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ORIGINAL ARTICLE

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Initiating a Fontan multidisciplinary clinic: Decreasing care variability, improving surveillance, and subsequent treatment of Fontan survivors

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Abstract

Background: Children with single ventricle (SV) heart disease who undergo Fontan operation are at risk for developing multiorgan dysfunction. Although survival has improved, significant comorbidities involving multiple organ systems may develop, requiring evaluation and management by many subspecialists. Using data from an internal survey, we documented high care variability for our Fontan population. We then developed a multidisciplinary clinic, designed and implemented a clinical care pathway to decrease variability of patient assessment.

Methods: After creating a multidisciplinary team and a clinical care pathway, we initiated a multidisciplinary clinic (MDC) where patients could see multiple subspecialists during a single encounter. We then monitored our effectiveness by retrospective chart review to determine care pathway adherence (process measure) and incidence of new diagnoses of end-organ injury (outcome measure) as well interventions implemented. Adherence was analyzed using statistical process control (SPC) charts.

Results: Eighty-six patients were seen in the MDC from January 2016 to September 2017. The proportion of patients with appropriate testing increased, related to strong care pathway adherence. A significant amount of novel pathology was diagnosed in all evaluated organ systems, both Fontan-associated comorbidities and general pediatric diagnoses. Subsequent interventions included cardiac catheterization n = 21 (31%) with more than half of these patients undergoing intervention n = 17 (20%), and liver biopsy n = 9 (10%). Additionally, 58 patients (67%) were referred to a neuropsychologist based on perceived clinical need, with n = 34 (40%) undergoing a neuropsychological evaluation.

Conclusions: Children who have undergone Fontan palliation are at risk for developing cardiac and noncardiac comorbidities. Use and adherence to an institutional care pathway resulted in the diagnosis of significant novel pathology and subsequently provided opportunity for intervention.

KEYWORDS

Fontan, multidisciplinary clinic, quality improvement

1 | INTRODUCTION

Survival after palliative surgery for single ventricle heart disease has improved dramatically over the last several decades and a higher proportion of these patients are able to advance to Fontan palliation.[1-3] Although postsurgical mortality has declined, there is mounting evidence for significant comorbidities involving multiple organ systems that lead to decreased quality of life and mortality in this population.[4] While the underlying pathophysiology is not yet fully understood, some aspects of subsequent pathology has been attributed to elevation in central venous pressure from total cavopulmonary anastomosis[5,6] (Figure 1). A multiyear approach was taken to better understand current treatment practice in caring for this patient population. In 2013, we conducted an internal survey to assess practice patterns in the care for children who had undergone Fontan operation, including the type and frequency of surveillance testing for cardiac and noncardiac morbidities. We found significant practice variation in screening for end-organ dysfunction. For example, a roughly equal proportion of providers referred patients with Fontan physiology routinely to a pediatric hepatologist for liver evaluation vs. never referred (unpublished data).

In 2014, we organized a multidisciplinary team of providers to see these patients in clinic in an effort to ensure subspecialist involvement, improve efficiency for patients and families, as well as decrease practice variation in care for this unique population.[7] We developed a key driver diagram (KDD) (Figure 2) to define the goals of the project. A time line has been included to show more clearly the steps that were taken to create the clinic (Figure 3). Our team is comprised of the following subspecialists: a nurse coordinator, cardiologists, pulmonologists, hepatologists, a neuropsychologist, and a pediatric psychologist; in addition, we have incorporated dieticians and a psychosocial wellness team, including social workers, chaplains, and child life specialists. Using this team's expertise, we created an evidence-based clinical care pathway to address variation in Fontan care identified from our 2013 survey data. An abbreviated version of the care pathway is presented as Table 1, as well as an accompanying synopsis in text form (Table 2).

We initiated a quality improvement project to evaluate adherence to our Fontan clinical care pathway, determine the incidence of new diagnoses of end-organ injury in this population, and catalog the interventions that were implemented based on these novel diagnoses. Our hypothesis was that a multidisciplinary team of providers following an evidence-based care pathway would result in less practice variability, increase identification of comorbidities and provision of interventions in a subset of identified comorbidities.

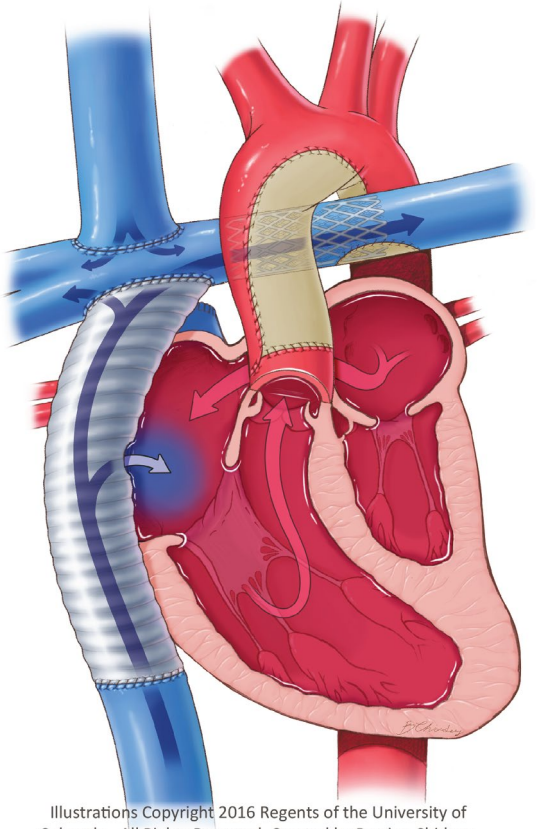

FIGURE 1 Hypoplastic left heart syndrome with extracardiac Fontan operation

2 | METHODS

2.1 | Setting

Children's Hospital Colorado (CHCO) is a quaternary care, freestanding children's hospital located in the Rocky Mountain region, drawing from a geographically diverse, 7-state catchment area. The cardiac surgical program performs >550 heart surgeries per year, with >350 requiring cardiopulmonary bypass. This study was approved by the CHCO Organizational Research Risk & Quality Improvement Review Panel.

2.2 | Patient population

The cohort included patients who had undergone Fontan palliation and are followed at our main campus in Aurora, Colorado. There are over 200 patients who have undergone Fontan operation meeting these criteria, with around 20 patients per year entering the population by undergoing Fontan surgery.

2.3 | Process measures

Fontan patients scheduled for clinic were identified and reviewed for pathway compliance (Tables 1 and 2) by our nurse coordinator who then helped setup subsequent testing. Determination of pathway compliance and identification of novel diagnoses was performed retrospectively.

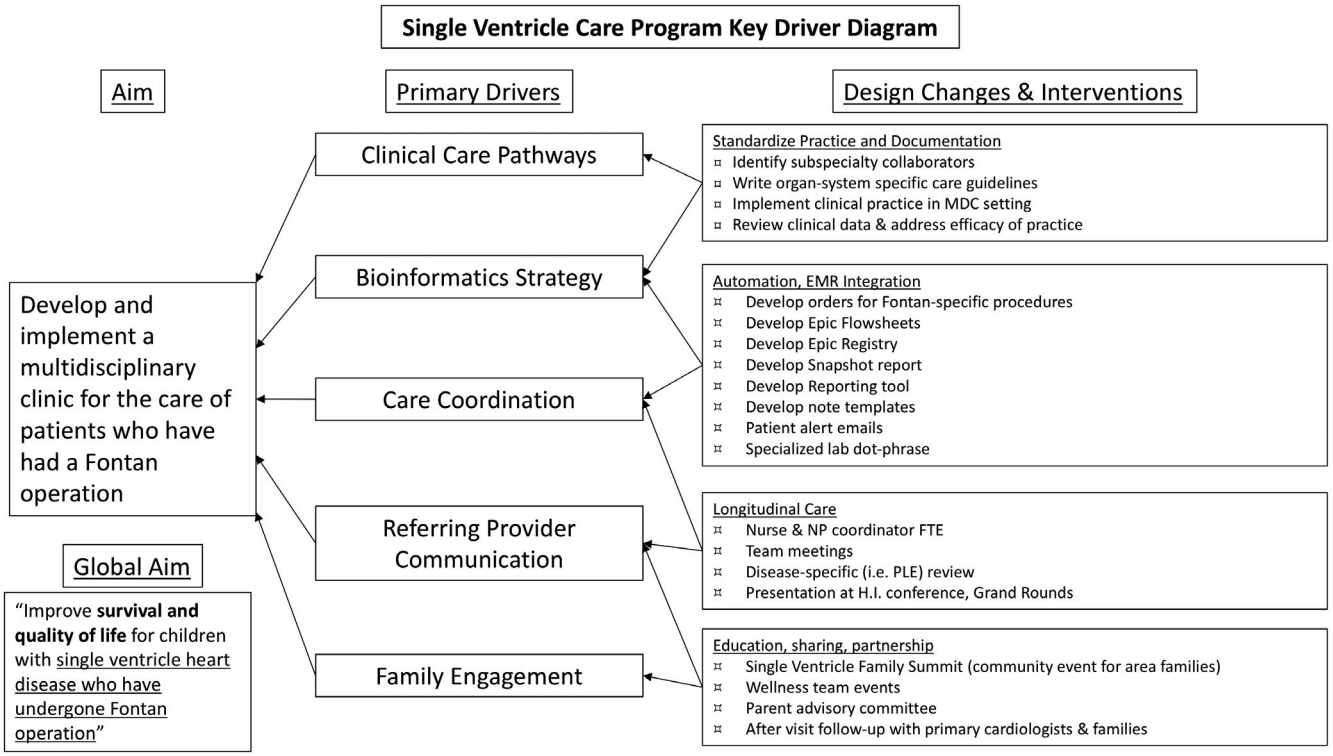

FIGURE 2 The single ventricle care program key driver diagram
Note: Global and specific aims are shown, with primary drivers and interventions.

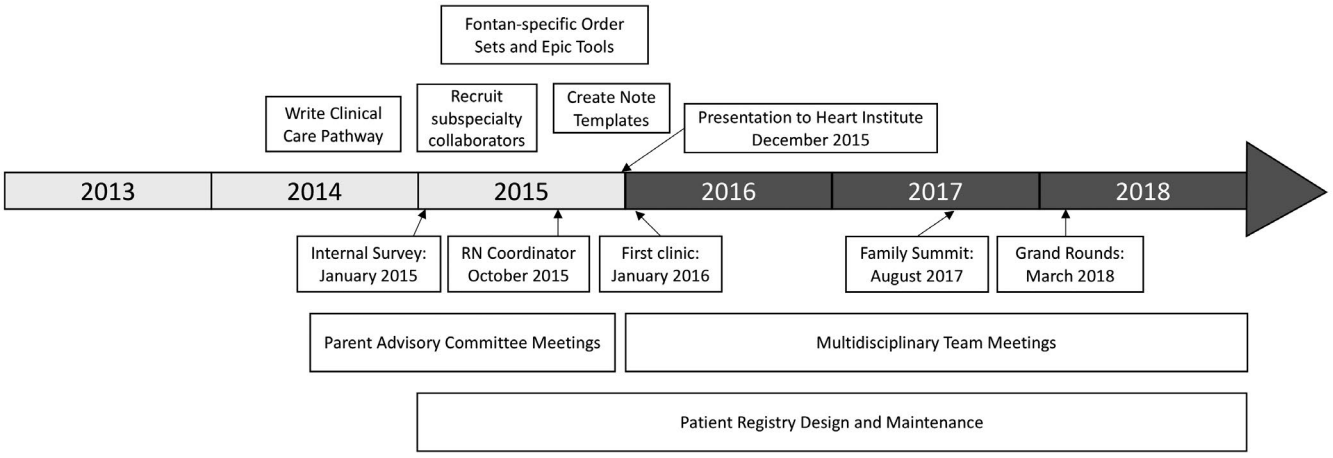

FIGURE 3 Single ventricle clinic timeline
Note: Much of the conceptual framework for the clinic as well as the supporting documentation for the care pathway were developed well before the clinic began. An internal survey of practice patterns and provider preferences was also helpful in creating a clinic that would serve the needs of the Heart Institute. Onboarding a nurse coordinator and recruiting subspecialty collaborators occurred in the latter half of 2015, along with creating note templates and other specialized components of the electronic medical record. A patient registry was created in parallel to the clinic and continues to be maintained. A family education and community building event, the Family Summit, was put on in the summer of 2017. The clinic results to date were presented at the Department of Pediatrics Grand Rounds in the spring of 2018.

2.4 | Outcome measures

Novel diagnoses were defined as pathology that was unknown prior to evaluation in the multidisciplinary clinic (MDC). Specifically, Holter monitors were reviewed for the presence of ectopy or pauses. Cardiopulmonary exercise test results were evaluated for both ectopy as well as sinus node dysfunction (reduced heart rate response to exercise). Echocardiograms, cardiac magnetic resonance imaging (CMR) studies, and cardiac catheterizations were reviewed for the presence of previously undocumented structural heart lesions.

TABLE 1 Recommendations for scheduled surveillance testing are indicated by diamonds in this table

Schedule for surveillance testing after Fontan operation													
	Labs*	ECG	Echo	Holter	CPET	cMRI	Cath	PFTs	Liver U/s	Liver Bx	Neuropsych Screening	Peds Psych Screening	MDC Visit
Fontan operation at 2-3 years of age													
4 y	◆	◆	◆	◆				◆				◆	
5 y	◆	◆	◆					◆	◆		◆	◆	◆
6 y	◆	◆	◆	◆				◆				◆	
7 y	◆	◆	◆					◆	◆			◆	◆
8 y	◆	◆	◆	◆	◆			◆			◆	◆	
9 y	◆	◆	◆					◆	◆			◆	◆
10 y	◆	◆	◆	◆	◆	◆	◆	◆		◆		◆	
11 y	◆	◆	◆					◆	◆		◆	◆	◆
12 y	◆	◆	◆	◆	◆			◆				◆	
13 y	◆	◆	◆					◆	◆			◆	◆
14 y	◆	◆	◆	◆	◆			◆			◆	◆	
15 y	◆	◆	◆					◆	◆			◆	◆
16 y	◆	◆	◆	◆	◆			◆				◆	
17 y	◆	◆	◆					◆	◆		◆	◆	◆
18 y	◆	◆	◆	◆	◆			◆				◆	
Transfer to adult congenital heart disease team													

Abbreviations: Bx, biopsy; Cath, cardiac catheterization; cMRI, cardiac magnetic resonance imaging; CPET, cardiopulmonary exercise test; ECG, electrocardiogram, Echo, echocardiogram, MDC, multidisciplinary; Neuropsych, neuropsychology; Peds, pediatric; PFTs, pulmonary function testing; U/s, ultrasound.
*Labs include a complete blood count, complete metabolic panel, Brain-type natriuretic peptide, Cystatin-C, and urinalysis.

TABLE 2 Recommendations for care of the patient with Fontan physiology

At the time of discharge after Fontan operation, the following should be implemented:
- ASA (3-5 mg/kg/day)
  - Therapeutic warfarin (goal INR 2-3) or enoxaparin (goal anti-10a activity 0.5-1) should be instituted for high-risk patients.

Routine follow-up visits on a semiannual or annual basis should include:
- History
  - Cough, expectorated casts, recurrent pneumonia, sudden respiratory distress, chest pain, lower extremity pain, and swelling
  - Abdominal fullness, peripheral edema, and diarrhea
  - Fractures, back pain, and scoliosis
  - Palpitations, exercise intolerance, and syncope
- Neuropsychological interview at any age and screening questionnaires in patients >4 years of age
- Health-Related Quality of Life screening questionnaire
- Broad screening questionnaire of Emotional and Behavioral Functioning
- Pulse oximetry
- Annual lab evaluation to include:
  - Brain-type natriuretic peptide
  - Complete blood count
  - Complete metabolic panel (including AST/ALT, GGT, alkaline phosphatase, albumin)
  - Prothrombin time
  - 25-(OH)-Vitamin D level
- Electrocardiogram
- Echocardiogram
- Spirometry, lung volumes

Every 2 years after Fontan operation:
- Holter monitor
- Stool alpha-1-antitrypsin
- Exercise testing with spirometry
- Liver ultrasound with Doppler and elastography

At the age of 8-12, if not already performed:
- Cardiac MRI
- Cardiac catheterization should be considered on an individual patient basis.
  Transjugular liver biopsy should be obtained during any cardiac catheterization and if there is evidence of significant hepatic fibrosis on imaging studies.

Cardiac catheterization reports were reviewed, and interventions performed were tallied.

Pulmonary function testing results were reviewed and the presence of obstructive or restrictive lung disease was noted, and clinical documentation was reviewed for sleep concerns and plastic bronchitis.

Abdominal ultrasound elastography results were reviewed, and those with a mean elastography value of >1.8 meters per second were deemed to have an abnormal result. In those who underwent liver biopsy, the surgical pathology reports and the congestive hepatic fibrosis score were reviewed. Portal hypertension was defined as an elevated hepatic venous wedge gradient on cardiac catheterization >3 mm Hg. Protein losing enteropathy was screened for on history in the electronic medical record and confirmed with a fecal alpha-1-antitrypsin. Malnutrition was defined as a weight for age z-score of −2.0. Anthropometrics were reviewed as a way of assessing for malnutrition and obesity.

Patients evaluated in the MDC undergo a consultation with a pediatric neuropsychologist to determine need for formal neuropsychology testing including a brief clinical interview, neurobehavioral examination, and parent-report rating measures of cognition and learning. For those patients seen for a formal neuropsychological evaluation, a diagnosis of developmental delay/intellectual disability, specific learning disability, language disorder, autism spectrum disorder, and/or attention deficit hyperactivity disorder were recorded.

Patients were seen for a health and behavior assessment as part of the consultation with the pediatric psychologist. Health-related quality of life (HRQoL) screening was conducted utilizing the Pediatric Quality of Life Inventory 3.0 Cardiac Module (PedsQL-Cardiac). Emotional and behavioral functioning was broadly assessed utilizing the Behavior Assessment System for Children, Third Edition (BASC-3). Clinical documentation was reviewed to indicate the number of patients screened, number of patients who scored in the impaired range on the PedsQL-Cardiac total score, and number of patients who scored in the at-risk or clinically significant range for difficulties on the BASC-3 scales.

2.5 | Analytic approach

The proportion of patients who had undergone Fontan operation who had undergone testing as described above were plotted on statistical process charts (SPC) P charts. Three sigma limits were used to set the upper and lower control limits; these were created using QI Charts Version 2.0.2.2 (Process Improvement Products, Austin, Texas). Standard SPC charting rules for determining special cause were used as evidence of improvement.[8] Of note, because these data are subject to autocorrelation, the only rule applied to determine special cause was a run of >8 points outside the control limits. Updated mean and control limits were plotted after special cause was detected.

3 | RESULTS

Eighty-six consecutive patients were included in the analysis, seen between January 1, 2016 and September 30, 2017. Subject characteristics are presented in Table 3. Novel pathology identified as a result of adherence to the care pathway and subsequent interventions that were performed is presented in Table 4.

3.1 | Process measure and outcome measures by system

3.1.1 | Cardiac

The proportion of patients who underwent cardiopulmonary exercise testing increased from 20.0% in the preintervention period to 48.9% post-care plan implementation (Figure 4A). There was no

TABLE 3 Demographics

Demographics	Total cohort *n* = 86
Female	40 (46)
Race	
African American	2 (2)
Asian	6 (7)
White	47 (55)
Other	16 (19)
Ethnicity	
Hispanic or Latino	31 (36)
Non-Hispanic	42 (85)
Missing	13 (15)
Prenatal diagnosis	42 (49) Missing 23 (27)
Genetic syndrome	
No	74 (86)
Yes	1 (1)
Unknown	9 (10)
Missing	1 (1)
Heterotaxy	8 (9)
Single ventricle type	
HLHS	31 (36)
Single RV (non-HLHS)	15 (17)
Single LV	40 (47)
HLHS type	
MS/AS	11 (13)
MS/AA	4 (5)
MA/AA	8 (9)
MA/AS	1 (1)
Initial shunt type	
No shunt	13 (15)
mBTS	31 (36)
RV-PA conduit	21 (24)
Other	2 (2)
Missing	19 (22)

significant change in the proportion of Fontan patients who had a cardiac MRI (Figure 4B). Fifteen subjects (17.6%) were found to have heart rhythm abnormalities and four (4.6%) previously undiagnosed structural heart disease. Twenty-seven patients underwent cardiac catheterizations (31.4%), resulting in 17 interventions (20.0%).

3.1.2 | Pulmonary

The proportion of patients who had PFTs more than doubled, from 20.7% in the preintervention era to 54.5% in the postintervention time frame (Figure 4C). Seven new diagnoses of obstructive lung disease (8%), and five cases of restrictive lung disease were identified (5.8%). Sleep-related abnormalities were discovered in 21 subjects (24.4%). No new diagnoses of plastic bronchitis were made. Interventions related to diagnoses of asthma and sleep abnormalities can be found in Table 4.

3.1.3 | Gastrointestinal (GI)

An increase in the number of patients that underwent liver ultrasound was noted (41.5% pre- and 79.5% postintervention) (Figure 4D). Liver ultrasound elastography was abnormal in 41 subjects (47.6%). For the 9 (10.4%) patients who underwent catheterization and liver biopsy, all had either liver fibrosis (8.1%) or cirrhosis (2.3%). Interestingly, none of the patients who underwent cardiac catheterization with measurement of hepatic venous wedge gradient had portal hypertension. Similarly, there were no new cases of protein losing enteropathy made based on historical screening or fecal alpha-1-antitrypsin measurement. Three (3.5%) diagnoses of malnutrition were made. Interventions for GI diagnoses can also be found in Table 4.

3.1.4 | Neuropsychological

The proportion of Fontan patients who had a formal evaluation by a neuropsychologist increased more than fivefold, from 3.7% to 19.1% postintervention (Figure 3E). Table 5 summarizes the results of neuropsychiatric screening . Fifty-eight subjects were referred for a formal neuropsychological evaluation (64.7%), and to date there have been 34 completed (39.5%). Intellectual disability or borderline IQ was diagnosed in nine patients (10.6%). Specific learning disability was diagnosed in 10 patients (11.6%). A diagnosis of language disorder was made in three cases (3.5%) and autism in three patients (3.5%). ADHD was diagnosed in eight cases (9.3%). Interventions pertaining to neuropsychological can be found in Table 4.

3.1.5 | Psychological

The proportion of patients evaluated by a pediatric psychologist increased from a mean of 11.5% to 50.7% after implementation of the MDC (Figure 4F). Fifty-five patients were administered the PedsQL-Cardiac during this visit, of which 28 (32.6%) scored in the impaired range for their total score (<71 out of 100). Fifty patients were administered the BASC-3, of which 15 (17.4%) scored in the at-risk range and 27 (31.4%) scored in the clinically significant range for difficulties on any of the subscale or composite scale scores. Psychological interventions in Table 4.

4 | DISCUSSION

Survival of patients with single ventricle heart disease palliated by way of a Fontan operation has increased, with an estimated 10-year survival greater than 95%; however, late complications, both cardiovascular and noncardiovascular, are frequent and decrease

TABLE 4 Novel pathology diagnosed at single ventricle multidisciplinary clinic and subsequent interventions

Novel pathology	*N* (%)		Intervention	*N* (%)
Cardiac				
Heart rhythm abnormality	15/86 (18%)			
			Medication change	1 (1%)
			Pacemaker	1 (1%)
Structural lesion	4/86 (5%)			
			Additional imaging	16 (19%)
			Underwent catheterization	38 (44%)
Pulmonary				
Asthma	7/86 (8%)			
			New medication	10 (12%)
			Asthma action plan	9 (10%)
Restrictive lung disease	5/86 (6%)			
Sleep abnormality	21/86 (24%)			
			Sleep study	13 (15%)
			Nasal rinse or steroids	10 (12%)
			ENT consult	3 (3%)
Liver/GI				
Abnormal elastography	41/86 (48%)			
			Referred for biopsy	17 (20%)
			Additional evaluation	2 (2%)
			New follow-up plan	6 (7%)
Protein losing enteropathy	0/86 (0%)			
			Medication change	5 (6%)
			Cardiac procedure	3 (3%)
			Dietician consult	10 (12%)
Malnutrition	3/86 (4%)			
			Dietician consult	10 (12%)
			GI Intervention	2 (2%)
Liver biopsy obtained	9/86 (10%)			
Neuropsychology				
	Total Cohort	Referred for Evaluation		
Learning disability	10/86 (12%)	10/34 (29%)	IEP or referral to learning specialist for intervention	16 (19%)
Intellectual disability or borderline IQ	9/86 (11%)	9/34 (26%)	IEP	6 (7%)
Attention deficit hyperactivity disorder	8/86 (9%)	8/34 (23%)	IEP, 504 plan, referral for medication, referral for behavioral intervention	7 (8%)
Language disorder	3/86 (3%)	3/34 (9%)	IEP, referral for outpatient speech/language therapy	7 (8%)
Autism	3/86 (3%)	3/34 (9%)	IEP, referral for behavioral intervention, outpatient speech/language therapy	2 (2%)
Psychology				
	Total Cohort	Referred for evaluation		
Impaired HRQoL	28/86 (32%)	28/55 (51%)	Referred for counseling	24 (28%)
Administered BASC-3	50/86 (58%)		Psychoeducation	86 (100%)

(Continues)

TABLE 4 (Continued)

Novel pathology	*N* (%)		Intervention	*N* (%)
	Total cohort	BASC-3	Therapeutic group	4 (5%)
At-risk difficulties (BASC-3)	15/86 (17%)	15/50 (30%)	Regimen adherence	3 (3%)
Clinically significant difficulties (BASC-3)	27/86 (31%)	27/50 (54%)	ACHD transition readiness	7 (8%)

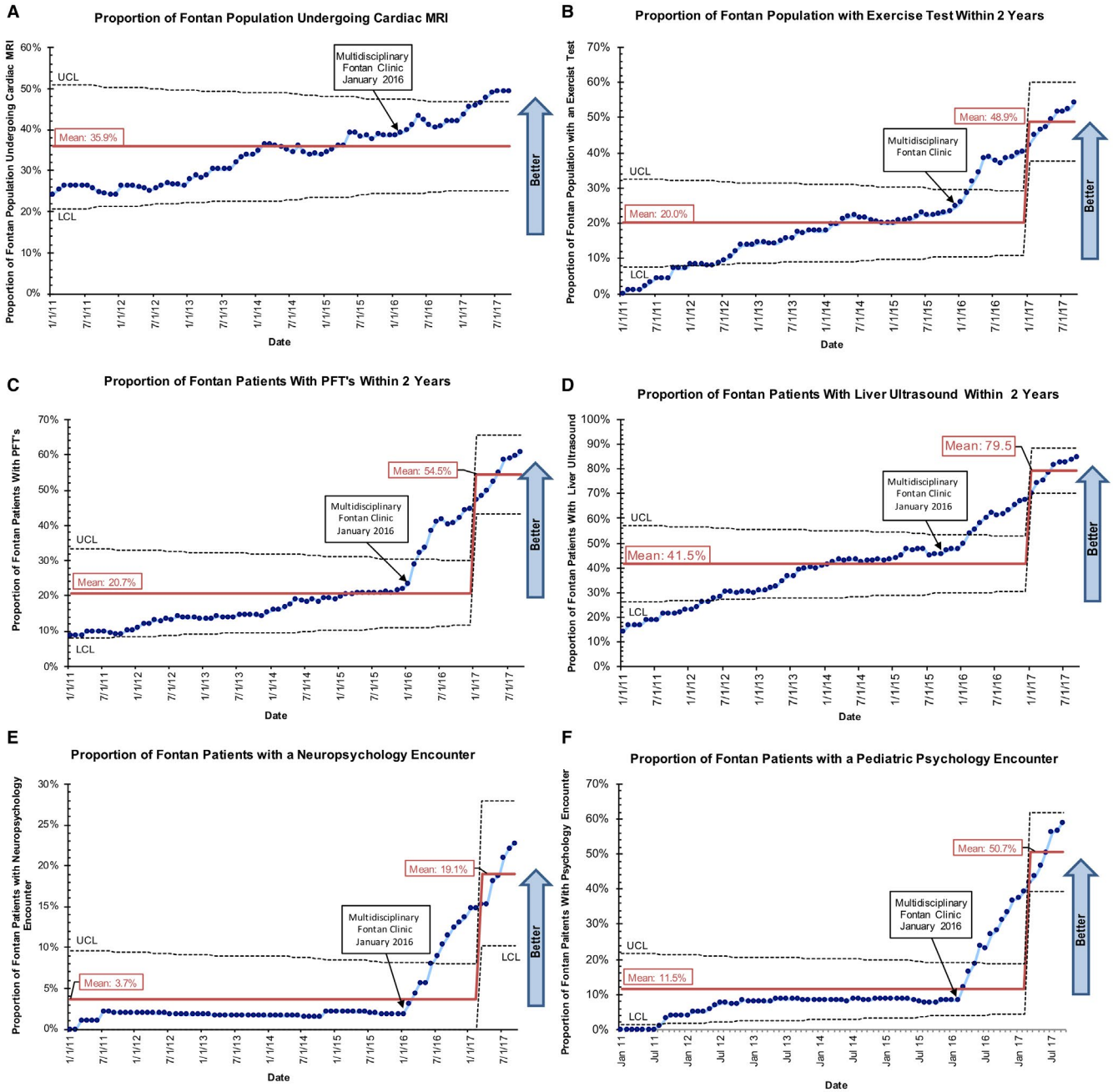

FIGURE 4 Composite SPC P chart showing the proportion of Fontan patients undergoing (A) cardiac magnetic resonance imaging, (B) cardiopulmonary exercise test, (C) pulmonary function testing, (D) liver ultrasound, (E) neuropsychology evaluation, and (F) psychology encounter

TABLE 5 Factors related to referral for neuropsychiatric evaluation

Referral for neuropsychological evaluation	Total patients (*n* = 86) *n* (%)
No referral	28 (32)
Referred and seen	34 (40)
Referred and not seen due to patient or family either not scheduling or keeping appointment	24 (28)

the quality of life for these patients, potentially contributing to premature death. We successfully implemented a care pathway, decreased practice variability, improved diagnosis of novel pathology, and facilitated interventions for cardiac and noncardiac end-organ dysfunction in this population. To our knowledge, this is the first description of clinical effectiveness of a dedicated multidisciplinary program in a relatively small, but medically complex, cohort of Fontan survivors.

Early identification of cardiac structural and cardiac rhythm abnormalities may improve overall cardiac function and other cardiac-related end-organ morbidities. We were able to increase compliance with cardiac imaging and exercise testing leading to cardiac catheterization interventions for 20% of our patients. Commensurate with the traditional care delivery model where children with SV heart disease were solely evaluated by their pediatric cardiologist every 6-12 months, discovery of novel cardiac pathology was rare. In contrast, diagnosis of previously unknown hepatic, pulmonary, neurocognitive, or psychological pathology was more prevalent, demonstrating the validity of this multidisciplinary approach. The inclusion of these particular subspecialists in the clinic was predicated on prior work.[9-11] Our care pathway also includes screening for renal, endocrine, and hematologic pathology, though these subspecialists do not personally participate in the MDC visits. The importance of institutional support for such an endeavor cannot be overstated, as the ability convene a multispecialty clinic draws heavily on institutional resources, and is a considerable time commitment for the involved subspecialists.

Patients with single ventricle physiology are faced with unique physical, emotional, and intellectual challenges that impact their psychosocial functioning and HRQoL across home, school, and social domains. Although identification of end-organ disease is important, equally important is identification of neuropsychological and behavioral challenges these patients and families are may be facing. Adolescents with SV show a threefold increase for psychiatric problems and have increased susceptibility to anxiety, disruptive behavior, and depressive disorders.[12] Indeed, neuropsychological difficulties are predictive of long-term outcomes in adulthood for individuals with SV, including delayed acquisition of independence, increased risk for comorbid psychiatric disorders, and suboptimal social and vocational outcomes.[13]

As depicted in our KDD, referring providers play a central role in patient flow into the MDC, and so creating an environment in which cardiologists in our group felt comfortable sending patients to the MDC continues to be paramount. Prior to our first clinic, the MDC was presented to the Heart Institute at a weekly didactic conference, with emphasis on the fact that we would operate as a consultant service, and not assume care of the patients unless desired by the referring provider. Subsequently, a strong effort to include referring providers in discussions regarding treatment decisions runs through every interaction with patient families. For families traveling from large distances away, we have made an effort to arrange testing in a way that accommodates travel schedules and clinic visits in other specialties. We have also reached out to pediatric cardiologists in surrounding communities to help determine regionally specific patient needs, disseminate information about our care pathway and clinic, as well as elicit feedback in an ongoing fashion. Many services and tests, such as ultrasound elastography and neuropsychological testing, are not available in local communities, placing an emphasis on care coordination and information exchange between our team and area cardiologists.

4.1 | Limitations

Due to CHCO's catchment area (7-state region in the mountain west), we were only able to implement these process and outcomes measures at our main campus in Aurora, Colorado. Challenges related to this distance include limited access to specialized equipment (eg, ultrasound elastography) and obtaining laboratory studies which may require special collection or analysis (eg, stool alpha-1-antitrypsin). Our approach for families traveling from out of town is to ask them to arrive a day early for testing, so that our team has access to test results on the morning of the MDC visit. The exceptions to this approach are the neuropsychology evaluations, which often require a full day of testing. The process for interacting with referring providers outside the Denver area is similar to those inside our institution.

Our MDC, located in a quaternary care hospital with multispecialty expertise available, is not typical of most community practices. Though the generalizability of this approach is problematic, use of telemedicine may allow referring providers to obtain recommendations for testing and appropriate follow-up. There was a trend toward improvement in the frequency of testing immediately prior to the initiation of the MDC in some cases, and it is likely that the Hawthorne effect played some role because the MDC was being discussed and created during this time. These trends did not meet criteria for special cause or an adjustment of the mean. Pulmonary function testing and cardiopulmonary testing could not be performed in children less than 4 years of age and in any children who were nonambulatory and/or could not follow directions. Some children did not complete all recommended testing due to time constraints. Administration of

HRQoL screener was limited in that the PedsQL-Cardiac, as other cardiac-specific HRQoL measures (eg, Pediatric Cardiology Quality of Life Inventory[14]) have not been validated in non-English speaking populations.

5 | CONCLUSION

Children and young adults who have undergone Fontan operation are at risk for developing cardiovascular, hepatic, pulmonary, neurodevelopmental, and psychological comorbidities, among others. A multidisciplinary clinic and development and adherence to a care pathway at our center encouraged a unified approach to testing and resulted in the diagnosis of significant novel pathology. Future work should focus on defining new quality metrics which can be applied broadly.

CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest with the contents of this article.

AUTHOR CONTRIBUTIONS

Concept/Design, *Data analysis/interpretation*, *Drafting article*, *Critical revision of article*, *Data collection:* Di Maria
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