

Surveillance and screening practices of New England congenital cardiologists for patients after the Fontan operation

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Abstract

Introduction: Surveillance and management guidelines for Fontan patients are lacking due to the paucity of evidence in the literature of screening efficacy on outcome measures.

Methods: The Fontan Working Group within the New England Congenital Cardiology Association designed an electronic survey to assess surveillance practices for patients with Fontan procedures among New England congenital cardiologists and to explore variability in screening low-risk vs high-risk Fontan patients across regional programs.

Results: Fifty-six cardiologists representing 12 regional programs responded to the survey, comprising ~40% of the total New England congenital cardiac physicians. The majority of desired testing and consultation was available within 50 miles of the patient's home institution with some limitations of cardiac catheterization and cardiac magnetic resonance imaging availability. Surveillance and screening were less frequent in low-risk Fontan patients compared to high-risk Fontan patients. Counseling practices were similar for both low-risk and high-risk Fontan patients. Aspirin monotherapy was recommended by 82% of providers for low-risk Fontan patients, while anticoagulation regimens were more varied for the high-risk population. Practitioners with ≤15 years of experience were more likely to provide quality of life testing in both low-risk and high-risk Fontan patients. There were no other major differences in testing frequencies by years of practice, quaternary vs nonquaternary care facility, or the number of Fontan patients in a practice.

Conclusion: This survey provides insight into regional practices of screening and surveillance of Fontan patients. These data may be used to design future research studies and evidence-based guidelines to streamline the approach to manage these complex patients.

KEYWORDS

congenital heart disease, Fontan, screening, single ventricle, survey

1 | INTRODUCTION

Single ventricle surgical palliation has become a widespread and accepted practice over the past 40 years.¹ Single ventricle patients after the Fontan operation are a vulnerable patient population with increasing morbidity and mortality as they age.²⁻⁵ Early identification of the long-term consequences of Fontan physiology may create opportunities to improve the lifespan of these patients.

Significant variability in outcomes exists within the Fontan population. Some patients require frequent hospitalizations and accumulate multiple complications, while others have an excellent quality of life with minimal need for medications or interventions.² Comorbidities such as arrhythmias, congestive heart failure, protein-losing enteropathy (PLE), plastic bronchitis, and liver fibrosis are associated with death and the need for cardiac transplantation.³⁻⁷ While there are increasing data about the risk factors for morbidity and mortality, there are fewer data about cardiologist surveillance and testing practices. There are limited evidence-based guidelines to inform the testing modalities and optimum testing frequencies in this patient population.^{8,9}

The New England Congenital Cardiology Association (NECCA) is a collaborative group of regional physicians and advanced practice clinicians focused on quality improvement, research, patient advocacy, and continuing education in the field of pediatric and adult congenital heart disease (CHD). The Fontan Working Group was created during the 2015 annual NECCA meeting to explore barriers for the development of guidelines and to initiate collaborative research strategies to inform the practice of cardiologists managing these patients. This group determined that an understanding of the current variability of practices of Fontan screening would help inform future research projects to streamline the management of these patients. The goals of this study were to assess regional access to testing and surveillance and to evaluate provider practices caring for Fontan

patients across a diverse group of cardiologists and medical groups in New England.

2 | METHODS

The Fontan Working Group created an electronic survey targeting pediatric and adult congenital cardiologists within New England to evaluate access to testing and current practices of screening and management of Fontan patients in the region. The content within the survey was based on highlighted topics discussed during the regional meeting; the questions and survey format were edited, revised, and tested by members of the Fontan Working Group prior to distribution. The survey included questions about the provider's institution, years of experience, number of Fontan patients within their practice, cardiology subspecialization, and local access to testing modalities.

Providers were asked to differentiate between their screening practices for low-risk and high-risk Fontan patient groups as defined by their individual assessment. Suggested examples of characteristics of a low-risk Fontan patient were provided including: normal systolic and diastolic function assessment, less than moderate systemic AV valve or semilunar valve regurgitation, mean Fontan pressures < 16 mm Hg, systemic oxygen saturation \geq 92%, no arrhythmias requiring pacemaker or implantable defibrillator, no history of sustained tachyarrhythmia, no history of thrombotic complications, absence of PLE, absence of plastic bronchitis, presence of varices, ascites, splenomegaly or thrombocytopenia score for portal hypertension of one or less, normal serum creatinine level, patient < 20 years post Fontan surgery, high functional status, and minimal symptoms or complications secondary to the cardiac condition.¹⁰ The suggested characteristics of a high-risk Fontan patient were individuals who did not meet the low-risk criteria as above. Providers were given the

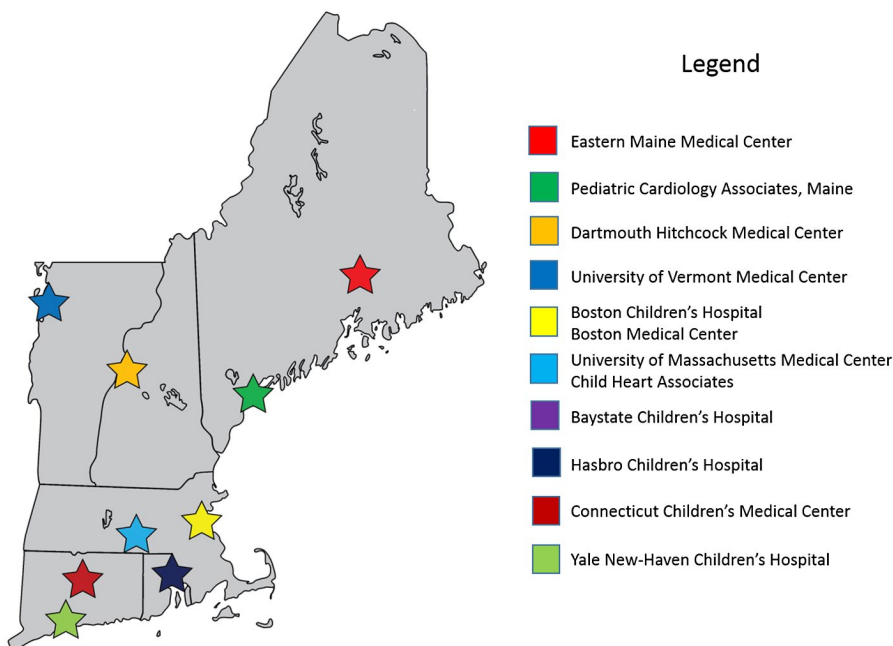


FIGURE 1 Map of New England institutions where cardiologists completed the regional survey

TABLE 1 Provider demographic information

	n = 55	(%)
Years practicing cardiology		
<5	12	21.8
5-10	8	14.5
10-15	5	9.1
>15	30	54.5
Number of Fontan patients in personal practice ^a		
<5	11	20.4
5-10	5	9.3
10-15	9	16.7
>15	29	53.7
Median age of patients ^a		
<12 years of age	19	35.2
12-18 years of age	19	35.2
>18 years of age	16	29.6
Primary cardiac specialty		
General pediatric cardiology	27	50.0
Adult congenital cardiology	8	14.8
Cardiac interventionalist	3	5.6
Cardiac imaging (Fetal, CMR, echo)	8	14.8
Heart failure/transplant	2	3.7
Electrophysiology	3	5.6
Other ^b	3	5.6

^aExcluding missing response (n = 1).

^bCardiac genetics (2) and exercise physiology (1).

opportunity to agree, agree with modifications or disagree with the suggested characteristics.

Screening questions included the frequency of outpatient follow-up, echocardiography, Holter monitoring, cardiac magnetic resonance imaging (MRI), exercise stress testing (EST), and cardiac catheterization. Additional questions assessed the frequency of sleep study, pulmonary function testing, blood work, abdominal ultrasound, liver MRI, hepatology consultation, neurodevelopmental assessment, adult congenital transition and risk assessment counseling, and anticoagulation. For analysis, the frequency of testing was categorized into a routine (regularly recurring schedule) and non-routine (performed once then as needed, only as needed or never). Additional subgroups included frequent routine testing (once a year or more) and infrequent routine testing (less than once a year). The survey questions were predominantly arranged in a multiple-choice format with an option to add comments for modifications to the high-risk and low-risk Fontan characteristics (see Online Appendix).

The survey was sent electronically to NECCA providers within the region who were then asked to advertise and distribute it to all congenital cardiologists within their institution. These cardiologists care for a large proportion of congenital cardiac patients who live in Connecticut, Massachusetts, Maine, New Hampshire, Rhode Island and Vermont. The survey was also publicized at the 2016 NECCA annual meeting and paper copies were provided that were later electronically entered into the database. Survey Monkey (Survey Monkey Inc, San Mateo, California, USA, www.surveymonkey.com) was used to create and process the survey. Descriptive statistical analysis was conducted to present the number and percentage of

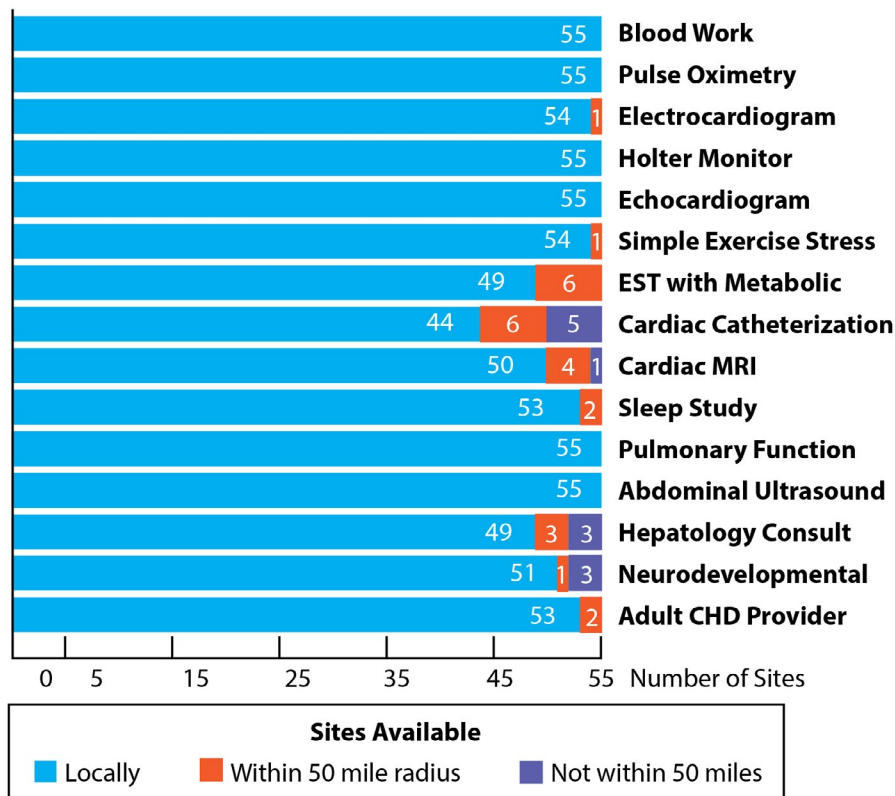


FIGURE 2 Local access to test modalities

TABLE 2 Provider responses for surveillance and testing in the low-risk and high-risk Fontan groups

	Low-risk group responses		High-risk group responses	
	<i>n</i>	%	<i>n</i>	%
<i>Average testing and surveillance frequency</i>				
Outpatient visit with EKG and pulse oximetry				
Every 6 months	9	20.9	34	82.9
Annually	34	79.1	7	17.1
Echocardiogram				
Every 6 months	2	4.7	23	56.1
Annually	35	81.4	18	43.9
Every 2 years or more	6	14.0	0	0.0
Holter monitor				
Every 6 months	0	0.0	3	7.3
Annually	6	14.0	22	53.7
Every 2 years or more	26	60.5	12	29.3
Once, then only repeat if concern arises	6	14.0	1	2.4
Only perform as needed if concern arises	5	11.6	3	7.3
Cardiac MRI				
Annually	1	2.4	3	7.5
Every 2 years or more	16	38.1	25	62.5
Once, then only repeat if concern arises	9	21.4	7	17.5
Only perform as needed if concern arises	16	38.1	5	12.5
Simple stress test (nonmetabolic)				
Every 6 months	0	0.0	1	2.6
Annually	2	5.1	5	13.2
Every 2 years or more	11	28.2	12	31.6
Once, then only repeat if concern arises	2	5.1	4	10.5
Only perform as needed if concern arises	16	41.0	7	18.4
Never	8	20.5	9	23.7
Metabolic stress test				
Every 6 months	0	0.0	1	2.4
Annually	2	4.9	6	14.6
Every 2 years or more	17	41.5	18	43.9
Once, then only repeat if concern arises	6	14.6	8	19.5
Only perform as needed if concern arises	13	31.7	7	17.1
Never	3	7.3	1	2.4
Cardiac catheterization				
Every 2 years or more	3	7.1	8	19.5
Once, then only repeat if concern arises	9	21.4	20	48.8
Only perform as needed if concern arises	30	71.4	13	31.7
Sleep study				
Every 6 months	0	0.0	1	2.4
Once, then only repeat if concern arises	2	4.7	8	19.5
Only perform as needed if concern arises	38	88.4	27	65.9
Never	3	7.0	5	12.2
Pulmonary function testing				
Every 6 months	0	0.0	1	2.4

(Continues)

TABLE 2 (Continued)

	Low-risk group responses		High-risk group responses	
	n	%	n	%
Annually	0	0.0	2	4.9
Every 2 years or more	6	14.0	8	19.5
Once, then only repeat if concern arises	8	18.6	8	19.5
Only perform as needed if concern arises	28	65.1	20	48.8
Never	1	2.3	2	4.9
Liver MRI				
Annually	1	2.5	0	0.0
Every 2 years or more	1	2.5	3	7.9
Every 5 years or more	3	7.5	6	15.8
Once, then only repeat if concern arises	3	7.5	7	18.4
Only perform as needed if concern arises	29	72.5	20	52.6
Never	3	7.5	2	5.3

a cardiologist for each variable. Practice variability by institution, years practicing cardiology, number of Fontan patients, and median age of Fontan patients were assessed using Fisher's exact test, with a *P* value of < .05 considered significant. All statistical analysis was conducted using Stata 14.2 (StataCorp, College Station, Texas).

3 | RESULTS

A total of 56 clinicians responded to the survey, which comprised approximately 40% of the pediatric and adult congenital cardiologists in New England. The NECCA group includes members of 15 institutions in New England and congenital cardiologists from 12 institutions participated in the survey (Figure 1). The largest group of participants came from Boston Children's Hospital, comprising 38% of the respondents, followed by Connecticut Children's Medical Center at 17% and Yale-New Haven Children's Hospital at 11%. We excluded 10 respondents with a missing data rate of > 70% in the analysis of the low-risk and high-risk Fontan populations. One respondent did not complete the demographic portion of the survey. Provider characteristics of 55 respondents who completed the demographic section are summarized in Table 1. The median age of Fontan patients cared for by providers was evenly distributed between those < 12 years of age, 12-18 years of age, and > 18 years of age.

Cardiologists reported that the vast majority of testing and consultation required by Fontan patients during their lifetime is available at their home institution or within a 50-mile radius (Figure 2). This included blood work, pulse oximetry, electrocardiography, Holter monitoring, echocardiography, simple exercise stress test, EST with metabolic assessment, sleep study, pulmonary function testing, abdominal ultrasound, and access to adult CHD providers. Some limitations to testing access were reported. Testing that was not available

within a 50-mile radius included cardiac catheterization for 9% of respondents and cardiac MRI for 2% of respondents. Three percent of practitioners reported that a hepatologist was not available within a 50-mile radius and another 3% did not have access to a neurodevelopmental specialist within a 50-mile radius.

The survey next addressed testing and surveillance practices for Fontan patients at low-risk vs those at high-risk as defined by the individual cardiologist. Agreement with no or some modifications for the low-risk and high-risk definition characteristics was reported for 98% of providers. Surveillance and screening patterns by providers differed in low-risk compared to high-risk Fontan patients. Cardiologists reported that most common cardiac tests, including echocardiography, Holter monitoring, cardiac MRI, and metabolic stress test, were more frequently performed in high-risk Fontan patients (Table 2). Simple stress tests and cardiac catheterizations were not routinely ordered for low-risk or high-risk groups. Testing and consultation related to other organ systems that can be affected by Fontan physiology, including abdominal ultrasound, liver MRI, hepatology consultation, sleep study, and pulmonary function testing, were also not ordered routinely for either group.

Surveillance blood tests, including a complete blood count (CBC), basic metabolic panel (BMP), liver function tests (LFTs), and gamma-glutamyl transferase (GGT), were recommended annually for both low-risk and high-risk Fontan patients (Table 3). Providers reported adding coagulation factors and a B-type natriuretic peptide (BNP) to this annual blood work for high-risk patients.

Counseling practices were similar for both low-risk and high-risk Fontan patients (Table 4). More than half of the providers did not provide quality of life survey testing for low-risk or high-risk Fontan patients. Practitioners with ≤ 15 years of practice experience were more likely to perform quality of life testing in their low-risk and high-risk Fontan patients in comparison to their counterparts

TABLE 3 Provider responses for surveillance and screening blood work in the low-risk and high-risk Fontan groups

	Low-risk group responses		High-risk group responses	
	<i>n</i>	%	<i>n</i>	%
<i>Surveillance and screening blood work</i>				
Complete Blood Count (CBC)				
Every 6 months	0	0.0	7	17.5
Annually	18	40.0	23	57.5
Every 2 years or more	14	31.1	3	7.5
Once, then only repeat if concern arises	2	4.4	3	7.5
Only perform as needed if concern arises	11	24.4	3	7.5
Never	0	0.0	1	2.5
Basic metabolic panel (BMP)				
Every 6 months	0	0.0	10	25.0
Annually	20	44.4	22	55.0
Every 2 years or more	15	33.3	3	7.5
Once, then only repeat if concern arises	1	2.2	4	10.0
Only perform as needed if concern arises	9	20.0	1	2.5
Liver function tests (LFTs)				
Every 6 months	0	0.0	7	17.5
Annually	21	46.7	26	65.0
Every 2 years or more	18	40.0	3	7.5
Once, then only repeat if concern arises	1	2.2	2	5.0
Only perform as needed if concern arises	5	11.1	2	5.0
Gamma-glutamyl transpeptidase (GGT)				
Every 6 months	0	0.0	5	12.8
Annually	18	40.9	24	61.5
Every 2 years or more	13	29.6	3	7.7
Once, then only repeat if concern arises	2	4.6	2	5.1
Only perform as needed if concern arises	10	22.7	3	7.7
Never	1	2.3	2	5.1
Reticulocyte count				
Every 6 months	0	0.0	5	12.5
Annually	7	16.3	11	27.5
Every 2 years or more	5	11.6	2	5.0
Once, then only repeat if concern arises	3	7.0	5	12.5
Only perform as needed if concern arises	23	53.5	12	30.0
Never	5	11.6	5	12.5
Iron level				
Every 6 months	0	0.0	2	5.1
Annually	7	15.6	11	28.2
Every 2 years or more	8	17.8	2	5.1
Once, then only repeat if concern arises	4	8.9	7	18.0
Only perform as needed if concern arises	24	53.3	12	30.8
Never	2	4.4	5	12.8
Magnesium				
Every 6 months	0	0.0	5	12.8
Annually	10	22.2	13	33.3

(Continues)

TABLE 3 (Continued)

	Low-risk group responses		High-risk group responses	
	<i>n</i>	%	<i>n</i>	%
Every 2 years or more	10	22.2	1	2.6
Once, then only repeat if concern arises	2	4.4	5	12.8
Only perform as needed if concern arises	21	46.7	10	25.6
Never	2	4.4	5	12.8
Coagulation factors (PT/INR, PTT)				
Every 6 months	0	0.0	4	10.3
Annually	8	17.8	17	43.6
Every 2 years or more	13	28.9	3	7.7
Once, then only repeat if concern arises	2	4.4	5	12.8
Only perform as needed if concern arises	21	46.7	9	23.1
Never	1	2.2	1	2.6
Quantitative IgG serum immunoglobulins				
Every 6 months	0	0.0	1	2.6
Annually	0	0.0	4	10.3
Every 2 years or more	2	4.4	3	7.7
Once, then only repeat if concern arises	6	13.3	7	18.0
Only perform as needed if concern arises	31	68.9	17	43.6
Never	6	13.3	7	18.0
Cystatin C				
Annually	1	2.2	2	5.1
Every 2 years or more	1	2.2	3	7.7
Once, then only repeat if concern arises	2	4.4	3	7.7
Only perform as needed if concern arises	23	51.1	14	35.9
Never	18	40.0	17	43.6
(Pro-) Brain natriuretic peptide (BNP)				
Every 6 months	0	0.0	7	18.4
Annually	8	18.2	16	42.1
Every 2 years or more	8	18.2	1	2.6
Once, then only repeat if concern arises	6	13.6	6	15.8
Only perform as needed if concern arises	22	50.0	7	18.4
Never	0	0.0	1	2.6
Vitamin D level				
Every 6 months	0	0.0	1	2.6
Annually	3	6.7	8	20.5
Every 2 years or more	5	11.1	2	5.1
Once, then only repeat if concern arises	5	11.1	5	12.8
Only perform as needed if concern arises	24	53.3	15	38.5
Never	8	17.8	8	20.5
Hepatitis C antibody				
Annually	0	0.0	2	5.3
Every 2 years or more	2	4.6	3	7.9
Once, then only repeat if concern arises	13	29.6	15	39.5
Only perform as needed if concern arises	26	59.1	15	39.5
Never	3	6.8	3	7.9

(Continues)

TABLE 3 (Continued)

	Low-risk group responses		High-risk group responses	
	<i>n</i>	%	<i>n</i>	%
Fibro test/FibroSure/ActTest liver fibrosis panel				
Every 6 months	0	0.0	1	2.6
Annually	1	2.2	6	15.4
Every 2 years or more	9	20.0	7	18.0
Once, then only repeat if concern arises	7	15.6	8	20.5
Only perform as needed if concern arises	17	37.8	9	23.1
Never	11	24.4	8	20.5

($P < .01$). Cardiologists with ≤ 15 patients in their practice were less likely to order frequent Holter monitors for low-risk patients ($P < .01$) but more likely to order frequent echocardiograms and neurodevelopmental assessments in high-risk patients ($P < .01$, $P = .045$). The neurodevelopmental assessment was more likely to be performed in patients between age 2-7 years in comparison to age > 7 years of age ($P = .03$). Otherwise with minor exceptions, no significant practice variation trends were observed based on institution, years in practice, patient volume or patient age.

The majority of cardiologists recommended influenza, pneumonia, hepatitis A and B, Haemophilus influenzae type b, and tetravalent meningococcal conjugate vaccines if age-appropriate for both low-risk and high-risk Fontan patients. Aspirin monotherapy was recommended by 82% of providers for low-risk Fontan patients. Responses were more varied for the high-risk population with 21% of providers using aspirin monotherapy, 42% of providers using warfarin monotherapy, 18% using a combination of aspirin and warfarin, and 18% using other agents or combinations of anticoagulants (Figure 3).

4 | DISCUSSION

This survey included approximately 40% of the population of pediatric and adult congenital cardiologists in New England, resulting in a good representation of programs with varying size and resources in the region. The data from this survey suggest that testing modalities are locally accessible for most routinely ordered surveillance evaluations of Fontan patients in New England, although access was limited for cardiac catheterization, cardiac MRI, hepatology consultation, and neurodevelopmental consultation in a small percentage of practices. Screening frequency was greater in high-risk Fontan patients, although counseling patterns were similar for both groups. Surveillance trends were similar based on provider institution, years in practice, patient volume, and median patient age.

Two other provider surveys of practice patterns in the management of Fontan patients are described in the medical literature. Thirty-nine practitioners from Australia and New Zealand comprising 50% of the cardiologists caring for Fontan patients in the region completed a survey to address the long-term care of Fontan

patients in their practice.¹¹ The survey focused primarily on medical management and exercise recommendations. A focus on the modal response of providers demonstrated that 36% of cardiologists used aspirin alone, while 49% used warfarin. Fifty-nine percent of providers used angiotensin-converting enzyme inhibitors in the presence of ventricular dysfunction and/or atrioventricular or semilunar valve regurgitation. Fifty-six percent used beta-blockers in the presence of ventricular dysfunction. Forty-four percent of providers encouraged medium-level competitive sports but restricted high-intensity exercise.

Physicians coordinating Fontan care programs from 11 academic institutions throughout the United States completed another recent survey.¹² Annual electrocardiograms and echocardiograms were recommended by most centers (73% and 64% modal response, respectively). Serum studies, including a CBC (73%), complete metabolic panel (73%), and BNP (54%) were recommended annually, while hepatic testing, typically consisting of liver ultrasound, was recommended biennially (45%) or less frequently (45%). Forty-six percent of respondents recommended a routine liver biopsy.

Although there are many similarities between the findings of this survey and those above, there are differences in the approach and structure of our survey. This survey was completed by providers from a variety of small, medium, and large practices throughout the New England region and explored differential access to test modalities to help inform which tests were utilized most. For example, the NECCA group includes large academic medical centers with over 70 cardiology providers as well as small group practices comprising five or fewer cardiologists. Our survey was completed by individual cardiologists rather than a single representative of an institution, enabling us to evaluate practice variation within as well as across institutions. A significant contribution of this survey is the focus on patient surveillance and testing in low-risk and high-risk Fontan patients. This reflects the real-world differences in the complexity of patients with Fontan physiology and highlights the differences observed in responses to questions about surveillance frequency and anticoagulation management in our results.

Although the majority of desired testing was available locally or within 50 miles, this in part may be a reflection of the compact

TABLE 4 Provider responses for counseling and assessments in the low-risk and high-risk Fontan groups

	Low-risk group responses		High-risk group responses	
	<i>n</i>	%	<i>n</i>	%
<i>Counseling and assessments</i>				
Neurodevelopmental assessment				
Age 2-7 years	26	59.1	28	71.8
Age 8-14 years	1	2.3	1	2.6
Age 15-18 years	0	0.0	0	0.0
At or beyond age 18 years	0	0.0	0	0.0
Only perform as needed if concern arises	15	34.1	8	20.5
Never	2	4.6	2	5.1
Adult congenital transition discussion				
Age 2-7 years	0	0.0	11	29.0
Age 8-14 years	9	20.5	19	50.0
Age 15-18 years	25	56.8	5	13.2
At or beyond age 18 years	7	15.9	1	2.6
Only perform as needed if concern arises	1	2.3	2	5.3
Never	2	4.6	0	0.0
Contraception/pregnancy counseling				
Age 2-7 years	0	0.0	0	0.0
Age 8-14 years	13	29.6	16	42.1
Age 15-18 years	26	59.1	20	52.6
At or beyond age 18 years	3	6.8	0	0.0
Only perform as needed if concern arises	2	4.6	2	5.3
Never	0	0.0	0	0.0
Exercise counseling with activity recommendations				
Age 2-7 years	22	50.0	18	47.4
Age 8-14 years	19	43.2	18	47.4
Age 15-18 years	2	4.6	1	2.6
At or beyond age 18 years	0	0.0	0	0.0
Only perform as needed if concern arises	1	2.3	1	2.6
Never	0	0.0	0	0.0
Tobacco, alcohol, illicit drug, and obesity avoidance discussion				
Age 2-7 years	3	7.0	5	13.2
Age 8-14 years	20	60.5	23	60.5
Age 15-18 years	13	30.2	9	23.7
At or beyond age 18 years	0	0.0	0	0.0
Only perform as needed if concern arises	1	2.3	1	2.6
Never	0	0.0	0	0.0
Quality of life testing				
Age 2-7 years	2	4.6	3	7.9
Age 8-14 years	1	2.3	5	13.2
Age 15-18 years	5	11.4	0	0.0
At or beyond age 18 years	0	0.0	0	0.0
Only perform as needed if concern arises	13	29.6	11	29.0
Never	23	52.3	19	50.0

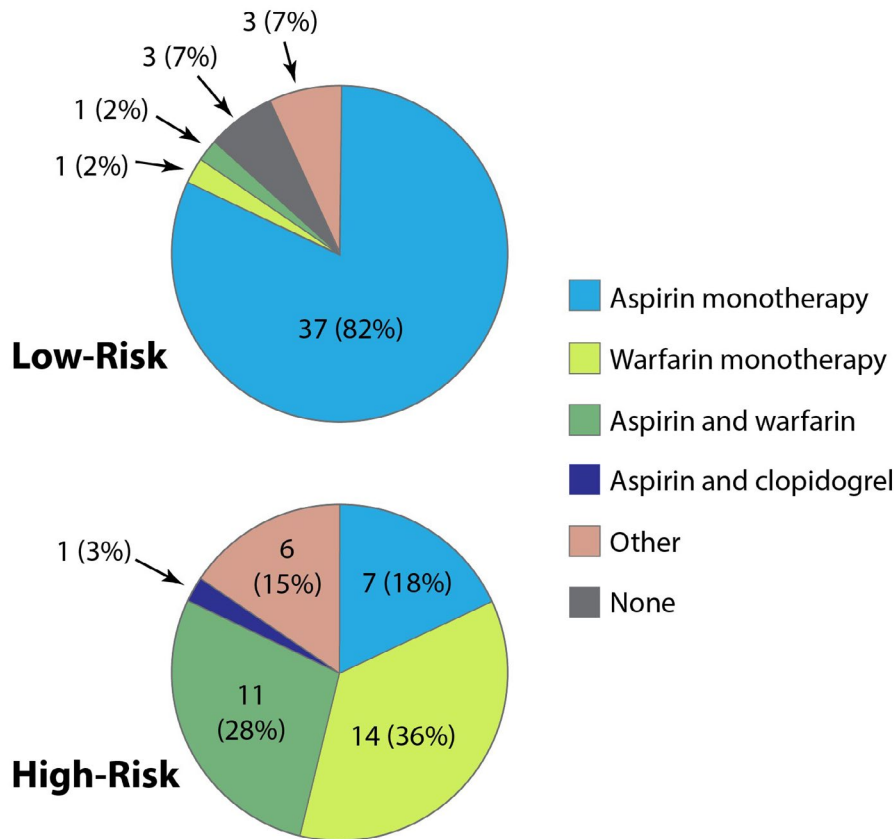


FIGURE 3 Anticoagulation recommendations by risk category

geography of New England. Many of the congenital cardiology practices in the region are located in major population centers and within a short distance from other centers, allowing access for patients who need additional testing. This may not be true in other regions of the country.

This survey of cardiologists studying their general approach to Fontan patient surveillance may not reflect actual practice patterns in a busy working outpatient clinic. Documentation and tracking of tests actually ordered in clinics would provide important data about current practices and provide a framework to study changes in practice if new guidelines are released. In addition, the quality of testing modalities, including expertise in interpretation in studies such as cardiac MRI, was not addressed. Differentiation of low-risk and high-risk Fontan patient groups was defined by the individual cardiologist in this survey; although most respondents agreed with the suggested characteristics, answers to survey questions could be influenced by differing definitions or interpretations of the characteristics of the two groups.

This survey highlights regional testing access and surveillance practices of physicians caring for Fontan patients which can aid in the development of future research within the field. With increasing focus by insurance companies on the necessity and frequency of testing, these data provide a framework to support the coverage of appropriate surveillance testing to prevent unnecessary out-of-pocket expenses for patients and potentially to avoid loss of

follow-up due to patients' financial constraints. Further studies are needed to evaluate the ability of practitioners to identify Fontan patients at higher risk and to determine whether the increased frequency of surveillance and testing result in differences in outcome measures such as quality of life, morbidity, and death. This can lead to evidence-based guideline development to promote consistent, high-quality care for complex patients with Fontan physiology.

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CONFLICT OF INTEREST

The authors report no disclosures to any relationship with industry or other relevant entities, financial or otherwise, within the past 2 years that might pose a conflict of interest in connection with the submitted article.

AUTHOR CONTRIBUTIONS

Dr Davey conceptualized and designed the study, designed the database for data collection, collected data, drafted the initial manuscript, and reviewed and revised the manuscript.

Drs Salazar, Gauthier, Valente, Elder, Wu, Berman, Pollack and Rathod conceptualized and designed the study, and reviewed and revised the manuscript.

Dr Lee performed the data analysis, drafted portions of the initial manuscript, and reviewed and revised the manuscript.

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SUPPORTING INFORMATION

Additional supporting information may be found online in the Supporting Information section.

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Surveillance and Screening of the Fontan Patient

Cardiologist and Practice Information

1. What is the name of your institution or practice?

Other (please specify)

2. How many years have you been practicing cardiology? (not including fellowship training)

- < 5 years
- 5-10 years
- 10-15 years
- > 15 years

3. How many Fontan patients are you currently following in your practice?

- <5
- 5-10
- 10-15
- >15

4. What is the median age of your Fontan cohort?

- Most <12 years of age
- Most 12-18 years of age
- Most 19 years of age or above

5. What is your *primary* cardiac specialty?

- General pediatric cardiology
- Adult congenital
- Cardiac imaging (Fetal, CMR, echo)
- Heart failure/Transplant
- Electrophysiology
- Cardiac Interventionalist
- Other
- N/A

Other (please specify)

6. What is the availability of testing and consultation?

	Locally available	Available within a 50 mile radius outside home institution	Not available within 50 miles
Bloodwork	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Pulse oximetry	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Electrocardiogram	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Holter monitor	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Echocardiogram	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Simple exercise stress testing without metabolic assessment	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Exercise stress testing with metabolic assessment	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Cardiac catheterization	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Cardiac MRI	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Sleep study	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Pulmonary function testing	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Abdominal ultrasound	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Hepatology specialist consultation	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Neurodevelopmental specialist consultation	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Adult congenital heart disease provider	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Surveillance and Screening of the Fontan Patient

Surveillance of 'Low Risk' Fontan Patients (as defined by individual cardiologist)

Low risk Fontan definition ~ 'Good' Fontan clinical characteristics *based on cardiologist's individual assessment*

Examples of characteristics of a low risk Fontan patient *may* include the following: Normal systolic and diastolic function assessment, less than moderate systemic AV valve or semilunar valve regurgitation, mean Fontan pressures <16mmHg, systemic oxygen saturation 92% or higher, no arrhythmias requiring pacemaker or ICD, no history of sustained tachyarrhythmia, no history of thrombotic complications, absence of PLE, absence of plastic bronchitis, VAST score 1, normal serum creatinine level, patient <20 years post Fontan surgery, high functional status, and minimal symptoms or complications secondary to the cardiac condition.

PLEASE PROVIDE ANSWERS REFLECTING YOU PERSONAL PRACTICE CARING FOR FONTAN PATIENTS >12 YEARS OF AGE

7. How would you define a 'low risk' or 'good' Fontan patient in your personal practice?

Agree with characteristics listed above: Type 'agree'

Agree with characteristics listed above but with the following additions and modifications:

Disagree with characteristics listed and would suggest the following:

11. In Fontan patients who you consider to be 'low risk', on average when do you begin the following Neuro/Behavioral evaluations?

	Age 2-7 years	Age 8-14 years	Age 15-18 years	At or beyond age 18 years	Only perform as needed if concern arises	Never
Neurodevelopmental assessment	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Adult Congenital transition discussion	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Contraception/pregnancy counseling	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Exercise counseling with activity recommendations	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Tobacco, alcohol, illicit drug, obesity avoidance discussion	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Quality of Life testing	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

12. In Fontan patients who you consider to be 'low risk', what are your recommendations on the following vaccinations?

	Recommended if age appropriate	Recommended if diagnosis of heterotaxy syndrome	Not regularly recommended
Influenza	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Pneumococcus	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Hepatitis A and B	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Hib and MCV4 boosters	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

13. In general terms, what do you use for primary anticoagulation prophylaxis in your 'low risk' Fontan patients?

- Aspirin monotherapy
- Warfarin monotherapy
- Aspirin and Warfarin
- Clopidogrel monotherapy
- Aspirin and Clopidogrel
- Factor Xa inhibitors
- None
- Other agents or combinations (please specify)

Surveillance and Screening of the Fontan Patient

Surveillance of 'High Risk' Fontan Patients (as defined by individual cardiologist)

High risk Fontan definition ~ 'Poor' Fontan clinical characteristics *based on cardiologist's individual assessment*

Examples of characteristics of a high risk Fontan *may* include one or more of the following:

Abnormal systolic and diastolic function assessment, moderate or more systemic AV valve or semilunar valve regurgitation, Fontan pressures 16mmHg or greater, systemic oxygen saturation 91% or less, arrhythmias requiring pacemaker or ICD, history of sustained tachyarrhythmia, history of thrombotic complications, PLE, plastic bronchitis, VAST score 2 or more, abnormal serum creatinine level, patient 20 years or more post Fontan surgery, diminished functional status, and/or frequent symptoms or complications secondary to the cardiac condition.

PLEASE PROVIDE ANSWERS REFLECTING YOUR PERSONAL PRACTICE CARING FOR FONTAN PATIENTS >12 YEARS OF AGE

Note: For the following questions, answer in broad terms of screening a high risk patient to avoid missing a complication rather than testing specific individual patients with known complications requiring diagnostic evaluation.

14. How would you define a 'high risk' or 'poor' Fontan patient in your personal practice?

Agree with characteristics listed above: Type 'agree'

Agree with characteristics listed above but with the following additions and modifications:

Disagree with characteristics listed and would suggest the following:

18. In Fontan patients you consider to be 'high risk', on average when do you begin the following Neuro/Behavioral evaluations?

	Age 2-7 years	Age 8-14 years	Age 15-18 years	At or beyond age 18 years	Only perform as needed if concern arises	Never
Neurodevelopmental assessment	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Adult Congenital transition discussion	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Contraception/pregnancy counseling	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Exercise counseling with activity recommendations	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Tobacco, alcohol, illicit drug, obesity avoidance discussion	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Quality of Life testing	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

19. In Fontan patients you consider to be 'high risk', what are your recommendations on the following vaccinations?

	Recommended if age appropriate	Recommended if diagnosis of heterotaxy syndrome	Not regularly recommended
Influenza	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Pneumococcus	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Hepatitis A and B	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Hib and MCV4 boosters	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

20. In general terms, what do you use for primary anticoagulation prophylaxis in your 'high risk' Fontan patients that require more aggressive anticoagulation?

- Aspirin monotherapy
- Warfarin monotherapy
- Aspirin and Warfarin
- Clopidogrel monotherapy
- Aspirin and Clopidogrel
- Factor Xa inhibitors
- None
- Other agents or combinations (please specify)

