Cardiology in the Young

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Developing an adolescent and adult Fontan Management Programme

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Abstract

Patients with single-ventricle CHD undergo a series of palliative surgeries that culminate in the Fontan procedure. While the Fontan procedure allows most patients to survive to adulthood, the Fontan circulation can eventually lead to multiple cardiac complications and multi-organ dysfunction. Care for adolescents and adults with a Fontan circulation has begun to transition from a primarily cardiac-focused model to care models, which are designed to monitor multiple organ systems, and using clues from this screening, identify patients who are at risk for adverse outcomes. The complexity of care required for these patients led our centre to develop a multidisciplinary Fontan Management Programme with the primary goals of earlier detection and treatment of complications through the development of a cohesive network of diverse medical subspecialists with Fontan expertise.

Given the growing numbers of patients with a Fontan circulation reaching adulthood, there is a need for standardised protocols for monitoring and management of the multiple cardiac and non-cardiac sequelae of the Fontan circulation. Further, there is a window of time during which intervention (e.g., medical or procedural) provides substantial benefit. Metastasis of hepatocellular carcinoma and progression of multi-organ disease that precludes transplantation are examples, where late recognition of disease is associated with limited treatment options and substantially worse outcomes.[1]

Adults with a Fontan commonly have other acquired health conditions requiring adult subspecialty care that make it difficult to care for these patients solely within a paediatric institution. Care delivery in an adult hospital setting, however, is complicated by the general lack of highly specialised, single-ventricle CHD caregivers. This fragmentation of care may contribute to Fontan patients getting referred too late for potential life-sustaining interventions, surgeries, mechanical circulatory support, or heart transplantation.

Over the past decade, several congenital heart centres have developed multidisciplinary clinics to provide standardised, subspecialty Fontan care.[2] Most have focused primarily on children. The burden of morbidity and mortality in single-ventricle disease peaks early around the time of the initial surgeries and then later increases again, more insidiously, in late adolescence and adulthood.[3] This risk starts to increase at the time patients are transitioning care from paediatric to adult providers, a time frequently associated with gaps in medical care.[4] To address this

shortcoming, we sought to formalise a multidisciplinary adolescent and adult Fontan Management Programme to ensure the best possible patient care and outcomes through continuity of care, high-quality cardiac and extra-cardiac organ surveillance, and comprehensive preventive care. We envisioned a Fontan management team composed of specialists from cardiology, hepatology, radiology, cardiac surgery, general surgery, haematology, nephrology, pulmonology, endocrinology, and pharmacy. These specialists would work together to develop screening protocols and care pathways to recognise early decline or Fontan-related complications, identify preventative mechanisms, facilitate prompt treatment, and streamline referral for transplant evaluation or advanced cardiac therapies. Ideally, diverse specialty care and testing could be provided with fewer clinic visits and reduced patient cost. The programme's initial goals over the first 3 years included: (1) to improve screening for complications of Fontan-associated liver disease, including portal hypertension and hepatocellular carcinoma; (2) to provide appropriately timed referrals for mechanical circulatory support and/or heart transplantation; and (3) to develop a combined heart and liver transplant programme.

Methods

Challenges in implementation of the programme

The establishment of our Fontan Management Programme and the achievement of our goals required significant start-up funding and institutional support. This support was provided through a Cincinnati Children's Hospital Medical Center Academic and Research Committee grant that supported the programme through its first 3 years until stability was achieved. At the outset, we aimed to become self-sustaining through the generation of new referrals, imaging, procedures/surgeries, and performance of combined heart and liver transplants.

Bridging the gap between paediatric and adult care was another significant challenge faced, both as it related to the routine care of complex adult patients largely within a paediatric institution and as it related to establishing the capability to perform combined heart–liver transplants. This required the identification of paediatric subspecialists who were comfortable caring for adults and required the identification of subspecialists from our affiliated adult hospital to be credentialed and able to consult and perform procedures at our children's hospital when needed.

Getting buy-in from local practitioners was another hurdle faced by the programme. Practitioners across multiple subspecialties were already caring for patients who had undergone the Fontan procedure and had established care patterns. Demonstrating the benefit of the multidisciplinary clinic and justifying proposed screening protocols were important efforts as the programme was being established. As the programme has grown, we have accommodated both the transition of care of patients entirely to the Fontan programme and single or periodic consultations while the patient maintains care with the primary cardiology or subspecialty team. Patients that are co-managed with a primary cardiologist often alternate visits every 6 months between the Fontan Management Programme and their primary cardiologists.

Design of the Fontan Management Programme

The proposed Fontan Management Programme was discussed with multiple adult patients with a Fontan circulation who gave valuable feedback on how such a programme could benefit them. With this feedback, the multidisciplinary Fontan Management Programme was designed around: (1) A core and diverse group of enthusiastic subspecialists with interest in the Fontan circulation and its complications who would: (A) Lead development of the clinical and research programs and (B) Develop surveillance recommendations, and diagnostic and therapeutic algorithms; (2) A multidisciplinary clinic supported by: (A) A dedicated Fontan nurse care manager and (B) Weekly Fontan care meetings where all team members review patients together to develop consensus on the best course of action; (3) Establishment of a comprehensive patient registry to track progress, support quality improvement, and research; and (4) Formalising a combined heart and liver transplant programme.

Implementation of the Fontan Management Programme

The Fontan Management Programme began in January, 2018, coincident with the award of the Cincinnati Children's Hospital Medical Center Academic and Research Committee grant. This provided protected time for core team members to develop key elements of the Fontan Management Programme, including standardised surveillance practices (Table 1), establishment of the physical clinic, and development of patient resources. The Academic and Research Committee grant also allowed the programme to fund a nurse care manager, which is identified as the most crucial role for the ultimate success of the clinical programme. This individual oversees the management of care for patients in the programme and organises multiple, same-day subspecialist visits, test scheduling, and follow-up with patients.

In January, 2018, we started to hold once monthly full-day dedicated clinics. Based on increased volume, the frequency of these clinics increased to twice per month in 2019. Simultaneously, we started weekly Fontan Clinic Operations meetings to streamline and standardise care. In this forum, we review and discuss upcoming clinic patients, current inpatients, strategies to improve clinical care, and ongoing research projects. This weekly meeting is led by the Fontan nurse care manager and is regularly attended by programme cardiologists (including a cardiologist with expertise in transplantation and mechanical circulatory support), hepatologists, radiologists, and cardiology fellows, with other subspecialists participating as needed (Table 2). In this meeting, the Fontan nurse care manager summarises and presents the patients scheduled to be seen in the near future, including an overview of cardiac anatomy, recent health problems, current symptoms, medications, and the results of recent laboratory testing. Cardiologists present results of recent echocardiograms, exercise testing, advanced cardiac imaging – CT/MRI, invasive haemodynamics, and angiography. Radiologists with expertise in Fontan-associated liver disease review recent (and historic as relevant) cross-sectional liver imaging, estimating the degree of congestion/fibrosis and presence of portal hypertension, and identifying liver lesions of potential concern. All presented results are discussed as a group to define the appropriate subspecialists to see the patient at the next clinic visit and suggested workup and testing, including which patients should undergo targeted liver lesion biopsy (Fig 1).[5]

On the day of a patient's Fontan clinic visit, they have same-day testing that typically includes cardiac studies, liver imaging, and laboratory testing. Patients see a cardiologist at every Fontan Management Programme clinic visit and a hepatologist at baseline and at all visits if they have any liver pathology that requires expert input such as a liver lesion of concern, rapidly developing fibrosis, or protein-losing enteropathy. If, during the weekly meetings, the team decides that a patient would benefit from additional

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Table 1. Cincinnati Children's Hospital working consensus screening for adolescent and adult patients with a Fontan circulation

Age (years)	Frequency of follow-up	ECG	Echo	Cardiac MRI/ CT	Holter	Cardiac Cath	Liver US with ARFI*	Liver MRI with MRE*	QOL	PFTs†	Labs‡	Aspirin resistance §	GXT with PVP
Below 12	q 1yr	q1yr	q1yr	One time	q3yr	One time post-Fontan	One time	One time	One time	As needed	One time	One time	One time
≥ 12	q1yr	q1yr	q1yr	q3yr	q3yr	10 yrs after last cath	q2yr	q2yr	q1yr	Baseline	q2yr	One time	q2yr
>18	q6 mo–q1yr	q1yr	q1yr	q3yr	q2yr	q10yr◇	q1–2yr	q2yr	q1yr	q3–5 yr	q1yr	One time	q2yr

ARFI=Acoustic Radiation Force Impulse; ASA=Aspirin; Cath=catheterisation; CT=computerised tomography; ECG=electrocardiogram; Echo=echocardiogram; GXT=graded exercise test; mo=months; MRE=Magnetic Resonance Elastography; MRI=magnetic resonance imaging; PVP=peripheral venous pressure monitoring; QOL=quality of life assessment; US=ultrasound; yr=year

*Alternate liver ultrasound and MRI. Liver ultrasound does not require Doppler and liver MRI should include contrast (e.g., Gadoxetate disodium). A CT with contrast may be required in patients with epicardial pacemaker leads. In this scenario, the frequency of liver imaging may need to be adjusted to limit radiation exposure

†Spirometry, lung volumes, respiratory muscle strength, and diffusion

‡Labs include complete blood count, renal profile, hepatic profile, GGT, cystatin C, alpha-fetoprotein, NT Pro BNP, stool alpha-1-antitrypsin

§Only inpatients taking aspirin

◇Consider a transjugular liver biopsy during the heart catheterisation

subspecialty care, these subspecialists will also see the patient alongside the cardiologist. This includes potential consultation with a heart failure/transplant cardiologist based on The Advanced Cardiac Therapies Improving Outcomes Network proposed criteria.[6] The cardiology and subspecialist visits are billed under the Heart Institute, which compensates the other divisions for the specialist's time. The patient is only charged one facility fee, rather than the two facility fees that they would incur if they were to have separate visits in other departments.

Patients followed by the Fontan Management Programme that require inpatient care are admitted to the paediatric cardiology service with consultation from the adult congenital heart team. The Fontan team is available for inpatient consultation of patients who are not followed in the Fontan Management Programme at the admitting cardiologist's discretion.

Starting in early 2018, multiple meetings were held between key stakeholders in a planned combined heart and liver transplant programme designed to care for patients who had undergone the Fontan procedure. This included members of the core Fontan Management team as well as practitioners from the affiliated adult hospital including cardiothoracic surgery, liver transplant surgery, and adult heart failure transplant cardiology. To enable this programme, a retainer, initially supported by ARC funding and subsequently by the Division of Cardiology, was established to support ongoing efforts by the adult care providers. Substantial effort was invested in establishing policies, processes, and procedures for heart–liver transplant in a Fontan patient that would facilitate seamless care between the paediatric and adult teams and that would facilitate transfer of care as needed. It was determined that, given the complexity of the cardiac anatomy in a patient with a Fontan, the combined heart and liver transplant surgery would occur within the children's hospital with the post-operative care in the Children's Hospital Cardiac ICU. The adult subspecialists were contracted to participate in the post-operative care of these adult patients at the children's hospital. As the combined heart and liver transplant recipients recovered and were ready to transfer from the Cardiac ICU, a case-by-case decision would be made to either remain in the children's hospital or transfer to the adult hospital, where they would complete their hospital course and begin their follow-up at the adult institution.

Results

Success of the programme

The Fontan management clinic experienced significant growth since initiation. Outpatient clinic visits increased from 71 in year one, to 118 in year two, and to 138 in year three (Fig 2a). The aforementioned standardised surveillance protocol was created and included most of the basic and in-depth surveillance suggestions made by the American Heart Association Scientific Statement (Table 1).[7] This surveillance protocol was presented to the Heart Institute faculty for widespread adoption. Consistent with this protocol, the percentage of patients seen in Fontan clinic with cross-sectional liver imaging within a year of their most recent appointment increased from 68 to 84% (Fig 2b), and the per cent of patients who had liver imaging in the prior 2 years increased from 79 to 90%. Between January, 2018 and January, 2020, there were 88 liver MRI examinations performed (≥12 years old) for screening of Fontan-associated liver disease and focal liver lesions. This is compared to 53 performed in the 2 years before the start of the Fontan programme. From January, 2018 to 1 June, 2020, there

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Table 2. Members of the Fontan management team and their roles.

Nurse care manager	- Leading weekly care meetings - Scheduling visits and testing - Creating an environment of a cohesive medical home
Cardiology	
Paediatric cardiologist	- Cardiac evaluation of patients aged < 18
Adult congenital cardiologist	- Cardiac evaluation of patients aged ≥ 18
Cardiac imaging	- Interpreting imaging (e.g., echo, cardiac CT, or MRI) - Guidance on protocols (e.g., pulmonary embolism evaluation)
Interventional cardiologist	- Cardiac catheterisations including haemodynamics and Fontan conduit and/or pulmonary artery stenting
Electrophysiologist	- Management of arrhythmias - Risk stratification for implantable cardioverter–defibrillators
Heart failure/transplant cardiologist	- Evaluating patients with Fontan circulatory failure for mechanical support or transplantation
Pulmonary hypertension specialist	- Management of pulmonary vasodilator medications
Cardiac surgeon	- Fontan conversion/revision surgery - Performing heart transplants
Hepatology/liver transplant team	- Evaluation of Fontan-associated liver disease - Management of liver lesions/hepatic neoplasms
Pathologist	- Interpreting liver biopsies within context of a Fontan circulation
Pulmonologist	- Management of respiratory muscle weakness, haemoptysis, and plastic bronchitis
Endocrinologist	- Management of abnormal bone density
Nephrologist	- Evaluation and management of chronic kidney disease
Maternal-Fetal Medicine physician	- Preconception counselling - Pregnancy management
Gynaecologist	- Counselling and management of contraception - Management of dysfunctional uterine bleeding
Pharmacist	- Anticoagulation management - Dosing, monitoring, and evaluating medications
Sleep medicine physician	- Evaluation for obstructive sleep apnea - Assessment of risks of positive airway pressure
Hematologist	- Management of coagulopathy
Dietitian	- Optimise weight gain/growth in paediatrics - Aid in limiting excessive weight gain in older patients
Exercise physiologist	- Improve exercise performance and alleviate symptoms of exercise intolerance - Improvement in sarcopenia
Psychologist /Psychiatrist	- Management of disease-related anxiety, post-traumatic stress disorder, depression, or other psychological and psychiatric diagnoses
Social worker	- Health insurance guidance - Aid in navigating the healthcare system
Business manager	- Keeping a dashboard of patient visits, testing, and billing - Measuring financial impact of the programme
Research coordinator	- Development of a data registry - Enrolling patients in research studies

were eight targeted liver biopsies for high-risk focal liver lesions performed in five patients. This is compared to two targeted liver biopsies in the prior two and a half years. Two patients were diagnosed with hepatocellular carcinoma, which was discovered during screening in both patients. One of these patients underwent Yttrium-90 radioembolisation treatment and successfully underwent *en bloc* heart and liver transplantation. The other patient underwent microwave ablation of the hepatocellular carcinoma and is currently listed for combined heart and liver transplants.

Prior to the development of our Fontan Management Programme, there were no patients evaluated at our hospital for combined heart and liver transplants. Between January, 2018 and June, 2020, six adult patients have been evaluated for combined heart and liver transplants, and two have been evaluated for heart transplant alone. All eight of these patients are still living; two have had a successful *en bloc* heart and liver transplant, one had a successful heart transplant, three patients remain on the waitlist, one patient was declined due to high

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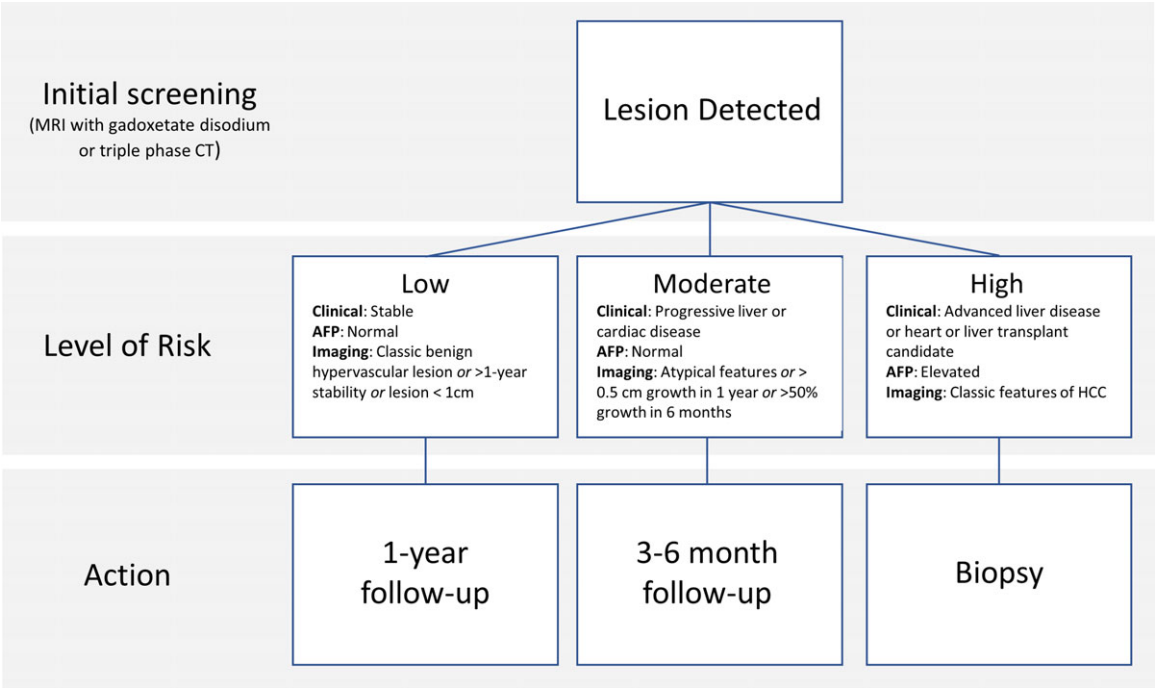

Figure 1. Local working guidelines for the management of focal liver lesions in patients with a Fontan circulation. Moderate risk lesions that show continued growth are referred for biopsy.
AFP = alpha-fetoprotein; HCC = hepatocellular carcinoma. Adopted from (Dillman et al, Imaging of Fontan-Associated Liver Disease, Pediatr Radiol (2020)).

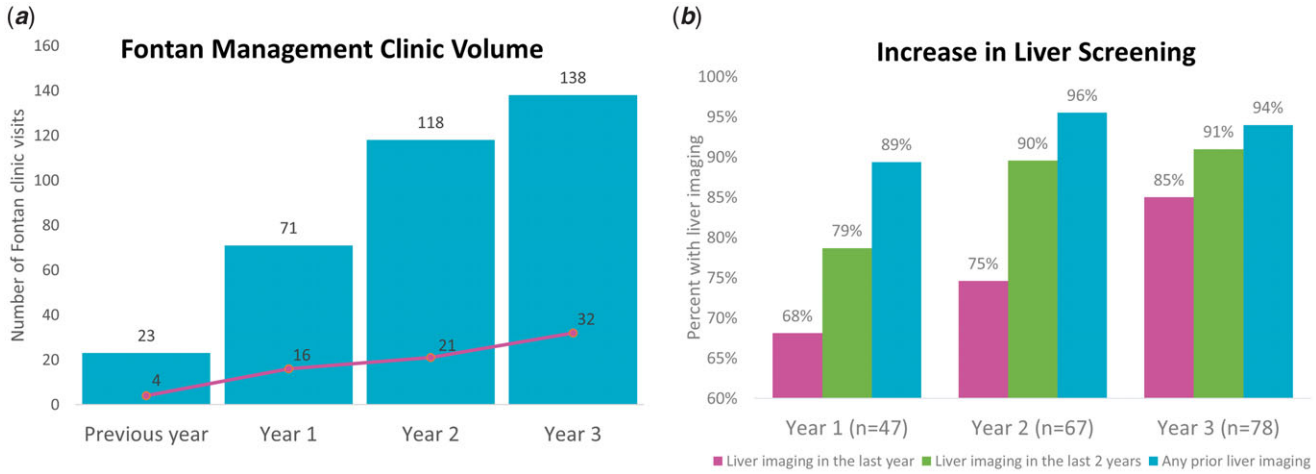

Figure 2. **(a)** Annual Fontan clinic outpatient visit volume. Year 1 refers to the first year of the funded Fontan Management Programme. The pink line depicts the number of regional/national patients seen outside of Cincinnati Children's primary or secondary service area. **(b)** Increase in liver screening since the start of the Fontan Management Programme. The first column (pink) represents the per cent of patients who had liver imaging within the last year at the time of their clinic visit, the second column (green) is the per cent with liver imaging within 2 years of their clinic visit, and the third column (blue) is the per cent of patients who have had any prior liver imaging regardless of the timeframe. Note that there were obstacles in performing routine outpatient follow-up and liver imaging during year 3 due to COVID-19.

panel-reactive antibodies, and one patient was declined due being too well for listing. In the 3 years preceding the Fontan programme, there were no ventricular assist devices implanted in adults with a Fontan. Since the Fontan programme began, four patients have been successfully discharged from the children's hospital with a HeartMate 3 ventricular assist device, and one patient is awaiting device placement. One of the adults with a ventricular assist device had a successful combined heart and liver transplant, one had a successful heart transplant, and the other two have a ventricular assist device as chronic therapy.

Discussion

CHD patients who have had a Fontan procedure have a circulation that, while it does not require a sub-pulmonary ventricle, does so at the expense of systemic venous hypertension and a relatively low cardiac output state. Consequently, this can lead to multi-organ dysfunction with an increased risk of early and late morbidity and mortality.[8] With the increasing age of the Fontan population, redesigning how clinical care is delivered may facilitate the management of the complex care needs of this population and

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eventually lead to improved quality of life. We described the steps used to develop the Fontan Management Programme at our paediatric institution to care for adolescents and adults with a Fontan circulation.

Summary of the experience and future directions

Our multidisciplinary adolescent and adult Fontan programme was developed within a paediatric hospital in close collaboration with key subspecialties and adult medicine colleagues. Initial start-up funding was necessary to provide dedicated time for expertise to develop across disciplines, decrease the barriers to collaboration, and allow the trust to develop amongst key members of the team. As a result, the patients had more streamlined care and an increase in surveillance testing. Since the start of the programme, we have standardised and increased screening for hepatocellular carcinoma and identified two cases of hepatocellular carcinoma early in the disease course, which would otherwise have been undiagnosed. We have referred six patients for evaluation, and successfully performed two *en bloc* heart and liver transplants and have implanted a VAD in four patients with progressive deterioration.

We anticipate patient volume will continue to grow as clinicians locally and regionally gain awareness. We aim to focus on improving care via developing additional quality metrics such as optimal frequency of cardiopulmonary exercise testing, quality of life assessments, and interventions to improve physical fitness. Additionally, we are one of the pilot centres that will soon begin contributing to the Fontan Outcomes Network, a United States Fontan registry and learning network.[9] We hope to harness the power of this multicentre collaboration to improve the care of our patients.

Translation to other settings

The steps in developing a Fontan Management Programme depend significantly on the local infrastructure (adult hospital versus paediatric hospital versus combined adult and paediatric hospital) and the availability of divisional support and/or intra or extramural funding to initiate such a programme. In programmes where start-up funding is limited, the development of a core group of enthusiastic care team members can be successful. Having baseline data and showing data of improvement in screening and revenue generation can be very helpful to demonstrate value. A Fontan programme champion (in this case, a nurse care manager) dedicated to the success of the programme is fundamentally important. We believe that this innovative care model can be implemented in different settings, but must be tailored to the characteristics of each practice and hospital, based on the available resources.

Conclusions

We developed an adolescent and adult Fontan Management Programme within a paediatric hospital and demonstrated increases in liver screening that led to the diagnosis of hepatocellular carcinoma and to the development and successful initiation of a heart and liver transplant programme. Ultimately, the value of this programme will be realised if we demonstrate improved outcomes and quality of life with lower healthcare costs.

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Conflicts of interest. None

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