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MRI Evaluation of Lymphatic Abnormalities in the Neck and Thorax after Fontan Surgery: Relationship with Outcome.

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ORIGINAL RESEARCH • PEDIATRIC IMAGING

Radiology

MRI Evaluation of Lymphatic Abnormalities in the Neck and Thorax after Fontan Surgery: Relationship with Outcome

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Conflicts of interest are listed at the end of this article.

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Background: The Fontan operation is performed for surgical palliation of single ventricle physiology. This operation is usually preceded by a superior cavopulmonary connection (SCPC); lymphatic abnormalities after SCPC may be demonstrated at MRI and prior to the Fontan operation.

Purpose: To determine if the degree of neck and thoracic lymphatic abnormalities at T2-weighted MRI in patients after superior cavopulmonary connection (SCPC) correlated with surgical outcomes from the Fontan procedure.

Materials and Methods: Patients for whom SCPC was performed for palliation of single ventricle disease who underwent chest MRI between July 2012 and May 2015 at a single institution were retrospectively reviewed. T2-weighted images were scored as lymphatic type 1 (little or no T2 mediastinal and supraclavicular signal) to type 4 (T2 signal into both the mediastinum and the lung parenchyma). Fontan takedown, duration of post-Fontan hospitalization and pleural effusion, postoperative plastic bronchitis, need for transplant, and mortality were tabulated. The relationship between lymphatic type and clinical outcomes was evaluated by using analysis of variance (ANOVA), the Kruskal-Wallis H test, and the Fisher exact test.

Results: A total of 83 patients (mean age, 7.9 years ± 2.6) were evaluated. Among these 83 patients, 53 (64%) were classified with type 1 or 2 lymphatic abnormalities, 17 (20%) with type 3, and 12 (16%) with type 4. The rate of failure of Fontan completion was higher in patients with type 4 than in type 1 or 2 (54% vs 2%, respectively; $P = .004$). Need for cardiac transplant (one of 13 [8%]) and death (three of 13 [23%]) occurred only in type 4. Median postoperative length of stay was longer for patients with type 4 than for those with types 1 or 2 (29 days vs 9 days, respectively; $P < .01$).

Conclusion: Greater MRI-based severity of lymphatic abnormalities in patients prior to planned Fontan procedure was associated with failure of Fontan completion and longer postoperative stay.

Online supplemental material is available for this article.

The spectrum of single ventricle disease and associated surgical palliation is one of the most complex subgroupings of congenital heart disease. This growing patient population has unique needs for clinical and imaging follow-up due to their susceptibility to a wide range of acute and late complications (1,2).

The total cavopulmonary connection or Fontan operation is the final planned stage of surgical palliation for patients with single ventricle physiology and is usually preceded by a superior cavopulmonary connection (SCPC). The lymphatic system is adversely affected by Fontan physiology (3–6). Increased central venous pressure, inherent in the Fontan circulation, leads to a shift in Starling forces and increased lymphatic production, especially by the liver (7–10). The increased central venous pressure also impedes lymphatic drainage, leading to compensatory lymphatic changes and in some cases complications of lymphatic failure, such as chylous effusions, plastic bronchitis, and protein-losing enteropathy (3–6). Lymphatic abnormalities in patients with single ventricle physiology are often present in fetal life in association with congenital cardiovascular anomalies and demonstrate early effects on neonatal outcomes (11). The observation of early lymphatic abnormalities has been demonstrated with SCPC prior to the Fontan operation (5).

Sometimes these cardiovascular and lymphatic complications can lead to the need for urgent Fontan takedown or patient death (12–14). Multiple clinical and imaging factors have been investigated as predictors of successful Fontan palliation, including ventricular systolic and diastolic function, favorable pulmonary vascular resistance, and unobstructed pulmonary vasculature (15); however, most cases of Fontan failure are still unexplained despite

Abbreviations

ANOVA = analysis of variance, ECMO = extracorporeal membrane oxygenation, HLHS = hypoplastic left heart syndrome, Qp/Qs = ratio of pulmonary to systemic blood flow, SCPC = superior cavopulmonary connection, 3D = three dimensional

Summary

Patients with single ventricle physiology and greater extension and distribution of lymphatic channels in the chest and neck at T2-weighted MRI had worse outcomes after planned Fontan surgery.

Key Points

- Two percent of patients classified with type 1 or 2 lymphatic channels (mild to moderate lymphatic distention) did not complete Fontan palliation versus 54% of patients with type 4 (severe distention) ($P < .001$).
- Patients classified with type 4 lymphatic channels (severe) have greater than three times the median length of hospital stay after Fontan surgery (29 days) in comparison to those classified with type 1 or 2 (mild to moderate, 9 days).

evaluation with echocardiography, cardiac catheterization, and cardiac MRI (12,13).

In this study, we hypothesized that greater neck and thoracic lymphatic abnormalities in patients who underwent SCPC would be associated with worse post-Fontan outcomes. Therefore, the purpose of this study was to correlate the extent of lymphatic abnormalities at T2-weighted MRI in patients after SCPC with surgical outcomes following the Fontan procedure.

Materials and Methods

The institutional review board at a tertiary children's hospital approved the study with waiver for informed consent.

Population

We performed a retrospective review of all consecutive patients with a history of surgical palliation with SCPC who underwent cardiac MRI with a three-dimensional (3D), T2-weighted sampling perfection with application-optimized contrasts using different flip-angle evolution (SPACE) sequence between June 1, 2012, and May 31, 2015, at the Children's Hospital of Philadelphia (Fig 1). At our institution, this T2-weighted sequence was routinely added to the standard protocol for cardiac MRI in all patients with single ventricle disease regardless of symptoms or indications beginning at the commencement of the study period.

MRI Protocol

All T2-weighted lymphatic imaging was performed during routine cardiac MRI examinations that were ordered for clinical indications in patients with single ventricle disease. Images were obtained in the coronal orientation on a 1.5-T MRI magnet (Siemens MAGNETOM Avanto, Malven, Penn) with the following parameters: matrix, 256 × 256; field of view, 300–450; repetition time, 2500 msec; echo time, 650 msec; flip angle, 140 degrees; and voxel size, 1.0 × 1.0 × 1.0 to 1.3 × 1.3 × 1.3 mm. All studies imaged the neck and chest. Depending on the patient's size, the abdomen was also included as caudally as possible. The average imaging time was approximately 5–6 minutes but this varied based on patient size.

MRI Analysis and Classification

The T2-weighted images were evaluated independently by a pediatric radiologist (D.M.B., with 4 years of experience subspecializing in lymphatic imaging) and a pediatric interventional cardiologist (Y.D., with 6 years of experience subspecializing in lymphatic imaging) blinded to the clinical outcomes of the patients. The images were reviewed in the coronal plane only and scored on a scale of 1 to 4 based on the anatomic location of prominent lymphatic channels within the neck and chest. Abnormal lymphatic perfusion appears to begin with extension of prominent lymphatic channels in the supraclavicular region usually adjacent to the venous angle before extending to additional anatomic regions. Images at T2-weighted MRI scored as type 1 were interpreted as having little or no presumed lymphatic channels within the supraclavicular region and mediastinum (Fig 2). Images scored as type 2 demonstrated abnormal increased lymphatic channels within the supraclavicular region without extension into the mediastinum (Fig 3). Images scored as type 3 demonstrated abnormal supraclavicular lymphatics with extension into the mediastinum (Fig 4). Images scored as type 4 demonstrated abnormal supraclavicular lymphatic channels with extension both into the mediastinum and in an interstitial pattern into the lung parenchyma (Fig 5, Fig E1 [online]). Following independent evaluation of the images, discrepancies between the two readers were solved by consensus.

Data Analysis

The medical record was retrospectively reviewed. Demographic information, including age at T2-weighted imaging, sex, age at SCPC, and age at Fontan surgery, was obtained. The underlying cardiac anatomic and functional echocardiographic diagnosis was also recorded. If a pre-Fontan cardiac catheterization was performed, hemodynamics including SCPC pressure, right atrial pressure, transpulmonary gradient, pulmonary vascular resistance, and the ratio of pulmonary to systemic blood flow (Qp/Qs) were recorded. From the Fontan operation, cardiopulmonary bypass time, cross clamp time, circulatory arrest times, and the type of Fontan (extracardiac conduit or intra-atrial lateral tunnel) were recorded.

The postoperative course following Fontan palliation was evaluated, including Fontan completion, Fontan takedown, duration of Fontan hospitalization, duration of pleural effusion determined by duration of chest tube placement, postoperative diagnosis of plastic bronchitis, need for transplant within 3 years, and 3-year mortality from the time of T2-weighted imaging.

Statistical Analysis

Distributions of demographic characteristics along with clinical and outcomes variables are presented by patient type. Demographic and clinical variables include sex, age at SCPC (continuous), age (in years) at imaging (continuous), age (in years) at Fontan completion (continuous), dominant ventricular morphology (hypoplastic left heart syndrome [HLHS] vs non-HLHS), ventricular dysfunction by echo (normal or mild

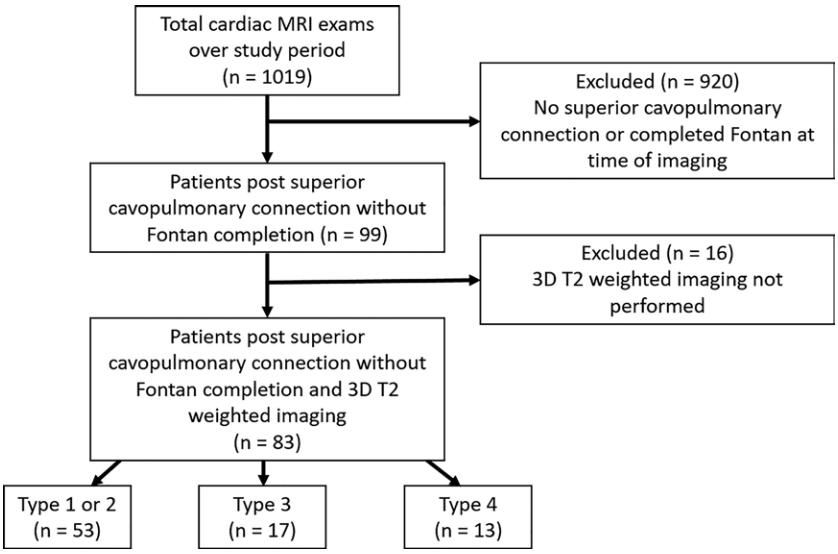
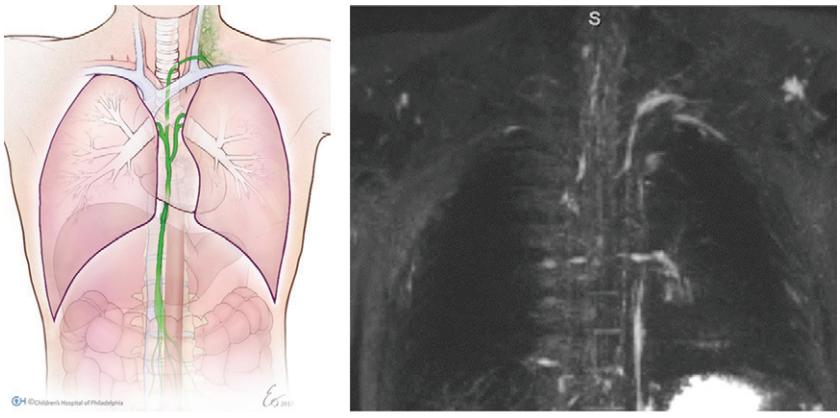


Figure 1: Flow diagram of study cohort.



a. b.

Figure 2: Type 1 classification schematic and T2-weighted MRI in a 39-month-old male patient. **(a)** Schematic shows minimal supraclavicular increased signal intensity. **(b)** A representative maximum intensity projection from T2-weighted MRI demonstrates minimal lymphatic channels in the mediastinum or neck.

vs moderate or severe), atrioventricular valve regurgitation by echo (qualitatively as normal or mild, moderate, severe), and length of Fontan operation in minutes (continuous; cardiopulmonary bypass time, cross-clamp time, circulatory arrest times). Additional continuous clinical variables include cardiac catheterization: SCPC pressure (mmHg), right atrial pressure (mmHg), transpulmonary gradient (mmHg), indexed pulmonary vascular resistance (Wood units), and Qp/Qs. Outcomes include Fontan completion, Fontan takedown, transplant within 3 years, mortality within 3 years, duration of effusions in days, and duration of hospital stay in days. Additional outcomes include atrioventricular valve regurgitation severity and a composite outcome of mortality, transplant, Fontan takedown, and failure to achieve Fontan. Associations between demographic or clinical and outcome variables were evaluated by using analysis of variance (ANOVA) and Kruskal-Wallis H test for normally distributed and non-normally distributed variables, respectively. The Fisher exact test was used for categorical variables. The Bonferroni correction and the Dunn test were used for pairwise comparisons when the overall test was statistically significant. Statistical significance was established at P values less than .05 for overall and pairwise adjusted tests of significance and P values less than .017 for pairwise tests that were not adjusted. The interrater reliability of the independent categorization of the imaging by the two readers was measured with Fleiss kappa coefficients. A kappa value of 0.40 or less indicates poor agreement, 0.41–0.59 indicates fair agreement, 0.60–0.74 indicates good agreement, and 0.75 or greater indicates excellent agreement (16). All statistical analyses were performed by using Stata v14.0 (Stata, College Station, Tex).

Results

Patient Demographics and Clinical History

Over 3 years, a total of 1019 cardiac MRI examinations were performed at our institution in children and young adults. Among these patients, 99 had undergone an SCPC without a Fontan palliation. Although the 3D T2-weighted sequence was included in our cardiac MRI protocol for single ventricle disease at the commencement of the study period, 16 patients were excluded because the 3D T2-weighted sequence was inadvertently not performed during the cardiac examination (Fig 1). The final study cohort was 83 patients (median age, 7.3 years [interquartile range {IQR}, 6.7–8.3 years]; mean age ± standard deviation [SD], 7.9 years ± 2.6). There were 48 male patients (median age, 7.4 years [IQR, 7.0–8.3 years]; mean age ± SD, 8.1 years ± 2.6) and 35 female patients (median age, 7.1 years [IQR, 6.1–8.3 years]; mean age ± SD, 7.6 years ± 2.4). There was no significant difference in age distribution between male and female patients (P = .124). Most of the patients (56%) had a diagnosis of HLHS, with others presenting with various diagnoses, such as pulmonary atresia with intact ventricular septum, double outlet right ventricle, or tricuspid atresia. Among all patients, the average age at SCPC completion was 0.68 years, the average age at T2-weighted imaging was 3.3 years, and the average age at Fontan surgery (if completed) was 3.6 years. None of the patients were lost to follow-up.

Lymphatic Classification Results

Based on the imaging classification noted above (Figs 2–5), among the 83 patients in the study, 53 (64%) were classified as having type 1 or 2 lymphatic channels, 17 (20%) were classified as having type 3, and 13 (16%) were classified as having type 4.

Interrater reliability was good using Fleiss kappa (κ = 0.62) and there was no disagreement between the readers in regards to classifying type 4 patients. Demographic and clinical variables are presented in Table 1. There was no statistical difference between the groups in relation to the age at SCPC completion or Fontan completion (if performed, *n* = 74), but there was a difference in the age when T2-weighted imaging was performed.

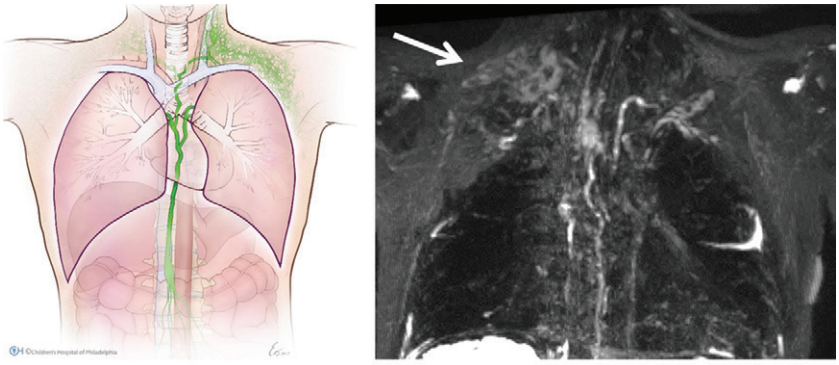
Lymphatic Classification and Catheterization Outcome

Prior to Fontan completion, 61 of 83 (73%) patients underwent cardiac catheterization. All patients characterized as type 4 underwent preoperative cardiac catheterization and 15 of 17 (88%) patients categorized as type 3 underwent cardiac catheterization, whereas only 33 of 53 (62%) patients characterized as type 1 or 2 underwent preoperative cardiac catheterization. Table 2 lists the differences in the preoperative cardiac catheterization values and T2-weighted classification. SCPC pressures were available in all patients who underwent cardiac catheterization and were significantly higher in type 4 patients (Table 2). No significant differences among the classifications were demonstrated for the indexed pulmonary vascular resistance (reported in 56 patients), the transpulmonary pressure gradient (reported in 59 patients), and the Qp/Qs based on cardiac catheterization oximetry data (reported in 55 patients) (Table 2).

Lymphatic Classification and Clinical Outcome

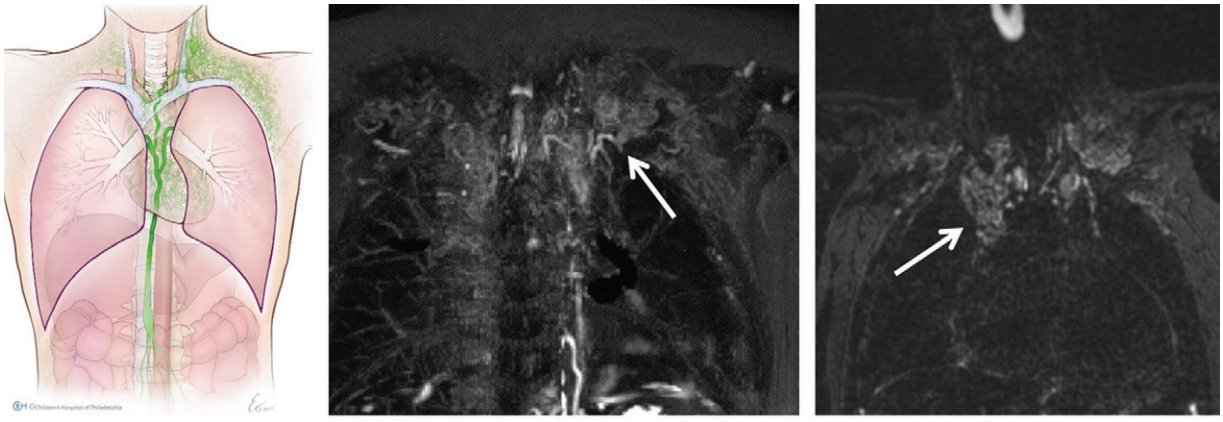
The associations between lymphatic classifications at MRI and clinical outcomes are listed in Table 3. Patients with type 1 and type 2 were grouped together since they had no difference in clinical course and outcomes. Significant differences were demonstrated when comparing rates of Fontan completion, 3-year mortality, duration of effusions (type 1 or 2, 6 days; type 3, 8 days; type 4, 15 days), and length of postoperative hospital stay (type 1 or 2, 9 days; type 3, 10 days; type 4, 29 days). These significant differences were also seen when directly cross comparing type 1 and 2 patients to type 4 patients (Table 3). When comparing type 3 patients to type 4 patients, a significant difference was demonstrated in Fontan completion. Death (*n* = 3), need for transplant (*n* = 1), extracorporeal membrane oxygenation (ECMO) (*n* = 3), plastic bronchitis (*n* = 2), and acute Fontan takedown (*n* = 1) were outcomes only in type 4 patients. Of the 13 patients categorized as having type 4, only six (46%) completed the Fontan operation and only two (15%) are alive with Fontan anatomy at last follow-up.

On echocardiography, there was no difference between the groups in ventricular function but type 4 patients were significantly more likely to have moderate to severe atrioventricular valve regurgitation compared with type 1 and type 2 patients (Table 1). If a Fontan operation was performed (*n* = 74),



a. b.

Figure 3: Type 2 classification schematic and T2-weighted MRI in a 38-month-old female patient. **(a)** Schematic depicts increased signal intensity within the bilateral supraclavicular region without extension into the mediastinum. **(b)** A representative maximum intensity projection demonstrates increased signal intensity within bilateral supraclavicular regions more on the right (arrow) without extension into the mediastinum.



a. b. c.

Figure 4: Type 3 classification schematic and T2-weighted MRI in a 34-month-old female patient. **(a)** Schematic demonstrates increased signal intensity in the supraclavicular regions and extending into the mediastinum. **(b)** A representative maximum intensity projection of a three-dimensional (3D), heavily T2-weighted MRI sequence demonstrates presumed lymphatic channels within the neck with extension into the mediastinum (arrow). **(c)** A coronal image from a 3D, heavily T2-weighted MRI sequence better demonstrates the extension of lymphatics within the mediastinum (arrow).

there was no difference between the groups in regards to circulatory arrest time, cross-clamp time, or cardiopulmonary bypass time. Additional analyses show that eight of 13 (60%) type 4 patients who had mild or moderate atrioventricular valve regurgitation experienced outcomes of death, transplant, Fontan takedown, or failure to achieve Fontan, whereas all type 4 patients with severe atrioventricular valve regurgitation experienced one of these outcomes.

Discussion

In this study, we aimed to determine if the grading of lymphatic abnormalities prior to Fontan surgery correlates with acute superior cavopulmonary connection (SCPC) outcomes using a T2-weighted MRI sequence. This sequence is available across multiple vendors with different names where a fast spin-echo sequence is performed and optimized for isotropic 3D imaging. We classified pre-Fontan lymphatic abnormalities into four types based on the anatomic location of prominent lymphatic channels within the neck and thorax. For analysis, patients with type 1 and 2 abnormalities were grouped together as these patients did not differ in their clinical course and outcome. Our results show that patients with these four types of T2 MRI signal abnormality had similar demographics and underlying cardiac anatomic categories. However, patients with type 4 abnormality had a higher risk for acute Fontan failure and adverse outcomes. Death (n = 3), need for transplant (n = 1), ECMO (n = 3), plastic bronchitis (n = 2), and need for acute Fontan takedown (n = 1) were only present in patients within this category.

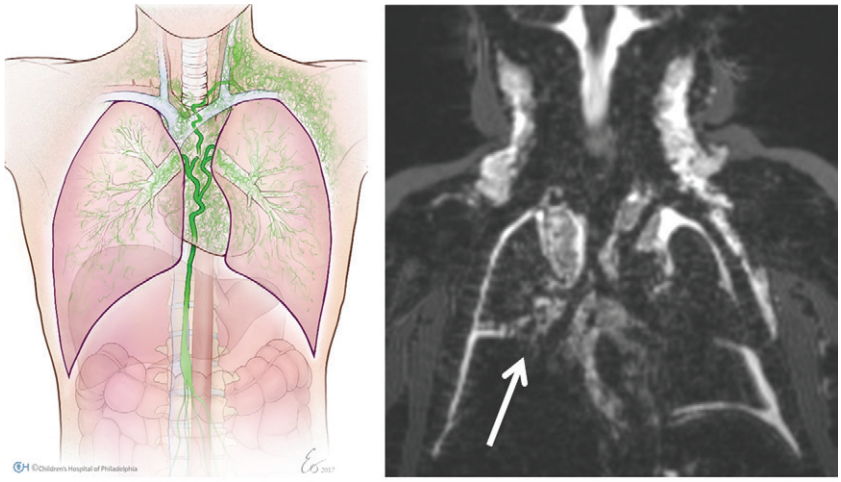


Figure 5: Type 4 classification schematic and T2-weighted MRI in a 7-month-old male patient. **(a)** Schematic shows increased abnormal signal intensity in the bilateral supraclavicular regions extending into the mediastinum and with an interstitial pattern into the lungs. **(b)** A coronal image from a three-dimensional, heavily T2-weighted MRI sequence demonstrates T2 abnormality within the bilateral supraclavicular regions with extension into the mediastinum, and into the interstitium of the lungs surrounding the right bronchus (arrow).

Table 1: Demographic and Clinical Variables in Children with SCPC with Type 1 or 2, Type 3, and Type 4 Lymphatic Abnormalities on T2-weighted MRI

Variable	Type 1 or 2 (n = 53)	Type 3 (n = 17)	Type 4 (n = 13)	P Value
Sex				.05
Male	31 (58.5)	8 (47.1)	9 (69.2)	
Female	22 (41.5)	9 (52.9)	4 (30.8)	
Age (y) at SCPC*	0.65	0.54	0.43	.07
Age (y) at T2-weighted imaging*	3.3	4.4	2.3	.04
Age (y) at Fontan completion, if performed*	3.5	4.4	3.0	.23
Dominant ventricular morphology				.96
Hypoplastic left heart syndrome	30 (56.6)	9 (52.9)	7 (53.8)	
Non–hypoplastic left heart syndrome	23 (43.4)	8 (47.1)	6 (46.2)	
Ventricular dysfunction echo				.12
Normal/mild	52 (98.1)	16 (94.1)	11 (84.6)	
Moderate/severe	1 (1.9)	1 (5.9)	2 (15.4)	
Atrioventricular valve regurgitation by echo				.004
Normal/mild	46 (86.8)	11 (64.7)	7 (53.8)	
Moderate	7 (13.2)	5 (29.4)	3 (23.1)	
Severe	0 (0.0)	1 (5.9)	3 (23.1)	
Fontan operation, if performed (n = 74)*				
Cardiopulmonary bypass time (min)	67.7	69.5	63.0	.67
Cross-clamp time (min)	28.1	31.2	29.8	.61
Circulatory arrest time (min)	24.5	21.3	12.5	.06

Note.—Unless otherwise indicated, data are number of patients. Data in parentheses are percentages. SCPC = superior cavopulmonary connection.

* Data are means.

Table 2: Cardiac Catheterization Data and T2-weighted MRI Classification in Children with SCPC

Variable	Lymphatic Abnormality			*P* Value			
	Type 1 or 2 (*n* = 53)	Type 3 (*n* = 17)	Type 4 (*n* = 13)	All	Type 1 or 2 vs Type 3*	Type 1 or 2 vs Type 4*	Type 3 vs Type 4*
SCPC pressure (mmHg) (*n* = 61)	10 (9,11)	10 (8,12)	12 (11,14)	.04	.94	.02	.10
Right atrium pressure (mmHg) (*n* = 60)	6 (4,8)	6 (4,8)	8 (6,9)	.10	1.00	.05	.16
Transpulmonary gradient (mmHg) (*n* = 59)†	3.8	4.1	4.5	.12	NA	NA	NA
PVRi (Wood units) (*n* = 58)†	1.62	1.64	1.54	.91	NA	NA	NA
Qp/Qs (*n* = 55)	0.60 (0.50, 0.70)	0.65 (0.60, 0.80)	0.65 (0.53, 0.77)	.33	NA	NA	NA

Note.—Unless otherwise indicated, data are medians. Data in parentheses are the interquartile ranges. NA = not applicable, PVRi = indexed pulmonary vascular resistance, Qp/Qs = ratio of pulmonary to systemic blood flow, SCPC = superior cavopulmonary connection.

* *P* values adjusted for pairwise comparison.

† Data are means.

Table 3: Surgical Outcomes and T2-weighted MRI Classification of Neck and Thoracic Lymphatic Abnormalities in Children with Superior Cavopulmonary Connection

Surgical Outcome	Lymphatic Abnormality			*P* Value			
	Type 1 or 2 (*n* = 53)	Type 3 (*n* = 17)	Type 4 (*n* = 13)	All	Type 1 or 2 vs Type 3	Type 1 or 2 vs Type 4	Type 3 vs Type 4
Fontan completion*				<.01	.43	<.001	.01
Yes	52 (98.1)	16 (94.1)	6 (46.2)				
No	1 (1.9)	1 (5.9)	7 (53.8)				
Fontan takedown				.07	NA	NA	NA
Yes	0 (0.0)	0 (0.0)	1 (7.7)				
No	53 (100)	17 (100)	12 (92.3)				
Transplant*				.16	NA	.20	.43
Yes	0 (0.0)	0 (0.0)	1 (7.7)				
No	53 (100)	17 (100)	12 (92.3)				
Mortality*				.003	NA	.006	.07
Alive	53 (100)	17 (100)	10 (76.9)				
Deceased	0 (0.0)	0 (0.0)	3 (23.1)				
Duration of effusions (d)†	6 (4, 8.5)	8 (6.5,17)	14.5 (9.5, 20)	.02	.11	.03	.29
Duration of hospital stay (d)†	9 (7, 11)	10 (8, 34)	28.5 (21.5, 29.5)	.003	.07	.003	.10

Note.—Unless otherwise indicated, data are number of patients. Data in parentheses are percentages. NA = not applicable.

* *P* value level of significance for pairwise comparisons = .017.

† Data are medians. Data in parentheses are first and third quartiles. *P* values adjusted for pairwise comparison.

Previously, we have demonstrated by dynamic contrast material–enhanced MR lymphangiography that the reticulated supraclavicular, mediastinal, peribronchial, and pulmonary interstitial findings correlate with abnormal lymphatic perfusion and dilated lymphatic networks (3,4). When extending into the lung parenchyma, as seen in type 4 patients, this abnormal perfusion has been called pulmonary lymphatic perfusion syndrome (4). Patients with type 3 and type 4 imaging findings appear to be clinically distinct, with patients with type 4 findings having poorer outcomes. However, it is unclear if these categories represent distinctly different congenital lymphatic abnormalities or whether these represent the severe end of a spectrum of the same abnormality. It is possible that the distinction between these categories is a result of varying progression of the same abnormality as a result of chronic lymphatic overload in the setting of chronically increased central venous pressure.

Several studies have shown that lymphatic abnormalities in patients with single ventricle anatomy can be present prenatally, possibly pointing toward a congenital etiology (11,17,18). It has also been shown that prenatal abnormalities correlate with poor neonatal outcome (11). Furthermore, it is known that increased innominate vein pressure inherent in SCPC physiology leads to increased lymphatic afterload and poor lymphatic drainage, perhaps supporting lymphatic overload as a potential etiology (9,19). Upon conversion to Fontan physiology, elevated central venous pressure leads to significant increased lymphatic production mainly by the liver, which could lead to early lymphatic failure and possibly Fontan failure; this explains why patients with severe pre-Fontan lymphatic abnormalities do not do well after the Fontan operation. It seems most likely that a combination of anatomic susceptibility with increased load is the etiology of the abnormalities described here.

Given the high mortality rate and Fontan failure rate in the type 4 patients, it would be beneficial to try to identify patient characteristics in this group that put these patients at risk for acute Fontan catastrophic events such as need for transplant, ECMO, Fontan takedown, or death. Of the five patients who died (three within 3 years), three had poor SCPC hemodynamics with SCPC pressures above 15 mmHg, three had severe valve regurgitation, and two had severe ventricular dysfunction. This would suggest that patients with lymphatic dysfunction combined with poor hemodynamics or cardiac function are at particularly high risk. New lymphatic interventional techniques could potentially alter the outcome of these patients but further studies will need to be conducted to better understand which of the patients with severe lymphatic abnormalities will ultimately be able to complete the Fontan operation successfully and which should be considered for transplant as an alternative strategy.

The main limitation of this study is that it is retrospective. In addition, there is a risk for selection bias as only patients referred for pre-Fontan cardiac MRI were included and they may not be representative of the entire Fontan population. Despite there being a possible selection bias in the patients chosen for MRI studies, this percentage of patients is consistent with the rate of acute Fontan failure reported in the literature when pleural effusions were considered as a criteria for acute Fontan failure (13). Although there was moderate interrater reliability, additional standardization with further experience may be beneficial. The follow-up of our cohort is also relatively short term; adverse outcomes such as plastic bronchitis, cardiac transplant, and mortality are time-dependent and would be influenced with a longer-term follow-up.

This study demonstrates an association between lymphatic abnormality grading on T2-weighted imaging and Fontan outcomes. Grading of lymphatic patterns on T2-weighted MRI may serve as a noninvasive means of risk assessment prior to Fontan sugery and warrants further investigation.

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