

GUIDELINE

Authors Insights on the Updated International Society for Heart and Lung Transplantation Guidelines for the Management of Pediatric Heart Failure (Update From 2014)



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Pediatric heart failure (HF) secondary to cardiomyopathies, acquired heart disease, and congenital heart disease is associated with significant morbidity and mortality. These guidelines represent an update from the International Society for Heart and Lung Transplantation guidelines for the management of pediatric HF that were published in 2014 and incorporate interval advancements in medical therapies and new approaches in the evaluation and management of children with HF. J Heart Lung Transplant 2025;44:1529–1534

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1. INTRODUCTION

Pediatric heart failure (HF) secondary to cardiomyopathies, acquired heart disease, and congenital heart disease remains associated with significant morbidity and mortality. These guidelines represent an update from the International Society for Heart and Lung Transplantation guidelines for the management of pediatric HF that were published in 2014. The purpose of this update is to incorporate advancements in pharmacologic therapies and new approaches in the evaluation and management of children with HF since the prior guideline publication. These guidelines are not intended for use in the management of HF related to left to right shunts and are primarily focused on the population of children with HF due to dysfunction of the myocardium. Because the indications for heart transplantation in children, selection and management of ventricular assist devices in children and patients with congenital heart disease, management of cardiovascular implantable electronic devices in children, the diagnosis and treatment of hypertrophic cardiomyopathy, and the diagnosis and management of cardiac complications of muscular dystrophies are covered by existing consensus statements or guidelines, they are largely not included in this document.³⁻⁷

2. TOP TAKEAWAYS

- 1. The 2022 Adult Heart Failure guidelines proposed a new mechanism for classifying HF by left ventricular ejection fraction (EF).⁸ There are no data from randomized controlled trials (RCTs) or even descriptive studies in pediatric HF displaying differing prognosis or response to treatment based on EF. However, there may be benefits in extrapolating this adult–based classification strategy into the pediatric population and a proposed classification of HF by EF in children is included in these updated guidelines.
- 2. Several Class III recommendations from the 2014 Pediatric Heart Failure Management Guidelines have been removed. These recommendations were removed due to a lack of sufficient evidence that definitively justifies Class III status, combined with the risk of excluding potentially beneficial therapies for certain subgroups as emerging evidence evolves.
- 3. There are new recommendations for the treatment of children with HF and reduced EF (HFrEF) based on the development of new medical therapies proven to be efficacious in adults. PANORAMA-HF was a RCT in children comparing the angiotensin receptor neprilysin inhibitor, sacubitril-valsartan, to the angiotensin–converting enzyme inhibitor, enalapril. While the results of PANORAMA-HF were neutral at study completion, sacubitril-valsartan received approval from the US Food and Drug Administration for the treatment of children over the age of 1 year with symptomatic left ventricular systolic dysfunction.
- 4. Ivabridine reduces heart rate by inhibiting I_f channels and slowing phase 4 depolarization. Ivabridine is recommended for the treatment of HFrEF in select adult patients. ^{8,10} In a RCT of ivabradine vs placebo in children with HFrEF, ivabradine met the primary end-point of heart rate reduction and was therefore approved by the US Food and Drug Administration for children older than 6 months of age with symptomatic chronic HF. ¹¹
- 5. Sodium-glucose cotransporter 2 (SGLT2) inhibitors have proven to be highly efficacious in the treatment of adults with HFrEF. 12,13 There are limited data and no RCTs for SGLT2 inhibitors for the treatment of children with HFrEF. 14,15 However, based on extrapolation of adult data, a pediatric recommendation is made.
- 6. The phase III VICTORIA study investigated oral vericiguat, a soluble guanylate cyclase inhibitor, vs placebo in adults with high-risk HF. Based on the results of this trial, vericiguat was assigned a Class 2b recommendation for adults with HFrEF, elevated natriuretic peptide levels, and a recent HF hospitalization or use of intravenous diuretics. A phase II/III randomized clinical trial (VALOR study) to investigate efficacy, safety, and the pharmacokinetic profile of vericiguat in children with stable systolic HF is in progress at the time of this publication. A recommendation for the use of vericiguat is made based on extrapolation of adult clinical trial data.
- 7. A new recommendation for the treatment of children with HF and preserved EF with SGLT2 inhibitors is made based on the extrapolation of adult clinical trial data. 12,17
- 8. The updated guidelines include new recommendations related to anesthetic considerations for children with HF, novel pharmacologic therapies (e.g., direct oral anticoagulants), expansion of recommendations for electrophysiology testing and treatment, and treatment approaches for special populations, including those

- with single ventricle congenital heart disease, prior exposure to cardiotoxic cancer therapy, a diagnosis of storage diseases or dystrophinopathies.
- 9. There are new sections with accompanying recommendations addressing advancements in technology (e.g., wearable devices) and nonpharmacologic interventions, such as nutrition and exercise.
- 10. New sections emphasize health equity, including addressing barriers to care and health literacy, the importance of evaluating goals of care, including the role of hospice, and the significance of monitoring quality metrics and reporting to optimize outcomes for children with HF.

3. CONCLUSION

These updated guidelines for the management of pediatric HF incorporate emerging evidence, novel pharmacologic therapies, and advancements in technology since the 2014 version. By integrating data from pediatric studies and extrapolating from adult clinical trials, these recommendations provide a framework to harmonize care, reduce practice variation, and promote evidence–based decision-making. However, despite increasing efforts to elucidate the mechanisms of pediatric HF and expand clinical research in this population, substantial gaps in evidence remain. These guidelines should not displace equipoise for therapies lacking definitive pediatric data to ensure that future trials can be conducted to further refine treatment strategies. Additionally, these guidelines emphasize the importance of health equity, multidisciplinary care models, and quality improvement initiatives to enhance outcomes for children with HF. As research continues to progress, sustained collaboration among clinicians, researchers, industry partners, and policymakers will be essential for driving advancements in the field and improving care for this vulnerable population.

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| Abbreviations: NIH/HNHLBI, National Institutes of Health/National Heart Lung and Blood Institute. | | | | | | | |

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References

- 1. Irving C, Azeka E, Adorisio R, et al. The International Society for Heart and Lung Transplantation guidelines for the management of pediatric heart failure (update from 2014). J Heart Lung Transplant 2025.
- 2. Kirk R, Dipchand AI, Rosenthal DN, et al. The International Society for Heart and Lung Transplantation Guidelines for the management of pediatric heart failure: executive summary [Corrected]. J Heart Lung Transplant 2014;33:888-909.
- 3. Canter CE, Shaddy RE, Bernstein D, et al. Indications for heart transplantation in pediatric heart disease: a scientific statement from the American Heart Association Council on Cardiovascular Disease in the Young; the Councils on Clinical Cardiology, Cardiovascular Nursing, and Cardiovascular Surgery and Anesthesia; and the Quality of Care and Outcomes Research Interdisciplinary Working Group. Circulation 2007;115:658-76.
- 4. Lorts A, Conway J, Schweiger M, et al. ISHLT consensus statement for the selection and management of pediatric and congenital heart disease patients on ventricular assist devices Endorsed by the American Heart Association. J Heart Lung Transplant 2021;40:709-32.
- 5. Writing Committee Members, Shah MJ, Silka MJ, et al. 2021 PACES expert consensus statement on the indications and management of cardiovascular implantable electronic devices in pediatric patients. Heart Rhythm 2021;18:1888-924.
- 6. Birnkrant DJ, Bushby K, Bann CM, et al. Diagnosis and management of Duchenne muscular dystrophy, part 2: respiratory, cardiac, bone health, and orthopaedic management. Lancet Neurol 2018:17:347-61.
- 7. Ommen SR, Ho CY, Asif IM, et al. 2024 AHA/ACC/AMSSM/HRS/PACES/SCMR guideline for the management of hypertrophic cardiomyopathy: a report of the American Heart Association/American College of Cardiology Joint Committee on Clinical Practice Guidelines. Circulation 2024;149:e1239-311.

- 8. Heidenreich PA, Bozkurt B, Aguilar D, et al. 2022 AHA/ACC/HFSA guideline for the management of heart failure: a report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. Circulation 2022;145:e895-1032.
- 9. Shaddy R, Burch M, Kantor PF, et al. Baseline characteristics of pediatric patients with heart failure due to systemic left ventricular systolic dysfunction in the PANORAMA-HF trial. Circ Heart Fail 2023;16:e009816.
- 10. McDonagh TA, Metra M, Adamo M, et al. 2021 ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure. Eur Heart J 2021;42:3599-726.
- 11. Bonnet D, Berger F, Jokinen E, Kantor PF, Daubeney PEF. Ivabradine in children with dilated cardiomyopathy and symptomatic chronic heart failure. J Am Coll Cardiol 2017;70:1262-72.
- 12. McMurray JJV, Solomon SD, Inzucchi SE, et al. Dapagliflozin in patients with heart failure and reduced ejection fraction. N Engl J Med 2019;381:1995-2008.
- 13. Santos-Gallego CG, Vargas-Delgado AP, Requena-Ibanez JA, et al. Randomized trial of empagliflozin in nondiabetic patients with heart failure and reduced ejection fraction. J Am Coll Cardiol 2021;77:243-55.
- 14. Konduri A, West C, Lowery R, et al. Experience with SGLT2 inhibitors in patients with single ventricle congenital heart disease and Fontan circulatory failure. Pediatr Cardiol 2023;46:81-8.
- 15. Newland DM, Law YM, Albers EL, et al. Early clinical experience with dapagliflozin in children with heart failure. Pediatr Cardiol 2023;44:146-52.
- 16 Efficacy, safety and pharmacokinetics of vericiguat in pediatric participants with heart failure due to left ventricular systolic dysfunction (MK-1242-036); 2023. Available at: www.clinicaltrials.gov.
- 17. Packer M, Anker SD, Butler J, et al. Cardiovascular and renal outcomes with empagliflozin in heart failure. N Engl J Med 2020;383:1413-24.