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ABSTRACT

Background: Hirschsprung's disease (HD) is a rare and complex malformation. The corrective operation is challenging and schedulable. The complete care situation for the corrective surgery for HD in Germany is uninvestigated.
Methods: For the years 2016–2022, the microdata of the diagnosis-related groups (DRG) -statistics provided by the Research Data Center of the German Federal Statistical Office were accessed. All hospital stays for corrective surgery of HD in patients aged 0–17 were analyzed for patient's comorbidities, treatment characteristics and hospital structures. The occurrence of severe early postoperative complications during the hospital stay were documented.
Results: The care structure for HD in Germany is decentralized with 109 hospitals performing 1199 corrective surgeries in 7 years. 75% of the participating hospitals performed three or less cases per year and 55 participating hospitals did not perform corrective surgery for HD each year. Early postoperative complications were common with at least one severe early complication in 18.6% of the cases. With an overall low case load per hospital, a volume outcome relationship cannot be established within Germany. Compared to international high volume centers the quality of outcomes for some of the investigated parameters was reduced. Despite the establishing of centers of expertise by the European reference network ERNICA for the treatment of HD no trend towards centralization occurred in Germany.
Conclusions: The corrective surgery for HD in Germany is decentralized and results in an overall high rate of early complications. The comparison with international studies from high-volume centers indicates potential for improvement for the corrective surgery of HD. Centralization remains essential for the improvement of care for patients with HD.

1. Background

Hirschsprung disease (HD) is a complex malformation with a birth rate of 1.28 (1.19–1.38) per 10,000 live births [1] diagnosed in the first year of life. Considering that only about 84–92% [2] of patients with HD are diagnosed in the newborn period, but milder cases are diagnosed later in childhood or adolescence, the total yearly HD incidence is estimated about 123 cases in Germany per year [3].

The failure of neural crest cells migration to the full length of the large intestine leaves an aganglionotic spastic segment from the anal canal orally varying in length, clinically manifesting as intestinal obstruction or Hirschsprung associated enterocolitis. HD can be associated with other congenital malformations and genetic anomalies [4].

The surgical treatment of HD aims to remove the aganglionotic segment of the intestine and anastomise the healthy intestine above the dentate line with the careful preservation of the anal canal. Treatment of HD is demanding, and many sources of errors have been reported for the diagnostics, the perioperative management, and the corrective surgery [5].

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Surgical treatment is often multi-staged, with the corrective surgery being the most challenging part. Different techniques have been described for the corrective surgery of HD, most of which are either performed transanally alone or transanally with an additional minimal invasive approach [6].

Due to the complexity of the management of HD, a minimum caseload for the corrective surgery has been promoted by the European reference network ERNICA and centralization of care is recommended [7]. Since 2017 centers of expertise regarding HD can become certified members of ERNICA in Germany. Despite the increased demand for quality standards for hospitals treating cases of HD, no mandatory quality criteria have been implemented yet.

Hospital discharge data are accurate for the coding of congenital anomalies in European health care systems [8]. Especially for rare conditions in decentralized healthcare systems, these secondary data analyses of routine practice data allow for virtually complete case numbers that are otherwise difficult to obtain.

The complete care situation for the corrective surgery of HD in Germany has not been analyzed before. The presented study aims to provide a basis for quality control and policy making on the centralization of care for HD in Germany in future.

2. Methods

2.1. Data

The microdata of the diagnosis-related groups (DRG-) statistics of the years from 2016 to 2022 were provided by the Research Data Center of the German Federal Statistical Office and were accessed by controlled remote data processing [9]. The DRG statistics encompass data records of all hospital cases billed according to the German DRG system. The data contain information on age, gender, diagnoses (ICD-10-GM), procedures (German operations and procedure code, OPS), length of stay, discharge mode, and all specialty departments that cared for the patient. Cases can be attributed to a particular institution by a pseudonymized institutional code.

2.2. Cases

Based on the coded diagnoses, the cases of HD were identified according to ICD-10. Only those cases were considered that had a corrective surgery for HD in the same hospital stay. Hence, units of analysis were inpatient cases with a principal or secondary diagnosis of HD aged up to 17 years, and a procedure code of corrective surgery. For the definition of the corrective surgery through procedure codes the specific OPS codes for corrective surgery (Duhamel, Rehbein, de la Torre) were used as well as procedure codes for general colorectal resections. Case selection flow is depicted in Fig. 1 and the exact definition of the units of analysis by diagnosis and procedure codes is shown in Table 1.

2.3. Variables

The cases were characterized by age and sex, length of stay, and mode of admission and discharge. Based on the coded diagnoses the presence of additional congenital anomalies was identified. The type of the corrective operation and the surgical approach were identified via procedure codes, as well as the placement of an enterostomy.

To describe the health care structure, we considered the annual number of treating hospitals as well as the annual hospital caseload of corrective surgeries for HD. The presence or absence of a pediatric or pediatric-surgical department was identified based on the specialty department codes of all inpatient cases treated by a hospital. If at least 10 cases with a code of a pediatric or, respectively, a pediatric-surgical department were present in a hospital in one year, we assumed the presence of the respective department in this year.

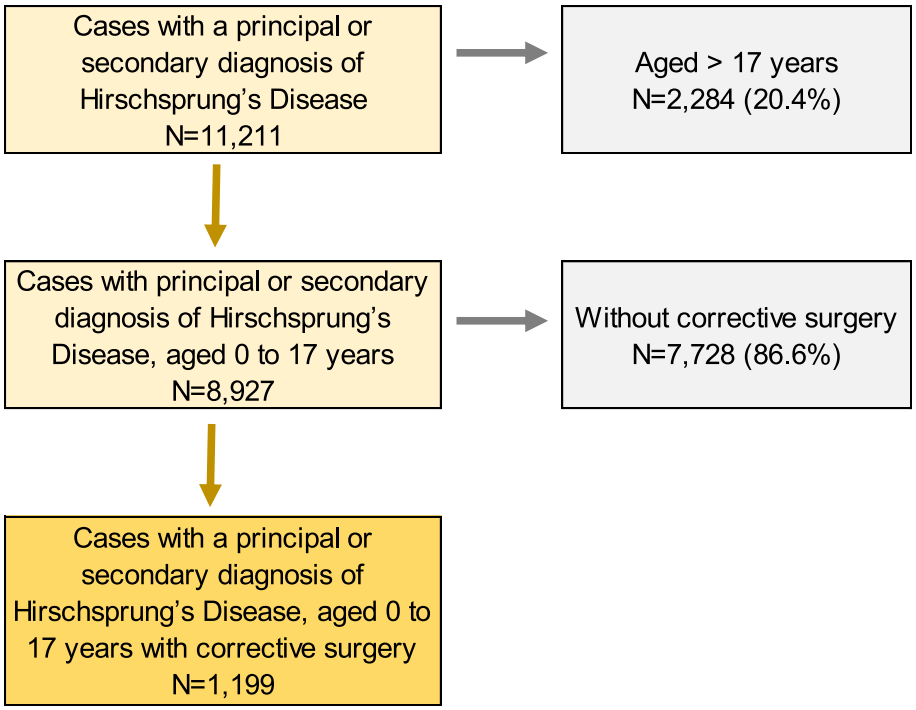

Fig. 1. Corrective surgery for Hirschsprung's Disease: Selection of cases. Note: Accumulated data from 2016 to 2022.

Table 1
Definition of units of analysis and study variables.

Units of analysis	
Cases with corrective surgery for Hirschsprung's Disease	(PD/SD Q431) AND (OPS 5484 548290 5482a 5482b0 5455 5456) AND (age ≤ 17 years)
Patient characteristics	
Additional malformation (multiple coding possible)	
Malformation of the cardiovascular system	PD/SD Q20 Q21 Q22 Q23 Q24 Q25 Q26 Q27 Q28
Malformation of the respiratory system	PD/SD Q30 Q31 Q32 Q33 Q34
Malformation of the esophagus	PD/SD Q39
Malformation of the small intestine	PD/SD Q41
Malformation of the urinary system	PD/SD Q60 Q61 Q62 Q63 Q64
Malformation of the genital system	PD/SD Q50 Q51 Q52 Q53 Q54 Q55
Malformation of the spine, limbs, or bony thorax	PD/SD Q70 Q71 Q72 Q73 Q74 Q75 Q76 Q77 Q78 Q79
Chromosomal anomalies	PD/SD Q90 Q91 Q92 Q93 Q95 Q96 Q97 Q98 Q99
At least one additional malformation	PD/SD Q20 Q21 Q22 Q23 Q24 Q25 Q26 Q27 Q28 Q30 Q31 Q32 Q33 Q34 Q39 Q41 Q50 Q51 Q52 Q53 Q54 Q55 Q60 Q61 Q62 Q63 Q64 Q70 Q71 Q72 Q73 Q74 Q75 Q76 Q77 Q78 Q79 Q90 Q91 Q92 Q93 Q95 Q96 Q97 Q98 Q99
Newborn	age ≤ 28 days
Treatment	
Corrective surgery	OPS 5484 548290 5482a 5482b0 5455 5456
Surgical approach of corrective surgery (hierarchical *)	
Open surgical (including conversion from laparoscopic approach)	(OPS 5484 6th digit 1 2 8 9) OR (OPS 5455 6th digit 1 2 3 4 7) OR (OPS 5456 6th digit 0 1 2 3 4 8)
Laparoscopic	(OPS 5484 6th digit 5 6) OR (OPS 5455 6th digit 5 6) OR (OPS 5456 6th digit 5 6 7) OR (OPS 5482a)
Transanal	(OPS 5484 6th digit 7) OR (OPS 548290) OR (OPS 5482b0)
Not specified	(OPS 5484 6th digit x) OR (OPS 5484y) OR (OPS 5455 6th digit x) OR (OPS 5455y) OR (OPS 5456 6th digit x) OR (OPS 5456y)
Duhamel, Rehbein, or La Torre procedure	OPS 54841 54840 54842 54829 5482a 5482b0
Enterostomy	OPS 5460 5461 5462 5463
Indicators of complications or complicated course	
Anastomotic leakage	SD K9183
Malfunction of stoma **, postoperative ileus	SD (K913 AND OPS 5460 5461 5462 5463) OR SD K914
Anal or rectal prolapse, or urethral stricture	SD K622 K623 N991 OR OPS 55501 55721 8137
Peritonitis or perforation of intestine	SD K630 K631 K65 P780 P781
Septicaemia or systemic inflammatory response syndrome	SD A41 R572 P36 R650 R651
Transfusion of whole blood or erythrocytes >5 units	OPS 88001 OR OPS 8800c 6th digit 1–9, a-r
Revision surgery during the same inpatient stay **	OPS 55413 55414 55450 55451 55411 OR (OPS 5464 AND OPS 5460 5461 5462 5463)
Invasive ventilation >6 h	Hours of ventilation >6 AND OPS 8701
At least one indicator of complication or complicated course	(SD K9183) OR (SD K913 AND OPS 5460 5461 5462 5463) OR (SD K914 K622 K623 N991 K630 K631 K65 P780 P781 A41 R572 P36 R650 R651) OR (OPS 88001) OR (OPS 8800c 6 ht digit 1–9, a-r) OR (OPS 55501 55721 8137 55413 55414 55450 55451 55411) OR (OPS 5464 AND OPS 5460 5461 5462 5463) OR (hours of ventilation >6 AND OPS 8701)
Hospital characteristics	
Pediatric department	At least 10 cases (out of all inpatient cases) per year with specialty department code 1000 1100 1200 1004 1006 1007 1009 1011 1014 1028 1051 3610 1136 1012 1050
Pediatric surgical department	At least 10 cases (out of all inpatient cases) per year with specialty department code 1300 1513

PD: Principal diagnosis. SD: Secondary diagnosis. OPS: Operationen-und Prozedurenschlüssel (German operations and procedure code). * Each treatment case was assigned to only one subgroup. Hierarchy levels correspond to the order as displayed in the table. ** Malfunction of stoma or revision of enterostomy were only considered in cases with enterostomy during the same inpatient stay.

Indicators of major early complications or a complicated course were defined through diagnosis codes (e.g., anastomosis leakage), or through procedure codes (e.g., blood transfusion). Because a single case could be affected by multiple complications, the occurrence of at least one indicator of complication was defined as the primary outcome parameter (Table 1).

3. Results

From 2016 to 2022, a total of 1199 corrective surgeries for HD were performed in 109 different German hospitals (Table 2). On average 63 (range 54–75) different hospitals annually had at least one case of corrective surgery for HD. Of those hospitals, 96.8% had a pediatric department and 80.7% had a pediatric surgical department. There was no trend towards a change of the numbers of participating hospitals during the investigated time period (Table 2).

Within the observation period on average 171 (range 157–192) corrective surgeries for HD were coded per year nationwide. The median (IQR) annual caseload per hospital was 2 [1–3] and the 95th percentile was 7. At least 55 hospitals performed less than 1 corrective surgery for HD per year (Table 2).

24.4% of patients were female. At least one additional malformation was coded in 16.4% of the cases, with chromosomal anomalies with 11.0% and cardiovascular anomalies with 6.7% being the most common ones. The median (IQR) age at the time of corrective surgery was 0 (0–1) years, with the 95th percentile being 10 years. 3.9% of patients had a corrective surgery during the newborn period (≤28 days of age).

In 40.6% of corrective surgeries, an HD specific code was used such as Duhamel, Rehbein or de La Torre procedure. In 59.4% a general code for a colorectal procedure was used. For corrective surgery in 25.3% of the cases a transanal approach alone was coded. A transanal with an additional laparoscopic approach was used in

Table 2
Corrective surgery for Hirschsprung's Disease (HD): Treating hospitals, patient characteristics, and treatment.

		2016	2017	2018	2019	2020	2021	2022	Total
Treating hospitals									
Hospitals with at least one case of corrective surgery for HD	**N (%)**	**61 (100)**	**75 (100)**	**60 (100)**	**66 (100)**	**54 (100)**	**61 (100)**	**64 (100)**	**109 (100)**
Hospitals with pediatric department per year	N (%)	58 (95.1)	72 (96.0)	57 (95.0)	63 (95.5)	53 (98.1)	61 (100)	63 (98.4)	**n.a.**
Hospitals with pediatric surgical department per year	N (%)	50 (82.0)	61 (81.3)	50 (83.3)	54 (81.8)	42 (77.8)	47 (77.0)	52 (81.3)	**n.a.**
Caseload per hospital	Median (IQR)	2 (1–3)	2 (1–3)	2 (1–3)	2 (1–3)	2 (1–3)	2 (1–3)	2 (1–3)	**2 (1–3)**
	P95	6	7	8	8	7	7	6	**7**
		2016	**2017**	**2018**	**2019**	**2020**	**2021**	**2022**	**Total**
Patient characteristics									
Cases with a diagnosis of HD and corrective surgery	**N (%)**	**169 (100)**	**192 (100)**	**167 (100)**	**182 (100)**	**157 (100)**	**164 (100)**	**168 (100)**	**1199 (100)**
Female	N (%)	50 (29.6)	38 (19.8)	35 (21)	44 (24.2)	29 (18.5)	46 (28)	51 (30.4)	**293 (24.4)**
Newborns (age ≤ 28 days)	N (%)	7 (4.1)	10 (5.2)	6 (3.6)	11 (6)	3 (1.9)	10 (6.1)	0 (0)	**47 (3.9)**
Age (years)	Median (IQR)	0 (0–2)	0 (0–1)	0 (0–2)	0 (0–1)	0 (0–2)	0 (0–1)	0 (0–2)	**0 (0–1)**
	P95	10	9	11	8	10	8	13	**10**
Additional malformation (multiple coding possible)									
Malformation of the cardiovascular system	N (%)	*	*	*	*	*	*	*	**80 (6.7)**
Malformation of the respiratory system	N (%)	*	*	*	*	*	*	*	**6 (0.5)**
Malformation of the esophagus	N (%)	*	*	*	*	*	*	*	**0 (0.0)**
Malformation of the small intestine	N (%)	*	*	*	*	*	*	*	**10 (0.8)**
Malformation of the urinary system	N (%)	*	*	*	*	*	*	*	**18 (1.5)**
Malformation of the genital system	N (%)	*	*	*	*	*	*	*	**16 (1.3)**
Malformation of the spine, limbs, or bony thorax	N (%)	*	*	*	*	*	*	*	**7 (0.6)**
Chromosomal anomalies	N (%)	*	*	*	*	*	*	*	**132 (11.0)**
At least one additional malformation	N (%)	31 (18.3)	32 (16.7)	19 (11.4)	36 (19.8)	21 (13.4)	25 (15.2)	33 (19.6)	**197 (16.4)**
		2016	2017	2018	2019	2020	2021	2022	Total
Treatment									
Cases with a diagnosis of HD and corrective surgery	**N (%)**	**169 (100)**	**192 (100)**	**167 (100)**	**182 (100)**	**157 (100)**	**164 (100)**	**168 (100)**	**1199 (100)**
Type of corrective surgery									
Duhamel, Rehbein, or La Torre procedure	N (%)	42 (24.9)	38 (19.8)	33 (19.8)	100 (54.9)	94 (59.9)	88 (53.7)	92 (54.8)	**487 (40.6)**
Other procedure	N (%)	127 (75.1)	154 (80.2)	134 (80.2)	82 (45.1)	63 (40.1)	76 (46.3)	76 (45.2)	**712 (59.4)**
Surgical approach of corrective surgery									
Transanal	N (%)	25 (14.8)	23 (12.0)	17 (10.2)	70 (38.5)	64 (40.8)	54 (32.9)	50 (29.8)	**303 (25.3)**
Laparoscopic	N (%)	25 (14.8)	22 (11.5)	28 (16.8)	20 (11.0)	19 (12.1)	29 (17.7)	39 (23.2)	**182 (15.2)**
Open surgical (including conversion from laparoscopic)	N (%)	72 (42.6)	87 (45.3)	65 (38.9)	72 (39.6)	62 (39.5)	59 (36.0)	65 (38.7)	**482 (40.2)**
Not specified	N (%)	47 (27.8)	60 (31.3)	57 (34.1)	20 (11.0)	12 (7.6)	22 (13.4)	14 (8.3)	**232 (19.3)**
Enterostomy	N (%)	17 (10.1)	19 (9.9)	17 (10.2)	18 (9.9)	16 (10.2)	22 (13.4)	18 (10.7)	**127 (10.6)**
Transfusion of whole blood or erythrocytes ≥ 1 unit	N (%)	35 (20.7)	47 (24.5)	34 (20.4)	32 (17.6)	38 (24.2)	40 (24.4)	48 (28.6)	**274 (22.9)**
Length of stay (days)	Median (IQR)	10 (8–15)	11 (8–15)	10 (8–15)	10 (8–15)	10 (7–15)	9 (7–15)	9 (7–12)	**10 (7–15)**
Indicators of complications or complicated course (multiple coding possible)									
Anastomotic leakage	N (%)	6 (3.6)	6 (3.1)	7 (4.2)	7 (3.8)	10 (6.4)	*	7 (4.2)	**43 (3.6)**
Malfunction of stoma[1] or postoperative ileus	N (%)	*	*	7 (4.2)	6 (3.3)	5 (3.2)	5 (3.0)	*	**35 (2.9)**
Anal or rectal prolapse, or urethral stricture	N (%)	4 (2.4)	5 (2.6)	*	*	*	4 (2.4)	6 (3.6)	**29 (2.4)**
Peritonitis or perforation of intestine	N (%)	15 (8.9)	17 (8.9)	14 (8.4)	8 (4.4)	10 (6.4)	5 (3.0)	10 (6.0)	**79 (6.6)**
Septicaemia or systemic inflammatory response syndrome	N (%)	12 (7.1)	12 (6.3)	*	10 (5.5)	4 (2.5)	6 (3.7)	9 (5.4)	**56 (4.7)**
Transfusion of whole blood or erythrocytes >5 units	N (%)	*	*	*	*	*	*	*	**11 (0.9)**
Revision surgery[1] during the same inpatient stay	N (%)	7 (4.1)	10 (5.2)	7 (4.2)	13 (7.1)	13 (8.3)	*	5 (3.0)	**56 (4.7)**
Invasive ventilation >6 h	N (%)	7 (4.1)	18 (9.4)	5 (3.0)	14 (7.7)	18 (11.5)	12 (7.3)	12 (7.1)	**86 (7.2)**
At least one indicator of complication or complicated course	N (%)	31 (18.3)	37 (19.3)	24 (14.4)	36 (19.8)	37 (23.6)	25 (15.2)	33 (19.6)	**223 (18.6)**

Notes: * not displayed due to small cell count. [1] Malfunction of stoma or revision of enterostomy were only considered in cases with enterostomy during the same inpatient stay. IQR: interquartile range. P95: 95th percentile. n.a.: Not available.

15.2% of cases. An either primary laparotomy or a laparotomy as conversion was used in 40.2% of cases. In 19.3% of cases an unspecified approach was used, that can be either transanal alone, laparoscopic or laparotomic.

An additional enterostomy during the hospital stay for corrective surgery was placed in 10.6% of cases (Table 2).

At least one indicator for a severe complication or complicative course was observed in 18.6% of cases, with the need for prolonged postoperative invasive ventilation, and peritonitis or intestinal perforation being the most common ones (7.2% and 6.6% respectively). An anastomotic leakage was documented in 3.6% of the cases. 4.7% of patients underwent a surgical revision in the same hospital stay. A prolapse of the anus or rectum, or an injury to the urethra occurred in 2.4% of cases. About 1% of cases received more than five blood transfusions.

In addition to these indicators for a complicative course, in 22.2% of cases 1–5 blood transfusions were coded during the hospital stay for corrective surgery for HD.

The median (IQR) length of stay was 10 days (7–15 days) (Table 2).

4. Discussion

This study represents a complete data analysis of all 1199 cases of corrective surgery for HD in Germany from 2016 to 2022 based on hospital discharge data. It reveals an extremely decentralized provision of care with 75% of the 109 hospitals participating in HD corrective surgery performing three or less cases per year and 55 participating hospitals not performing corrective surgery for HD each year. Severe early postoperative complications were common and no trend towards centralization could be demonstrated during the investigated time period.

This study supports the usability of routine practice data for the investigation of the care situation of HD, due to the plausibility of the results. The occurrence of associated anomalies (16.4%), as well as the male-to-female ratio of 3:1 is in accordance with the literature [10]. Furthermore, the incidence of corrective surgeries is consistent with the expected number of cases.

The difference between the average cases of corrective surgeries for HD per year of 171 in the present study compared to the expected incidence of 123 cases can be explained by either patients born in other countries, who came to Germany for the corrective surgery, or, probably to a larger extend, by patients with HD receiving a re-do of corrective surgery during a different hospital stay. Furthermore, the applicability of the EUROCAT registry to the German population in light of overall very low incidences might be limited [1,2].

The presented results show an extremely decentralized care situation for corrective surgery in HD. The number of corrective surgeries as well as the number of hospitals performing corrective surgeries per year remained stable throughout the investigated time period. The certification process by the European reference Network ERNICA in 2017, in which 4 hospitals in Germany were certified for the treatment of HD [11], had no impact on centralization of care on a national level. Despite aspired centralization by the ERN there is no restriction for non-certified hospitals with very low case numbers to perform corrective surgery for HD. The high percentage of nearly 20% of hospitals performing corrective surgery for HD without having a pediatric surgical unit adds to the controversy of how expert treatment can be achieved in this setting. As individual hospitals cannot be identified by the pseudonymized institutional code provided in this data, the development of caseloads in Germany's certified ERNICA centers cannot be analyzed. Similarly this study cannot evaluate the outcomes for individual hospitals or individual surgeons.

Establishing a volume–outcome relationship is impossible when caseloads are below ten cases per year, as with these very low case numbers a difference in quality cannot be detected [12]. Consequently, previous studies with extremely low overall operative volumes failed to find a volume–outcome relationship for the treatment of HD [13]. With the scattered distribution of cases of corrective surgery for HD in Germany, the evaluation of a volume–outcome relationship is not feasible. Any clinical research on the perioperative outcome of corrective surgery for HD fall under the same requirement for minimal case numbers and are equally affected by the decentralized care situation [14].

Only few studies from hospitals with a caseload of more than 20 corrective surgery for HD per year exist. Compared to published data from these centers there are some striking findings of this work that point towards a negative effect of the decentralized care structure on early postoperative outcomes, and a possible volume-outcome-relationship for some of the investigated parameters.

In at least 40.2% of corrective surgeries for HD in Germany a laparotomy was used as surgical approach. As in 19.3% the surgical approach was not specified, the rate of laparotomies may even be higher. Since the 1970s methods have been described to avoid a laparotomy by either a transanal approach alone or a transanal approach assisted by a minimal invasive approach [15]. While laparotomy might be necessary in some cases, the rate of laparotomies performed in Germany is comparably high. In the only study from a high-volume center published in the literature with a case volume of more than 20 corrective surgeries per year, out of 75 minimally invasive assisted surgeries, no conversion to laparotomy occurred and no primary laparotomy was necessary in primary HD corrective surgeries during the investigated time period of 1.5 years [16].

The superiority of laparoscopically assisted corrective surgery for HD versus laparotomy has been clearly shown for the outcomes length of hospital stay, estimated blood loss, and number of complications in a large meta-analysis [17]. A trend towards increased use of laparoscopy versus laparotomy has been noted in centralized care situations [6].

The lack of an increase in the use of minimal-invasive techniques during the investigated time period in Germany does not reflect this tendency, and decentralization might be the decisive factor. The significant discrepancy between the length of stay during corrective surgery for HD in Germany with a mean 10 days as compared to a mean of 7.5 days in a high-volume center [16], may be a consequence of the higher rate of laparotomies.

In 10.6% of cases an enterostomy was placed during the hospital stay for corrective surgery of HD. This does not include cases in which an enterostomy had been placed during a previous hospital stay. Placement of an enterostomy during the hospital stay for corrective surgery are either performed simultaneously during the corrective surgery for HD or after corrective surgery in case of the occurrence of a complication. The necessity of the placement of an enterostomy during corrective surgery can be reduced if patient preparation for corrective surgery through sufficient transanal irrigation, or an enterostomy placed prior to the hospital stay for corrective surgery for treatment of the intestinal obstruction is performed [18]. The rate of enterostomy placement during the hospital stay for corrective surgery is high compared to the data from a high-volume center with 4% enterostomy placement [16].

The incidence and risk factors for a perioperative blood transfusion in infants undergoing corrective surgery for HD has been explored by Reppucci et al. and were shown to be neglectable with 0 out of 40 cases of corrective procedures for HD needing blood products [19]. Intraoperative blood loss was shown to be minimal (3 ml–5.1 ml on average) in the collective by Zhang et al. [16]. The German procedure code only differentiates a code for 1–5 blood

units, and a code for more than 5 units. Since anemia in children with HD is common and a blood transfusion during the hospital stay might not necessarily correlate with a bleeding complication, we only included those cases in the index for a complication or complicated course, that received more than 5 units of blood during the stay for corrective surgery for HD. However, the reasons for 22.2% of children needing 1–5 units of blood during the hospital stay for corrective surgery for HD should be explored further and should be taken into account for perioperative management. 1% of children needing 6 or more units of blood during the hospital stay for elective surgery for HD points toward a serious bleeding complication and has not been described as a typical complication for corrective surgery in HD in the literature before.

Rectal prolapse, anal prolapse or urethral stricture were summarized in the analysis, as they have the commonality of being avoidable complications if preventive measures are taken [5]. The circumferential prolapse of the neorectum or anal canal in the immediate postoperative course results from the damage of relevant sphincteric structure. If the anal canal is intact after corrective surgery, it is highly unlikely that the pulled through colon, which is under tolerable tension, should prolapse. In a similar way, urethral stricture in patients with HD corrective surgery should not occur if the urethra remains undamaged during the operative procedure. The careful preservation of the anal canal and the sphincteric structures, as well as avoiding damage to the urethra are of utmost importance for the long-term outcome of patients with HD and we consider the frequency of 2.4% of these avoidable complications as high.

Further coded complications were peritonitis or perforation of the intestine in 6.6% of cases and anastomotic leakage in 3.6% of cases. In 4.7% of cases a surgical revision was coded during the same hospital stay. The rate of these complications differs widely in the literature. In a high-volume center, anastomotic leakage occurred in 2.4% of cases and no surgical revision was necessary during the same hospital stay [16].

3.9% of corrective surgeries for HD were performed in the neonatal period ($\leq$28 days of age). General risks of surgery and anesthesia during this vulnerable period of life should be carefully weighed against the perceived advantage of early correction [20].

For the successful management of HD, the correct diagnosis of the extent of the malformation as well as the correct time of surgery and the follow-up care were shown to be essential [21]. These aspects of the patient journey cannot be evaluated using the DRG statistics' hospital discharge data, as different hospital cases cannot be linked to the same patient. For this very reason long-term outcomes such as anastomotic stricture, incontinence, Hirschsprung associated enterocolitis, or the assessment of quality of life could not be evaluated in this study. Furthermore, only those outcomes that can be documented with a diagnosis code or procedure code could be evaluated.

5. Conclusions

The surgical care structure for HD in Germany is extremely decentralized. Major early complications after corrective surgeries are common. Evidence of a volume–outcome relationship cannot be investigated within Germany because of the overall low case volume per hospital.

The comparison of the analyzed data from Germany to international studies from high-volume centers indicates potential for improvement for the corrective surgery of HD, with regard to the outcome parameters length of hospital stay, need for blood transfusions, need for revision and need for placement of an enterostomy. The high rate of laparotomies for corrective surgery and no increase in the use of minimal invasive techniques during the investigated time period point towards a damaging effect of decentralized care on the development of the specialty of pediatric surgery. The certification process of ERNICA had no influence on the number of hospitals performing these complex operations. Meaningful steps towards centralization should be undertaken to improve surgical outcomes of HD corrective surgery in Germany.

Conflicts of interest

Prof R. Busse is a member of the German government commission for modern and needs-based hospital care (Regierungskommission für eine moderne und bedarfsgerechte Krankenhausversorgung).

The other authors have no conflict of interest.

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