 **Informed consent** Informed consent of all patients and their parents/
337 legal guardians was verbally obtained before inclusion.

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