Selected revised recommendations (R) and new recommendations (N)						
New or revised	**Recommendation in 2015 version**	**Class**[a]	**Recommendation in 2022 version**	**Class**[a]	**Arbejdsgruppens anbefaling** **-Endorsement** **-Ændring**	**Arbejdsgruppens kommentarer**
Right heart catheterization and vasoreactivity testing – Recommendation Table 1						
N			It is recommended that RHC comprises a complete set of haemodynamics and is performed following standardized protocols	**I**	**Endorsement**	
R	Adenosine should be considered for performing vasoreactivity testing as an alternative	**IIa**	Inhaled nitric oxide, inhaled iloprost, or i.v. epoprostenol are recommended for performing vasoreactivity testing	**I**	**Endorsement**	
	Inhaled iloprost may be considered for performing vasoreactivity testing as an alternative					
Diagnostic strategy – Recommendation Table 2						
N			It is recommended to assign an echocardiographic probability of PH, based on an abnormal TRV and the presence of other echocardiographic signs suggestive of PH (see *Table 10*)	**I**	**Endorsement**	**Vi foreslår inkorporering af guideline figur 4, figur 5 og tabel 10 i guidelines på ekkokardiografi.dk afsnittet om RV vurdering.**
N			It is recommended to maintain the current threshold for TRV (>2.8 m/s) for echocardiographic probability of PH according to the updated haemodynamic definition	**I**	**Endorsement**	
N			Based on the probability of PH by echocardiography, further testing should be considered in the clinical context (i.e. symptoms and risk factors or associated conditions for PAH/CTEPH)	**IIa**	**Endorsement**	
N			In symptomatic patients with intermediate echocardiographic probability of PH, CPET may be considered to further determine the likelihood of PH	**IIb**	**Endorsement**	
Screening and improved detection of pulmonary arterial hypertension and chronic						

thrombo-embolic pulmonary hypertension – Recommendation Table 3						
N			In patients with SSc, an annual evaluation of the risk of having PAH is recommended	**I**	**Endorsement**	
R	Resting echocardiography is recommended as a screening test in asymptomatic patients with SSc, followed by annual screening with echocardiography, DLCO, and biomarkers	**I**	In adult patients with SSc of >3 years' disease duration, an FVC ≥40%, and a DLCO <60%, the DETECT algorithm is recommended to identify asymptomatic patients with PAH	**I**	**Endorsement**	**Dette varetages i samarbejde med reumatologiske og dermatologiske afdelinger**
N			In patients with SSc, where breathlessness remains unexplained following non-invasive assessment, RHC is recommended to exclude PAH	**I**	**Endorsement**	
N			Assessing the risk of having PAH, based on an evaluation of breathlessness, in combination with echocardiogram or PFTs and BNP/NT-proBNP, should be considered in patients with SSc	**IIa**	**Endorsement**	
N			Policies to evaluate the risk of having PAH should be considered in hospitals managing patients with SSc	**IIa**	**Endorsement**	
R	RHC is recommended in all cases of suspected PAH associated with CTD	**I**	In symptomatic patients with SSc, exercise echocardiography or CPET, or CMR may be considered to aid decisions to perform RHC	**IIb**	**Fjernes**	**Det er arbejdsgruppens holdning at for danske forhold er RHC den primære undersøgelse hvis der ved TTE og øvrig paraklinik er mistanke om PH. Derfor fastholdes oprindelige anbefaling**
N			In patients with CTD with overlap features of SSc, an annual evaluation of the risk of PAH may be considered	**IIb**	**Endorsement**	
R	In PE survivors with exercise dyspnoea, CTEPH should be considered	**IIa**	In patients with persistent or new-onset dyspnoea or exercise limitation following PE, further diagnostic evaluation to assess for CTEPH/CTEPD is recommended	**I**	**Endorsement**	
N			For symptomatic patients with mismatched perfusion lung defects beyond 3 months of anticoagulation for acute PE, referral to a	**I**	**Endorsement**	

			PH/CTEPH centre is recommended after considering the results of echocardiography, BNP/NT-proBNP, and/or CPET			
N			Counselling regarding the risk of PAH, and annual screening is recommended for individuals who test positive for PAH-causing mutations and in first-degree relatives of patients with HPAH	**I**	**Nedgraderes til klasse II rekommandation**	**Årlig screening er ikke velunderbygget og bør vurderes individuelt**
N			In patients referred for liver transplantation, echocardiography is recommended as a screening test for PH	**I**	**Endorsement**	
N			Further tests (echocardiography, BNP/NT-proBNP, PFTs, and/or CPET) should be considered in symptomatic patients with CTD, portal hypertension, or HIV to screen for PAH	**IIa**	**Endorsement**	
Evaluating the disease severity and risk of death in patients with pulmonary arterial hypertension – Recommendation Table 4						
N			For risk stratification at the time of diagnosis, the use of a three-strata model (low, intermediate, and high risk) is recommended, taking into account all available data including haemodynamics	**I**	**Endorsement**	
N			For risk stratification during follow-up, the use of a four-strata model (low, intermediate–low, intermediate–high, and high risk) based on WHO-FC, 6MWD, and BNP/NT-proBNP is recommended, with additional variables taken into account as necessary	**I**	**Endorsement**	
R	Achievement/maintenance of an intermediate-risk profile should be considered an inadequate treatment response for most patients with PAH	**IIa**	In some PAH aetiologies and in patients with comorbidities, optimization of therapy should be considered on an individual basis while acknowledging that a low-risk profile is not always achievable	**IIa**	**Endorsement**	

General measures and special circumstances – Recommendation Table 5						
R	Supervised exercise training should be considered in physically deconditioned PAH patients under medical therapy	**IIa**	Supervised exercise training is recommended in patients with PAH under medical therapy	**I**	**Endorsement**	
R	Immunization of PAH patients against influenza and pneumococcal infection is recommended	**I**	Immunization of patients with PAH against SARS-CoV-2, influenza, and *Streptococcus pneumoniae* is recommended	**I**	**Endorsement**	
R	Correction of anaemia and/or iron status may be considered in PAH patients	**IIb**	In the presence of iron-deficiency anaemia, correction of iron status is recommended in patients with PAH	**I**	**Endorsement**	
N			In the absence of anaemia, iron repletion may be considered in patients with PAH with iron deficiency	**IIb**	**Endorsement**	
R	Oral anticoagulant treatment may be considered in patients with IPAH, HPAH, and PAH due to use of anorexigens	**IIb**	Anticoagulation is not generally recommended in patients with PAH but may be considered on an individual basis	**IIb**	**Endorsement**	
R	The use of angiotensin-converting enzyme inhibitors, angiotensin-2 receptor antagonists, beta-blockers, and ivabradine is not recommended in patients with PAH unless required by comorbidities (i.e. high blood pressure, coronary artery disease, or left HF)	**III**	The use of ACEis, ARBs, ARNIs, SGLT-2is, beta-blockers, or ivabradine is not recommended in patients with PAH unless required by comorbidities (i.e. high blood pressure, coronary artery disease, left HF, or arrhythmias)	**III**	**Endorsement**	
R	In-flight O_2 administration should be considered for patients in WHO-FC III and IV and those with arterial blood O_2 pressure consistently <8 kPa (60 mmHg)	**IIa**	In-flight O_2 administration is recommended for patients using oxygen or whose arterial blood oxygen pressure is <8 kPa (60 mmHg) at sea level	**I**	**Endorsement**	
R	In elective surgery, epidural rather than general anaesthesia should be preferred whenever possible	**IIa**	For interventions requiring anaesthesia, multidisciplinary consultation at a PH centre to assess risk and benefit should be considered	**IIa**	**Endorsement**	
Women of childbearing potential – Recommendation Table 6						
R	It is recommended that PAH patients avoid pregnancy	**I**	It is recommended that women of childbearing potential with PAH are counselled at the	**I**	**Endorsement**	

			time of diagnosis about the risks and uncertainties associated with becoming pregnant; this should include advice against becoming pregnant, and referral for psychological support where needed			
N			It is recommended that women of childbearing potential with PAH be provided with clear contraceptive advice, considering the individual needs of the woman but recognizing that the implications of contraceptive failure are significant in PAH	**I**	**Endorsement**	
N			It is recommended that women with PAH who consider pregnancy or who become pregnant receive prompt counselling in an experienced PH centre to facilitate genetic counselling and shared decision-making, and to provide psychological support to the patients and their families where needed	**I**	**Endorsement**	
N			For women with PAH having termination of pregnancy, it is recommended that this be performed in PH centres, with psychological support provided to the patient and her family	**I**	**Endorsement**	
N			For women with PAH who desire to have children, where available, adoption and surrogacy with pre-conception genetic counselling may be considered	**IIb**	**Endorsement**	
N			As teratogenic potential has been reported in preclinical models for endothelin receptor antagonists and riociguat, these drugs are not recommended during pregnancy	**III**	**Endorsement**	
Treatment of vasoreactive patients with idiopathic, heritable, or drug-associated pulmonary arterial hypertension –						

Recommendation Table 7						
R	Continuation of high doses of CCBs is recommended in patients with IPAH, HPAH, and DPAH in WHO-FC I or II with marked haemodynamic improvement (near normalization)	**I**	Continuing high doses of CCBs is recommended in patients with IPAH, HPAH, or DPAH in WHO-FC I or II with marked haemodynamic improvement (mPAP <30 mmHg and PVR <4 WU)	**I**	**Endorsement**	
N			In patients with a positive vasoreactivity test but insufficient long-term response to CCBs who require additional PAH therapy, continuation of CCB therapy should be considered	**IIa**	**Endorsement**	
Treatment of non-vasoreactive patients with idiopathic, heritable, or drug-associated pulmonary arterial hypertension who present without cardiopulmonary comorbidities[b] **– Recommendation Table 8**						
N			In patients with IPAH/HPAH/DPAH who present at high risk of death, initial combination therapy with a PDE5i, an ERA, and i.v./s.c. prostacyclin analogues should be considered[c]	**IIa**	**Endorsement**	
N			In patients with IPAH/HPAH/DPAH who present at intermediate–low risk of death while receiving ERA/PDE5i therapy, the addition of selexipag should be considered	**IIa**	**Endorsement**	
N			In patients with IPAH/HPAH/DPAH who present at intermediate–high or high risk of death while receiving ERA/PDE5i therapy, the addition of i.v./s.c. prostacyclin analogues and referral for lung transplantation (LTx) evaluation should be considered	**IIa**	**Endorsement**	
N			In patients with IPAH/HPAH/DPAH who present at intermediate–low risk of death while receiving ERA/PDE5i	**IIb**	**Endorsement**	

			therapy, switching from PDE5i to riociguat may be considered			
Initial oral drug combination therapy for patients with idiopathic, heritable, or drug-associated pulmonary arterial hypertension without cardiopulmonary comorbidities – Recommendation Table 9						
R	Ambrisentan + tadalafil	**I**	Initial combination therapy with ambrisentan and tadalafil is recommended	**I**	**Endorsement**	
N			Initial combination therapy with macitentan and tadalafil is recommended	**I**	**Endorsement**	
R	Other ERA + PDE-5i	**IIa**	Initial combination therapy with other ERAs and PDE5is should be considered	**IIa**	**Endorsement**	
N			Initial combination therapy with macitentan, tadalafil, and selexipag is not recommended	**III**	**Endorsement**	
Sequential drug combination therapy for patients with idiopathic, heritable, or drug-associated pulmonary arterial hypertension – Recommendation Table 10						
N			It is recommended to base treatment escalations on risk assessment and general treatment strategies (see treatment algorithm)	**I**	**Endorsement**	
R	Macitentan added to sildenafil	**I**	The addition of macitentan to PDE5is or oral/inhaled prostacyclin analogues is recommended to reduce the risk of morbidity/mortality events	**I**	**Endorsement**	
N			The addition of oral treprostinil to ERA or PDE5i/riociguat monotherapy is recommended to reduce the risk of morbidity/mortality events	**I**	**Fjernes**	**Ikke indregistreret i DK**
R	Bosentan added to sildenafil	**IIb**	The addition of bosentan to sildenafil is not recommended to reduce the risk of	**III**	**Endorsement**	**COMPASS-2 studiet viste ikke effekt på morbiditet/mortalitet**

			morbidity/mortality events			
R	Riociguat added to bosentan	**I**	The addition of riociguat to bosentan should be considered to improve exercise capacity	**IIa**	**Endorsement**	
Treatment of non-vasoreactive patients with idiopathic, heritable, or drug-associated pulmonary arterial hypertension who present with cardiopulmonary comorbiditiesb **– Recommendation Table 11**						
N			In patients with IPAH/HPAH/DPAH and cardiopulmonary comorbidities, initial monotherapy with a PDE5i or an ERA should be considered	**IIa**	**Endorsement**	
N			In patients with IPAH/HPAH/DPAH with cardiopulmonary comorbidities who present at intermediate or high risk of death while receiving PDE5i or ERA monotherapy, additional PAH medication may be considered on an individual basis	**IIb**	**Endorsement**	
Efficacy of intensive care management for pulmonary arterial hypertension – Recommendation Table 12						
N			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 vasopressors, fluid management, and PAH drugs as appropriate	**I**	**Endorsement**	
N			Mechanical circulatory support may be an option for selected patients as a bridge to transplantation or to recovery, and interhospital transfer should be considered if such resources are unavailable on site	**IIa**	**Endorsement**	

Lung transplantation – Recommendation Table 13						
R	Lung transplantation is recommended soon after inadequate clinical response on maximal medical therapy	**I**	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**	**Endorsement**	
N			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**	**Endorsement**	**Varetages af højt specialiseret lungemedicin i DK**
Pulmonary arterial hypertension associated with drugs or toxins – Recommendation Table 14						
N			It is recommended to make a diagnosis of drug- or toxin-associated PAH in patients who had relevant exposure and in whom other causes of PH have been excluded	**I**	**Endorsement**	
N			In patients with suspected drug- or toxin-associated PAH, it is recommended to discontinue the causative agent immediately whenever possible	**I**	**Endorsement**	
N			Immediate PAH therapy should be considered in patients who present with intermediate/high-risk PAH at diagnosis	**IIa**	**Endorsement**	
N			Patients with low-risk PAH should be re-evaluated 3–4 months after discontinuing the suspected drug or toxin, and PAH therapy may be considered when the haemodynamics have not normalized	**IIb**	**Endorsement**	
Pulmonary arterial hypertension associated with connective tissue disease – Recommendation Table 15						

N			In patients with PAH associated with CTD, treatment of the underlying condition according to current guidelines is recommended	**I**	**Endorsement**	
Pulmonary arterial hypertension associated with human immunodeficiency virus infection – Recommendation Table 16						
N			In patients with PAH associated with HIV infection, antiretroviral treatment according to current guidelines is recommended	**I**	**Endorsement**	
N			In patients with PAH associated with HIV infection, initial monotherapy should be considered, followed by sequential combination if necessary, taking into consideration comorbidities and drug–drug interactions	**IIa**	**Endorsement**	
Pulmonary arterial hypertension associated with portal hypertension – Recommendation Table 17						
R	Echocardiographic assessment for signs of PH is recommended in symptomatic patients with liver disease or portal hypertension and in all candidates for liver transplantation	**I**	Echocardiography is recommended in patients with liver disease or portal hypertension with signs or symptoms suggestive of PH, and as a screening tool in patients evaluated for liver transplantation or transjugular portosystemic shunt	**I**	**Endorsement**	
R	It is recommended that the treatment algorithm for patients with other forms of PAH should be applied to patients with PAH associated with portal hypertension, taking into account the severity of liver disease	**I**	In patients with PAH associated with portal hypertension, initial monotherapy should be considered, followed by sequential combination if necessary, taking into consideration the underlying liver disease and indication for liver transplantation	**IIa**	**Endorsement**	
R	Liver transplantation may be considered in selected patients responding well to PAH therapy	**IIb**	Liver transplantation should be considered on an individual basis in patients with PAH associated with portal hypertension, as long as PVR is normal or near	**IIa**	**Endorsement**	**Varetages i regi af levermedicinsk speciale i DK**

			normal with PAH therapy			
N			Drugs approved for PAH are not recommended for patients with portal hypertension and unclassified PH (i.e. elevated mPAP, high CO, and a normal PVR)	**III**	**Endorsement**	
Shunt closure in patients with pulmonary–systemic flow ratio >1.5:1 based on calculated pulmonary vascular resistance– Recommendation Table 18						
N			In patients with ASD, VSD, or PDA and a PVR <3 WU, shunt closure is recommended	**I**	**Endorsement**	**Varetages i DK hovedsageligt i regi af ACHD subspeciale i dialog med PH teams.**
N			In patients with ASD, VSD, or PDA and a PVR of 3–5 WU, shunt closure should be considered	**IIa**	**Endorsement**	
N			In patients with ASD and a PVR >5 WU that declines to <5 WU with PAH treatment, shunt closure may be considered	**IIb**	**Endorsement**	
N			In patients with VSD or PDA and a PVR >5 WU, shunt closure may be considered after careful evaluation in specialized centres	**IIb**	**Endorsement**	
N			In patients with ASD and a PVR >5 WU despite PAH treatment, shunt closure is not recommended	**III**	**Endorsement**	
Pulmonary arterial hypertension associated with adult congenital heart disease – Recommendation Table 19						
N			Risk assessment is recommended for patients with persistent PAH after defect closure	**I**	**Endorsement**	**Varetages i DK hovedsageligt i regi af ACHD subspeciale i dialog med PH teams.**
N			Risk assessment should be considered in patients with Eisenmenger syndrome	**IIa**	**Endorsement**	
R	Bosentan is recommended in WHO-FC III patients with Eisenmenger syndrome	**I**	Bosentan is recommended in symptomatic patients with Eisenmenger syndrome to improve exercise capacity	**I**	**Endorsement**	
R	The use of supplemental iron treatment may be considered in	**IIb**	Supplemental iron treatment should be considered in	**IIa**	**Endorsement**	

	patients with low ferritin plasma levels		patients with iron deficiency			
R	Combination drug therapy may be considered in patients with Eisenmenger syndrome	**IIb**	In patients with PAH after corrected adult CHD, initial oral combination therapy with drugs approved for PAH should be considered for patients at low and intermediate risk, while initial combination therapy including i.v./s.c. prostacyclin analogues should be considered for patients at high risk	**IIa**	**Endorsement**	
R	Combination drug therapy may be considered in patients with Eisenmenger syndrome	**IIb**	In patients with adult CHD, including Eisenmenger syndrome, sequential combination therapy should be considered if patients do not meet treatment goals	**IIa**	**Endorsement**	
N			In women with Eisenmenger syndrome, pregnancy is not recommended	**III**	**Endorsement**	
R	If symptoms of hyperviscosity are present, phlebotomy with isovolumic replacement should be considered, usually when the haematocrit is >65%	**IIa**	In patients with Eisenmenger syndrome, routine phlebotomy to lower elevated haematocrit is not recommended	**III**	**Endorsement**	
Pulmonary arterial hypertension with signs of venous/capillary involvement – Recommendation Table 20						
R	A combination of clinical findings, physical examination, bronchoscopy, and radiological findings is recommended to diagnose PVOD/PCH	**I**	A combination of clinical and radiological findings, ABG, PFTs, and genetic testing is recommended to diagnose PAH with signs of venous and/or capillary involvement (PVOD/PCH)	**I**	**Endorsement**	
N			In patients with PVOD/PCH, the use of drugs approved for PAH may be considered with careful monitoring of clinical symptoms and gas exchange	**IIb**	**Endorsement**	
N			Lung biopsy is not recommended to confirm a diagnosis of PVOD/PCH	**III**	**Endorsement**	
Paediatric pulmonary hypertension – Recommendation Table 21						
N			It is recommended to perform the diagnostic work-up, including RHC and acute vasoreactivity testing, and treat	**I**	**Endorsement**	**Varetages i DK i samarbejde mellem primært pædiatrisk kardiologisk speciale med input fra PH teams**

			children with PH at centres with specific expertise in paediatric PH			
R	A PH diagnostic algorithm work-up is recommended for diagnosis and definition of the specific aetiology group in paediatric PH patients	**I**	In children with PH, a comprehensive work-up for confirming diagnosis and specific aetiology is recommended (similar to that in adults, but adapted for age)	**I**	**Endorsement**	
N			For confirming PH diagnosis, RHC is recommended, preferably before initiating any PAH therapy	**I**	**Endorsement**	
N			In children with IPAH/HPAH, acute vasoreactivity testing is recommended to detect those who may benefit from calcium channel blocker therapy	**I**	**Endorsement**	
N			It is recommended to define a positive response to acute vasoreactivity testing in children similar to adults by a reduction of mPAP ≥10 mmHg to reach an absolute value of mPAP ≤40 mmHg, with an increased or unchanged CO	**I**	**Endorsement**	
R	A PAH-specific therapeutic algorithm is recommended in paediatric PH patients	**I**	In children with PAH, a therapeutic strategy based on risk stratification and treatment response is recommended, extrapolated from that in adults but adapted for age	**I**	**Endorsement**	
R	Specific paediatric determinants of risk should be considered	**IIa**	It is recommended to monitor the treatment response in children with PAH by serially assessing a panel of data derived from clinical assessment, echocardiographic evaluation, biochemical markers, and exercise tolerance tests	**I**	**Endorsement**	
N			Achieving and maintaining a low-risk profile should be considered as an adequate treatment response for children with PAH	**IIa**	**Endorsement**	
N			It is recommended to screen infants with bronchopulmonary dysplasia for PH	**I**	**Endorsement**	
N			In infants with (or at risk of) bronchopulmonary dysplasia and PH, treating lung disease, including hypoxia, aspiration, and structural airway disease, and	**I**	**Endorsement**	

			optimizing respiratory support is recommended before initiating PAH therapy			
N			In neonates and infants, a diagnostic and therapeutic approach to PH distinct from that in older children and adults should be considered, given the frequent association with developmental vascular and parenchymal lung disease	**IIa**	**Endorsement**	
Pulmonary hypertension associated with left heart disease – Recommendation Table 22						
N			RHC is recommended for suspected PH in patients with LHD, if it aids management decisions	**I**	**Endorsement**	
N			RHC is recommended in patients with severe tricuspid regurgitation with or without LHD prior to surgical or interventional valve repair	**I**	**Endorsement**	
R	Patients with PH-LHD and a severe pre-capillary component as indicated by a high DPG and/or high PVR should be referred to an expert PH centre for a complete diagnostic work-up and an individual treatment decision	**IIa**	For patients with LHD and suspected PH with features of a severe pre-capillary component and/or markers of RV dysfunction, referral to a PH centre for a complete diagnostic work-up is recommended	**I**	**Endorsement**	
N			In patients with LHD and CpcPH with a severe pre-capillary component (e.g. PVR >5 WU), an individualized approach to treatment is recommended	**I**	**Endorsement**	
N			When patients with PH and multiple risk factors for LHD, who have a normal PAWP at rest but an abnormal response to exercise or fluid challenge, are treated with PAH drugs, close monitoring is recommended	**I**	**Endorsement**	
N			In patients with PH at RHC, a borderline PAWP (13–15 mmHg) and features of HFpEF, additional testing with exercise or fluid challenge may be considered to uncover post-capillary PH	**IIb**	**Endorsement**	

Pulmonary hypertension associated with lung disease and/or hypoxia – Recommendation Table 23						
R	Echocardiography is recommended for the non-invasive diagnostic assessment of suspected PH in patients with lung disease	**I**	If PH is suspected in patients with lung disease, it is recommended that echocardiographyd b e performed and the results interpreted in conjunction with ABG, PFTs including DLCO, and CT imaging	**I**	**Endorsement**	
R	Optimal treatment of the underlying lung disease, including long-term O_2 therapy in patients with chronic hypoxaemia, is recommended in patients with PH due to lung diseases	**I**	In patients with lung disease and suspected PH, it is recommended to optimize treatment of the underlying lung disease and, where indicated, hypoxaemia, sleep-disordered breathing, and/or alveolar hypoventilation	**I**	**Endorsement**	
R	Referral to an expert centre is recommended in patients with echocardiographic signs of severe PH and/or severe right ventricular dysfunction	**I**	In patients with lung disease and suspected severe PH, or where there is uncertainty regarding the treatment of PH, referral to a PH centre is recommendede	**I**	**Endorsement**	
N			In patients with lung disease and severe PH, an individualized approach to treatment is recommended	**I**	**Endorsement**	
N			It is recommended to refer eligible patients with lung disease and PH for LTx evaluation	**I**	**Endorsement**	
R	RHC is not recommended for suspected PH in patients with lung disease, unless therapeutic consequences are to be expected (e.g. LTx, alternative diagnoses such as PAH or CTEPH, and potential enrolment in a clinical trial)	**III**	In patients with lung disease and suspected PH, RHC is recommended if the results are expected to aid management decisions	**I**	**Endorsement**	
N			Inhaled treprostinil may be considered in patients with PH associated with ILD	**IIb**	**Fjernes**	**Inhaleret treprostenil er ikke tilgængeligt i DK**
N			The use of ambrisentan is not recommended in patients with PH associated with IPF	**III**	**Endorsement**	
N			The use of riociguat is not recommended in patients with PH associated with IIP	**III**	**Endorsement**	
Chronic thrombo-						

embolic pulmonary hypertension and chronic thrombo-embolic pulmonary disease without pulmonary hypertension – Recommendation Table 24						
R	Lifelong anticoagulation is recommended in all patients with CTEPH	**I**	Lifelong therapeutic doses of anticoagulation are recommended in all patients with CTEPH	**I**	**Endorsement**	
N			Antiphospholipid syndrome testing is recommended in patients with CTEPH	**I**	**Endorsement**	
N			In patients with CTEPH and antiphospholipid syndrome, anticoagulation with VKAs is recommended	**I**	**Endorsement**	
R	It is recommended that all patients with CTEPH receive assessment of operability and decisions regarding other treatment strategies made by a multidisciplinary team of experts	**I**	It is recommended that all patients with CTEPH are reviewed by a CTEPH team for the assessment of multimodality management	**I**	**Endorsement**	
R	Surgical PEA in deep hypothermia circulatory arrest is recommended for patients with CTEPH	**I**	PEA is recommended as the treatment of choice for patients with CTEPH and fibrotic obstructions within pulmonary arteries accessible by surgery	**I**	**Endorsement**	
R	Interventional BPA may be considered in patients who are technically inoperable or carry an unfavourable risk:benefit ratio for PEA	**IIb**	BPA is recommended in patients who are technically inoperable or have residual PH after PEA and distal obstructions amenable to BPA	**I**	**Endorsement**	
R	Riociguat is recommended in symptomatic patients who have been classified as having persistent/recurrent CTEPH after surgical treatment or inoperable CTEPH by a CTEPH team including at least one experienced PEA surgeon	**I**	Riociguat is recommended for symptomatic patients with inoperable CTEPH or persistent/recurrent PH after PEA	**I**	**Endorsement**	**Der er i DK gode erfaringer med brug af PDE5i hos denne pt. gruppe**
N			Long-term follow-up is recommended after PEA and BPA, as well as for patients with CTEPH established on medical therapy	**I**	**Endorsement**	
N			A multimodality approach should be considered for patients with persistent PH after PEA and for patients	**IIa**	**Endorsement**	

			with inoperable CTEPH			
N			In patients with CTEPD without PH, long-term anticoagulant therapy should be considered on an individual basisf	**IIa**	**Endorsement**	
N			PEA or BPA should be considered in selected symptomatic patients with CTEPD without PH	**IIa**	**Endorsement**	
N			Treprostinil s.c. may be considered in patients in WHO-FC III–IV who have inoperable CTEPH or persistent/recurrent PH after PEA	**IIb**	**Endorsement**	
R	Off-label use of drugs approved for PAH may be considered in symptomatic patients who have been classified as having inoperable CTEPH by a CTEPH team including at least one experienced PEA surgeon	**IIb**	Off-label use of drugs approved for PAH may be considered in symptomatic patients who have inoperable CTEPH	**IIb**	**Endorsement**	
N			In patients with inoperable CTEPH, a combination of sGC stimulator/PDE5i, ERA, or parenteral prostacyclin analogues may be considered	**IIb**	**Endorsement**	
N			BPA may be considered for technically operable patients with a high proportion of distal disease and an unfavourable risk:benefit ratio for PEA	**IIb**	**Endorsement**	
Pulmonary hypertension centres – Recommendation Table 25						
N			It is recommended that PH centres maintain a patient registry	**I**	**Endorsement**	
N			It is recommended that PH centres collaborate with patient associations	**I**	**Endorsement**	
N			Accreditation of the PH centres should be considered (e.g. https://ec.europa.eu/health/ern/assessment_en)	**IIa**	**Endorsement**	
R	It should be considered that a referral centre follow at least 50 patients with PAH or CTEPH and should receive at least two new referrals per month with documented PAH or CTEPH	**IIa**	PH centres should follow-up a sufficient number of patients to maintain expertise (at least 50 patients with PAH or CTEPH and at least two new referrals per month with documented PAH or CTEPH) and consider establishing	**IIa**	**Endorsement**	

			collaborations with high-volume centres			

[a]

Class of recommendation.

[b]

Cardiopulmonary comorbidities are predominantly encountered in elderly patients and include risk factors for HFpEF, such as obesity, diabetes, coronary heart disease, a history of hypertension, and/or a low DLCO.

[c]

Initial triple-combination therapy including i.v./s.c. prostacyclin analogues may also be considered in patients presenting at intermediate risk but severe haemodynamic impairment (e.g. RAP ≥20 mmHg, CI <2.0 L/min/m2, SVI <31 mL/m2, and/or PVR ≥12 WU).

[d]

Assessments should ideally be made when the patient is clinically stable, as exacerbations can significantly raise PAP.

[e]

This recommendation does not apply to patients with end-stage lung disease who are not considered candidates for LTx.

[f]

Long-term anticoagulant therapy is recommended when the risk of PE recurrence is intermediate or high, or when there is no history of VTE.

GRADE				
	Quality of evidence	Strength of recommendation	**Class**	**Bemærkning**
In patients with IPAH/HPAH/DPAH who present at low or intermediate risk of death, initial combination therapy with a PDE5i and an ERA is recommended	Low	I	**B**	**Endorsement**
The use of PDE5i in patients with HFpEF and isolated post-capillary PH is not recommended	Low	II	**C**	**Endorsement**
PDE5i may be considered in patients with severe PH associated with ILD (individual decision-making in PH centres)	Very low	**IIb**	**C**	**Endorsement**
The use of PDE5i in patients with ILD and non-severe PH is not recommended	Very low	**III**	**C**	**Endorsement**
n patients with CTEPH who are candidates for BPA, medical therapy should be considered prior to the intervention	Very low	**IIa**	**B**	**Endorsement**

[a]