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Original Article

Long-term management of patients with hypoplastic left heart syndrome: the diagnostic approach at All Children's Hospital

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Abstract Improved survival in children with hypoplastic left heart syndrome has created a sub-population of children and young adults who are living with functionally univentricular physiology. Routine surveillance with comprehensive screening for structural cardiac disease, functional cardiac disease, arrhythmias, thromboembolic disease, and associated dysfunction of end organs is important. Future directives will better define the plans of care for routine surveillance in patients with hypoplastic left heart syndrome.

Keywords: Fontan; single ventricle; functionally univentricular heart; hypoplastic left heart syndrome; follow-up; adult congenital cardiac disease; Norwood operation

Advances in paediatric cardiac surgery and paediatric cardiology have improved survival in children with hypoplastic left heart syndrome and created a sub-population of children and young adults who are living with functionally univentricular physiology. This group will require comprehensive specialised cardiac follow-up to care for their unique disease. Hypoplastic left heart syndrome accounts for 1% of all congenital cardiac disease and is the most common severe functionally univentricular anomaly.[1] It accounts for 9% of all critically ill newborns with congenital cardiac disease, causing the largest number of cardiac deaths in the first year of life.[2,3] Hypoplastic left heart syndrome exists within a continuum and may include any or all of the following:

- hypoplasia of the left ventricle,
- stenosis/atresia of the aortic valve,
- stenosis/atresia of the mitral valve, and
- varying degrees of hypoplasia of the ascending aorta and aortic arch.

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Hypoplastic left heart syndrome is uniformly fatal without surgical intervention. In the early 1980s, before the initiation of surgical palliation, 90% of patients died within the first 30 days of life. At present, surgical palliation with a staged approach can be achieved with numerous surgical variations:

- Stage 1: Norwood operation – with Blalock–Taussig shunt or Sano shunt – versus Hybrid procedure.
- Stage 2: Bidirectional Glenn – bidirectional cavopulmonary anastomosis – or hemi-Fontan operation versus comprehensive Stage 2 with arch reconstruction and bidirectional Glenn – bidirectional cavopulmonary anastomosis – or hemi-Fontan operation.
- Stage 3: Fontan operation with or without fenestration that may be performed via an atriopulmonary connection – the "classic Fontan" – or more commonly via a total cavopulmonary connection – intra-atrial "lateral tunnel" or extracardiac external conduit.

Physicians who provide the long-term management of patients with hypoplastic left heart syndrome require a thorough understanding of the inherent

morbidities associated with the native cardiac disease and the acquired morbidities that can develop over a lifetime. Surgical palliation with a Fontan operation is not a cure – it is just a palliative operation for an otherwise lethal disease. Clinicians caring for patients with hypoplastic left heart syndrome therefore need to be astute to the development of

- structural cardiac disease,
- functional cardiac disease,
- electrical conduction disorders,
- thromboembolic disease, and
- associated dysfunction of end organs.

The early recognition of these findings may allow for early medical or surgical intervention that may improve the overall quality of life. The development of centres of excellence with a comprehensive team of medical providers dedicated to caring for children and adults with congenital cardiac disease will be required. In the future, the development of plans of care that allows for routine surveillance of these issues is needed to standardise care throughout the country. Below, we describe our centre's multi-speciality team's diagnostic approach to the long-term care of patients with hypoplastic left heart syndrome. Separate articles within this Supplement to *Cardiology in the Young* from HeartWeek 2011 will address the surgical considerations, role of transplantation, and transitional issues related to these children entering adulthood.

Structural cardiac disease

The anatomical surveillance of a patient with hypoplastic left heart syndrome starts with a thorough understanding of the prior surgical and catheterisation procedures performed. All old reports are requested before any visit to the clinic. Important factors to be aware of in the management of these complex patients include:

- the location and type of prior shunts,
- the type of Fontan operation – atriopulmonary connection, atrioventricular connection, lateral tunnel, or external conduit,
- the placement of coils or stents, and
- previously noted venous or arterial thrombosis.

Medical providers always need to consider the flow of blood in patients with functionally univentricular hearts in order to determine the possible structural abnormalities. Typically, after the Fontan operation in a patient with hypoplastic left heart syndrome:

- unoxygenated blood returns via the superior caval vein and inferior caval vein through a version of a Fontan baffle to the pulmonary arteries,
- unoxygenated blood then proceeds through the lungs and returns as oxygenated blood via the pulmonary veins,
- this oxygenated blood then travels across the atrial septum into the right atrium, tricuspid valve, and right ventricle – systemic ventricle, and
- then, this oxygenated blood is ejected across the neo-aortic valve – native pulmonary valve – into the aorta.

Haemodynamically significant structural abnormalities may exist at any or all of these locations. Structural abnormalities can also develop over a lifetime and may include:

- thrombosis,
- development of stenosis or leak of the Fontan baffle, and
- development of collaterals.

Multiple imaging modalities may be required to provide a complete anatomical surveillance of the patient with hypoplastic left heart syndrome, including

- transthoracic echocardiography,
- transoesophageal echocardiography,
- three-dimensional echocardiography,
- cardiac magnetic resonance imaging,
- cardiac computed axial tomographic angiography, and
- cardiac catheterisation with angiography.

As described in the manuscript by Nguyen et al elsewhere in this Supplement to *Cardiology in the Young* from HeartWeek 2011, we utilise transthoracic echocardiography with a complete congenital segmental assessment in all patients with hypoplastic left heart syndrome every 6 months.

Table 1 reviews the segmental approach to surveillance of structural cardiac defects in children or adults with hypoplastic left heart syndrome who have undergone the Fontan operation. Table 2 reviews the diagnostic tests utilised.

The superior caval vein, inferior caval vein, and Fontan baffle – extracardiac or intracardiac – can develop venous thrombosis because of stasis from low velocity flow. Stenosis of the intracardiac baffle or leaks may occur, especially in those with the lateral tunnel variety of the Fontan operation. In addition, the presence or absence of a residual fenestration needs to be assessed, especially in patients with clinical cyanosis. Approximately 40% of fenestrations will spontaneously close, and residual shunts can lead to progressive cyanosis or be a source of embolic stroke.[4] The systemic veins, Fontan baffle, and fenestration can be difficult to delineate in older children and adults by transthoracic imaging. In these cases,

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Table 1. Segmental approach to assess for residual structural cardiac anomalies in the long-term surveillance of patients with hypoplastic left heart syndrome who have undergone the Fontan operation.

Cardiac segment	Anomaly
Systemic veins	Thrombus in the superior caval vein or inferior caval vein
	Residual fenestration
	Stenosis or thrombosis of the baffle of the Fontan
Pulmonary arterial branches	Right or left peripheral pulmonary stenosis
Pulmonary veins	Pulmonary venous stenosis
Atrial septum	Restrictive atrial septum
Right atrium	Right atrial dilation due to lateral tunnel Fontan or progressive tricuspid regurgitation
Tricuspid valve	Tricuspid regurgitation
Right ventricle	Systolic dysfunction
	Diastolic dysfunction
	Asynchrony from disturbances of electrical conduction
Neoaortic valve	Regurgitation
	Stenosis
Neoaortic root	Dilation of the neoaortic root
	Formation of aneurysm
Aorta	Residual coarctation
Collaterals	Venovenous collaterals
	Arteriovenous collaterals

transoesophageal echocardiography and magnetic resonance imaging may be useful.

Stenosis of the pulmonary arterial branches may be secondary to suture lines or tenting from previously placed shunts. Stenosis of the right pulmonary artery may be seen following placement of a right-sided Blalock–Taussig shunt. Stenosis of the left peripheral pulmonary artery may be seen in patients because of the constriction of ductal tissue or secondary to compression from dilation of the aortic root. The pulmonary arteries are usually not well seen by transthoracic imaging. Alternative non-invasive imaging modalities include cardiac magnetic resonance angiography or computed axial tomographic angiography to better delineate the peripheral pulmonary arteries and pulmonary vasculature.

Pulmonary venous stenosis in patients with hypoplastic left heart syndrome is a rare but often fatal disorder. Usually, the atrial septum has been surgically resected during the prior surgeries. In some cases, the atrial septum may become restrictive as patients age. Pulmonary venous stenosis and a restrictive atrial septum cause reflexive pulmonary hypertension and may lead to failure of the Fontan. The recognition of these disorders can be achieved by transthoracic echocardiography, but advanced cardiac imaging may also be needed to be confirmatory.

Tricuspid valvar regurgitation may be caused by

- congenital dysplasia of the tricuspid valve,
- septal shift due to hypoplasia of the left ventricle,
- right ventricular dysfunction, or
- progressive valvar degeneration with age.

Risk factors for tricuspid regurgitation include mitral atresia – smaller left ventricle size – and length of myocardial ischaemia during the Norwood operation.[5] Regular assessment of the tricuspid valve and quantification of the degree of regurgitation is essential. Transthoracic echocardiography and three-dimensional echocardiography are useful for following the degree of tricuspid regurgitation.

Neo-aortic valvar regurgitation or stenosis may also develop in patients with hypoplastic left heart syndrome. Factors that may lead to the development of neo-aortic valvar regurgitation or stenosis include:

- congenital dysplasia of the native pulmonary valve or native aortic valve if it exists,
- positioning of the patch used to reconstruct the aorta during the Norwood operation, and
- dilation of the neo-aortic root.

Transthoracic echocardiography or three-dimensional echocardiography can also image the neo-aortic valve.

Dilation of the neo-aortic root may develop because of several potential aetiologies:

- the systemic vasculopathy,
- non-laminar flow across the reconstructed aorta,
- weakening of the aortic wall by placement of the patch used to reconstruct the aorta during the Norwood operation, and/or
- the true native pulmonary root subjected to lifelong systemic pressure and impedance.

Dilation of the aortic root may lead to

- progressive neo-aortic valvar regurgitation,
- compression of the pulmonary arterial branches, or
- rupture and sudden cardiac death.

Aortic root dilation can be followed by transthoracic echocardiography; however, additional imaging with cardiac magnetic resonance angiography or cardiac computed axial tomographic angiography might be beneficial for further delineation.

Some patients will have residual or recurrent coarctation of the aorta, hypoplasia of the transverse aortic arch, or aortic aneurysms develop after the Norwood operation. The aortic arch is interrogated by transthoracic echocardiography; however, full delineation of the aortic arch usually requires cardiac magnetic resonance angiography, computed axial tomographic angiography, and, occasionally, cardiac catheterisation with angiography.

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Table 2. Diagnostic testing used in the long-term surveillance in patients with hypoplastic left heart syndrome who have undergone the Fontan operation.

Mode of surveillance	Advantages	Frequency
Office visit/physical exam		Every 6 months
Electrocardiogram	Screen for abnormalities of conduction	Every 6 months
Echocardiogram	Screen for structural anomalies, formation of thrombus, and changes in cardiac function	Every 6 months
Three-dimensional echocardiogram	Helpful in the assessment of valvar regurgitation	As clinically indicated
Transesophageal echocardiogram	Assessment of systemic veins, fenestration, pulmonary veins, atrial septum, valvar lesions, and formation of thrombus	As clinically indicated
Cardiac magnetic resonance imaging/ magnetic resonance angiography	• Good assessment of thoracic vascular structures such as branches of the pulmonary arteries, pulmonary veins, and aortic arch • Also able to give functional assessment of the right ventricle • May not be able to be performed in patients with pacemakers or patients who have previously had implantation of stents and/or coils	As clinically indicated
Cardiac computerized axial tomographic scan/angiography	• Good assessment of thoracic vascular structures such as branches of the pulmonary arteries, pulmonary veins, and aortic arch • Good assessment of pulmonary parenchyma • May be performed in patients with pacemakers or patients who have previously had implantation of stents and/or coils	As clinically indicated
Laboratory values	Lipid panel	Annual
Additional laboratory values to consider	Liver function tests (AST/ALT) Basic chemistry Brain natriuretic peptide D-dimer Thyroid function tests	As clinically indicated
Pulmonary function tests	Specially consider in patients with significant scoliosis or deformity of the wall of the chest, or in patients taking amiodarone for arrhythmias	As clinically indicated

ALT = alanine aminotransferase; AST = aspartate aminotransferase

Patients with functionally univentricular physiology may also be at risk of development of aortopulmonary collaterals, veno-venous collaterals, and/or pulmonary arteriovenous malformations. Other than aortopulmonary collaterals, these collateral vessels may lead to progressive cyanosis from right-to-left shunting. These collateral vessels may be detected by cardiac magnetic angiography or cardiac computed axial tomographic angiography, but are best delineated and addressed by cardiac catheterisation if suspected.

The surveillance of structural cardiac disease following a Fontan operation in patients with hypoplastic left heart syndrome requires an experienced team trained in congenital cardiac disease that includes:

- physicians,
- nurses,
- echocardiographic sonographers, and
- technicians who specialise in advanced cardiac imaging.

Transthoracic echocardiography is a useful non-invasive method of routine surveillance for most of the cardiac structures in patients following a Fontan operation. Transoesophageal echocardiography and three-dimensional echocardiography may also be helpful adjuncts. Cardiac magnetic resonance imaging and computed axial tomographic angiography are best for periodic assessment of extracardiac anomalies in locations such as[6]

- the systemic veins,
- the Fontan baffle,
- the pulmonary arteries,
- the pulmonary veins, and
- the aorta.

Functional cardiac disease

Ventricular dysfunction is a significant risk factor for early and late morbidity and mortality in patients with hypoplastic left heart syndrome. Patients may develop right ventricular systolic dysfunction or diastolic dysfunction. The primary issues that lead to right ventricular systolic function are:

- idiopathic developmental issues with the myocardium,
- increased work load due to progressive obstruction of the outflow tract,

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- volume loading from tricuspid regurgitation or neo-aortic regurgitation, or
- myocardial ischaemic injury associated with an abnormality of the coronary arteries.

Patients with hypoplastic left heart syndrome with aortic atresia and mitral stenosis are particularly prone to abnormalities of the coronary arteries because of the development of coronary arterial fistulas from the hypertensive left ventricle. Acquired forms of right ventricular dysfunction include:

- the repetitive myocardial insults associated with cardiopulmonary bypass during the three stages of surgical palliation,
- prior right ventriculotomy if a Sano shunt was used during Stage 1 surgical palliation,
- insufficiency of the tricuspid valve,
- electrical conduction disorders – chronic tachyarrhythmias or chronic bradyarrhythmias, and/or
- the development of atherosclerotic disease of the coronary arteries as these children enter adulthood.

Systolic dysfunction leads to decreased cardiac output and may create symptoms of fatigue, exercise intolerance, ventricular arrhythmias, and diastolic dysfunction.

Diastolic dysfunction is common in hypoplastic left heart syndrome. Diastolic dysfunction is an inability of the ventricle to relax because of the loss of compliance. A progressive change in diastolic function occurs after the Fontan operation. In a study of 546 children who had undergone the Fontan operation, diastolic dysfunction was found in 72% with right ventricular morphology of the systemic ventricle.[7] Those with right ventricular morphology of the systemic ventricle were more likely to have diastolic dysfunction and atrioventricular valvar insufficiency than those with dominant left ventricle.[8] This finding may be secondary to intrinsic abnormalities within the myocardium, such as endocardial fibroelastosis, chronic coronary ischaemia, and/or hypoxic ischaemic injury from cardiopulmonary bypass. Diastolic dysfunction causes

- increased atrial filling pressures,
- atrial dilation,
- reflexive pulmonary hypertension,
- elevated central venous pressure, and
- increased dysfunction end organ(s) including cirrhosis of the liver and protein-losing enteropathy.

The evaluation of systolic and diastolic right ventricular dysfunction is traditionally done via transthoracic echocardiography. Systolic function can be assessed by utilising a shortening fraction or ejection fraction. Better assessments of right ventricular systolic function may be performed by three-dimensional assessment of the volume of the heart and cardiac magnetic resonance imaging. Recent studies have shown that decreased right ventricular ejection fraction as calculated by magnetic resonance imaging was predictive of interstage mortality.[9] Diastolic dysfunction may be assessed by Tei index, Dp/Dt, or tissue Doppler assessment. The use of two-dimensional or three-dimensional assessment of strain may also help in the assessment of ventricular function. In 2011, Petko et al[10] demonstrated that strain acquired through two-dimensional speckle tracking predicted regional differences in myocardial deformation and intraventricular dyssynchrony in patients with hypoplastic left heart syndrome with aortic atresia and mitral atresia – smallest left ventricular size – as compared with other subgroups of patients with hypoplastic left heart syndrome. Right ventricular myocardial deformation has also been observed to be reduced at the site of the ventriculotomy in patients who have previously undergone placement of a Sano shunt from the right ventricle to the pulmonary artery.[11] The long-term effect of these diastolic changes remains unclear.

Arrhythmias

Patients with hypoplastic left heart syndrome are at risk of the development of tachyarrhythmias and/or bradyarrhythmias. The incidence of arrhythmias in patients with hypoplastic left heart syndrome has been estimated to range from 40% to –57%.[12,13] Early tachyarrhythmias typically consist of supraventricular tachycardia, which are most common during Stages 1 and 2, and have been associated with increased interstage mortality.[12] Late postoperative tachyarrhythmias occur at mean age of 6–11 years after the Fontan operation.[14] Such tachyarrhythmias include intra-atrial re-entry tachycardia and atrial ectopic tachycardia. Intra-atrial re-entry tachycardia is the most common arrhythmia that develops after the Fontan operation, with a frequency of 10–40%.[15] The risk factors for developing intra-atrial re-entry tachycardia include[15]

- older age at the time of the Fontan operation,
- female gender,
- moderate to severe atrioventricular valvar regurgitation, and
- history of resection of the atrial septum.

Sustained arrhythmias with heartbeat rates greater than 100 beats per minute may result in congestive heart failure within 12–36 hours, and require acute termination with medical or electrical cardioversion. Care should be taken to exclude the presence of an

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intracardiac thrombus before cardioversion. Patients with chronic atrial tachycardia will require anticoagulation with Warfarin (Coumadin) and consideration for transcatheter ablation. Rates of success are reported from 33% to 100%, with recurrences in 33–100%, and these rates are worse than ablation done in patients with other types of congenital cardiac disease.[14–18] Frequent tachyarrhythmias refractory to pharmacological management or associated with significant haemodynamic problems may require Fontan revision or conversion – Re-do Fontan – with arrhythmia surgery, an operation that involves[14–18]

- debulking the right atrium,
- conversion to a total cavopulmonary connection,
- removal of thrombus,
- excising right atrial scarring,
- a modified right-sided Maze procedure, and
- implanting a permanent epicardial pacemaker.
- If the patient has a history of atrial fibrillation, a left-sided Maze procedure is done as well.

Khairy and colleagues[16] found that after Fontan conversion with arrhythmia ablation, 50% of patients were able to stop antiarrhythmic medication and 30% with recurrence were all controlled with medication.

Bradyarrhythmias typically consist of dysfunction of the sinus node or high-grade atrioventricular block. These bradyarrhythmias have a poor prognosis and have had mortality estimated as high as 73%. Bradyarrhythmias may occur secondary to multiple aetiologies, including

- damage of the sinus node or atrioventricular node during surgery,
- hypoxic injury during cardiopulmonary bypass, or
- chronic fibrosis.

Most such arrhythmias occur in the first postoperative week. A permanent pacemaker is likely necessary in patients with sustained dysfunction of the sinus node, sustained cardiac block, or severe symptoms. Aboulhosn and Child found that chronic ventricular pacing was more effective with multi-site epicardial pacing in comparison with single-site epicardial pacing.[19] Chronic pacing may lead to progressive ventricular dysfunction due to dyssynchrony.

Thrombus

Formation of thrombus is a frequent complication after the Fontan operation, with an incidence of 20–30%.[20–22] Aetiologies of thrombus formation include

- low rate of flow throughout the cavopulmonary circuit,
- a hypercoagulable state,
- atrial arrhythmia,
- suture lines,
- scarring and clotting,
- loss of factors in pleural effusion early postoperatively or later with protein-losing enteropathy.

An almost equal predilection exists for formation of thrombus in the right atrium (48%) versus the pulmonary venous circulation (44%).[20] The median time to develop thrombus is 22–23 months.[21,22] Symptoms of thrombus include signs of venous obstruction, progressive cyanosis, paradoxical emboli, and atrial tachycardia.

Currently, no consensus exists on thromboprophylaxis. Kaulitz et al[23] recommend against routine anticoagulation in all patients after the Fontan operation. Patients with venous pathways composed of completely autologous tissue may be at less risk for formation of thrombus, and therefore may have less need for anticoagulation. Those with pathways that include prosthetic material are usually placed on aspirin. Warfarin (Coumadin) is indicated in patients with specific risk factors including

- prior formation of thrombus,
- previous thromboembolic event,
- residual fenestration,
- residual intra-atrial shunt,
- dilated right ventricle with "slow blood flow phenomenon", or
- recurrent atrial arrhythmias.

Recently, longitudinal assessment of profiles of coagulation demonstrated an increase in factor VIII after the Fontan operation, which may indicate a subset of patients at risk for thrombosis.[24] At our institution, all patients are maintained on aspirin from Stage 1 to through completion of the Fontan operation and beyond, unless a hypercoagulable state is suspected, in which case Warfarin (Coumadin) is initiated.

Additional surveillance of end organs

Assessment of the function of end organs may be just as important as the assessment of ventricular function in patients with hypoplastic left heart syndrome. Progressive injury to end organs may result from any of several potential aetiologies:

- the effect of cavopulmonary anastomosis with elevated central venous pressure,
- multiple exposures to cardiopulmonary bypass,
- risk of chronic thromboembolic disease,
- associated systemic vasculopathy with lesions of the left ventricular outflow tract, and
- chronic administration of medications.

The surveillance for dysfunction of the end organs in patients with hypoplastic left heart syndrome

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Table 3. Dysfunction of end organs other than the heart associated with hypoplastic left heart syndrome and/or Fontan circulation.

Part of the body	Complications
Central nervous system	Seizure, stroke, motor delays, learning disabilities
Head–eyes–ears–neck–throat	Dysfunction of the vocal cord(s)
	Chronic otitis media/hearing loss
	Sinusitis
	Intolerance of oral feeding
Respiratory	Chronic lung disease
	Reactive airway disease
	Restrictive lung disease due to scoliosis
	Pulmonary fibrosis from medications such as amiodarone
	Tracheobronchomalacia
Gastro-intestinal	Gastro-oesophageal reflux
	Oesophageal varices if portal hypertension develops from cirrhosis of the liver
	Protein losing enteropathy
	Failure to thrive
Endocrine	Thyroid dysfunction
Musculoskeletal	Scoliosis
Haematology	Hypercoagulable state
	Thromboemboli

requires physicians to use a systematic approach (Table 3). Medical providers can think of this approach as an assessment of the body from head to toe.

The central nervous system can be affected in patients with hypoplastic left heart syndrome due to congenital disease, seizure, stroke, and global ischaemic injury. Chronic seizures and developmental delays, both motor and learning, may result. The "head–neck–throat" can be affected because of dysfunction of the vocal cord(s) from chronic intubation or injury to the left recurrent laryngeal nerve during surgery. Dysfunction of the vocal cord(s) may lead to problems with speech or swallowing. The thyroid gland may also have dysfunction and should be assessed at regular intervals. Pulmonary disease is also common and may consist of

- chronic lung disease,
- reactive airway disease,
- thromboembolic disease, or
- restrictive disease due to musculoskeletal abnormalities such as scoliosis and/or other deformities of the wall of the chest, or medication administration, for example, with amiodarone.

Elevated central venous pressure may cause cirrhosis of the liver or protein-losing enteropathy, which may be followed by assessment of

- tests of blood for hepatic function,
- levels of protein and albumin in the serum, and
- the presence of alpha 1-antitrypsin in the stool.

Renal dysfunction may occur because of intrinsic disease of the kidney, systemic vascular issues, or multiple hypoxic episodes during cardiopulmonary bypass.

Conclusion

As our patients with hypoplastic left heart syndrome live into adulthood, we are learning more about the complications of long-term life after the Fontan operation. Routine surveillance with comprehensive screening for structural cardiac disease, functional cardiac disease, arrhythmias, thromboembolic disease, and associated dysfunction of end organs is important. Future directives will better define the plans of care for routine surveillance in patients with hypoplastic left heart syndrome.

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