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The use of stomas in the early management of Hirschsprung disease: Findings of a national, prospective cohort study☆,☆☆

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ABSTRACT

Background/purpose: Primary pull-through without a stoma has become preferred practice in managing Hirschsprung disease (HD). The aims of this study were to establish stoma rate and identify factors associated with stoma formation in a population-based cohort in the UK and Ireland.
Methods: Live-born infants with HD were prospectively identified in all 28 specialist pediatric surgical units in the UK and Ireland between October 2010 to September 2012. Method of colonic decompression was recorded and multivariable logistic regression was used to identify factors associated with stoma formation.
Results: 305 infants with HD were identified. Rectal washouts were initially used in 86% (263) with a defunctioning stoma formed as the primary management in 13% (39). Ultimately, 36% (111) required a stoma prior to definitive surgery. Compared to infants managed with rectal washouts alone; infants managed with a stoma were more likely to have a transition zone proximal to the splenic flexure, Down (or another) syndrome, and HD diagnosis established more than 28 days after presentation.
Conclusions: Although rectal washouts are commonly employed, a stoma prior to definitive surgery was required in 36% of infants in a national cohort. Delayed diagnosis, aganglionosis proximal to the splenic flexure and presence of other anomalies are associated with stoma formation.
Type of study and level of evidence: Prognosis study (high-quality prospective cohort study with 80% follow-up and all patients enrolled at same time point in disease).

Hirschsprung disease (HD) classically presents during the neonatal period or early infancy [1] and management involves initial colonic decompression, followed by a pull-through procedure. Most pediatric surgeons advocate rectal washouts, and aim to perform a primary pull-through without a preceding stoma [2]. However, some infants fail to decompress adequately with this management and require a stoma to achieve satisfactory colonic decompression, and some are considered to be unsuitable for rectal washouts from the outset.

There are little published data describing factors that influence surgeons' decision-making in choosing between rectal washouts or an initial stoma; which infants are most likely to need a stoma; the proportion of infants who will ultimately require a stoma prior to definitive surgery; or the resource implications for primary and secondary care of either management strategy. The majority of studies examining the early management of HD are retrospective case series [3,4], voluntary reporting surveys [1,5,6], surveys of intended practice [2,7,8], or meta-analyses and systematic reviews of retrospective case series [9–11]. To date there are very few prospective, population-based observational studies of HD anywhere in the world [1,5], and none that provide representative data in a cohort of children born during a short time period.

The British Association of Paediatric Surgeons Congenital Anomalies Surveillance System (BAPS-CASS) identified a national cohort of infants diagnosed with HD over a 2-year period from October 2010 [12]. Using this cohort, we aimed to describe the proportion of infants managed with a stoma prior to definitive surgery, and factors associated with an increased likelihood of stoma formation.

1. Methods

We identified a national cohort of live-born infants diagnosed with HD up to 6 months of age from 1st October 2010 to 30th September

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2012. The BAPS-CASS methodology has already been described [12,13]. Briefly, nominated reporting clinicians in all 28 pediatric surgical units in the UK and Ireland identified infants diagnosed with HD, and a data collection form was completed including demographic and clinical data, early management prior to definitive surgery and site of pathological transition zone (TZ) in infants who had undergone definitive surgery. Up to five reminders were sent if the data collection form was not returned.

All data were double-entered into a centrally held, customized database. Duplicate reports were eliminated by comparing hospital of birth, gestation at birth and date of notification and follow-up with the reporting clinicians.

1.1. Statistical analyses

Descriptive statistics were used to describe the basic demographics of the cohort, the early management prior to definitive surgery and the proportion of infants undergoing stoma formation. To investigate the factors associated with stoma formation before definitive surgery, we used univariable logistic regression analysis to examine the association of 25 independent variables with stoma formation (identified from existing literature or clinical hypothesis). The association of pathological TZ and stoma formation was examined initially by comparing infants with TZs proximal or distal to the splenic flexure of the colon. We used this categorization, as it was recognized that definitive surgery in this "TZ proximal to splenic flexure" group would be complicated by the need to take down the middle colic vessels during the colonic mobilization and we were keen to evaluate the impact of this requirement on surgical decision-making. Thirteen variables found to be associated with 'stoma performed before surgery' at a p-value of ≤0.01 were evaluated for inclusion in a multivariable model using stepwise forward regression. Likelihood ratio test (LR-test) was used to assess the incremental fit of the model with each added variable. Variables 'associated anomalies' and 'cardiac anomaly' were not included in the model because of their co-linearity with the 'syndromic HD' variable ($r > 0.99$, $p < 0.001$), however, separate sensitivity analyses were conducted to test their association with the outcome. Similarly, birth weight and gestational age were highly collinear ($r = 0.70$, $p < 0.001$), therefore, birth weight was tested in a separate model. Five variables (pathological TZ-site distal to splenic flexure versus proximal to splenic flexure, washout performed at home, syndromic HD, days between presentation to diagnosis, and contrast enema performed) that significantly affected the fit of the model were retained. In addition, we tested the association between 'pathological TZ-site recto-sigmoid versus long-segment or total colonic TZ site' and 'stoma performed before surgery' by replacing the 'pathological TZ-site distal to splenic flexure versus proximal to splenic flexure variable" with this variable in a separate model.

The continuous variables (gestational age, birth-weight, age at presentation, age at diagnosis, days between presentation and diagnosis and center case volume) were tested for their deviations from linearity, by fitting functional polynomials in the univariable regression analyses with multiple transformations of the continuous variables [14]. All variables demonstrated a linear relationship with the outcome; however, the variables were included categorically in the final model for ease of presentation, with the exception of gestational age, which was used as a continuous variable. The relationship between center volume and the likelihood of stoma formation was investigated by dichotomizing infants into two groups, based on whether they were managed at a center performing more or less than the median number of cases per center per year of 8.5.

In the cohort there were 111 children in whom a stoma was performed before surgery and 194 without stoma. With this sample size, our analysis had 80% power to detect an odds ratio of 6.8 or greater associated with a stoma being performed before definitive surgery at $p < 0.05$ (two-tailed) for the risk factor that had the highest prevalence among children without a stoma (93.8% for abdominal distension at presentation) and an odds ratio of 8.7 or greater for perforation (0.5%) that had the lowest prevalence.

The proportions of children with missing information for the included variables were low, except for passage of first meconium (12%), and both pathology TZ-site variables (6% each). We assumed the data to be not missing at random and included a proxy variable for the missing group. We also conducted a series of sensitivity analysis by redistributing the missing observations into the different categories of the variables. This did not alter the findings. All statistical analyses were performed using STATA v14.

2. Results

Between 1st October 2010 and 30th September 2012, 347 infants were reported, although 25 did not meet the case definition. Data were unavailable for a further 17 infants, leaving 305 infants with HD identified as the national cohort.

Rectal washouts were initially utilized in 263 (86%) infants, and 179 (68%) infants had this treatment at home. A stoma was formed in 39 (13%) infants without initial trial of rectal washouts, and a further 72 infants subsequently underwent stoma formation, having initially been managed with rectal washouts. In total, 111 (36%) infants received a stoma prior to definitive surgery, at a median age of 13 days (range 1–367). Table 1 lists the 114 indications that were described in 109 infants undergoing stoma formation. In 2 infants, the indication for stoma formation was not described.

Table 2 summarizes the results of a multivariate model used to investigate the factors associated with stoma formation. Compared to infants managed with rectal washouts alone; infants managed with a stoma were more likely to have a transition zone proximal to the splenic flexure, Down syndrome, another identifiable syndrome, and to have a diagnosis of HD established more than 28 days after presentation. They were less likely to have received a contrast enema or home rectal washouts. These five variables when included in the final model explained 43% of the variance in the outcome. After controlling for all other variables, the odds of a stoma being performed before surgery was almost nine-fold higher among children in whom the pathological TZ-site was 'long-segment or total colonic' compared to children with a recto-sigmoid TZ site (adjusted OR = 8.75; 95% CI = 4.21 to 18.20).

Table 3 summarizes the surgical management of all 305 infants in the cohort. At 1 year after diagnosis, definitive surgery had been performed in 270 infants (89%); 26 patients had not yet undergone definitive surgery, and 9 patients (3%) had died (none following definitive surgery). A primary pull-though (no preceding stoma) was employed in 191 (71%) infants and 79 (29%) had a staged pull-through (preceded by a stoma). The median age at definitive surgery was 67 days (range 8–322) and 159 days (range 31–383) for infants undergoing primary and staged procedures respectively. For those infants who have completed definitive surgery, the pathological transition zone (TZ) was rectosigmoid in 198 (73.3%), long segment (proximal to the sigmoid colon) in 60 (22.2%), total colonic in 8 (3.0%) and unknown in 4 (1.5%).

Table 1
Indications for stoma formation in 111 infants.

Indication	*n* (%)
Failure to decompress with rectal washouts	42 (36.8)
Emergency laparotomy	25 (21.9)
Suspected long segment disease	16 (14.0)
Enterocolitis	10 (8.8)
Consultant preference	7 (6.1)
Comorbidity	7 (6.1)
Failure to manage rectal washouts	6 (5.3)
Delayed presentation	1 (0.9)
Unknown	2 (1.8)

*114 indications were given for 109 infants. In 2 cases, the reason for stoma formation was not listed.