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ISHLT GUIDELINES

The International Society for Heart and Lung Transplantation Guidelines for the management of pediatric heart failure: Executive summary

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These guidelines have been produced to bring together current recommendations on the evaluation and management of pediatric heart failure (HF) and update the previous guideline.[1]

The writing group was chosen from the membership of the International Society for Heart and Lung Transplantation (ISHLT), the Association of European Pediatric and Congenital Cardiology (AEPC), and the Pediatric and Congenital Electrophysiology Society (PACES) across health care disciplines to achieve representation of HF practice throughout the world. Overall, 90 contributors from 13 counties across 4 continents (Appendix 1) were assigned various aspects of HF according to their expertise. A comprehensive review of the available published evidence for HF management was undertaken. The strength and level of evidence was assessed according to standard practice.[2] The recommendations were achieved by consensus with the contributors; however, it is recognized that the evidence base for many of the recommendations is Level C due to the lack of trials in children. In some areas, there is such a lack of information that no recommendations can be made: it is hoped that by recognizing these deficiencies, research will be stimulated to address them. External review was undertaken by 8 international experts invited from adult advanced HF, pediatric cardiology, and congenital cardiovascular surgery (Appendix 2). The final guidelines were reviewed and approved by the ISHLT Board and Standards & Guidelines Committee, endorsed by the AEPC, and those guidelines pertinent to electrophysiology, by the PACES Executive Committee.

The background information for these guidelines and complete references have been published in the monograph series, Volume 8 by the ISHLT.[3] The abbreviations used are listed in Appendix 3.

Definition and documentation recommendation[4,5]

Chairs: Richard Kirk, Anne I. Dipchand, and David N. Rosenthal

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HF in children is a clinical and pathophysiologic syndrome that results from ventricular dysfunction, volume, or pressure overload, alone or in combination. It leads to characteristic signs and symptoms, such as poor growth, feeding difficulties, respiratory distress, exercise intolerance, and fatigue, and is associated with circulatory, neurohormonal, and molecular abnormalities. HF has numerous etiologies that are a consequence of cardiac and non-cardiac disorders, either congenital or acquired.

Class I

1. The documentation of HF severity and when appropriate, staging facilitates monitoring of disease progression and patient management (Table 1): **Level of Evidence C**

Genetic testing recommendations

Chairs: Richard Kirk and Jeffery Towbin

Dilated cardiomyopathy[6–10]

Class I

1. Comprehensive or targeted DCM genetic testing (*LMNA* and *SCN5*) is recommended for patients with DCM and significant cardiac conduction disease (i.e., first-, second-, or third-degree heart block) and/or a family history of premature unexpected sudden death. **Level of Evidence C**
2. Mutation-specific genetic testing is recommended for first-degree family members after the identification of a DCM-causative mutation in the index case. **Level of Evidence C**

Class IIa

1. Genetic testing can be useful for patients with familial DCM to confirm the diagnosis, to recognize those who are at highest risk of arrhythmia and syndromic features, to facilitate cascade screening within the family, and to help with family planning. **Level of Evidence C**

Table 1 Heart Failure Severity Classifications

Class	NYHA	Ross
I	No limitations of physical activity	No limitations or symptoms
II	May experience fatigue, palpitations, dyspnea, or angina during moderate exercise but not during rest	Mild tachypnea or diaphoresis with feeding
III	Symptoms with minimal exertion that interfere with normal daily activity	Infants with growth failure and marked tachypnea or diaphoresis with feedings, older children with marked dyspnea on exertion
IV	Unable to carry out any physical activity because they typically have symptoms of HF at rest that worsens with any exertion	Symptoms at rest such as tachypnea, retractions, grunting, or diaphoresis

Expansions for the abbreviations used in Table 1 are provided in Appendix 3.

Hypertrophic cardiomyopathy[9,11–13]

Class IIa

1. Genetic testing is indicated in the most clinically affected individual to facilitate screening. **Level of Evidence A**
2. Mutation-specific genetic testing is recommended for first-degree family members after the identification of an HCM-causative mutation in the index case. **Level of Evidence A**

Restrictive cardiomyopathy[14,15]

Class I

1. Mutation-specific genetic testing is recommended for first-degree family members after the identification of an RCM-causative mutation in the index case. **Level of Evidence C**

Class IIb

1. RCM genetic testing may be considered for patients in whom a cardiologist has established a clinical index of suspicion for RCM based on examination of the patient's clinical history, family history, and ECG/echocardiographic phenotype. **Level of Evidence C**

LV non-compaction cardiomyopathy[16]

Class I

1. Mutation-specific genetic testing is recommended for first-degree family members after the identification of an LVNC-causative mutation in the index case. **Level of Evidence C**

Class IIa

1. LVNC genetic testing can be useful for patients in whom a cardiologist has established a clinical diagnosis of LVNC based on an examination of the patient's clinical history, family history, and ECG/echocardiographic phenotype. **Level of Evidence C**

Arrhythmogenic ventricular cardiomyopathy[17,18]

Class I

1. Mutation-specific genetic testing is recommended for first-degree family members after the identification of the AVC-causative mutation in an index case. **Level of Evidence C**

Class IIa

1. Comprehensive or targeted AVC genetic testing (*DSC2*, *DSG2*, *DSP*, *JUP*, *PKP2*, and *TMEM43*) can be useful

for patients who satisfy task force diagnostic criteria for AVC. **Level of Evidence C**

Diagnostic recommendations

Chairs: Anne I. Dipchand, Michael Burch, and Luc Mertens

B-Type natriuretic peptide[19–23]

Class IIb

1. BNP/NT-proBNP can be used as an adjunctive marker in the integrated evaluation and monitoring of patients with known HF to further define severity, response to therapy, and its progression. **Level of Evidence B**
2. BNP/NT-proBNP can be used as an adjunctive marker, not a stand-alone test, to aid in the diagnosis of new HF in symptomatic patients. **Level of Evidence B**

Imaging[24–30]

Table 2 compares the imaging modalities.

Echocardiogram

Class I

1. Measurement of LV dimensions and LV wall thickness is an essential part of every echocardiographic functional LV assessment in patients with HF. The recent American Society of Echocardiography pediatric recommendations propose the use 2-D imaging instead of M-mode, but this was not based on actual data proving its clinical superiority. Normal values are more readily available for M-mode measurements. **Level of Evidence B**
2. LV remodeling should be monitored during serial follow-up. This consists of measuring cavity dimensions, wall thickness, and where clinically relevant, LV mass. **Level of Evidence B**
3. For patients with HF, assessing LV function by calculating LVEF based on a well-standardized 2-D method (biplane Simpson's or area-length method) should be undertaken. **Level of Evidence B**

Class IIa

1. Fractional shortening can be used for sequential assessment of LV function, although caution is required in patients with abnormal or paradoxical septal motion. **Level of Evidence C**
2. No recommendations can be made about the use of automated 2-D methods or 3-D echocardiography. These are emerging techniques for EF calculation that still need further validation in pediatric HF. **Level of Evidence C**

Class IIb

1. LVEF is load-dependent. The use of methods correcting for afterload have been proposed that do not improve diagnostic accuracy and predictive value. **Level of Evidence C**
2. Although measured and reported, the value of blood pool Doppler parameters, such as dP/dt, myocardial performance index, and the ratio of systolic-to-diastolic duration, in the assessment of LV systolic function in pediatric patients with HF is probably limited. (Problems with reliability of measuring time intervals, loading, and heart rate dependency of the methods, and their uncertain prognostic value, all limit their clinical use.) **Level of Evidence C**
3. Although measured and reported, the role of tissue Doppler imaging (TDI) and strain imaging in pediatric patients with HF is still uncertain. Pulsed TDI at the annulus has the benefit of published normal ranges and may assist in the early detection of myocardial dysfunction. Apart from the potential additional predictive information, its use in the description of mechanical dyssynchrony and identification of candidates for CRT seems to be important but requires further investigation. **Level of Evidence C**

Echo diastolic function[31]

Class IIa

1. Diastolic function, including mitral inflow patterns, mitral annular TDI, and pulmonary venous Doppler flow patterns should be assessed by echocardiography in children with HF. The tracings should be interpreted to define the type of diastolic abnormality (relaxation abnormality, reduced compliance, restrictive filling) as well as to attempt to diagnose the presence of elevated filling pressure (Table 3 summarizes typical changes in progressive LV diastolic dysfunction). **Level of Evidence C**

Cardiac MRI[32–34]

Class IIb

1. Cardiac MRI can be used to assess LV function in children. Use can be limited due to the requirement for general anesthesia in younger children, the presence of arrhythmia, and the availability and cost associated with cardiac MRI. **Level of Evidence C**
2. Balanced steady-state-free precession cine analysis has become the reference standard for volumetric and myocardial mass assessment of the LV. Where feasible, it should be used to assess children with HF. However, accuracy depends on standardization of analysis protocol and acquisition of adequate temporal and spatial resolution specific to the patient's heart rate and size. **Level of Evidence C**

RV systolic function[24]

Class IIa

1. For the quantitative assessment of RV function by 2-D echocardiography, it is reasonable to use fractional area of change from the apical 4-chamber view together with tricuspid annular planar systolic excursion. **Level of Evidence C**

Table 2 Comparison of Imaging Modalities

Modality	Advantages	Limitations	Use in pediatric HF
Echocardiography	First-line technique for all patients in acute/chronic HF; anatomic + functional assessment possible	Limited by acoustic windows	First-line technique in the assessment of HF
M-mode	High temporal resolution	Only works if global remodeling/ dysfunction	Serial measurement of LV dimensions, wall thickness, and fractional shortening
	Normal pediatric values available	Not valid if paradoxical septal motion	
2-D echocardiography	Assessment of 2-D anatomy, identification of structural disease	Dependent on acoustic windows	Identification of structural disease as cause for HF
2-D EF	Good parameter for global performance	Geometrical assumptions	Recommended technique for assessment of LV performance
	Can be calculated in patients with regional dysfunction	Load dependency	
3-D EF	No geometric assumptions	Highly dependent on acoustic windows Lower temporal resolution	Emergent technique replacing 2-D EF
Blood Doppler techniques (MPI, dP/dt, S/D ratio)	Geometry-independent	Variability of measuring time intervals	Limited use in heart failure patients
	High temporal resolution	No spatial information Load dependency of different indices	
TDI	Geometry-independent	Angle-dependent	Assessment of longitudinal function, early detection of dysfunction, assessment of diastolic function
	High temporal resolution	Limited information on its use in children	Dyssynchrony detection
	Quantification of regional function	Load dependency	
Strain imaging	Geometry-independent	Software dependency and differences between vendors	Emergent technique
	Images myocardial mechanics	Load dependency Limited normal data	Dyssynchrony assessment
Cardiac MRI	Not limited by imaging windows, no radiation, good for imaging extracardiac structures, flow quantification, tissue characterization (fibrosis imaging)	Accessibility, requirement for general anesthesia, cost, not compatible with many devices (pacemaker, assist devices)	Limited use in HF patients; quantification of RV function, patients with limited acoustic windows, fibrosis detection
Steady-state free precession imaging	Clinical reference technique for quantification of LV and RV volumes + mass	Lower temporal resolution (ECG-gated)	Mainly calculation of volumes and EF
Phase contrast	Reference technique for quantifying flows	Lower temporal resolution than Doppler techniques	Calculation of CO, quantification of valve regurgitation
MR tagging	Allows quantifying myocardial mechanics	Difficult post-processing; lower frames rates compared with echocardiography	Limited clinical use in heart failure patients. Research tool
Late gadolinium enhancement	Identification of regional fibrosis	Limited spatial resolution, only detects regional fibrosis	Clinical use still uncertain; possible prognostic value in HCM, detection of EFE in DCM or obstructive lesions
Angiography	Detection of extracardiac abnormalities	Spatial resolution is less good than cardiac CT (coronary imaging)	Can be used to detect extracardiac abnormalities causing heart failure (arch, coronary anomalies)
Cardiac CT	Not limited by imaging windows; high resolution, allows studying spatial relationships between cardiovascular structures and the airway, anesthesia often not required	Exposure to radiation, limited functional information, no flow quantification, coronary imaging influenced by high heart rates	Mainly used for coronary artery imaging if uncertain about coronary artery origins on echocardiography; limited role in follow-up

Expansions for the abbreviations used in Table 2 are provided in Appendix 3.

Table 3 Summary of Typical Changes in Progressive Left Ventricular Diastolic Dysfunction

Variable	Early relaxation abnormality	Progressive decreasing compliance	Restrictive filling
Mitral e′	↓	Persistent ↓	↓↓
Mitral E velocity	↓	↑	↑
Mitral A velocity	↑	↔	↓
Mitral E/A ratio	↓	Pseudonormalizes	↑
Deceleration time	↑	Progressive ↓	↓↓
Pulmonary vein D	↓	↑	
Pulmonary vein S	↑	↓	↓↓
Other		Abnormal pulmonary vein S/D ratio; increased pulmonary vein A wave reversal	Prominent A reversal in pulmonary veins

D, diastole; S, systole. Expansions for the other abbreviations used in Table 3 are provided in Appendix 3.

2. TDI assessment of tricuspid annular motion is a useful technique for the assessment of RV longitudinal function and should be added in the quantitative assessment of RV function. **Level of Evidence C**

Class IIb

1. Due to methodologic variability and the load-dependency, the clinical use of RV myocardial performance index in the assessment of right HF is limited. **Level of Evidence C**
2. RV strain analysis of RV longitudinal deformation is an emerging technique and requires further validation before routine use for the assessment of RV function. **Level of Evidence C**

MRI RV function[35–37]

Class IIa

1. Assessment of RV size and EF by MRI is considered the clinical reference for the assessment of RV function in patients with RV failure. Restricted access, the need for general anesthesia or sedation in infants and young children, and costs remain important limitations. **Level of Evidence C**

RV diastolic function

Class IIb

1. Doppler echocardiography can be used for the assessment of RV diastolic function, although criteria for grading and assessment of filling pressures are still poorly validated. **Level of Evidence C**
2. Different MRI techniques are available for studying RV diastolic dysfunction, but their clinical utility still needs to be evaluated. **Level of Evidence C**

Single-ventricle echo size and function

Class IIb

1. Quantitative techniques, such as fractional area of change, EF, annular excursion, and TDI for serial follow-up may be used, but good data regarding the prognostic value of observed changes are not available. **Level of Evidence C**

Single-ventricle MRI size and function[38]

Class IIa

1. Cardiac MRI is the reference standard for volumetric analysis of the single ventricle. Larger studies are required to define acceptable ranges of end-diastolic or end-systolic volume in these patients. **Level of Evidence B**

Exercise testing[39,40]

Class IIa

1. Metabolic exercise testing with a measurement of peak Vo_2 consumption$_2$, if feasible, should be part of the assessment of cardiomyopathy patients with HF. **Level of Evidence C**
2. A peak Vo_2 consumption of $< 50\%$ predicted for age and sex in patients with Stage C HF associated with severe limitation in exercise and activity may form the basis for consideration of heart transplantation. **Level of Evidence C**

Sleep study[41]

Class IIa

1. Children with HF and a history consistent with sleep apnea should undergo evaluation for sleep disorder breathing. **Level of Evidence C**

Ambulatory monitoring[42–44]

Class I

1. In a pediatric HF patient who presents with palpitations or syncope, some form of ambulatory monitoring should be considered to achieve a specific diagnosis and drive further management decisions. Less frequent symptoms may require use of longer-term event monitors. **Level of Evidence C**

Class IIa

1. In pediatric HF patients with a high risk of developing atrial or ventricular arrhythmias or heart block, regular

ambulatory monitoring should be considered. This would include patients after a Fontan palliation, any form of atrial switch procedure, heterotaxy (isomeric) syndromes, congenitally corrected transposition of the great arteries, or cardiomyopathy (HCM, DCM, RCM, and LVNC). **Level of Evidence C**
2. In asymptomatic pediatric HF patients, there are no data to support the timing and frequency of regular ambulatory monitoring for arrhythmias; however, intermittent screening for asymptomatic arrhythmias should be considered. **Level of Evidence C**

Cardiac catheterization[1,45–48]

Class IIa

1. EMB should not be performed in clinically diagnosed myocarditis with minimal symptoms and mild dysfunction or rapid normalization of function. **Level of Evidence C**
2. It is reasonable for cardiac catheterization to be performed if increased pulmonary resistance is suspected to measure the PVR and reversibility of pulmonary hypertension in patients with CHD and HF. **Level of Evidence B**

Class IIb

1. Cardiac catheterization and EMB can be considered for the assessment of unexplained cardiomyopathy or myocarditis if non-invasive testing does not yield a diagnosis. **Level of Evidence C**
2. Cardiac catheterization and EMB is reasonable in the setting of suspected AVC. **Level of Evidence C**
3. Cardiac catheterization and EMB is reasonable in pediatric patients presenting with HF when a specific diagnosis is suspected that would influence therapy. **Level of Evidence C**
4. Cardiac catheterization may be considered to determine PVR and reversibility after medical therapy has been optimized in patients with cardiomyopathy if they are stable enough to undergo testing, but the necessity is controversial outside of consideration for transplantation. **Level of Evidence C**
5. It is reasonable for EMB to be performed in the setting of unexplained new-onset HF associated with hemodynamic compromise, ventricular arrhythmia, or heart block where there is failure to respond to medical therapy. **Level of Evidence B**

Assessment of PVR[49,50]

Class IIa

1. Doppler-derived estimations of right heart pressures (right ventricular systolic pressure and pulmonary artery pressures) can be obtained for assessment and longitudinal follow-up of HF patients to monitor response to treatment, progression of disease, and contribute to decision making about more invasive assessment of PVR for the purpose of decision-making around medical and/or surgical interventions including transplantation. **Level of Evidence C**
2. It is reasonable for cardiac catheterization to be performed to assess PVR and reversibility in patients with CHD and HF. **Level of Evidence B**

Class IIb

1. The role of MRI in the assessment of PVR in children with HF requires further study. **Level of Evidence C**

Tachycardia-induced cardiomyopathy[51,52]

Class IIa

1. Electrophysiology testing or long-term monitoring can be useful in pediatric patients with presyncope or syncope with at least moderately impaired LV function. **Level of Evidence C**
2. Tachycardia-induced cardiomyopathy should be considered in any patient presenting with DCM because it is potentially reversible with appropriate intervention. **Level of Evidence B**

Class IIb

1. Programmed ventricular stimulation may be helpful in specific situations, such as in patients with HF and syncope, but does not appear to have a routine role in risk stratification in the pediatric patient with HF. **Level of Evidence C**

Pharmacologic treatment of chronic, reduced EF (systolic HF) recommendations

Chairs: David N. Rosenthal and Robert Shaddy

Diuretics[53,54]

Class I

1. Patients with fluid retention associated with ventricular dysfunction (HF Stage C) should be treated with diuretics to achieve a euvolemic state. **Level of Evidence C**

ACE inhibitors[55–59]

Class1

1. For the treatment of symptomatic left ventricular dysfunction (HF Stage C), ACE inhibitors should be routinely used unless there is a specific contraindication. These medications should be started at low doses and should be up-titrated to a maximum tolerated safe dose. **Level of Evidence B**

Class IIa

1. For the treatment of asymptomatic left ventricular dysfunction (HF Stage B), ACE inhibitors should be routinely used unless there is a specific contraindication. **Level of Evidence B**

2. ACE inhibitor therapy should be considered for individuals with a diagnosis of Duchenne muscular dystrophy unless there is a specific contraindication, although the optimal age of institution of therapy is unclear. **Level of Evidence B**

Class IIb

1. ACE inhibitor therapy should not be routinely instituted for all patients with single-ventricle CHD, but could be considered in specific cases such as in situations of valve regurgitation or ventricular dysfunction. **Level of Evidence B**

β-Receptor antagonists[60–62]

Class IIa

1. Following adult HF guidelines, it is reasonable to consider β-blockers in symptomatic children with systemic LV systolic dysfunction, particularly if the systemic ventricle has a LV morphology. Therapy should start at a small dose and slowly up-titrate. **Level of Evidence B**
2. Following adult HF guidelines, it is reasonable to consider β-blockers in asymptomatic children with systemic LV systolic dysfunction. Therapy should start at a small dose and slowly up-titrate. **Level of Evidence B**

Mineralocorticoid antagonists[63]

Class I

1. Following adult HF guidelines, it is reasonable to consider aldosterone antagonists in children with systemic LV dysfunction. **Level of Evidence C**

Angiotensin receptor antagonists[64]

Class IIa

1. Similar to adults, angiotensin receptor blockers are generally reserved for those children with systemic ventricular systolic dysfunction who would benefit from renin-angiotensin-aldosterone– system blockade but are intolerant of ACE inhibitors. **Level of Evidence C**

Digoxin and cardiac glycosides[65,66]

Class I

1. Digoxin is not recommended for children with asymptomatic LV dysfunction because no survival benefit was seen with digoxin in adults with HF and low EF. **Level of Evidence C**

Class IIa

1. Digoxin may be used to relieve symptoms in children with symptomatic HF and low EF. Doses targeting lower serum digoxin concentrations (e.g., 0.5–0.9 ng/ml) should be considered with attention to dose reductions in patients on carvedilol and amiodarone or those who have or are at risk for renal dysfunction. **Level of Evidence C**

Hydralazine combination with isosorbide dinitrate[67]

Class III

1. The use of combination therapy of hydralazine and isosorbide dinitrate is not recommended in pediatric HF. **Level of Evidence C**

Anti-arrhythmic medications[65,68]

Class IIb

1. The use of anti-arrhythmic medications may be warranted in select cases where arrhythmias persist after normalization of electrolyte disturbances or metabolic issues (i.e., hyperthyroidism) and the arrhythmias are poorly tolerated. **Level of Evidence C**

Class III

1. Anti-arrhythmic medications should not be used routinely in the management of children with HF with low EF. **Level of Evidence C**

Statin therapy

Class III

1. Treatment of HF with statin therapy is not indicated in pediatric HF patients. **Level of Evidence C**

Renin inhibitors

Class III

1. Direct renin inhibitors cannot be recommended for the treatment of HF in children. **Level of Evidence C**

Anticoagulants[69,70]

Class I

1. Patients with intracardiac thrombus should receive anticoagulation with heparin or warfarin. **Level of Evidence B**

Class IIa

1. For patients with a history of thrombus or a thromboembolic event who have an EF $<$ 25% (fractional shortening $<$ 15%), anti-coagulation with heparin or warfarin should be considered. **Level of Evidence C**
2. Extrapolating from the strong data in the adult HF population, children with low EF and persistent or

uncontrolled paroxysmal atrial fibrillation or flutter should receive anti-coagulation with heparin or warfarin. **Level of Evidence C**

Class III

1. No recommendation can be made regarding the use of anti-coagulation or anti-platelet therapy in patients with reduced EF and no history of thrombus or thromboembolic event, because there is insufficient evidence to justify a recommendation. **Level of Evidence C**

Nesiritide[71,72]

Class IIb

1. The use of nesiritide cannot be recommended for routine use in acute HF in children, although it may be considered in select situations where other interventions to lower central venous pressure have been unsuccessful. **Level of Evidence C**

Positive inotropic agents[73,74]

Class IIa

1. Inotropic therapy may be considered for symptomatic relief in the palliative setting. **Level of Evidence C**

Class III

1. On the basis of a lack of any pediatric data and lack of data supporting improved outcomes in adults, use of intermittent or chronic inotropic therapy, other than as a bridge to transplant, is not recommended. **Level of Evidence C**

Vasopressin receptor antagonists[75,76]

Class III

1. Vasopressin receptor antagonists cannot be recommended for the routine treatment of chronic HF in children. **Level of Evidence C**

Pharmacologic treatment of "preserved" EF (diastolic heart failure) recommendations

Chairs: David N Rosenthal and Robert Weintraub

Diuretics[77]

Class I

1. Use of diuretics to establish a clinically euvolemic state is recommended for children with HFpEF. **Level of Evidence C**
2. In patients with HFpEF, close monitoring of renal function and blood pressure should be performed during initiation and up-titration of diuretic therapy. **Level of Evidence C**

Class IIa

1. Treatment of systemic hypertension in patients with HFpEF is recommended to prevent disease progression. Although no particular class of medication is favored, diuretics may be considered for this purpose. **Level of Evidence C**

ACE inhibitors and angiotensin receptor blockade[78–80]

Class IIb

1. Routine use of renin-angiotensin antagonists is not recommended in HFpEF, unless there is an additional indication for use of these classes of medications such as hypertension. **Level of Evidence C**
2. Renin-angiotensin antagonists may be used for control of hypertension in HFpEF, but careful monitoring of hemodynamics and renal function is indicated due to the enhanced risk of hypotension and renal toxicity. **Level of Evidence C**

Calcium channel antagonists[81,82]

Class III

1. Use of calcium channel blockers is not recommended for treatment of HFpEF in children, unless there is an additional indication. **Level of Evidence C**

Mineralocorticoid/aldosterone receptor antagonists[83]

Class IIb

1. In children with HFpEF, aldosterone blockade with either spironolactone or eplerenone is not recommended. **Level of Evidence C**

Phosphodiesterase inhibitors[84,85]

Class IIb

1. Use of phosphodiesterase inhibitors is not recommended for treatment of HFpEF in children, unless there is an additional indication for use of these classes of medications such as pulmonary hypertension. **Level of Evidence C**

Digoxin and other digitalis glycosides

Class III

1. Use of digoxin is not recommended for treatment of HFpEF in children, unless there is an additional indication such as arrhythmia requiring atrial rate control. **Level of Evidence C**

Positive inotropic agents

Class III

1. Intravenous β-agonists, such as dopamine, dobutamine, and epinephrine, are not indicated for treatment of HFpEF. **Level of Evidence C**

Pulmonary vasodilators[86]

Class III

1. The use of prostaglandins and endothelin receptor antagonists to treat secondary pulmonary hypertension in children with HFpEF is not supported by current evidence. **Level of Evidence C**

Electrophysiology intervention recommendations

Chairs: David N. Rosenthal and Anne M. Dubin

Pacemaker therapy[87,88]

Class I

1. Permanent pacemaker implantation is recommended for advanced second- or third-degree atrioventricular block associated with ventricular dysfunction. **Level of Evidence B**

Class IIa

1. LV apical pacing can be useful in epicardial ventricular pacing systems. Technical considerations may require alternate ventricular lead placement. **Level of Evidence B**

Cardiac resynchronization therapy[89–91]

Class IIa

1. CRT can be useful for pediatric patients with a systemic LV with an EF $<$ 35%, complete left bundle branch block pattern, QRS duration (native or paced) $>$ ULN for age, NYHA Class II-IV on GDMT. **Level of Evidence B**

Class IIb

1. CRT may be considered for pediatric patients with a systemic RV, with an EF $<$ 35%, complete right bundle branch block pattern, QRS duration (native or paced) $>$ ULN for age, NYHA Class II-IV on GDMT. **Level of Evidence C**
2. CRT may be considered for pediatric patients with a single ventricle, with an EF $<$ 35%, complete bundle branch pattern, QRS duration (native or paced) $>$ ULN for age, NYHA Class II-V on GDMT. **Level of Evidence C**

ICD therapy[68,92–94]

Class I

1. ICD implantation is recommended in the pediatric survivor of cardiac arrest after evaluation to define the cause of the event and to exclude any reversible/treatable causes. **Level of Evidence B**

Class IIa

1. ICD implantation can be useful in the pediatric patient with unexplained syncope and at least moderate LV dysfunction and DCM. **Level of Evidence C**
2. ICD implantation can be useful for adolescent patients with HCM who have 1 or more major risk factors for SCD. In younger patients, the risk/benefit ratio must be considered due to technical considerations. **Level of Evidence C**
3. ICD implantation can be useful for the prevention of SCD in adolescent patients with AVC who have 1 or more risk factors for SCD. In younger patients, the risk/benefit ratio must be considered due to technical considerations. **Level of Evidence C**
4. ICD therapy can be useful in adolescent patients with a familial cardiomyopathy associated with SCD. In younger patients, the risk/benefit ratio must be considered due to technical considerations. **Level of Evidence C**

Class IIb

1. ICD therapy may be considered in pediatric patients with DCM who have an LVEF $<$ 35% and who are in NYHA Class II or III. **Level of Evidence C**
2. ICD implantation may be considered for patients with CHD with syncope in the presence of ventricular dysfunction. **Level of Evidence C**
3. ICD therapy may be considered in adolescent patients with LVNC and moderately depressed ventricular function. In younger patients, the risk/benefit ratio must be considered due to technical considerations. **Level of Evidence C**
4. ICD therapy may be considered in non-hospitalized pediatric patients with non-sustained or sustained ventricular tachycardia who required a VAD. **Level of Evidence C**

Ablation therapy

Class I

1. Ablation therapy is recommended in the pediatric patient with tachycardia-induced cardiomyopathy when medical therapy fails. **Level of Evidence B**

Class IIa

1. Ablation therapy can be useful as primary therapy in the adolescent patient with tachycardia-induced cardiomyopathy. **Level of Evidence B**

Class IIb

1. Ablation therapy may be considered in the pediatric patient with frequent premature ventricular contraction and cardiomyopathy of unknown etiology when medical therapy fails. **Level of Evidence B**

Mechanical circulatory support recommendations

Chairs: David N. Rosenthal and David Morales

Durable VAD system[95–97]

Class I

1. Implantation of a durable VAD system should be considered as a bridge to transplantation for children who are unable to be weaned from inotropic support and are showing early, reversible dysfunction of at least 1 other major organ system. **Level of Evidence C**

Class IIa

1. Management of chronic VAD devices in children, including anticoagulation medications, should be performed by a specialized team with appropriate expertise and focus. **Level of Evidence C**

Class IIb

1. Implantation of a chronic VAD system for children who are not eligible for transplantation may be considered as long-term support (destination therapy), provided a system is available that permits discharge to home with regular outpatient follow-up. **Level of Evidence C**

Temporary circulatory support[98–100]

Class IIa

1. For a child in cardiac arrest or cardiogenic shock with pulmonary compromise, ECMO should be considered for emergency cardiovascular support, as a temporizing measure as a bridge to recovery of function. **Level of Evidence C**
2. For a child with isolated cardiac failure that is believed to be reversible, ECMO or a temporary VAD may be considered as a temporizing measure as a bridge to recovery of function. If recovery does not occur, then transition to a chronic VAD for bridge to transplantation or for destination therapy (if child can receive a second- or third-generation VAD), is reasonable. **Level of Evidence C**
3. For a child with cardiogenic shock that is not believed to be due to a reversible underlying cause, consideration should be given to use of a temporary VAD or ECMO for resuscitation of end-organ function rather than directly implanting a chronic VAD system. **Level of Evidence C**

BiVAD support[95,101,102]

Class IIb

1. Use of BiVAD support should be minimized, reserving BiVAD support for patients who appear unlikely to achieve adequate hemodynamics from LVAD support alone. However, decision making for this remains highly individualized, with no broadly useful risk-stratification scheme available. **Level of Evidence C**

VAD support in the univentricular heart[103–106]

Class I

1. For neonates with univentricular circulation who require circulatory support for a reversible cause, ECMO should be considered. **Level of Evidence C**

Class IIb

1. For neonates with univentricular circulation who require long-term support as a bridge to transplantation, ECMO should only be deployed after careful evaluation of anticipated waiting list times and with consideration to donor scarcity. **Level of Evidence B**
2. Use of a long-term VAD for circulatory support may be considered in a neonate or older child with failed univentricular circulation, but outcomes with this support are poor for neonates and moderate for older children. **Level of Evidence B**

Class III

1. Use of a long-term VAD may be considered for circulatory support in a neonate with univentricular circulation who has failed to wean from cardiopulmonary bypass after palliative surgery but is not routinely recommended due to poor outcomes with this support. **Level of Evidence B**

Cardiac recovery[107]

Class IIb

1. Children who are supported on a chronic VAD system may be considered for a recovery protocol and weaning from VAD if recovery of cardiac function is documented. **Level of Evidence C**

Comorbidity recommendations

Chairs: David N. Rosenthal and Melanie Everitt

Anemia[108–110]

Class I

1. At the time of diagnosis and at ongoing regular intervals thereafter, the presence of anemia should be determined through measurement of plasma hemoglobin or hematocrit levels. **Level of Evidence C**
2. Iron deficiency as a treatable cause of anemia in the patient with HF should be sought by obtaining iron studies (ferritin with transferrin saturation). **Level of Evidence C**
3. It is reasonable to consider the use of intravenous iron to treat iron-deficiency anemia in pediatric patients with HF. **Level of Evidence C**

Class IIa

1. It is reasonable to consider the use of erythropoiesis-stimulating agents to treat anemia in pediatric patients with HF if anemia persists once iron stores are replete. **Level of Evidence C**

2. Restrictive transfusion thresholds appear to have no detrimental effect on outcomes and should be considered in the stable patient to reduce overall blood product exposure. **Level of Evidence C**

Class III

1. There is no evidence to support the use of intravenous iron or erythropoiesis-stimulating agents for the prevention of HF-associated anemia in children. **Level of Evidence C**

Renal dysfunction[111–113]

Class I

1. At the time of the HF diagnosis and at ongoing regular intervals, including after changes in medical therapy, the presence and severity of renal dysfunction should be determined. **Level of Evidence B**
2. Management of acute kidney injury should include strict recording of fluid balance, daily weight measurements, and calculation of the blood urea nitrogen/creatinine ratio to avoid poor renal perfusion secondary to dehydration**. Level of Evidence C**
3. Worsening renal function should prompt a review and possible adjustment of medications known to impair renal perfusion or function and medications that are renally excreted. **Level of Evidence A**

Class IIa

1. The modified Schwartz formula is reasonable to use in calculating glomerular filtration rate in pediatric patients aged older than 2 years. **Level of Evidence B**

Airway and parenchymal respiratory morbidity[114,115]

Class I

1. Chest X-ray imaging should be obtained in the pediatric HF patient who presents with new or worsening respiratory symptoms to evaluate for treatable pulmonary complications. **Level of Evidence C**
2. Flexible bronchoscopy is a safe and effective first-line approach to diagnosing bronchial compression due to cardiomegaly as a cause for recurrent pneumonia or persistent atelectasis in the child with HF. **Level of Evidence C**
3. Polysomnography should be performed in patients with signs and symptoms of sleep-related breathing disorders, especially when pulmonary hypertension is present. **Level of Evidence C**
4. Pulmonary function testing should be used for detection and reversibility of obstructive-related breathing disorders, especially when pulmonary hypertension is present. **Level of Evidence C**
5. Pulmonary function testing should be used for detection and reversibility of obstructive defects as well as for the diagnosis of restrictive lung disease. **Level of Evidence C**
6. Exercise testing with metabolic studies should be used to determine a component of respiratory limitation to exercise in symptoms with HF. **Level of Evidence C**

Class III

1. During acute intercurrent pulmonary illness (i.e., pneumonia, asthma exacerbation, bronchitis), the abrupt discontinuation of β-blockers and ACE inhibitors is not indicated unless there is a life-threatening complication related to their use. **Level of Evidence B**
2. Classical asthma medications, such as bronchodilators or corticosteroids, are generally not effective in the treatment of cardiac asthma. **Level of Evidence B**

Infectious diseases[116,117]

Class I

1. Pediatric HF patients aged $\leq$ 24 months, who meet criteria for prophylaxis, should receive palivizumab to reduce the risk of respiratory syncytial virus lower respiratory tract disease using published guidelines**. Level of Evidence A**
2. Immunizations reduce the incidence of respiratory infection, including community-acquired pneumonia and influenza. Children with HF should receive complete and age-appropriate immunizations, including pneumococcal conjugate/polysaccharide vaccines and an annual influenza vaccine. Annually updated vaccine schedules are provided by the Centers for Disease Control and Prevention. **Level of Evidence A**
3. Children with HF who might require transplantation should have a pre-transplant assessment of routine vaccinations, including rotavirus and other live virus vaccines. Catch-up and/or accelerated vaccination schedules should be used to ensure complete immunization before transplant. **Level of Evidence A**
4. Families, care givers, and close contacts of children with HF should receive all recommended vaccines, including a tetanus/diphtheria/pertussis immunization/booster and a yearly influenza vaccine. **Level of Evidence A**
5. Because pediatric patients with HF are at increased risk for health care-associated infections due to severity of illness and treatment interventions, validated infection prevention measures to reduce the risk of nosocomial infections should be encouraged. **Level of Evidence A**

Malnutrition and cachexia[118–120]

Class IIa

1. In the absence of clinical parameters to define cardiac cachexia in pediatrics, children plotting below the third percentile (–2 standard deviation) require further assessment, referral, or intervention for cachexia. **Level of Evidence C**
2. Energy and nutrient intake and barriers to intake should be individually assessed regularly. **Level of Evidence C**

Class III

1. There is no evidence to support the routine use of vitamin and mineral supplements in children unless indicated to treat a specific, documented deficiency. **Level of Evidence C**

Metabolic syndrome[121,122]

Class I

1. The presence of obesity in pediatric patients with heart disease should prompt specific evaluation for metabolic syndrome and all other cardiovascular risk factors, including hypertension, dyslipidemia, insulin resistance, and liver disease. **Level of Evidence A**
2. An intensive, multidisciplinary weight-reduction program and management of other identifiable risk factors should be initiated in pediatric patients with metabolic syndrome. **Level of Evidence B**

Depression and psychologic functioning[123–125]

Class I

1. Children with HF should be screened for mood disorders, including anxiety, depression, adjustment disorder, and sleep disorder. **Level of Evidence B**
2. The need for psychologic support should be discussed and provided as deemed appropriate before, immediately after, and during follow-up of children undergoing ICD and/or VAD implantation. **Level of Evidence C**

Cognitive and psychosocial performance[13,126–128]

Class I

1. Assessments of overall intelligence, speech/language, and motor development should be performed at least yearly in all patients with chronic HF. Early intervention and school accommodations are recommended for those with social and/or cognitive deficits**. Level of Evidence C**
2. Referral to a developmental specialist is recommended in all patients with chronic HF who are not meeting developmental milestones or who are demonstrating deficits in social or cognitive development. **Level of Evidence C**
3. Infants with chronic HF should be referred to appropriate early intervention programs, especially those with coexisting CHD. **Level of Evidence C**
4. School attendance and age-appropriate developmentally stimulating activities should be provided for medically stable patients with congestive HF, both in the inpatient and outpatient setting. **Level of Evidence C**

Class IIa

1. Brain imaging should be considered in patients with chronic HF and deficits in social and/or cognitive performance, but does not need to be performed routinely. **Level of Evidence B**
2. Brain imaging should be considered in patients with destination VAD therapy and deficits in social and/or cognitive performance, but does not need to be performed routinely. **Level of Evidence C**

Exercise training and activity recommendations[129,130]

Class I

1. Pre-participation health screening and risk stratification should be performed before initiating a program of exercise training to identify children at risk for adverse events during exercise. **Level of Evidence C**
2. Pediatric patients with HF should undergo cardiopulmonary exercise testing (age $\geq$ 6–8 years) before initiating exercise training to determine exercise capacity, assess risk for adverse events, and determine suitability for exercise training. **Level of Evidence C**
3. If deemed safe, exercise training in a supervised setting should be prescribed as an adjunctive approach to improve clinical status in ambulatory patients with current or prior symptoms of HF. **Level of Evidence C**

Class IIa

1. An exercise-training program should be individualized to the patient's ability and the patient's response to exercise, with an emphasis on safety. Recommendations for a supervised exercise program should include the frequency, intensity, time, and type of exercise. **Level of Evidence C**
2. Medical contraindications to an exercise-training program should be assessed. Detection of children at risk for sudden death and appropriate recommendations for a defibrillator as a primary or secondary intervention is essential before initiating an exercise program or increasing the frequency, intensity, or duration of a current program. **Level of Evidence C**
3. Informed consent from parents and/or assent from the child should be obtained at each session for exercise training. **Level of Evidence C**
4. Exercise training for children with HF should be performed by personnel with expertise in pediatric exercise physiology and in a facility with the ability to monitor vital signs and perform cardiopulmonary resuscitation. **Level of Evidence C**

Acute HF recommendations

Chairs: Anne I. Dipchand and Elfriede Pahl

Initial assessment[131–133]

Class I

1. The diagnosis of acute HF should be based on signs and symptoms of HF combined with supportive evidence from chest X-ray imaging, ECG, echocardiography, and laboratory evaluations, including BNP. **Level of Evidence B**

Table 4 Categories of Acute Decompensated Heart Failure

Variable	No congestion	Congestion
Adequate perfusion	"Warm and dry" Optimal profile: focus on prevention of disease progression and decompensation	"Warm and wet" Diuresis with continuation of standard therapy
Critical hypoperfusion	"Cold and dry" Limited further options for therapy	"Cold and Wet" Diuresis and redesign of regimen with other standard therapies

2. An assessment should be made of the severity of HF, including degree of congestion and adequacy of perfusion (Table 4). **Level of Evidence C**
3. An evaluation for the etiology of HF should be performed, with special attention to identifying reversible causes (e.g., repairable CHD, myocarditis, tachycardia-induced cardiomyopathy, and hypothyroidism). **Level of Evidence C**

Monitoring[134]

Class I

1. In children hospitalized with acute HF, initial observation in an intensive care unit setting should be considered. **Level of Evidence C**
2. In children hospitalized with acute HF, evaluation and monitoring for arrhythmias with continuous ECG monitoring/telemetry is warranted. **Level of Evidence C**
3. Manual blood pressure determination to validate oscillometric determinations during initial evaluations is warranted in the absence of an intra-arterial catheter. **Level of Evidence C**
4. It is reasonable to transfer patients with severe acute HF to a center with pediatric HF specialists and the expertise and ability to optimize medical therapy, evaluate for heart transplant, and if necessary, provide mechanical support. **Level of Evidence C**

Class IIa

1. In children with decompensated HF, it is reasonable to place intra-arterial catheters for continuous blood pressure monitoring in an acute care setting. **Level of Evidence C**
2. Central venous catheters should be considered in decompensated HF in an acute care setting to allow for measurement of central venous pressure and/or mixed venous saturations and to administer medications and fluids. **Level of Evidence C**

Class III

1. Pulmonary artery catheterization in children with acute HF is not recommended for routine use, but may be appropriate in selected patients. **Level of Evidence C**

Near-infrared spectroscopy

Class IIb

1. Trends in cerebral or somatic regional oxygen saturation and/or venous oxygen saturation levels may be useful in unstable patients with acute heart failure. **Level of Evidence C**

Alternative methods for CO/hemodynamic assessment[135]

Class III

1. Alternative methods for CO/hemodynamic assessment in acute HF are not recommended for routine use at this time. **Level of Evidence B**

Indications for cardiac catheterization[45]

Class I

1. Coronary angiography is indicated for patients with acute HF if coronary ischemia is suspected in the setting of other potential abnormalities that cannot be definitely excluded by non-invasive imaging. **Level of Evidence C**
2. Cardiac catheterization is indicated in patients with palliated or repaired CHD who present with acute HF if a non-invasive evaluation fails to establish a definitive diagnosis. **Level of Evidence C**

Serial testing

Class I

1. Serial testing to monitor for electrolyte abnormalities, hemoglobin levels, end-organ perfusion, and response to therapy are indicated for patients with acute HF. **Level of Evidence C**

Inotropes[136–138]

Table 5 summarizes the inotropic characteristics

Class I

1. Intravenous inotropic support may be temporarily used in patients with acute HF presenting as cardiogenic shock with poor systemic and end-organ perfusion. **Level of Evidence C**
2. Vasodilators may be used in pediatric patients with acute HF in the absence of hypotension. Vasodilators may be used in combination with diuretics for symptomatic relief in patients with pulmonary edema. **Level of Evidence C**

Class IIa

1. Intravenous inotropic support may be temporarily used in patients with acute HF presenting as hypotension with evidence of low CO and compromised end-organ perfusion. **Level of Evidence C**

Table 5 Inotropes Characteristics

Inotrope	α1	β1	β2	DAR	Half-life	CO	HR	SBP	PCWP	Myocardial O_2 consumption
Dobutamine	+	+ + + + +	+ + +	N/A	2–3 min	↑	↑	↔	↓	↑
Epinephrine	+ + + + +	+ + + +	+ + +	N/A	2–7 min	↑	↑	↑	↔	↑
Dopamine	+ + +	+ + + +	++	+ + + + +	2–20 min	↑	↑	↑	↔	↑
Milrinone	N/A	N/A	N/A	N/A	1-4 hours	↑	↑	↓	↓	↔
Levosimendan	N/A	N/A	N/A	N/A	1–1.5 hours	↑	↑	↓	↓	↔

Expansions for the abbreviations used in Table 5 are provided in Appendix 3.

2. The choice of inotropic agent(s) for a child in acute decompensated HF will depend on clinical presentation. Milrinone and/or dobutamine can be used as first-line rescue therapy, with epinephrine playing a role in the face of refractory hypotension and poor end-organ perfusion. **Level of Evidence C**

Class IIb

1. Levosimendan may be considered in children with acute decompensated HF unresponsive to traditional inotropic therapy. **Level of Evidence C**

Class III

1. Use of intravenous inotropic agents in the absence of clinical evidence of hypotension, impaired perfusion, low CO, and/or decreased end-organ perfusion is potentially harmful. **Level of Evidence B**

Corticosteroids[65]

Class IIb

1. In patients with hemodynamic compromise secondary to HF, consideration may be given to evaluation and treatment of adrenal insufficiency. **Level of Evidence C**

Anti-coagulation

Class IIa

1. In patients with severe cardiac dysfunction, prophylactic anti-coagulation may be considered, especially in the acute care setting and in the presence of indwelling intravascular catheters. **Level of Evidence C**

Thyroid hormone replacement[139]

Class IIa

1. Evaluation of a critically ill patient's systemic thyroid homeostasis is reasonable, and if hypothyroidism is identified, thyroid hormone replacement may be considered. **Level of Evidence C**

Class IIb

1. There is no indication for the routine use of thyroid hormone to treat acute HF in the absence of documented abnormal thyroid function. **Level of Evidence C**
2. Thyroid hormone can be considered for use in the acutely unwell post-cardiac surgery patient. **Level of Evidence B**

Immunomodulation[140,141]

Class IIb

1. The evidence in the literature does not support the routine use of corticosteroids in children with myocarditis. **Level of Evidence C**
2. The evidence in the literature does not support the routine use of intravenous immunoglobulin in children with myocarditis. **Level of Evidence C**

Ventilation[41,131]

Class I

1. Invasive ventilation should be considered in patients with acute decompensated HF and respiratory compromise in the setting of pulmonary edema and/or low CO. **Level of Evidence C**

Class IIa

1. Non-invasive ventilation may be considered for the treatment of children with acute HF, pulmonary edema, and significant increased work of breathing as an adjunct to other medical therapies. **Level of Evidence C**

Class IIb

1. Treatment with non-invasive ventilation might be reasonable in children with symptomatic HF in the absence of pulmonary edema as an adjunctive measure. **Level of Evidence C**

Fluid management[65,142–144]

Class 1

1. An initial and ongoing assessment of fluid status should be performed in all patients admitted to the hospital with acute HF. **Level of Evidence C**
2. Diuretics are the first-line therapy for patients admitted with evidence of fluid overload. **Level of Evidence C**
3. Careful monitoring for side effects of anti-congestive therapies, including renal function, electrolytes, and hypotension, should be performed. **Level of Evidence C**

Class IIa

1. Fluid restriction is reasonable for patients with acute HF, regardless of serum sodium level. **Level of Evidence C**
2. A low-sodium diet is reasonable for hospitalized patients. **Level of Evidence C**

Class III

1. Sodium supplementation is not recommended in children with HF. **Level of Evidence C**

Nutrition[145]

Class I

1. A nutritional assessment should be performed on hospitalized patients with HF. **Level of Evidence C**

Considerations in the treatment of acute right HF[146]

Class IIa

1. It is reasonable to use dobutamine, dopamine, or low-dose epinephrine in the setting of right HF. **Level of Evidence C**
2. It is reasonable to use milrinone as a pulmonary vasodilator in the setting of right HF. **Level of Evidence C**

Peri-operative acute HF[147–150]

Class I

1. Prompt evaluation and treatment of residual hemodynamically significant lesions should be performed when possible for those with early post-operative HF. **Level of Evidence C**

Class IIa

1. It is reasonable to use milrinone after cardiac surgery in infants and children to reduce the risk of LCOS. **Level of Evidence B**
2. Mechanical circulatory support (ECMO or VAD) is reasonable to use as a bridge to recovery or transplant for infants and children unable to wean from bypass or with refractory LCOS after cardiac surgery. **Level of Evidence C**

Transplantation listing recommendations

Chairs: Richard Kirk and Robert Gajarski

Pediatric listing guidance was published in 2007, and the ISHLT is currently updating its guidance, including both pediatric and adult congenital listing criteria.[151,152] Accordingly, there are no listing recommendations in this document.

Table 6 Proposed Metrics for Heart Failure

Appointments	A. Patients are provided with a calendar of scheduled visits (primary cardiologist, HF clinic, outpatient therapies, etc) before hospital discharge B. Calendar is refreshed with visits as cycles come to a close C. Non-attendance is documented D. Electronic application for e-mail and remote scheduling available
Program adherence to guidelines	A. HF management protocols are documented B. Patient variation from protocols are documented with reasons C. Documentation of HF severity and staging.
Frequency or number of unplanned readmissions	A. HF readmissions and emergency department visits B. Non-HF–related hospital admissions and emergency department visits
Monitoring of biomarkers, end-organ function, etc	A. BNP, NT-pro-BNP measurements B. General metabolic panel for electrolytes, renal function C. Hepatic function D. Drug levels (if appropriate)
Remote monitoring	A. Physical parameters (feeding, weight gain) B. Pacemaker/ICD interrogation if implanted C. Loop or event recorders regarding arrhythmias
Exercise testing	A. Metabolic exercise testing over time
Performance of activities of daily living	A. Feeding, sleep, daily activity journal B. Age-appropriate participation: school attendance, peer activity participation C. Meeting age-appropriate developmental milestones D. Cognitive development
Growth	A. Growth percentiles and maintenance over time
Nutrition	A. Age-appropriate nutritional plan provided B. Tracking intake at home with calorie estimation during visits or remotely
Home monitoring program	C. Home monitoring program availability and structure. D. Oversight provided by experienced nurse/nurse practitioner. E. Tracking of interventions invoked as result of home surveillance.

Expansions for the abbreviations used in Table 6 are provided in Appendix 3.

Health care delivery recommendations

Chairs: Richard Kirk and Maryanne Chrisant

Readiness for discharge[153,154]

Class I

1. Discharge criteria (Table 6), if possible, should be met before hospital discharge. **Level of Evidence C**
2. Discharge planning should address medication regimen, fluid and sodium restriction, recommended activity levels, and establishment of early follow-up. **Level of Evidence C**

Home surveillance and monitoring[155–157]

Class IIb

1. Non-invasive home surveillance and monitoring may have a place in HF management. **Level of Evidence C**

Class III

1. Invasive home monitoring is not recommended because hospitalization is not reduced in adult trials. **Level of Evidence C**

Transition[158,159]

Class I

1. A formal transition program, beginning several years before transfer of care from the pediatric to the adult team, should be a part of routine HF care. **Level of Evidence C**

Palliative care[160]

Class IIb

1. Pediatric palliative care should form part of the multidisciplinary care of all patients with potentially end-stage HF, including patients receiving ECMO, VAD support, and patients listed for transplantation. **Level of Evidence C**

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Appendix 3 Abbreviations

2-D	two-dimensional
3-D	three-dimensional
Mitral A velocity	mitral atrial wave velocity
ACE	angiotensin converting enzyme
AVC	arrhythmogenic ventricular cardiomyopathy
BiVAD	biventricular assist device
BNP	B-type natriuretic peptide
CHD	congenital heart disease
CRT	cardiac resynchronization therapy
CT	computed tomography
CO	cardiac output
DAR	dopaminergic receptor
DCM	dilated cardiomyopathy
dP/dt	echocardiographic measure of change in pressure over change in time
DSC2	Desmocollin-2
DSG2	Desmoglein-2
DSP	Desmoplakin
e′	early mitral annulus descent velocity
E velocity	early mitral inflow velocity
ECG	electrocardiogram
ECMO	extracorporeal membrane oxygenation
EF	ejection fraction
EFE	endomyocardial fibroelastosis
EMB	endomyocardial biopsy
GDMT	guideline-determined medical therapy
HCM	hypertrophic cardiomyopathy
HF	heart failure
HFpEF	heart failure with preserved ejection fraction
HR	heart rate
ICD	implantable cardiac defibrillator
JUP	junctional plakoglobin
LCOS	low cardiac output syndrome

LMNA	lamin A/C
LV	left ventricle
LVEF	left ventricular ejection fraction
LVNC	left ventricular non-compaction
MCS	mechanical circulatory support
MPI	myocardial performance index
MR	magnetic resonance
MRI	magnetic resonance imaging
N/A	not applicable
NT-proBNP	N-terminal pro-B-type natriuretic peptide
NYHA	New York Heart Association
PCWP	pulmonary capillary wedge pressure
PKP2	plakophillin-2
PVR	pulmonary vascular resistance
RCM	restrictive cardiomyopathy
RV	right ventricle
SBP	systolic blood pressure
SCD	sudden cardiac death
S/D ratio	ratio of systolic-to-diastolic duration
SCN5	sodium channel, voltage-gated, type V, alpha subunit
TDI	tissue Doppler imaging
TMTM43	transmembrane protein 43
ULN	upper limit normal
VAD	ventricular assist device
Vo_2	volume of oxygen consumption

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