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Original research

Childhood outcomes in children with Hirschsprung disease: a population-based data linkage study in England

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ABSTRACT

Objective Hirschsprung disease (HSCR) is a rare congenital intestinal condition that, despite corrective surgery, can lead to recurrent hospitalisations and reduced quality of life throughout childhood. There is limited population-level evidence on the causes of these admissions or associated educational needs.

Design/methods Using linked health and education data from the Education and Child Health Insights from Linked Data database, we created a cohort of all children born in England between 2002 and 2020. We compared admission rates, mortality, surgical procedures and causes of admission for children with and without HSCR at ages 0–4, 5–9 and 10–14 years. We also assessed the prevalence of recorded Special Educational Needs and Disability (SEND) by Year 1 of primary school (age 6).

Results Among 11 261 227 children, 3227 (0.03%) had HSCR. By age 4, 95.0% of children with HSCR had been readmitted, compared with 40.2% without. Common admission reasons included constipation, gastroenteritis, intestinal infection, abdominal pain and nausea or vomiting. 81.4% of children with HSCR had two or more surgical procedures between ages 0 and 4, compared with 2.9% in children without HSCR. Mortality by age 4 was 3.2% for HSCR versus 0.5% for non-HSCR children. By age 6, 44.0% of children with HSCR had recorded SEND compared with 17.9% of those without.

Conclusion Children with HSCR experience substantially higher hospital readmission rates, more surgeries and greater mortality up to age 14 than their peers. They are also more likely to require educational support, independent of comorbidities such as Down syndrome. Improvements in surgical and long-term care, including novel or complementary approaches, alongside enhanced educational and psychosocial support, are needed to improve outcomes and quality of life across childhood and adolescence.

WHAT IS ALREADY KNOWN ON THIS TOPIC

⇒ Hirschsprung disease (HSCR) is a rare congenital condition treated surgically in infancy, but long-term outcomes remain poorly understood. Existing studies have provided limited insight into broader population impacts of HSCR. Hence, comprehensive national data are needed to clarify ongoing health and developmental challenges in children with HSCR.

WHAT THIS STUDY ADDS

⇒ Using linked national health and education data from over 11 million children in England, this study provides the first population-level evidence on long-term outcomes in HSCR. Children with HSCR had significantly higher rates of hospital admissions, mortality and Special Educational Needs compared with peers. These findings highlight the persistent health and developmental burdens of HSCR despite early surgical intervention.

HOW THIS STUDY MIGHT AFFECT RESEARCH, PRACTICE OR POLICY

⇒ Our study emphasises the need for coordinated long-term, multidisciplinary care for children with HSCR. Our data can inform clinical pathways and policy planning to better address ongoing medical and educational needs for children and families living with HSCR.

INTRODUCTION

Hirschsprung disease (HSCR) is a rare congenital intestinal condition that

affects approximately 1 in 5000 live births.[1] The hallmark pathology is absence of ganglion cells within the enteric nervous system (the intrinsic innervation of the gastrointestinal tract) in a variable length of the distal bowel and occasionally even part of the small intestine.[2]

Most affected individuals are diagnosed in the neonatal period, though a proportion of patients may present in childhood with typical symptoms of severe constipation with abdominal distension and failure to thrive.[3] Treatment for the condition is surgical, involving the removal of the aganglionic bowel and performing a pull-through and anastomosis between ganglionic proximal bowel and the anorectum.[4 5] This is achieved most commonly using either Duhamel or endorectal pull-through.[6] Although primary surgery is generally the first choice among surgeons, there may be cases where a defunctioning stoma is necessary to safely decompress the bowel prior to elective surgery. Additionally, a proportion of affected individuals may have associated anomalies that mean faecal continence is likely to be poor and, in such cases, a definitive levelling stoma may be preferred.[7]

Although survival is high (>97%), children with HSCR are known to have higher morbidity than the general population, experiencing more frequent hospitalisations and longer inpatient stays and increased requirement for surgery or procedures under general anaesthesia compared with children with other congenital anomalies or those without congenital anomalies.[8] In the longer term, children with HSCR often experience lower quality of life than other children and are commonly affected by ongoing bowel dysfunction, including faecal incontinence, chronic constipation and soiling. One-third of patients will require a further unplanned abdominal procedure.[9] While bowel function improves with age, >10% of children ultimately require a permanent stoma, the need for which has been identified as a core outcome of importance in determining the success of treatment for HSCR.[10–12] A third of patients report social problems related to bowel function into adulthood.[13]

The developmental outcomes of children with HSCR are less well studied. There is some evidence to suggest that children with gastrointestinal anomalies, including HSCR, have impaired developmental and cognitive outcomes, which may be in part explained by frequent, extended hospitalisation and numerous surgical procedures.[14 15] There are concerns that multiple prolonged exposure to general anaesthesia during the neonatal period and infancy may affect brain development.[16 17] Additionally, chromosomal anomalies such as Trisomy 21, or syndromic conditions such as Waardenburg-Shah syndrome, which can be associated with sensorineural deafness, have been shown to be associated with HSCR.[18] Such conditions may impact on developmental outcomes and impact on learning outcomes.

Although the epidemiology of HSCR has been well described in different clinical cohorts across Europe and North America, there is relatively little population-level research to understand the longer-term outcomes, including the need for educational support, of HSCR.[1] Hence, the aim of this study was to assess the clinical and educational experience of children with HSCR to better define ongoing needs. In this study, we used national administrative data from ECHILD (Education and Child Health Insights from Linked Data) to describe characteristics and outcomes of children with HSCR, in England, including hospitalisation outcomes up to 14 years of age and recording of Special Educational Needs by age 6.

METHODS

Data source

We used ECHILD, which links administrative data from Hospital Episode Statistics (HES) and the National Pupil Database (NPD) for all children in England.[19] Education data from the NPD contains information on children attending state-funded schools in England, including Special Educational Needs and Disability (SEND).[20] Health data from HES analysed in this study included records of inpatient hospital attendances (admitted patient care) at all National Health Service (NHS) funded hospitals in England.[21] HES records contain basic demographic information (eg, sex, ethnicity), area-level deprivation measured by the Index of Multiple Deprivation (IMD) of residential postcode, and clinical information based on International Classification of Diseases 10th revision (ICD-10) diagnostic codes and Office of Population Censuses and Surveys Classification of Interventions and Procedures fourth revision (OPCS-4) procedure codes. Information on birth characteristics such as gestational age, birth weight and maternal age is also included through a mother–baby link.[22] Approximately 97% of all children born in NHS-funded hospitals in England have a birth record in HES. We also used linked mortality data from the Office for National Statistics.

Study population

Our study included all singleton children born in NHS-funded hospitals between 1 April 2002 and 31 March 2020. Higher-order births (eg, twins) were excluded due to less reliable linkage of hospital records over time. Hospital admission records from birth were used to identify a cohort of children with HSCR. For each admission, 1 primary diagnosis and up to 19 secondary diagnoses are recorded. Any recording of an ICD-10 diagnostic code for HSCR (Q43.1) was used to identify the cohort of children with the disease. This code has been shown to have 96% sensitivity and >99.99% specificity for identifying children with HSCR.[23] All other children (including those with other health conditions) were included in the comparison cohort.

Aligning with a previous study of HSCR using HES, we also identified the presence of common comorbidities based on ICD-10 codes: major cardiac anomalies (Q20–Q26) and chromosomal abnormalities (Q90–93, Q96–Q99, excluding Q93.6).[24]

Outcomes

Our primary outcome of interest was admissions between ages 0–4, 5–9 and 10–14 years (excluding the birth episode). We also explored reasons for admissions, number of surgical procedures (identified through OPCS codes, excluding minor procedures, diagnostic imaging procedures and codes describing subsidiary procedures/body parts, online supplemental table 1), and mortality. Focus groups and stakeholder work contributing to the development of the Children's Surgery Outcome Reporting (CSOR) programme have highlighted how the total number of operations a child undergoes is important to key stakeholder groups, not simply the number of operations that are related to a specific aspect of the child's care or performed in a specific setting. Development of this programme has also highlighted how, without individual case review, it is not currently reliably possible to differentiate where potentially unrelated operations, such as reconstructive vascular procedures, are genuinely unrelated, and where they are necessary due to a complication of a previous operation. To reliably capture the full burden of operative intervention for a child, and to reflect what is important to stakeholders, our analysis has therefore captured information about every operation a child has undergone, including operations other than those related directly to the definitive treatment of the child's HSCR. However, as multiple separate OPCS codes are frequently used to describe different parts of a single procedure, where multiple surgical procedures are recorded as occurring on the same day, these have been considered as one event.

To understand the extent to which children with HSCR needed learning support, we examined SEND recorded in school records by the end of Year 1 (age 6).[25] In England, children are entitled to receive SEND provision if they have 'a significantly greater difficulty in learning than the majority of others of the same age, or have a disability which prevents them from making use of facilities generally provided by mainstream schools'. Children likely to need SEND provision include those with disability or chronic physical or mental health conditions that affect learning.[26] In this study, we include both SEND support (classroom-based support arranged and funded by schools) and education health and care plans (arranged and funded by local authorities, for children whose needs cannot be met by SEND support) under our SEND grouping, and do not differentiate between support for physical versus learning needs.

Statistical analysis

We described the characteristics of children with and without HSCR, including year of birth, sex, gestational age (<32 weeks, 32–36 weeks, 37+weeks), birth weight (<1500 g, 1500–2500 g, 2500 g+), small or large for gestation (<10th or >90th percentile of birth weight for gestation derived from national birth weight percentiles),[27] ethnic group (black, white, Asian, mixed or other), maternal age (≤20, 21–30, 31–40 and 40+years) and quintile of deprivation (IMD of residential postcode). We quantified the number of children with and without HSCR with major cardiac anomalies (ICD-10 codes Q20–Q26) and chromosomal abnormalities including Down syndrome (ICD-10 codes Q90–93, Q96–Q99, excluding Q93.6).[24] For those with HSCR, we described age at diagnosis.

Our study population was stratified into 5-year birth cohorts (April 2002 to March 2007; April 2007 to March 2012; April 2012 to March 2016) with follow-up to age 14, age 9 and age 4, respectively (online supplemental figure 1). Since the majority of children with HSCR are diagnosed neonatally, and to ensure comparable time periods for children with and without HSCR, we evaluated the proportion of children who had an admission, a surgical procedure or death following discharge from their birth episode, up to the full extent of follow-up (or death). We then explored the total number of admissions and surgical procedures, and the most frequently occurring reasons for admission according to ICD-10 codes and the high-level chapter of the primary diagnosis code, stratified by sex. We also conducted a stratified analysis for children with and without Down syndrome.

Analysis of SEND outcomes in Year 1 (age 6) was conducted for the subset of children who were linked to their education record in NPD. We quantified SEND separately for the cohort of children without major cardiac anomalies or chromosomal abnormalities (since many of these children will require extra support at school, irrespective of having HSCR).

For admissions and SEND, we used multivariable generalised linear models to estimate the relative risk of having the outcome, adjusting for sex, ethnicity, maternal age, deprivation, gestational age and comorbidities. To explore whether the effect of HSCR on SEND differed according to sex or deprivation, we included interaction terms within the models. Analysis was conducted in Stata V.17.

RESULTS

Cohort characteristics

Of the 11 261 227 live births in England, between April 2002 and March 2020, 3227 (0.03%) had a diagnosis of HSCR coded in their hospital record (corresponding to ~1 in 3500 births). Of the 3227 children with HSCR, the majority (65.6%) were diagnosed in the first 28 days of life; an additional 10.4% were diagnosed between 29 and 90 days; 9.5% were diagnosed age 2 or older, and 4.3% were diagnosed at age 5 or older.

Compared with children without the condition, children with HSCR were more likely to be male, have been born to younger mothers, be living in more deprived areas, preterm, low birth weight, small for gestational age and have major cardiac anomalies or chromosomal abnormalities (table 1). 9.2% of children with HSCR also had Down syndrome compared with 0.1% of children with no HSCR (2.0% of children in the cohort with Down syndrome had HSCR).

Rates of hospital admissions and surgical procedures

Children with HSCR were more likely than those without to be admitted for any reason than those without HSCR: 95.0% of children with HSCR were admitted at least once by age 4 compared with 40.2% of those without (table 2). Children with HSCR had a median of 6 admissions (IQR 3–12) by age 4, compared with a median of 0 admissions (IQR 0–1) for those without HSCR. Over a quarter of children with HSCR had 10 or more admissions by age 4 (online supplemental table 2).

Admission rates decreased with age but were still substantially higher by age 14 in children with HSCR compared with those without. Boys were more likely to be admitted than girls. For children with Down syndrome, admission rates were still higher among the group with HSCR: 95.5% of those with HSCR were admitted by age 4 compared with 85.8% of those without HSCR; 65.0% versus 60.7% were admitted between ages 5 and 9, and 64.1% versus 48.7% were admitted between ages 10 and 14 (online supplemental table 3).

Of children with HSCR, 81.4% had two or more surgical procedures between ages 0 and 4, compared with 2.9% in children without HSCR (table 3, online supplemental table 4). Overall numbers of surgical procedures remained high at ages 5–9 and 10–14, with differences between children with and without HSCR persisting. The most commonly recorded surgical procedures were H41.2 (peranal excision of lesion of rectum; 73.7%) and H41.8 (other operations on rectum through anus; 61.1%) for ages 0–4, H56.8 (other operations on anus; 6.3%) for ages 5–9 and G45.1 (fibreoptic endoscopic examination of upper gastrointestinal tract and biopsy of lesion of upper gastrointestinal tract; 3.6%) for ages 10–14 (online supplemental table 5).

In analyses controlling for key risk factors (sex, ethnicity, deprivation, gestational age at birth, comorbidities), children with HSCR were 42% more likely to have been admitted between ages 0 and 4 (incidence rate ratio (IRR) 1.47; 95% CI 1.40 to 1.53), 40% more likely to have been admitted between ages 5 and 9 (IRR 1.40; 95% CI 1.29 to 1.51) and 35% more likely to have been admitted between ages 10 and 14 (IRR 1.35; 95% CI 1.17 to 1.55), compared with those without HSCR.

Reasons for admission

Across all age groups, children with HSCR were more likely than those without to be admitted with diagnoses falling under virtually all ICD-10 diagnosis chapters (online supplemental figure 2, online supplemental table 6). The largest differences in admission rates were seen in diagnoses relating to diseases of the digestive system. Reasons for admission were relatively similar for girls and boys (online supplemental tables 7 and 8 for ages 0–4 and ages 5–9; numbers were too small to stratify for the age 10–14 cohort).

The most frequently occurring diagnoses in admissions within the HSCR group across all age groups and sexes were constipation (affecting ~35% of children; figure 1, online supplemental table 9), followed by gastroenteritis and intestinal infections. Around 15% of children across all age groups were readmitted for nausea and vomiting, and 8–10% were readmitted for abdominal pain. Across all age groups, 8–11% of children were readmitted with a primary diagnosis of 'flatulence and related conditions' (ICD-10 code R14, which covers various digestive and abdominal symptoms, including an inability to pass flatus (ie, functional sphincter achalasia) as well as bloating, belching and gas pain).

Mortality

Mortality rates were substantially higher in children with HSCR: 86/2700 (3.2%) of children with HSCR had died by age 4 compared with 0.5% in children without HSCR; 10/1768 (0.6%) of children with HSCR died between ages 5 and 9 compared with 0.03% of children without HSCR. Excluding those with chromosomal abnormalities or major cardiac anomalies, 39/2217 (1.8%) children with HSCR had died by age 4 compared with 35 804/9 294 385 (0.4%) of children without HSCR. Numbers for deaths between ages 10 and 14, or deaths aged 5–9 for those without comorbidities, were too low to report. Taken together, these data suggest that while mortality rates are higher in children with HSCR throughout childhood, the early infancy (0–4 years) is a primary period of vulnerability.

Special Educational Needs at school age

Within the 8 046 292 (71.5%) of children who were linked to a school record in Year 1 (age 6) NPD, 892/2025 (44.0%) of those with HSCR had recorded Special Educational Needs compared with 1 213 133/6 777 687 (17.9%) of those without HSCR. For the cohort of children without chromosomal anomalies, 680/1807 (37.6%) of those with HSCR had recorded Special Educational Needs compared with 1 195 761/6 757 836 (17.7%) of those without HSCR. In adjusted analysis controlling for sex, ethnicity, maternal age, deprivation, gestational age and comorbidities, children with HSCR were 34% more likely to have recorded SEND by Year 1 compared with those without HSCR (IRR 1.34; 95% CI 1.24 to 1.45). Interaction terms

Table 1 Characteristics of the study cohort (births between April 2002 and March 2020)

	Total		Hirschsprung diagnosis		No Hirschsprung diagnosis	
	N	%*	N	%*	N	%*
Total	11 261 227		3227	0.03	11 258 000	99.97
Sex						
Male	5 766 904	51.2	2353	72.9	5 764 551	51.2
Female	5 480 929	48.7	873	27.1	5 480 056	48.7
Missing						
Ethnicity*						
White	7 657 457	76.0	2438	76.7	7 655 019	76.0
Black	533 469	5.3	185	5.8	533 284	5.3
Asian	1 076 733	10.7	274	8.6	1 076 459	10.7
Mixed	466 551	4.6	155	4.9	466 396	4.6
Other	344 087	3.4	126	4.0	343 961	3.4
Unknown	1 182 930	10.5	49	1.5	1 182 881	10.5
Maternal age*						
<20	539 059	5.2	166	5.9	538 893	5.2
20–29	4 681 567	44.9	1351	47.8	4 680 216	44.9
30–39	4 821 427	46.2	1188	42.0	4 820 239	46.2
40+	394 848	3.8	122	4.3	394 726	3.8
Missing	824 326	7.3	400	12.4	823 926	7.3
Quintile of deprivation (IMD)*						
Most deprived	2 231 324	25.6	936	29.4	2 230 388	25.6
2	1 776 098	20.4	645	20.2	1 775 453	20.4
3	1 489 401	17.1	554	17.4	1 488 847	17.1
4	1 316 385	15.1	477	15.0	1 315 908	15.1
Least deprived	1 888 650	21.7	576	18.1	1 888 074	21.7
Missing	2 512 370	22.3	38	1.2	2 515 332	22.3
Gestational age (weeks)*						
<32	104 176	1.2	79	3.3	104 097	1.2
32–36	534 113	6.0	248	10.4	533 865	6.0
37+	8 248 788	92.8	2064	86.3	8 246 724	92.8
Missing	2 374 150	21.1	836	25.9	2 373 314	21.1
Birth weight (g)*						
<1500	93 895	1.0	63	2.5	93 832	1.0
1500–2500	554 010	5.9	238	9.5	553 772	5.9
>2500	8 727 676	93.1	2214	88.0	8 725 462	93.1
Missing	1 885 646	16.7	712	22.1	1 884 934	16.7
Size for gestation*						
Small (<10th percentile)	605 184	7.0	177	7.6	605 007	7.0
Normal	7 304 306	84.5	1965	84.4	7 302 341	84.5
Large (>90th percentile)	739 402	8.5	187	8.0	739 215	8.5
Missing	2 612 335	23.2	898	27.8	2 611 437	23.2
Comorbidities						
Major cardiac anomalies (any)	159 871	1.4	501	15.5	159 370	1.4
…of cardiac chambers and connections	10 608	0.1	406	12.6	95 218	0.8
…of cardiac septa	95 624	0.8	55	1.7	16 190	0.1
…of pulmonary and tricuspid valves	16 245	0.1	40	1.2	14 352	0.1
…Other congenital malformations of heart	14 392	0.1	66	2	22 089	0.2
…of great arteries	22 155	0.2	278	8.6	92 993	0.8
…of great veins	93 271	0.8	19	0.6	5114	0
Chromosomal abnormalities (any)	31 357	0.3				
…Down syndrome	13 230	0.1	271	8.4	12 959	0.1

Continued

Table 1 Continued

	Total		Hirschsprung diagnosis		No Hirschsprung diagnosis	
	N	%*	N	%*	N	%*
…Edwards syndrome and Patau syndrome	1571	0.0	<10		†	
…Other trisomies and partial trisomies of the autosomes, NEC	3216	0.0	22	0.7	3194	0
…Monosomies and deletions from the autosomes, not elsewhere classified	8938	0.1	49	1.5	8889	0.1
…Turner syndrome	1256	0.0	<10		†	
…Other sex chromosome abnormalities, female phenotype, NEC	649	0.0	<10		†	
…Other sex chromosome abnormalities, male phenotype, NEC	1280	0.0	<10		†	
Primary or secondary pulmonary hypertension	7717	0.1	35	1.1	7682	0.1

*Percentages are of complete records.
†Cell sizes <10 and those which would allow small cell sizes to be derived have been supressed due to statistical disclosure control.
IMD, Index of Multiple Deprivation; NEC, not elsewhere classified.

indicated that there was no difference in the effect of HSCR on SEND by quintile of deprivation. However, the impact of HSCR on SEND was greater for females (IRR 1.57; 95% CI 1.33 to 1.85) compared with males (IRR 1.36; 95% CI 1.25 to 1.49; p value for interaction <0.0001).

DISCUSSION

Our study aimed to fill a gap in evidence on longitudinal outcomes for children with HSCR at a population level, describing characteristics and outcomes of children with HSCR including need for hospitalisation (up to 14 years of age) and educational needs. To the best of our knowledge, this represents the first time that a national database spanning multiple decades has been used to evaluate the frequency of hospital admissions in an HSCR cohort, as well as the specific health problems precipitating admissions. Our study highlights that irrespective of common comorbidities such as Down syndrome, children with HSCR have a significantly higher mortality rate and are admitted to hospital frequently, and more often than children without HSCR, at least up to age 14. Importantly, these admissions are often for constipation, enterocolitis and abdominal pain, suggesting ongoing severe sequelae despite surgical intervention. Our data also

Table 2 Number and percentage of children with and without Hirschsprung disease admitted to hospital, by age and sex

	Hirschsprung diagnosis			No Hirschsprung diagnosis		
		Children admitted			Children admitted	
	Total	N	%	Total	N	%
All						
Admission aged 0–4 years Cohort 1: born between 2002/2003 and 2016/2017	2700	2566	95.0	9 435 790	3 790 361	40.2
Admission aged 5–9 years Cohort 2: born between 2002/2003 and 2011/2012	1768	920	52.0	6 200 844	1 210 703	19.5
Admission aged 10–14 years Cohort 3: born between 2002/2003 and 2006/2007	855	320	37.4	2 910 568	427 511	14.7
Girls						
Admission aged 0–4 years **Cohort 1:** born between 2002/2003 and 2016/2017	733	680	92.8	4 593 676	1 672 581	36.4
Admission aged 5–9 years **Cohort 2:** born between 2002/2003 and 2011/2012	469	234	49.9	3 018 246	533 228	17.7
Admission aged 10–14 years **Cohort 3:** born between 2002/2003 and 2006/2007	225	73	32.4	1 418 725	197 343	13.9
Boys						
Admission aged 0–4 years **Cohort 1:** born between 2002/2003 and 2016/2017	1966	1886	95.9	4 829 738	2 117 780	43.8
Admission aged 5–9 years **Cohort 2:** born between 2002/2003 and 2011/2012	1299	686	52.8	3 170 930	677 475	21.4
Admission aged 10–14 years **Cohort 3:** born between 2002/2003 and 2006/2007	630	247	39.2	1 487 034	230 168	15.5

Table 3 Number of surgical procedures for children with and without Hirschsprung disease admitted to hospital, by age and sex

	Total				Boys				Girls			
	HSCR		No HSCR		HSCR		No HSCR		HSCR		No HSCR	
N procedures	N	%	N	%	N	%	N	%	N	%	N	%
Age 0–4	2700		9 435 790		1966		4 829 738		733		4 593 676	
1	300	11.1	919 105	9.7	191	9.7	551 895	11.4	109	14.9	367 073	8.0
2–3	976	36.1	206 444	2.2	709	36.1	130 867	2.7	266	36.3	75 555	1.6
4–5	582	21.6	33 039	0.4	437	22.2	19 624	0.4	145	19.8	13 411	0.3
6–9	449	16.6	17 343	0.2	356	18.1	10 062	0.2	93	12.7	7279	0.2
10+	191	7.1	13 265	0.1	144	7.3	7421	0.2	47	6.4	5843	0.1
Age 5–9	1768		6 200 844		1299		3 170 930		469		3 018 246	
1	175	9.9	1 023 620	16.5	111	8.5	583 739	18.4	64	13.6	439 782	14.6
2–3	563	31.8	337 656	5.4	416	32.0	207 774	6.6	147	31.3	129 863	4.3
4–5	377	21.3	47 712	0.8	271	20.9	28 970	0.9	106	22.6	18 738	0.6
6–9	338	19.1	21 848	0.4	269	20.7	12 757	0.4	69	14.7	9089	0.3
10+	227	12.8	17 723	0.3	178	13.7	10 004	0.3	49	10.4	7718	0.3
Age 10–14	855		2 910 568		630		1 487 034		225		1 418 725	
1	88	10.3	536 308	8.6	55	8.7	299 974	20.2	33	14.7	236 270	16.7
2–3	249	29.1	225 927	3.6	178	28.3	136 539	9.2	71	31.6	89 370	6.3
4–5	171	20.0	36 801	0.6	137	21.7	22 388	1.5	34	15.1	14 409	1.0
6–9	173	20.2	16 534	0.3	137	21.7	9705	0.7	36	16.0	6827	0.5
10+	127	14.9	13 124	0.2	94	14.9	7412	0.5	33	14.7	5711	0.4

HSCR, Hirschsprung disease.

highlight shifts in admission of children with HSCR for specific diagnosis according to age. Interestingly, we observed that ~10% of children with HSCR were admitted with diagnosis codes for flatulence and related conditions across all age groups. Although this ICD-10 code covers various digestive and abdominal symptoms, including bloating, belching and gas pain, this finding points towards ongoing issues surrounding bowel habits which potentially impact early schooling (through disruption caused by issues with toileting) and quality of life during adolescence.[28 29] These findings support previously published work that showed 49% of children with HSCR scored one SD below the reference population in terms of quality-of-life measures, and 42% of children with HSCR reporting significantly reduced psychological well-being.[30]

While surgery for HSCR is lifesaving, patients often display significant downstream issues affecting their quality of life.[31–34] The aetiology of these ongoing symptoms is currently incompletely understood despite substantial efforts.[24 35–40] For example, the NETS^{2HD} UK-based study, a prospective cohort study conducted from 2010 to 2012, detailed specific outcomes that could be tracked following surgical intervention to better inform surgical practice.[30] This included data from 28 UK and Irish paediatric surgical centres. The related NETS^{1HD} study developed a core outcome set to standardise surgical success metrics, such as 'long-term faecal incontinence' and a 'need for a permanent stoma'.[12] However, this has not yet been fully implemented or evaluated, and there is currently no clear consensus on the most effective surgical technique, further highlighting the importance of evaluating long-term functional results.

Our findings support evidence from several other studies that have conclusively demonstrated ongoing HSCR-related symptoms persisting post-surgery, into adult life.[41 42] NETS^{2HD}, which examined outcomes in children with HSCR at 5–8 years of age in the UK, found that 44% of children in this cohort had undergone at least one unplanned reoperation, with 27% classified as major or complex procedures.[30] However, previous studies have often been focused on specific outcomes, such as diarrhoea and constipation, with many studies relying on patient self-reporting QoL outcomes.[31 43–46]

In addition to increased mortality and ongoing health issues, we also found that children with HSCR were significantly more likely to have recorded Special Educational Needs at the start of compulsory schooling. A recent systematic review has outlined reductions in school functioning in children with HSCR.[18] This review particularly highlighted issues surrounding peer relationships, though other indicators such as school performance, teacher relationships and need for special education services varied widely and were less consistently measured, pointing to the need for large longitudinal studies with empirical metrics to better understand and address the educational needs in HSCR. While it is well established that HSCR can be associated with chromosomal abnormalities (eg, Trisomy 21), our study shows that even after

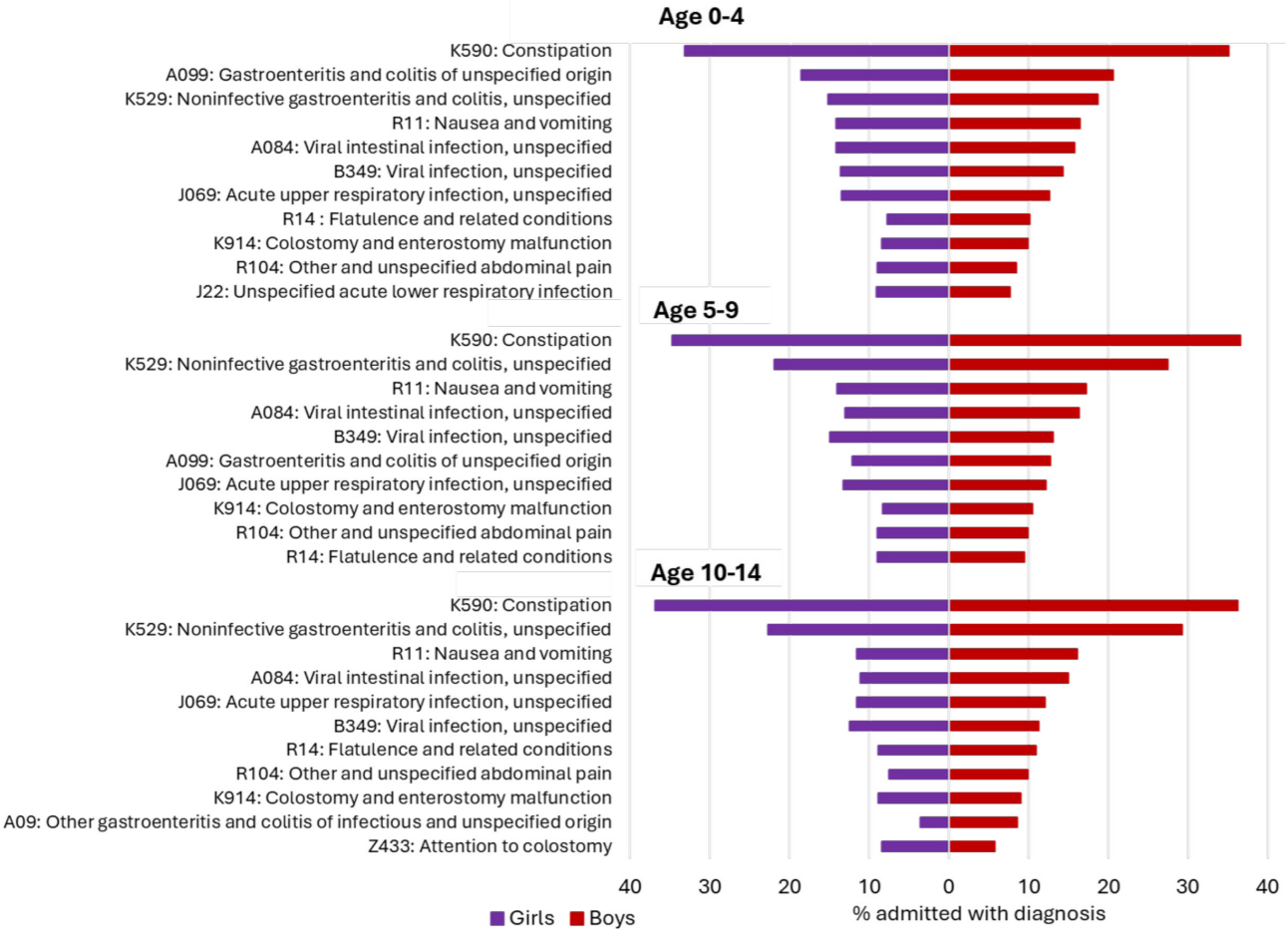

Figure 1 Most frequently occurring ICD 10 codes in primary diagnosis fields for children with Hirschsprung disease, by age and sex.

controlling for these conditions, children with HSCR remained 34% more likely to have recorded SEND by Year 1 compared with those without HSCR.[1 47 48] More research is needed to understand the extent to which this is explained by organic cognitive issues, ongoing needs to manage their condition or increased hospitalisations which may impact on learning. Interestingly, a Swedish study found that the adult HSCR population (aged 16–49) achieved comparable earning potential and educational qualifications compared with age and sex-matched individuals in the general population.[49] This may suggest that HSCR does not itself impact on learning potential, but perhaps that recorded SEND in the current study represents the impact of extended educational absences due to frequent hospital admissions or the need for practical assistance within school to deal with ongoing symptoms. Of note, our findings suggest the impact of HSCR on SEND was greater for females who have been shown to have a higher risk of long segment aganglionosis and may therefore be more likely to have a stoma or increased issues with soiling, which may require practical SEND assistance with toileting habits.[50] Importantly, the impact of HSCR on educational outcomes is highly likely to be country specific, as the ability of national education sectors to incorporate children with HSCR into school and keep them in education is likely to vary significantly country to country.[51] However, regardless of cause, our data highlight a critical ongoing need for children with HSCR and their families, particularly during formative years.

A major strength of our study is the population-level coverage of our data, capturing ~97% of births in NHS hospitals in England. Our findings are comparable with other literature using administrative health records to analyse outcomes of HSCR: a previous study using HES found that 85.3% of those with a diagnosis had an operation within the first year of life (in our study, this was 87.4%).[24] The longitudinal nature of the dataset allowed us to evaluate longer-term outcomes and to link with outcomes outside of health, in order to start to understand the experiences of children with HSCR in school. We specifically focused on children aged up to 14, before the transition from paediatric to adult services at the age of 16. Difficulties in the management of disease are often experienced at this transition period, and future work will look separately at older adolescents, including the impact that HSCR has on their mental health.[52 53]

A limitation of our study is the nature of recording of ICD-10 diagnosis codes and OPCS procedure codes; these codes are used primarily for billing purposes and there may be some misclassification. We relied on diagnosis dates as recorded in inpatient records. However, diagnoses may not always be made as an inpatient (eg, if a biopsy is returned after discharge) and in this case, would not be coded until a later hospital admission (in which case the true diagnosis date would have been earlier than we identified). Of note, 9.5% of the current cohort were recorded as being diagnosed aged 2 or above, which likely represents a higher than anticipated proportion of infants diagnosed at this age, even with a delay in recording of diagnosis. In order to investigate this further, we are proposing in future work to look in detail at the prior admissions for these infants to identify those who have a definitive late diagnosis (ie, those having their first rectal biopsy performed outside of the neonatal period), and those where the late appearance of the coded diagnosis is more likely to be due to delayed recording. In addition, some missing data on maternal age, deprivation and ethnicity means that we were unable to fully evaluate the influence of these factors. A further limitation is that we were not able to distinguish between different types or severities of HSCR, and the genetic basis for HSCR in individual patients is not recorded. Nevertheless, our study is clinically relevant and provides an improved understanding of HSCR burden, highlighting the real-world impact on children and their families/support groups. We could only evaluate recorded SEND for those with a linked educational record; those attending schools outside of England (eg, Wales, Scotland) or attending private schools or being home schooled were excluded. By including all children in our comparator group (including those who may have other significant health conditions), we may have underestimated the differences in outcomes between children with Hirschsprung's disease and 'healthy' children.

Clinical management of HSCR is primarily based on the success of early surgical interventions. However, despite well-established treatment regimens for HSCR, children with this condition remain at higher risk of chronic clinical complications and potential developmental delays. Combining administrative data that provide the power to generate population-level insights, with greater detail from dedicated follow-up of clinical cohorts, such as through CSOR, may help guide targeted interventions, tailored multidisciplinary follow-up and potentially earlier intervention for these children. Additionally, our data highlights the need for ongoing support of children with HSCR throughout childhood and adolescence, providing healthcare systems and carers with crucial information which can be used to anticipate and plan long-term needs and resources. Indeed, evidence suggests that families with a new diagnosis of HSCR often feel under-informed as to the potential impacts of the condition on their child's development.[45] For example, newly diagnosed patients with relatives affected by HSCR report better outcomes, possibly suggesting that better information is linked to higher quality of life.[46]

Moreover, our study highlights that while surgical intervention is essential, it does not provide complete resolution of the condition, with outcomes varying widely. Hence, there is a critical need for novel approaches to improve treatment success and quality of life for people living with HSCR. In addition, further research is also needed to understand the impact of HSCR and its treatment at the transition from paediatric to adult services, and to evaluate child development, educational attainment and other psychosocial factors including mental health.

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