10-1-2022

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Cardiology in the Young

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Review

Cite this article: Ronai C, Batlivala SP, Nguyen Q-T, Renno MS, Arthur L, Glickstein J, Johnson JN, Miller JR, Altman CA, and Selamet Tierney ES (2022) Paediatrician's guide to post-operative care for functionally univentricular CHD: a review. *Cardiology in the Young* **32**: 1547–1553. doi: 10.1017/S1047951122002943

Received: 11 January 2022
Revised: 8 August 2022
Accepted: 25 August 2022
First published online: 23 September 2022

Keywords:
Cardiac surgery; primary care; post-operative care; single ventricle

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Abstract

Importance Single ventricle CHD affects about 5 out of 100,000 newborns, resulting in complex anatomy often requiring multiple, staged palliative surgeries. Paediatricians are an essential part of the team that cares for children with single ventricle CHD. These patients often encounter their paediatrician first when a complication arises, so it is critical to ensure the paediatrician is knowledgeable of these issues to provide optimal care. *Observations* We reviewed the subtypes of single ventricle heart disease and the various palliative surgeries these patients undergo. We then searched the literature to detail the general paediatrician's approach to single ventricle patients at different stages of surgical palliation. *Conclusions and relevance* Single ventricle patients undergo staged palliation that drastically changes physiology after each intervention. Coordinated care between their paediatrician and cardiologist is requisite to provide excellent care. This review highlights what to expect when these patients are seen by their paediatrician for either well child visits or additional visits for parental or patient concern.

Single ventricle CHD encompasses a wide variety of anatomic and subsequent treatment sequelae. Single ventricle cardiac conditions include hypoplastic left heart syndrome, tricuspid atresia, double inlet left ventricle, unbalanced atrioventricular canal defects, and hypoplastic right heart lesions. Increasingly, these lesions are diagnosed prenatally, and standardised newborn CHD screening has helped improve outcomes.[1–3] Children with single ventricle anatomy and physiology are managed through a series of staged palliative interventions with the ultimate goal of separating the pulmonary and systemic circulations.[4] These interventions typically take place immediately after birth and continue over the course of the child's life. Children with these conditions see their paediatrician and cardiologist frequently and, while there is much information published about the importance of multi-disciplinary care and empowering caregivers, there is a dearth of information targeted specifically for their primary paediatric providers, especially following each palliative intervention.[5] Care for these children requires a coordinated approach between their cardiologist and paediatrician – to ensure they have an effective medical home.

Overview of care for children with single ventricle physiology

1. Initial post-natal interventions (Norwood, Ductal Stent, Blalock-Taussig-Thomas shunt, Pulmonary Artery Band)

All unpalliated single ventricle heart disease patients share the following physiologic characteristics. First, there is mixing of the systemic and pulmonary venous blood such that oxygen saturation is 75–85%. Second, the single ventricle pumps blood to both the body and lungs and thus is working harder than a biventricular circulation. This increased strain and decreased reserve is the primary reason why children with unpalliated single ventricle physiology are so fragile. And finally, the circulatory physiology is most stable when the systemic and pulmonary blood flows are appropriately balanced (typically reflected by oxygen

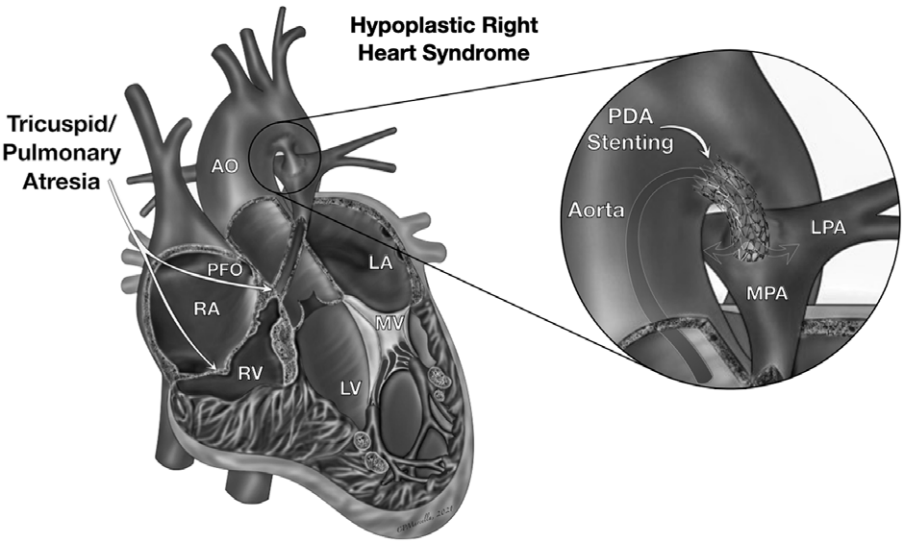

Figure 1. The anatomy of a hypoplastic right heart with an underdeveloped right heart, with pulmonary atresia and/or tricuspid atresia with a stent in the ductus arteriosus (PDA stenting). RA= right atrium, RV = right ventricle, PFO = patent foramen ovale, AO = aorta, LA = left atrium, MV = mitral valve, LV = left ventricle, LPA = left pulmonary artery. MPA = main pulmonary artery.

saturations of 75–85%). Too much or too little flow to one circuit places the child at risk of decompensation.

Depending on the cardiac anatomy, the first intervention for a newborn with single ventricle anatomy focuses on establishing reliable and balanced sources of pulmonary and systemic flow. If the native left ventricle and aorta are adequate, the infant usually requires only a stable source of pulmonary blood flow, achieved by placing either a stent in the ductus arteriosus or by surgically placing an aorta-to-pulmonary artery shunt (e.g., Blalock-Taussig-Thomas or central shunt) (Fig 1).

If the native left heart and/or aorta are inadequate (as in hypoplastic left heart syndrome), a Norwood operation with either a Blalock-Taussig-Thomas or Sano shunt (right ventricle to pulmonary artery) is performed to establish adequate pulmonary and systemic blood flow (Fig 2). Infants often require a prolonged recovery in the ICU and then cardiac ward. The "Hybrid Stage 1" is another technique that involves placing a ductus arteriosus stent for systemic blood flow and applying bands to each pulmonary artery to balance the pulmonary and systemic circulations.[6] This procedure is done without cardiopulmonary bypass, so some centres routinely perform it while other utilise this technique for patients with significant co-morbidities that may render them too fragile to undergo the more extensive Norwood operation.[6]

Some newborns with single ventricle anatomy have adequate systemic and pulmonary outflow tracts at birth, but only one side is capable of pumping effectively (e.g., tricuspid valve atresia with a large ventricular septal defect and normal aortic/pulmonary valves). As a result, these babies can have "unrestrictive" flow to their lungs which places them at risk for pulmonary over-circulation and potentially inadequate systemic flow. For these children, pulmonary blood flow needs to be restricted, typically with a pulmonary artery band, to balance the systemic and pulmonary circulations.

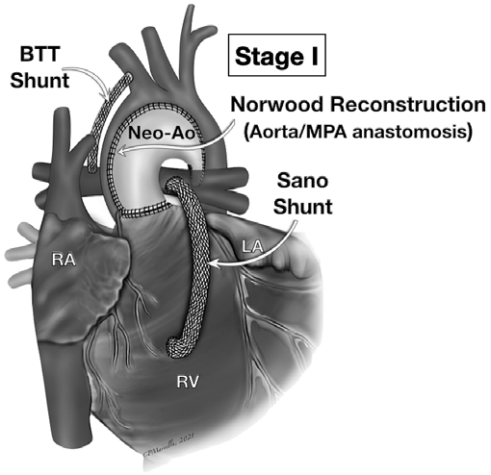

Figure 2. Norwood operation diagram.
Stage 1 operation depicting the Norwood operation pulmonary blood flow being supplied by either a right ventricle (RV) to pulmonary artery conduit (Sano Shunt) or Blalock-Taussig-Thomas (BTT) shunt from the innominate artery to the right pulmonary artery. RA = right atrium, RV = right ventricle, LA= left atrium, Neo-Ao = neo aorta, MPA = main pulmonary artery

2. Interstage phase

Home monitoring

The "interstage" period between the first (when performed) and second major operations typically lasts until 4–5 months of age and is a time associated with significant morbidity and mortality.[7] Those risks are related to the burden placed upon the single ventricle to maintain both the systemic and pulmonary circulations after the initial palliation. So, infants in the interstage period are often monitored closely by multi-disciplinary interstage teams, which has led to improved outcomes.[7–9] Home monitoring teams closely follow numerous data points such as daily weight and oxygen saturation. Families remain in close contact with the home monitoring team and are trained to identify markers of adverse outcomes, such as oxygen saturations that are persistently out of range (e.g., <75% or > 90%), feeding difficulties, or inadequate weight gain among others.

In addition, interstage patients are seen frequently by their cardiologist, as often as every 1–2 weeks in the first months after hospital discharge. Visits may be scheduled to alternate with paediatrician appointments, so that the infant is seen weekly. Follow-up may be spaced to 2–4 weeks, depending on the team's assessment of the infant's overall condition.[10]

https://doi.org/10.1017/S1047951122002943 Published online by Cambridge University Press

Feeding

Healthy growth is a crucial issue in the interstage period.[11] These infants may be discharged home with supplemental enteral feeds or a nasogastric tube, depending upon swallow/feeding evaluations and the infants' ability to take adequate oral feeds. Some centres prefer to place a surgical gastrostomy tube. Many interstage teams include dieticians who optimise caloric intake and growth prior to the next intervention. In addition, speech or occupational therapists may be consulted to improve oral motor skills.

Sternal Precautions and Wound Care

Most centres recommend sternal precautions for at least 6 weeks after an operation requiring a sternotomy. Sternal precautions include avoiding lifting the infant from under their arms since this places stress on the healing sternum and can potentially lead to sternal fracture and instability. Rather, infants should be lifted in a cradle position, supporting their legs, back, neck, and head. If signs of surgical site infection arise, the cardiologist and/or cardiothoracic surgeon will want to be notified immediately. Concerning findings include significant redness, warmth, discharge, and fever without a source.

Medications Including Antithrombotics

Common cardiac medications include digoxin for infants with hypoplastic left heart syndrome,[12,13] angiotensin-converting enzyme inhibitors for children with significant valve regurgitation or ventricular dysfunction,[14] and beta-blockers for infants with history of arrhythmia. Many infants are on a diuretic (e.g., furosemide) due to some degree of pulmonary over-circulation. The cardiologist will monitor these medications closely as the child's clinical status evolves over time.[15,16] Most interstage patients are also on some form of antithrombotic therapy to ensure that the ductal stent, Blalock-Taussig-Thomas shunt, or Sano remains patent. Antithrombotic agents vary and typically include aspirin, clopidogrel, and enoxaparin.

Immunisations

Infants with single ventricle CHD are eligible for palivizumab. In addition, most centres advocate that infants receive their 2- and 4-month immunisations per scheduled guidelines as recent evidence demonstrated that vaccine titers were not significantly affected by cardiopulmonary bypass.[16]

Neurodevelopmental Evaluation

Ideally, infants with single ventricle heart disease are enrolled in an Early Intervention Program at the time of initial hospital discharge and are seen in a dedicated Neurodevelopment Clinic at least once prior to their next cardiac surgery.[17] Understanding of neurodevelopmental outcomes for these patients is rapidly evolving; neurodevelopment concerns and delays are related to both the underlying CHD as well as the myriad anaesthetic episodes and interventional procedures they undergo in early life during critical periods. Over their lifetime, patients may struggle with cognitive impairment, impaired social interaction, inattention, impulsive behaviours and impaired executive function. Motor problems are present in up to 42% of school age children who have undergone surgical intervention for heart disease in the first year of life.[18] In some cases, CHD may be a marker of an underlying genetic or syndromic condition, further increasing the risk of disability. Undergoing a thorough evaluation in a dedicated programme, when possible, is especially important to establish a baseline and ensure that these children receive appropriate services.[19]

Cardiology Follow-up

Any concerning home monitoring data or issues noted at follow-up clinic visits may warrant hospital admission to ensure that systemic and pulmonary blood flow are adequate. Saturations below 75% can indicate inadequate pulmonary blood flow due to a problem with the ductal stent/Sano/Blalock-Taussig-Thomas shunt. Conversely, higher than expected saturations (>90%) may be a sign of pulmonary over-circulation or the development of aortic coarctation. Additional testing, such as a cardiac catheterisation, may be obtained if the team is concerned about oxygen saturations or the adequacy of systemic or pulmonary blood flow.

A cardiac catheterisation is routinely performed prior to the second surgical intervention to assess the pulmonary vascular resistance. This is important because the Stage 2 operation – discussed below – requires passive blood flow from the upper body through the lungs for gas exchange. In addition, if a Norwood surgery was performed, the reconstructed aortic arch will be evaluated to ensure there is no residual coarctation. This "pre-Glenn" cardiac catheterisation is generally performed within a month of the second-stage operation, usually between 3 and 5 months of age depending on the infant's weight, oxygen saturations, gestational age, and other clinical factors discussed above.

3. Stage 2: Glenn and Hemi-Fontan (superior cavopulmonary anastomosis)

Stage 2 palliation involves anastomosing the superior caval vein to the pulmonary artery, technically known as a superior cavopulmonary anastomosis. Two variations of this procedure are common, the bidirectional Glenn which involves direct end-to-side connection of the superior caval vein to the pulmonary artery (Fig 3) and the hemi-Fontan, which involves a more complex connection of the superior caval vein and pulmonary artery; the hemi-Fontan is typically done at institutions that subsequently perform intracardiac (i.e., lateral tunnel) Fontan procedures (discussed below). In addition, the ductal stent/Blalock-Taussig-Thomas shunt/Sano/pulmonary artery band is removed if present. Pulmonary blood flow is now entirely passive from the superior caval vein through the pulmonary arteries. This passive flow requires low pulmonary vascular resistance and is the primary reason for waiting until ~ 3–5 months of age to perform the Stage 2, since the pulmonary vascular resistance typically reaches its nadir after the first few months of life. In fact, the high pulmonary vascular resistance in the newborn period would not allow for effective Glenn flow, which is why ductal stents, Blalock-Taussig-Thomas shunts, or pulmonary artery bands are initially used to secure stable pulmonary flow in children with adequate systemic flow.

One important point is that Stage 2 circulation is more stable than after initial palliation because the pulmonary and systemic circulations are separated, resulting in a lower burden on the single ventricle which is dedicated to the systemic circulation. As a result, most infants require a shorter post-operative hospital stay following this second stage operation and families usually return the scales and other monitoring devices since the infants no longer require "interstage" home monitoring. Oxygen saturations are usually in the 80% range immediately after the operation, though typically slowly decrease over time. One important consideration is that the second stage circulation is "preload-dependent," so oxygen saturations may decrease in the setting of dehydration. Children with this physiology need special monitoring if they develop a gastrointestinal illness with significant vomiting and/or diarrhoea.

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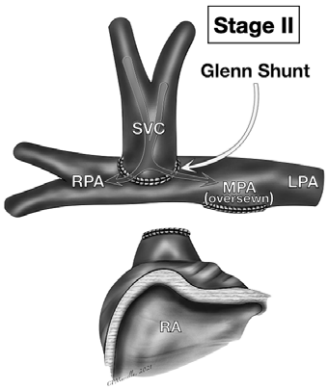

Figure 3. Glenn operation diagram.
Stage II Glenn operation depicting the superior caval vein (SVC) anastomosis to the right pulmonary artery (RPA) with flow going into both the RPA and left pulmonary artery (LPA). The main pulmonary artery (MPA) is oversewn and the right atrium (RA) no longer connected to the SVC.

Sternal Precautions and Wound Care

Post-operative issues are fairly standard after open-heart surgery and generally do not depend on the specific operation. As such, these issues are handled similar to after the first intervention.

Medications including Antithrombotics

Similar to after the first intervention, most are on some form of antithrombotic agent to prevent thrombus development in the cavopulmonary pathway. Aspirin is commonly utilised, with clopidogrel and enoxaparin prescribed frequently by many centres as well. Similarly, angiotensin-converting enzyme inhibitors, digoxin, sildenafil, beta-blockers, and diuretics may be used if certain other conditions are present. Again, the cardiologist will prescribe and adjust these medications as needed. As discussed above, given the pre-load dependence of the Stage 2 circulation, attention should be paid to children on diuretics who develop a gastrointestinal illness as the medication may need to be held temporarily.

Immunisations

Infants with Stage 2 palliation are recommended to receive 6- and 12-month immunisations as scheduled per guidelines. Most centres also strongly recommend the influenza vaccine when appropriate. In addition, depending upon age and time of year, these patients are also eligible for palivizumab.

Dental care and subacute bacterial endocarditis prophylaxis

Toddlers with single ventricle heart disease are recommended to visit a paediatric dentist for an initial visit at approximately 12–18 months of age as recommended by the American Academy of Pediatrics.[20] Spontaneous bacterial endocarditis prophylaxis is indicated for dental procedures, given the presence of "repaired" CHD with a residual shunt.[21]

School/daycare

Infants and young children with single ventricle heart disease are generally safe to attend daycare once discharged from the hospital and feeling well. This decision should be reviewed with the cardiologist on a case-by-case basis given the increased likelihood of respiratory and other infections in those settings. In addition, each daycare centre has unique requirements and experience caring for children with CHD so engaging with the centre as early as possible may facilitate attendance.

Activity restrictions

No specific activity limitations are indicated after sternal precautions have been lifted. Given the ages of patients in this stage (i.e., ~6 months to ~ 3–5 years), these children will tend to limit themselves as necessary.

Neurodevelopment evaluation

Infants and toddlers with Stage 2 circulation should continue to be seen by early intervention and neurodevelopment teams to ensure their development is appropriately supported.

Cardiology follow-up

Patients are usually seen a few weeks after discharge to ensure they are recovering from surgery adequately and oxygen saturations are appropriate. The regularity of follow-up is then based on overall clinical status, co-morbidities, and presence of residual cardiac defects. Typically, after a good surgical outcome in a patient with few other major conditions, the child is seen a few months after surgery, then ultimately spaced out to every 6 months. Families often miss the frequency of visits they had during the interstage period, specifically the intensive emotional, medical, and behavioural support.[9,22] However, the patients and families soon appreciate that fewer visits are a sign of a more stable child and enjoy the time spent out of the hospital and doctors' offices.

4. Stage-3: Fontan Operation (inferior cavopulmonary anastomosis)

The Fontan operation is the third and final stage of single ventricle palliation. This operation is called the inferior cavopulmonary anastomosis and, as the name implies, involves connecting the venous blood from the IVC/hepatic veins to the pulmonary circulation. The surgery is usually performed between 3 and 5 years of age, based on centre preference. Timing may also be influenced by oxygen saturations, exercise capacity, or other symptoms. Like the Stage 2, most patients undergo a pre-Fontan cardiac catheterisation to reevaluate the anatomy and pulmonary vascular resistance, ensuring that additional passive pulmonary blood flow from the lower half of the body will be well tolerated.

The Fontan operation is currently performed with one of two variations, either extracardiac or intracardiac (i.e., lateral tunnel). In addition, many centres "fenestrate" the Fontan by creating a small communication between the Fontan circuit and atrium. This fenestration decompresses the Fontan circuit, by providing a route for systemic venous blood to bypass the lungs (i.e., a right-to-left shunt) if the pulmonary resistance is temporarily high (e.g., immediately after surgery, during periods of respiratory illness). This decompression prevents sluggish flow and high venous pressures in the Fontan, thereby improving circulation overall. The pulmonary and systemic circulations are fully separated after the Fontan, so oxygen saturations can be in the upper 90% range with no fenestration and in the mid-80 to 90% range if fenestrated (Fig 4).

Post-operative hospital length of stay varies, primarily related to pleural drain output. Some Fontan patients have significant pleural drain output that takes days-to-weeks to clear, leading to a delay in tube removal. In fact, some centres may discharge patients with chest tubes if the output persists.[23] Occasionally, pleural effusions re-accumulate after discharge, so primary paediatric providers should observe for increased respiratory effort, decreased breath sounds, and change in oxygen saturations. If any of these are

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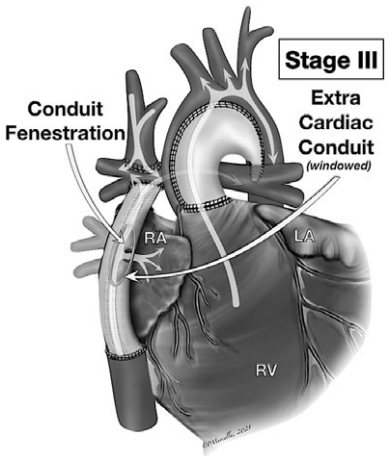

Figure 4. Fontan operation with fenestration diagram.
Stage III Fontan operation depicting the extracardiac Fontan conduit which brings the inferior caval vein blood up to the pulmonary arteries. Many centres will place a fenestration (i.e. "windowed") shown here between the conduit and the right atrium (RA). RV = right ventricle, LA = left atrium.

present, consider a chest X-ray and prompt call to the primary cardiologist.

Sternal precautions and wound care

As noted above, post-operative issues are standard after open-heart surgery and generally do not depend on the specific operation. So, these restrictions will be handled like the first- and second-stage palliations.

Medications including antithrombotics

Like prior stages, most patients are maintained on some form of antithrombotic to prevent thrombus formation in the Fontan pathway. Typical agents include aspirin, clopidogrel, and even warfarin. Special consideration is typically given to patients with a patent fenestration as well as for 6 months after fenestration device closure since they are at higher risk for a systemic (a.k.a. "paradoxic") embolism. Similarly, angiotensin-converting enzyme inhibitors, digoxin, sildenafil, beta-blockers, and diuretics may be used if certain other conditions are present. Over time, some patients develop significant ventricular dysfunction requiring additional heart failure medication. Again, the cardiologist will follow and adjust these medications as needed.

Vaccines

Children with Fontan circulation are recommended to receive all appropriate immunisations, per the Centers for Disease Control and Prevention schedule. Most centres also strongly recommend the influenza vaccine when appropriate. Data on the safety and efficacy of the COVID-19 vaccine do not exist in this population, so please refer to Centers for Disease Control and Prevention guidelines when available.

Dental care and subacute bacterial endocarditis prophylaxis

Excellent dental hygiene is important for these patients. Patients are recommended to visit a dentist every 6 months. As with the Stage 2 patients, spontaneous bacterial endocarditis prophylaxis is indicated for any patients with residual shunting (e.g., fenestration). In fact, many centres recommend spontaneous bacterial endocarditis prophylaxis for all Fontan patients.

School/daycare

Similar to the Stage 2 patients, children with Fontan circulation are safe to return to daycare or kindergarten once discharged from the hospital and feeling well. Given the differing requirements and experience among educational centres, paediatricians are encouraged to communicate the issues and needs of the child as soon as possible.

Activity restrictions

Once sternal precautions have been lifted, many cardiologists advise no activity limitations, as patients will typically limit themselves as necessary. Some cardiologists restrict from specific sports based on exercise stress testing or other cardiac test results. Many cardiologists reinforce the benefits of exercise, especially aerobic exercise, given the crucial interaction between pulmonary blood flow and general lung health. Depending on the needs and ongoing issues, some patients may also benefit from referral to a dedicated cardiac rehabilitation program[24] if available.

Neurodevelopment evaluation

Ongoing neurodevelopmental evaluation is recommended by both the American Academy of Pediatrics and American Heart Association.[17] Many centres refer according to those recommendations and evaluate all patients routinely at 4–5 years of age and then again at 11–12 years of age, with additional visits as indicated. Many school-age children benefit from an Individualized Education Plan under the Individuals with Disabilities Education Act, as studies show that children with CHD often have a higher rates of language, cognitive, and attentional difficulties.[25,26]

Cardiology follow-up

Patients are usually seen a few weeks after discharge to assess overall clinical condition and monitor for recurrence of pleural effusions. Subsequent cardiology visits are often spaced to every 6 months for a few visits and then yearly if the patient is clinically well with normal cardiac function. If a fenestration was placed, many centres perform a cardiac catheterisation one year post-Fontan to evaluate the Fontan circulation and determine if the fenestration can closed percutaneously. In addition, a growing number of centres are developing multi-disciplinary Fontan clinics to address the non-cardiac consequences of Fontan physiology.[26,27]

Long-term considerations

Though the staged palliation of single ventricle heart disease has significantly improved the quality and length of single ventricle patients' lives, numerous longterm issues arise.[28] For this reason, many centres have multi-disciplinary care clinics for ongoing care of Fontan circulation patients. A detailed review is beyond the scope of this document, but some of the more common significant issues affect the lungs, gastrointestinal tract, and liver as well as mental health.

A major pulmonary complication is plastic bronchitis, a respiratory condition that occurs due to proteinaceous material that accumulates in the lungs. It is marked by a productive cough without evidence of an infection (though super infections can develop), so be aware if patients develop chronic cough. Some patients may cough up "casts" of their bronchial tree. Plastic bronchitis is often an insidiously progressive condition and can be lethal. Many

https://doi.org/10.1017/S1047951122002943 Published online by Cambridge University Press

centres consider the development of plastic bronchitis to be an indication to list for heart transplantation.

Gastrointestinal complications include protein-losing enteropathy and hepatic disorders. Protein-losing enteropathy is a gastrointestinal issue that is marked by protein loss in stool. As expected, this typically presents with oedema/ascites and diarrhoea. Like plastic bronchitis, the natural history of protein-losing enteropathy is often associated with a poor long-term prognosis so some centres similarly consider it to be an indication to list for heart transplantation. From a liver standpoint, all Fontan patients are at risk of developing cirrhosis related to elevated hepatic venous pressures and congestion intrinsic to the Fontan circulation. Along those lines, Fontan patients are at increased risk of developing hepatocellular carcinoma.

Patient mental health is a field with emerging knowledge. Numerous recent studies have evaluated the long-term mental health and quality of life of patients with Fontan physiology.[29] Fontan patients are at risk for developing depression, anxiety, and attention deficit disorder so routine mental health screening is indicated.[30]

Last, though the 2nd and 3rd stages of single ventricle palliation separated the circulations so that the single ventricle is only pumping to the body *directly*, the overall workload on the heart is still increased because it has to drive the passive flow to the lungs. As a result, Fontan patients often develop significant cardiac dysfunction and a fair number ultimately require heart transplant.[28] Patients are also at risk of developing arrhythmias so routine evaluation is indicated. Long term there is evidence that lateral tunnel (intracardiac) Fontan patients have a higher arrhythmia burden.

Conclusions

Patients with single ventricle CHD are complex and include a spectrum of various anatomic lesions. They represent a vulnerable patient population as they undergo several procedures in infancy and early childhood, spending significant time in the hospital and clinics. As a result, patients and their families also shoulder a heavy burden of stress and anxiety. Free and open communication between their primary paediatric provider and cardiologist will achieve better coordinated care. Regular, multi-disciplinary care with the paediatrician at the helm of the child's medical home is the bedrock to maintain the health and wellness of these children.

Acknowledgements. The authors thank Christopher Marcella, RDCS, FASE, for drawing the included figures.

Financial support. This research received no specific grant from any funding agency, commercial, or not-for-profit sectors.

Conflicts of interest. None.

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https://doi.org/10.1017/S1047951122002943 Published online by Cambridge University Press