2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension: Developed by the task force for the diagnosis and treatment of pulmonary hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS). Endorsed by the International Society for Heart and Lung Transplantation (ISHLT) and the European Reference Network on rare respiratory diseases (ERN-LUNG).

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Section on Mechanical Circulatory Support

6.3.7.2. Mechanical circulatory support

In specialist centres, various forms of mechanical circulatory support are available for managing RV failure, with veno- arterial extracorporeal membrane oxygenation (ECMO) being the most widely used approach. Mechanical circulatory support has become an established bridging tool to transplantation in patients with irreversible right HF, but is occasionally used as a bridge to recovery in patients with treatable causes and potentially reversible RV failure.[468] No general recommendations can be made regarding the indication for mechanical circulatory support, which needs to be individualized, considering patient factors and local resources.[469,470] Long-term mechanical support analogous to left ventricular assist devices (LVADs) is not yet available for patients with PH and end- stage right HF.

Section on Mechanical Circulatory Support

Recommendation Table 12

Recommendations for intensive care management for pulmonary arterial hypertension

Recommendations	Class[a]	Level[b]
When managing patients with right HF in the ICU, it is recommended to involve physicians with expertise, treat causative factors, and use supportive measures, including inotropes and vasopresors, fluid management, and PAH drugs, as appropriate	I	C
Mechanical circulatory support may be an option for selected patients as a bridge to transplantation or recovery, and interhospital transfer should be considered if such resources are unavailable on site	IIa	C

Section on Lung and Heart-Lung Transplant

Lung transplantation remains an important treatment option for patients with PAH refractory to optimized medical therapy. In patients with PAH, referral to an LTx centre should be considered early (*Table 20*): (1) when they present with an inadequate response to treatment despite optimized combination therapy; (2) when they present with an intermediate–high or high risk of death (i.e. 1-year mortality >10% when estimated with established risk-stratification tools)471 (see *Section 6.2.7*), which exceeds the current mortality rate after LTx;472 (3) when patients have a disease variant that poorly responds to medical therapy, such as PVOD or PCH.

Section on Lung and Heart-Lung Transplant

Both heart–lung and bilateral LTx have been performed for PAH. Currently, most patients receive bilateral LTx, while combined heart–lung transplantation is reserved for patients who have additional non-correctable cardiac conditions.473 With the introduction of the lung allocation score (LAS), waiting list mortality has decreased and the odds of receiving a donor organ have increased.474 In some countries, an 'exceptional LAS' can be obtained for patients with severe PH. Some other countries not using the LAS have successfully implemented high-priority programmes for these patients.475 The patient and their next of kin should be fully engaged in the transplant assessment process and informed of the risks and benefits, and the final decision should be jointly made between the patient and medical team (see *Section 6.3.1.8*). For patients with PAH who survive the early post-transplant period, long-term outcomes are good. A study found that for primary transplant patients with IPAH who survived to 1 year, conditional median survival was 10.0 years.476

Section on Lung and Heart-Lung Transplant

Recommendation Table 13

Recommendations for lung transplantation

Recommendations	Class[a]	Level[b]
It is recommended that potentially eligible candidates are referred for LTx evaluation when they have an inadequate response to oral combination therapy, indicated by an intermediate–high or high risk or by a REVEAL risk score >7	I	C
It is recommended to list patients for LTx who present with a high risk of death or with a REVEAL risk score ≥10 despite receiving optimized medical therapy including s.c. or i.v. prostacyclin analogues	I	C