Heterotaxy Syndrome Infants Are at Risk for Early Shunt Failure After Ladd Procedure

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Background. **Cardiac-specific risks and complications after a Ladd procedure in patients with heterotaxy syndrome (HS) and intestinal rotational anomalies (IRA) are unknown. We sought to (1) describe rates of hospital mortality and early systemic-to-pulmonary (S-P) artery shunt failure after the Ladd procedure in patients with HS and (2) explore risk factors associated with early shunt failure in patients with HS with single ventricle (SV).**

Methods. **This retrospective study included all Ladd procedures performed from January 1999 to December 2012 in patients with HS at a single center. Risk factors investigated for early S-P artery shunt failure included birth weight, gestational age, sex, age at and timing of Ladd procedure relative to cardiac operations, and shunt type.**

Results. **Ladd procedure was performed on 54 infants with HS and congenital heart disease. Hospital mortality for the entire cohort was 5.6% (3 of 54 patients). Early shunt failure occurred in 19% (4 of 21) of HS infants with SV. Mean preoperative blood urea nitrogen (BUN) levels were higher in HS infants with early shunt failure (20 versus 12.5 mg/dL; $p = 0.054$).**

Conclusions. **Patients with SV and HS with S-P artery shunts are at risk for early shunt failure after a Ladd procedure. A higher mean preoperative BUN level is noted in patients with HS and early shunt failure. Careful risk-benefit analysis is indicated before recommending routine elective Ladd procedures in patients with HS.**

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Failure of embryonic lateralization and left-right asymmetry results in an abnormal arrangement of thoracic and abdominal viscera, referred to as heterotaxy syndrome (HS) [1]. Forty percent to 90% of patients with HS have intestinal rotational anomalies (IRAs) [2, 3]. Patients with IRAs are at risk for proximal small bowel obstruction, midgut volvulus, and bowel necrosis [2–5].

Ladd procedure is widely accepted as the treatment for symptomatic IRA [6]. It includes detorsion of the bowel when volvulus is present, division of congenital fibrous bands, broadening the mesentery of the small bowel to potentially reduce future risk of bowel torsion, and placement of small and large bowel in a nonrotated state.

Prophylactic Ladd procedure has been advocated by some centers for patients with HS with IRA in an attempt to decrease the potential risk of midgut volvulus [7, 8]. Early studies on patients with HS with IRA reported low mortality and morbidity risk after elective Ladd procedures [8–10]. Therefore many centers, including ours, perform an elective Ladd procedure on patients with HS with IRA even if they are asymptomatic [7, 11]. A more recent study reports a higher complication rate after the Ladd procedure and cautions against this practice. This study, however, predominantly focused on gastrointestinal (GI) complications, particularly small bowel obstruction [11].

Unlike non-HS patients with IRA, HS patients frequently have complex congenital heart disease [1]. A palliative cardiac surgical procedure often precedes the Ladd procedure. Patients with HS, particularly those with systemic-to-pulmonary (S-P) artery shunts and single ventricle (SV) physiology have a tenuous circulation, which may be perturbed by additional noncardiac operations.

This single-center retrospective study was therefore conducted with the following objectives:

1. Describe rates of hospital mortality and early S-P artery shunt failure after a Ladd procedure in patients with HS and SV.
2. Explore risk factors associated with early shunt failure.

Patients and Methods

Study Design, Site, and Participants

This retrospective study was conducted at Columbia University Medical Center, a tertiary care academic center in New York, New York, on infants with HS and congenital heart disease who underwent a Ladd procedure between January 1, 1999 and December 31, 2012. Study participants were primarily admitted to our neonatal intensive care unit.

It is our center's practice to screen all patients with HS for IRAs and to perform an elective Ladd procedure on

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those who screen positive, usually during the same hospitalization or rarely after discharge and when clinically stable. Patients who experience signs of intestinal obstruction or volvulus undergo an emergent Ladd procedure.

Patients with HS who underwent a Ladd procedure were identified by cross-referencing administrative databases from the Divisions of Neonatology and Pediatric Surgery. Included participants were diagnosed with HS and congenital heart disease, underwent screening for anomaly of rotation by an upper gastrointestinal (UGI) contrast study with positive results, and underwent a Ladd procedure by pediatric surgeons at our institution. Excluded patients were infants with HS without congenital heart disease and those who did not receive a Ladd procedure at our institution. Also excluded were patients in whom IRAs were associated with other GI anomalies, eg, omphalocele. The Institutional Review Board of Columbia University Medical Center approved this study with a waiver of informed consent.

Definitions

For the purpose of this study, congenital heart disease was defined as an anatomic abnormality of the heart present from birth. Isolated vascular abnormalities that did not require surgical intervention, eg, interrupted inferior vena cava were not considered congenital heart disease. Lesions with SV anatomy and following a univentricular pathway were defined as SV.

IRAs were identified by an UGI contrast study. A single pediatric radiologist (BL) reviewed all UGI studies to confirm the presence of IRAs. S-P artery shunt failure was defined as an urgent need for a transcatheter or surgical revision of an S-P artery shunt because of severe hypoxemia.

Operative Management

At our institution, S-P artery shunts are placed either by lateral thoracotomy or median sternotomy, based on anatomy and surgeon preference. A Ladd procedure is usually performed by open laparotomy, and standard techniques are used. General pediatric anesthesiologists, and rarely cardiac anesthesiologists, provide anesthetic management during Ladd procedures.

Perioperative Management

At our center, patients receiving S-P artery shunts are given intravenous maintenance fluids (100 mL/kg) with dextrose and standard electrolytes in the perioperative period. Intraoperative fluid management includes continuation of maintenance fluids or the addition of normal saline or Ringer's lactate, or both, as determined necessary by the anesthesia team.

Our current practice is to initiate oral acetylsalicylic acid (ASA) on postoperative day 1 after shunt placement at a dose of 20 mg/d. Earlier in the study period, ASA at the same dose was given only after full enteral feeding was established. Subcutaneous enoxaparin is provided only if enteral feedings are contraindicated, eg, in necrotizing enterocolitis or if anticoagulation therapy is indicated for a concomitant morbidity, eg, venous or arterial thrombosis. Anticoagulation strategy is often but not uniformly modified after noncardiac operations based on surgeon preference: ASA is replaced with subcutaneous enoxaparin on the day of the surgical procedure and continued until enteral feeding is reinitiated when ASA is reintroduced.

Outcomes

The primary end point was hospital mortality or S-P artery shunt failure requiring an urgent surgical or transcatheter intervention within 72 hours of the Ladd procedure, or both.

Study Procedures

Hospital medical records and operative, radiologic, and echocardiographic reports were reviewed for patient demographics, hospital mortality, description of cardiac lesions and operations, laboratory values, medication use, and postsurgical complications after the Ladd procedure.

Statistical Analysis

Descriptive statistics were performed to explore the distribution of variables of interest, such as birth weight, gestational age, age at Ladd procedure, time from first cardiac operation to Ladd procedure, sex, race, preoperative diuretic and anticoagulant use, perioperative blood urea nitrogen (BUN) and serum creatinine levels, S-P artery shunt size and type, and whether a Ladd procedure was performed emergently or electively.

Tabular analysis was performed to evaluate the relationship between variables of interest and infants with the outcome of early shunt failure postoperatively and those without. Wilcoxon rank-sum tests were used for continuous variables and Fisher's exact tests were used for categorical variables, as appropriate. Analyses were conducted using SAS, version 9.3 (SAS Institute, Cary, NC).

Results

Eighty-six infants were admitted to our institution with a diagnosis of HS during the study period (Fig 1). Excluded infants (n = 32) were those in whom congenital heart disease was not identified (n = 5), a UGI screening was not performed (n = 10), an IRA was ruled out (n = 8), or a Ladd procedure was not performed at our institution (n = 9). Fifty-four infants with HS and congenital heart disease underwent a Ladd procedure for IRA at our center from January 1, 1999 to December 31, 2012 and were included in the final analysis.

Sixty-one percent (33 of 54) of participants in this series were boys. The median age at Ladd procedure was 21 days (interquartile range [IQR], 15–57 days). Cardiac lesions and operative characteristics of the entire cohort are shown in Table 1. The most common cardiac defect was a right-sided obstructive lesion (35%), followed by partial or total anomalous pulmonary venous return (28%). Forty-five (83%) infants in this cohort had a cardiac surgical intervention before the Ladd procedure.

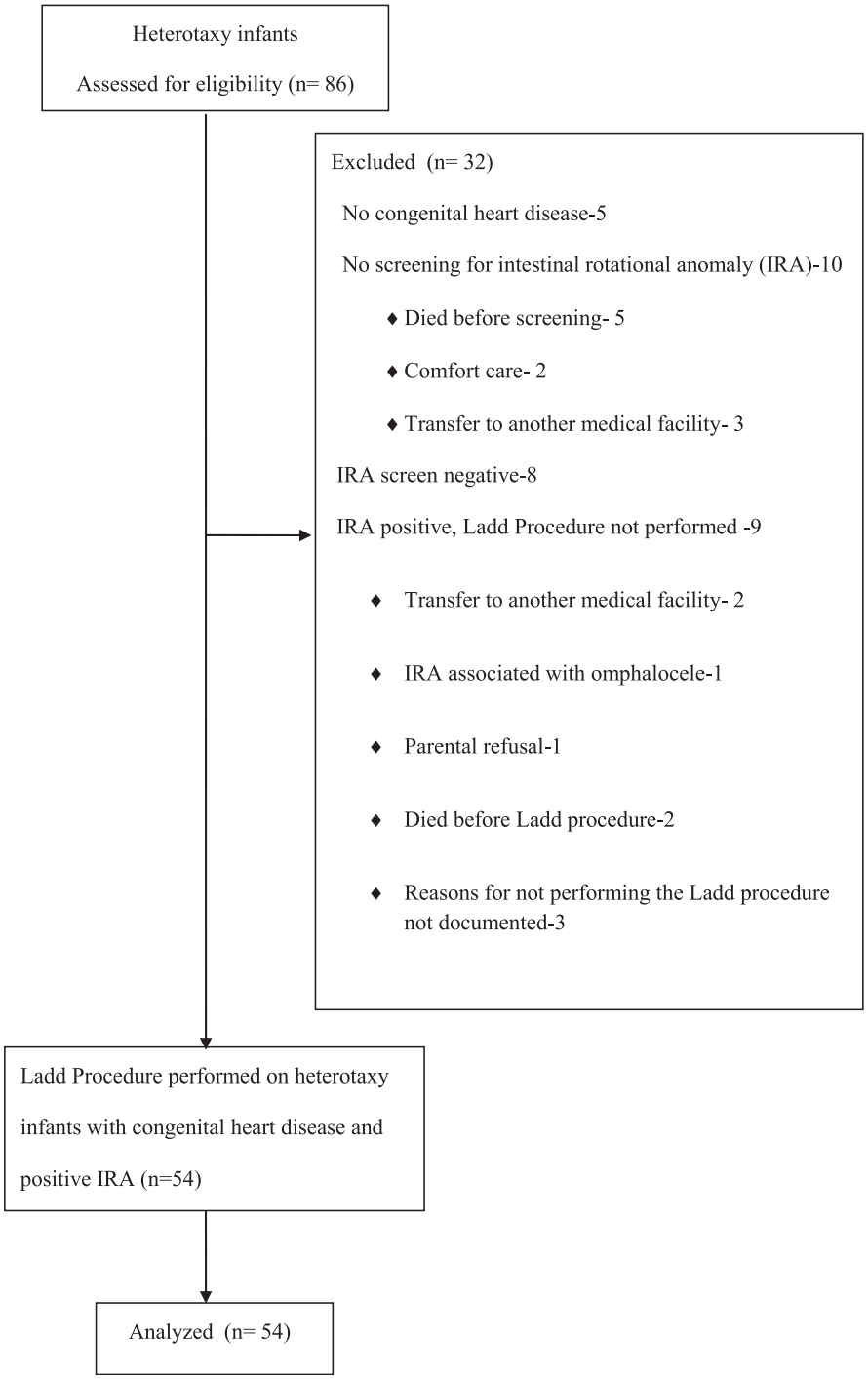

Figure 1. Flow diagram of heterotaxy syndrome (HS) patient population during the study period.

Twenty-six infants (48%) had S-P artery shunt placement, 21 of which followed an SV palliation pathway.

Three of 54 (5.5%) patients had an emergent Ladd procedure performed for symptoms concerning for volvulus. One of the 3 infants had confirmed volvulus and underwent partial bowel resection. Two patients manifested symptoms within the first week of life, including the patient with volvulus; the third patient was nearly 4 years old when symptoms appeared. The remaining 51 infants underwent a Ladd procedure electively.

Three infants (5.6%) in this cohort died during the initial hospitalization (Table 2). Two infants died of necrotizing enterocolitis, fulminant sepsis, and multi-organ failure. One infant sustained cardiopulmonary arrest after acute profound hypoxemia during transport

Table 1. Cardiac Lesions and Operations Before Ladd Procedure

Primary Cardiac Defect Class (No.) n = 54	Specific Cardiac Lesion (No.)	Most Recent Cardiac Surgical Intervention Before Ladd Procedure (No.) n = 45
AVC and septal defects (8)	Common AVC (4)	Pulmonary artery banding (1)
		No cardiac operation before Ladd procedure (3)
	ASD (1)	ASD closure (1)
	VSD and ASD (3)	Pulmonary artery banding (1)
		No cardiac operation before Ladd procedure (2)
Right ventricular outflow tract obstruction (19)	Pulmonary valve stenosis (7)	S-P artery shunt (6)
		No cardiac operation before Ladd procedure (1)
	Pulmonary atresia (12)	S-P artery shunt (10)
		Glenn procedure (1)
		Fontan procedure (1)
Left ventricular outflow tract obstruction (11)	Hypoplastic aortic arch (2)	Aortic arch reconstruction (2)
	Common AVC/coarctation of aorta (2)	Coarctation repair/pulmonary artery banding (2)
	Subaortic stenosis (1)	Ross-Konno operation (1)
	Hypoplastic left heart syndrome (4)	Norwood-Sano procedure (2)
		Norwood- BT shunt (2)
	DORV,TGA, hypoplastic aortic arch (1)	Arterial switch procedure, aortic arch reconstruction, VSD closure (1)
	Coarctation of aorta and pulmonary valve stenosis (1)	No cardiac operation before Ladd procedure (1)
Anomalous pulmonary venous drainage (3)	TAPVR (2)	TAPVR repair (2)
	PAPVR (1)	PAPVR repair (1)
Anomalous pulmonary venous drainage with right ventricular outflow tract obstruction (12)	TAPVR + PS (5)	TAPVR repair + S-P artery shunt (1)
		S-P artery shunt (1)
		TAPVR repair (1)
		Glenn procedure (1)
		No cardiac operation before Ladd procedure (1)
	TAPVR + PA (7)	TAPVR repair + S-P artery shunt (6)
		Glenn procedure (1)
Other (1)	L-TGA, Ebstein's anomaly (1)	No cardiac operation before Ladd procedure (1)

ASD = atrial septal defect; AVC = atrioventricular canal; B-T = Blalock-Taussig; DORV= double-outlet right ventricle; L-TGA = levo-transposition of the great arteries; PA = pulmonary atresia; PAPVR = partial anomalous pulmonary venous return; PS = pulmonary stenosis; S-P = systemic to pulmonary; TAPVR = total anomalous pulmonary venous return; TGA = transposition of the great arteries; VSD = ventricular septal defect.

to the intensive care unit after an elective Ladd procedure. Death in this patient was presumed to be secondary to S-P artery occlusion, although an autopsy was never performed to confirm. Three additional infants with HS and SV underwent surgical revision of S-P artery shunts for shunt occlusion. Thus early S-P artery shunt failure occurred after a Ladd procedure in 4 of 21 infants with HS (19%) with SV physiology. In all 4 patients, a Ladd procedure was performed electively.

Subgroup analysis of the 21 patients with SV and an S-P artery shunt is shown in Table 3. In this sample, infants who had early shunt failure had a higher preoperative BUN level (20 versus 12.5 mg/dL; $p = 0.054$; odds ratio, 1.2, 95% confidence interval, 0.99–1.48). Neither postoperative BUN nor preoperative or postoperative creatinine levels were significantly different between those with and without this outcome.

Four of 54 infants (7%) in our series experienced small bowel obstruction requiring operation at a median of 126 days after the Ladd procedure (range, 30–503 days).

Comment

To our knowledge, this is the largest study to date to investigate the risk of early S-P artery shunt failure after Ladd procedures in children with HS.

We evaluated the outcomes of hospital mortality and early S-P artery failure after the Ladd procedure in patients with HS. The mortality rate after an elective Ladd procedure in patients with HS is variable (0%–22%), with higher rates noted in patients with HS and functional SVs [7–12]. The 5.6% mortality rate in our series is within the range reported previously; however, our study includes both elective and emergent Ladd procedures.

Table 2. Case Summaries of Patients With Heterotaxy Syndrome and Primary Outcome of Hospital Mortality or Early Shunt Failure, or Both

Patient No.	Cardiac Lesion	Cardiac Operation	IRA and Ladd Procedure Summary	Complications	Hospital Mortality
1	RAI, CAVC, DORV, TGA, PS	3.5-mm right modified B-T shunt on DOL 4	Elective Ladd procedure 12 d after cardiac operation	Cardiopulmonary arrest 72 h after Ladd procedure; BT shunt occlusion was diagnosed by echocardiography and ductus arteriosus was reopened with PGE-1; a 3.5-mm central shunt was placed 9 d later; thrombus was not noted in original shunt	No
2	RAI, CAVC, DORV, TGA, PA, TAPVR	TAPVR repair, 3.5-mm central shunt on DOL 2	Elective Ladd procedure 10 d after cardiac operation	Cardiopulmonary arrest on transport from operating room after Ladd procedure	Yes
3	LAI, CAVC, small left ventricle, juxtaductal coarctation, PS	Listed for cardiac transplantation	Ladd procedure at 5 d of age for bilious aspirates; no volvulus was identified	Necrotizing enterocolitis and multiorgan failure secondary to *Citrobacter koseri* sepsis 14 d after Ladd procedure	Yes
4	RAI, CAVC, DORV, TGA, PS, TAPVR	TAPVR repair, 3. 5-mm central shunt on DOL 1	Elective Ladd procedure 27 d after cardiac operation	Shunt occlusion diagnosed on arrival to ICU; emergent 3.5-mm central shunt revision performed; thrombus identified in original shunt	No
5	RAI, CAVC, DORV, TGA, PA	4-mm right modified B-T shunt on DOL 15	Elective Ladd procedure 14 d after cardiac operation	Shunt occlusion diagnosed on arrival to ICU; emergent 3.5-mm right modified B-T shunt revision performed; thrombus identified in original shunt	No
6	RAI, CAVC, DORV, TGA, PS	3.5-mm right modified B-T shunt on DOL 16	Elective laparoscopic Ladd procedure 7 d before cardiac operation	Fulminant necrotizing enterocolitis and multisystem organ failure on DOL 19	Yes

B-T = Blalock-Taussig; CAVC = common atrioventricular canal; DOL = day of life; DORV = double-outlet right ventricle; ICU = intensive care unit; IRA = intestinal rotational anomaly; LAI = left atrial isomerism; PA = pulmonary atresia; PGE-1 = prostaglandin E1; PS = pulmonary stenosis; RAI = right atrial isomerism; TGA = transposition of the great arteries; TAPVR = total anomalous pulmonary venous return.

Table 3. Risk Factors for Early S-P Artery Shunt Failure in Infants With HS and Single Ventricle

Variable	Outcome		OR	*p* Value
	Yes, n = 4	No, n =1 7		
Male sex, n (%)	2 (50)	15 (71)	NA	0.57
African American race, n (%)	0 (0)	3 (18)	NA	0.7
Birth weight, g, mean (± SD)	2,757 (± 265)	3,250 (± 420)	NA	0.08
Gestational age, wk, mean (± SD)	39 (± 1)	38.5 (± 1.5)	NA	0.06
Age at Ladd procedure, d, median (IQR)	21.5 (14–28)	18 (15-31)	NA	0.86
Time between cardiac operation and Ladd procedure, d, median (IQR)	13 (11–20.5)	12 (8–19)	NA	0.62
Preoperative serum BUN, mg/dL, mean (± SD)	20 (± 7.3)	12.5 (± 5.7)	1.2; 95% CI, 0.99–1.48	0.054
Preoperative serum Cr, mg/dL, mean (± SD)	0.55 (± 0.25)	0.46 (± 0.16)	NA	0.4
Postoperative serum BUN, mg/dL, mean (± SD)	17.7 (± 4.7)	13 (± 4.5)	NA	0.088
Postoperative serum Cr, mg/dL, mean (± SD)	0.5 (± 0.17)	0.38 (± 0.13)	NA	0.19
Preoperative oxygen saturation, %, median (IQR)	73 (62–80.5)	83 (77–85)	NA	0.07
Shunt size, 3.5 mm versus ≥ 4 mm, n (%)	3 (75)	6 (35)	NA	0.42
Central shunt versus BT shunt, n (%)	2 (50)	7 (41)	NA	1
Preoperative exposure to diuretic agents, n (%)	4 (100)	15 (88)	NA	1
Anticoagulation held 24 h before operation	2 (50)	11 (65)	NA	0.65

B-T = Blalock-Taussig; BUN = blood urea nitrogen; Cr = creatinine; HS = heterotaxy syndrome; IQR = interquartile range; S-P = systemic to pulmonary.

Our study's primary finding is a 19% early S-P artery shunt failure rate after a Ladd procedure in patients with HS and SV. Only 1 study has previously described S-P artery shunt failure associated with a Ladd procedure in patients with HS, but this study included only 2 patients with SV, both of whom had S-P artery shunt thrombosis [12]. Our institution's S-P artery shunt occlusion rate of 8% in all patients with S-P artery shunts makes our findings all the more relevant.

An S-P artery shunt is placed in infants whose cardiac anatomy, age, or weight may not permit a 1-stage complete repair [13]. Pulmonary blood flow is dependent on shunt patency, and hence occlusion of an S-P artery shunt can cause life-threatening hypoxemia. S-P artery shunt occlusion is not uncommon; an in-hospital occlusion rate of 6.5% to 13.5% has previously been reported [14, 15]. Various factors increase the risk for shunt occlusion: lower weight, younger age, smaller shunt size, and surgical approach through a thoracotomy [14, 15].

Lowering the pressure gradient or increasing resistance across an S-P artery shunt may potentially decrease flow across the shunt and promote thrombus formation. Because preoperative BUN levels were higher in the patients with early shunt failure in our study, we speculate that shunt flow may have been decreased because of suboptimal hydration status. Perioperative fluid balance was not available in all patients, and postoperative serum BUN and creatinine levels were measured after shunt revision in the patients with shunt failure. Hence, it is not possible to determine conclusively, based on the available data, whether suboptimal perioperative hydration status contributed to shunt failure.

Neonates exposed to cardiopulmonary bypass, and especially those with SV physiology, are at elevated risk for postoperative thrombotic events [16–18]. Immature hemostatic mechanisms, developmental deficiencies, and postoperative depletion of circulating procoagulant and anticoagulant protein levels result in a diminished capacity to suppress thrombin generation after cardiac operations [16–19]. Hence a hypercoagulable state exists after cardiac operations that may exhibit resistance to anticoagulation [20, 21]. We speculate that additional inciting events such as surgical trauma may exacerbate a baseline prothrombotic state and increase the risk for S-P artery shunt thrombosis in the perioperative period. Coagulation profile and hypercoagulability markers were not measured in our series, and hence we were unable to confirm the role of hypercoagulability in patients who experienced early shunt failure.

There is broad agreement that patients with HS and symptomatic IRAs require a Ladd procedure [7–11]. However, there is no consensus for the management of asymptomatic IRAs in patients with HS. As a result, considerable controversy exists and widespread practice variation is noted [10]. Some centers screen all patients with HS for IRAs and perform an elective Ladd procedure on those who test positive [7–10]. Others adopt a conservative approach and defer from screening for IRAs unless the patient with HS develops symptoms [22]. This divergence of practice results from unclear benefits of elective Ladd procedures in asymptomatic patients with HS. A wide spectrum of IRAs exist, not all of which pose an equivalent risk of volvulus [23, 24]. Hence, the benefit of an elective Ladd procedure in an asymptomatic patient with an IRA variant that has low-risk potential for volvulus is likely minimal. However, the risks and complications of electively or emergently performed Ladd procedure are clear and have been previously reported

[25]. The most common complication after a Ladd procedure includes a significant risk of recurrent small bowel obstruction requiring surgical intervention in 5% to 14% of children [8, 25–27]. Furthermore, the risk of recurrent midgut volvulus, although rare, is not abolished after the Ladd procedure [27, 28].

Our experience underscores the importance of assessing the risk-benefit of an elective Ladd procedure in asymptomatic patients with HS, particularly in those with S-P artery shunts and SV. Our results indicate that an individualized approach in these patients is warranted. Our current practice is to perform a screening UGI contrast study in all patients with HS when stable. Patients who screen positive for IRAs and who do not have an S-P artery shunt undergo an elective Ladd procedure when they are stable from a cardiovascular standpoint and usually during the same hospitalization. Patients who have an S-P artery shunt undergo an elective Ladd procedure 3 to 4 months after S-P artery shunt takedown or a Glenn procedure, or both. The Glenn procedure, performed at 4 to 6 months of age increases oxygen saturation, decreases ventricular volume load, and permits a more stable circulation.

The risk of volvulus is greatest in neonates and declines with age but is not eliminated [28–30]. Hence, our center follows a conservative approach and offers an elective Ladd procedure to infants with HS despite a diminishing risk of volvulus in later infancy and childhood. Although our institution favors an elective Ladd procedure in all IRA variants, further risk stratification and asymptomatic patients with low-risk IRA variants can probably be managed with close surveillance and without the need for an elective Ladd procedure.

There are several limitations to this study, including its retrospective design with its inherent disadvantages. Our study may not have been powered to detect variables of interest that could potentially influence the primary end points. The extended study period may not reflect recent improvements in the performance of noncardiac procedures in patients with HS, including anesthetic delivery, fluid management in the perioperative period, and a better appreciation of circulatory balance and anticoagulation in patients with S-P artery shunts. Anticoagulant use and whether it was given before the Ladd procedure was not available for all patients and hence we were not able to identify if perioperative anticoagulant use was protective. This study was limited to Ladd procedures in patients with HS and does not answer if similar risks apply when patients with HS undergo other noncardiac operations, eg, gastrostomy tube placement. This study does not answer whether there is a heightened risk of shunt failure in patients with HS compared with non-HS patients, nor does it answer whether it is safe to defer Ladd procedures until after S-P artery shunt takedown and a Glenn procedure.

In conclusion, patients with SV and HS with S-P artery shunts are at risk for early shunt failure after a Ladd procedure. Careful risk-benefit analysis is indicated before recommending routine elective Ladd procedures in patients with HS.

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