Selected revised re	ecommendations (R) and	new recon	nmendations (N)			
New or revised	Recommendation in 2015 version	Classa	Recommendation in 2022 version	Classa	Arbejdsgruppens anbefaling -Endorsement -Ændring	Arbejdsgruppens kommentarer
Right heart catheterization and vasoreactivity testing – Recommendati on 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 Inhaled iloprost may be considered for performing vasoreactivity testing as an alternative	lla	Inhaled nitric oxide, inhaled iloprost, or i.v. epoprostenol are recommended for performing vasoreactivity testing	I	Endorsement	
Diagnostic strategy – Recommendati on 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 <i>Table 10</i>)	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 rany characteristic 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 – Recommendati on 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	lla	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	B
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 – Recommendati on 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/mainte nance of an intermediate-risk profile should be considered an inadequate treatment response for most patients with PAH	Ila	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 – Recommendati						
on Table 5						
R	Supervised exercise	IIa	Supervised exercise	I	Endorsement	
	training should be		training is			
	considered in physically		recommended in patients with PAH			
	deconditioned PAH		under medical			
	patients under		therapy			
D	medical therapy	I	In the second	T		
R	Immunization of PAH patients against	1	Immunization of patients with PAH	I	Endorsement	
	influenza and		against SARS-CoV-2,			
	pneumococcal		influenza,			
	infection is recommended		and <i>Streptococcus</i> pneumoniae is			
	recommended		recommended			
R	Correction of	IIb	In the presence of	Ι	Endorsement	
	anaemia and/or iron status may be		iron-deficiency anaemia, correction			
	considered in PAH		of iron status is			
	patients		recommended in			
N			patients with PAH	113		
Ν			In the absence of anaemia, iron	IIb	Endorsement	
			repletion may be			
			considered in			
			patients with PAH			
R	Oral anticoagulant	IIb	with iron deficiency Anticoagulation is	IIb	Endorsement	
	treatment may be		not generally			
	considered in		recommended in			
	patients with IPAH, HPAH, and PAH due		patients with PAH but may be			
	to use of anorexigens		considered on an			
-			individual basis			
R	The use of angiotensin-	III	The use of ACEis, ARBs, ARNIs, SGLT-	III	Endorsement	
	converting enzyme		2is, beta-blockers, or			
	inhibitors,		ivabradine is not			
	angiotensin-2		recommended in			
	receptor antagonists, beta-blockers, and		patients with PAH unless required by			
	ivabradine is not		comorbidities (i.e.			
	recommended in		high blood pressure,			
	patients with PAH unless required by		coronary artery disease, left HF, or			
	comorbidities (i.e.		arrhythmias)			
	high blood pressure,					
	coronary artery disease, or left HF)					
R	In-flight	Ila	In-flight	I	Endorsement	
	O ₂ administration		O2 administration is			
	should be considered for patients in WHO-		recommended for			
	FC III and IV and		patients using oxygen or whose			
	those with arterial		arterial blood oxygen			
	blood O ₂ pressure		pressure is <8 kPa			
	consistently <8 kPa (60 mmHg)		(60 mmHg) at sea level			
R	In elective surgery,	IIa	For interventions	IIa	Endorsement	
	epidural rather than		requiring			
	general anaesthesia		anaesthesia, multidisciplinary			
	should be preferred whenever possible		consultation at a PH			
	F F F F F S S S S S S S S S S S S S S S		centre to assess risk			
			and benefit should be			
Women of			considered			
childbearing						
potential –						
Recommendati on Table 6						
R	It is recommended	I	It is recommended	I	Endorsement	
	that PAH patients		that women of			
	avoid pregnancy		childbearing			
			potential with PAH are counselled at the			
	i	l		i		1

N Its recommended that women of childbearing percention of childbearing percention of contraceptive advice. Considering the individual needs of contraceptive failure are significant in PAH PAH who consider pregnancy or who become pregnant receive prompt consider pregnant or who become pregnant receive prompt consider pregnant or who percention and shared decision- making, and their families where analytic base for women whe PAH this be performed in Pictoreme where analytic market and bear performed in Pictoreme where analytic and bear family Its recommended that women with PAH this bear of the preference analytic and their family Its and their family N For women with PAH this bear of the preference analytic and bear family Its and their family N For women with PAH this bear of the preference analytic and bear family Its bear the preference analytic and bear family N For women with PAH the patients and here family Its bear the preference analytic and bear family					
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NFor women with PAH support provided to the patient and her familyIIIEndorsementNFor women with PAH who desire to have children, where available, adoption and surrogacy with pre-conception genetic counselling may be consideredIIIEndorsementNAs teratogenic potential has beenIIIEndorsement Endorsement				having termination of	
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Image: Normal base in the second s				support provided to	
NFor women with PAH who desire to have children, where available, adoption and surrogacy with pre-conception genetic counselling may be consideredIIbEndorsementNAs teratogenic potential has beenIIIEndorsement	l				
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N As teratogenic III Endorsement N As teratogenic III Endorsement	l	Lindor Schiefte	110		
N As teratogenic III Endorsement N As teratogenic III Endorsement	l				
and surrogacy with pre-conception genetic counselling may be considered III Endorsement potential has been	l				
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genetic counselling may be considered Endorsement N As teratogenic potential has been III	l			pre-conception	
N As teratogenic III Endorsement potential has been	l			genetic counselling	
potential has been	 ļ				
	l	Endorsement	III		Ν
	l				
reported in	l				
preclinical models for	l				
endothelin receptor	l				
antagonists and riociguat, these drugs	l			antagonists and	
are not	1				
recommended during	l				
pregnancy	1				
Treatment of					Treatment of
vasoreactive					
patients with					patients with
idiopathic,					idiopathic,
heritable, or					heritable, or
drug-associated					drug-associated
pulmonary					
arterial					
hypertension –					nypertension -

Recommendati on 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 cardiopulmona ry comorbiditiesb - Recommendati on 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 consideredc	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	Па	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	

	[r	thorony mitching	r		,
			therapy, switching from PDE5i to			
			riociguat may be			
			considered			
Initial oral drug			considered			
combination						
therapy for						
patients with						
idiopathic,						
heritable, or						
drug-associated						
pulmonary						
arterial						
hypertension						
without cardiopulmona						
ry						
comorbidities –						
Recommendati						
on Table 9						
R	Ambrisentan + tadala	I	Initial combination	I	Endorsement	
	fil	-	therapy with	-		
			ambrisentan and			
			tadalafil is			
			recommended			
N			Initial combination	I	Endorsement	
			therapy with			
			macitentan and			
			tadalafil is			
R	Other ERA + PDE-5i	IIa	recommended Initial combination	IIa	Endorsement	
к	Other ERA + PDE-51	па	therapy with other	na	Endorsement	
			ERAs and PDE5is			
			should be			
			considered			
N			Initial combination	III	Endorsement	
			therapy with			
			macitentan, tadalafil,			
			and selexipag is not			
			recommended			
Sequential drug						
combination						
therapy for						
patients with						
idiopathic,						
heritable, or						
drug-associated pulmonary						
arterial						
hypertension –						
Recommendati						
on Table 10						
N			It is recommended to	Ι	Endorsement	
			base treatment			
			escalations on risk			
			assessment and			
			general treatment			
			strategies (see	ĺ		
			treatment			
D	Magitartar add. 1.	T	algorithm)	T	Endorcoment	
R	Macitentan added to	I	The addition of macitentan to PDE5is	I	Endorsement	
	sildenafil		or oral/inhaled			
			prostacyclin			
			analogues is			
			recommended to			
			reduce the risk of			
			morbidity/mortality			
			events			
N			The addition of oral	I	<mark>Fjernes</mark>	Ikke indregistreret i
			treprostinil to ERA or			DK
			PDE5i/riociguat			
			monotherapy is			
			recommended to			
			reduce the risk of			
			morbidity/mortality events			
R	Bosentan added to	IIb	The addition of	III	Endorsement	COMPASS-2 studiet
IX.	sildenafil	110	bosentan to sildenafil		Liuoi sement	viste ikke effekt på
			is not recommended			morbiditet/mortalite
			to reduce the risk of			t

			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 cardiopulmona ry comorbiditiesb						
Recommendati on Table 11						
N			In patients with IPAH/HPAH/DPAH and cardiopulmonary comorbidities, initial monotherapy with a PDE5i or an ERA should be considered	lla	Endorsement	
Ν			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 – Recommendati on Table 12						
Ν			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						
-						
Recommendati on 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 - Recommendati on 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	lla	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 – Recommendati on Table 15						

		1				
Ν			In patients with PAH	I	Endorsement	
			associated with CTD,			
			treatment of the			
			underlying condition			
			according to current			
			guidelines is			
Dulmanan			recommended			
Pulmonary arterial						
hypertension						
associated with						
human						
immunodeficie						
ncy virus						
infection -						
Recommendati						
on Table 16						
N			In patients with PAH	Ι	Endorsement	
			associated with HIV			
			infection,			
			antiretroviral			
			treatment according			
			to current guidelines			
			is recommended			
N			In patients with PAH	IIa	Endorsement	
			associated with HIV			
			infection, initial			
			monotherapy should			
			be considered,			
			followed by			
			sequential			
			combination if			
			necessary, taking into			
			consideration			
			comorbidities and			
			drug–drug interactions			
Pulmonary			Interactions			
arterial						
hypertension						
associated with						
associated with portal						
associated with portal hypertension –						
associated with portal hypertension – Recommendati						
associated with portal hypertension –	Echocardiographic	I	Echocardiography is	I	Endorsement	
associated with portal hypertension – Recommendati on Table 17	Echocardiographic assessment for signs	I	Echocardiography is recommended in	I	Endorsement	
associated with portal hypertension – Recommendati on Table 17	Echocardiographic assessment for signs of PH is	I	recommended in	I	Endorsement	
associated with portal hypertension – Recommendati on Table 17	assessment for signs	I	recommended in patients with liver	I	Endorsement	
associated with portal hypertension – Recommendati on Table 17	assessment for signs of PH is	I	recommended in	I	Endorsement	
associated with portal hypertension – Recommendati on Table 17	assessment for signs of PH is recommended in	I	recommended in patients with liver disease or portal	I	Endorsement	
associated with portal hypertension – Recommendati on Table 17	assessment for signs of PH is recommended in symptomatic patients with liver disease or portal hypertension	I	recommended in patients with liver disease or portal hypertension with signs or symptoms suggestive of PH, and	I	Endorsement	
associated with portal hypertension – Recommendati on Table 17	assessment for signs of PH is recommended in symptomatic patients with liver disease or portal hypertension and in all candidates	I	recommended in patients with liver disease or portal hypertension with signs or symptoms suggestive of PH, and as a screening tool in	I	Endorsement	
associated with portal hypertension – Recommendati on Table 17	assessment for signs of PH is recommended in symptomatic patients with liver disease or portal hypertension and in all candidates for liver	I	recommended in patients with liver disease or portal hypertension with signs or symptoms suggestive of PH, and as a screening tool in patients evaluated	I	Endorsement	
associated with portal hypertension – Recommendati on Table 17	assessment for signs of PH is recommended in symptomatic patients with liver disease or portal hypertension and in all candidates	I	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	I	Endorsement	
associated with portal hypertension – Recommendati on Table 17	assessment for signs of PH is recommended in symptomatic patients with liver disease or portal hypertension and in all candidates for liver	I	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	I	Endorsement	
associated with portal hypertension – Recommendati on Table 17	assessment for signs of PH is recommended in symptomatic patients with liver disease or portal hypertension and in all candidates for liver	I	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	I	Endorsement	
associated with portal hypertension – Recommendati on Table 17 R	assessment for signs of PH is recommended in symptomatic patients with liver disease or portal hypertension and in all candidates for liver transplantation		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			
associated with portal hypertension – Recommendati on Table 17	assessment for signs of PH is recommended in symptomatic patients with liver disease or portal hypertension and in all candidates for liver transplantation	I	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 In patients with PAH	I	Endorsement	
associated with portal hypertension – Recommendati on Table 17 R	assessment for signs of PH is recommended in symptomatic patients with liver disease or portal hypertension and in all candidates for liver transplantation		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 In patients with PAH associated with			
associated with portal hypertension – Recommendati on Table 17 R	assessment for signs of PH is recommended in symptomatic patients with liver disease or portal hypertension and in all candidates for liver transplantation It is recommended that the treatment algorithm for		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 In patients with PAH associated with portal hypertension,			
associated with portal hypertension – Recommendati on Table 17 R	assessment for signs of PH is recommended in symptomatic patients with liver disease or portal hypertension and in all candidates for liver transplantation It is recommended that the treatment algorithm for patients with other		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 In patients with PAH associated with portal hypertension, initial monotherapy			
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associated with portal hypertension – Recommendati on Table 17 R	assessment for signs of PH is recommended in symptomatic patients with liver disease or portal hypertension and in all candidates for liver transplantation It is recommended that the treatment algorithm for patients with other forms of PAH should be applied to patients		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 In patients with PAH associated with portal hypertension, initial monotherapy should be considered, followed			
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associated with portal hypertension – Recommendati on Table 17 R	assessment for signs of PH is recommended in symptomatic patients with liver disease or portal hypertension and in all candidates for liver transplantation It is recommended that the treatment algorithm for patients with other forms of PAH should be applied to patients with PAH associated with portal		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 In patients with PAH associated with portal hypertension, initial monotherapy should be considered, followed by sequential combination if			
associated with portal hypertension – Recommendati on Table 17 R	assessment for signs of PH is recommended in symptomatic patients with liver disease or portal hypertension and in all candidates for liver transplantation 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		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 In patients with PAH associated with portal hypertension, initial monotherapy should be considered, followed by sequential combination if necessary, taking into			
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associated with portal hypertension – Recommendati on Table 17 R	assessment for signs of PH is recommended in symptomatic patients with liver disease or portal hypertension and in all candidates for liver transplantation 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		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 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			
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associated with portal hypertension - Recommendati on Table 17 R	assessment for signs of PH is recommended in symptomatic patients with liver disease or portal hypertension and in all candidates for liver transplantation 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 Liver transplantation may be considered in selected patients	I	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 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 should be considered on an individual	IIa	Endorsement	levermedicinsk
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associated with portal hypertension - Recommendati on Table 17 R	assessment for signs of PH is recommended in symptomatic patients with liver disease or portal hypertension and in all candidates for liver transplantation 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 Liver transplantation may be considered in selected patients responding well to	I	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 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 should be considered on an individual basis in patients with PAH associated with	IIa	Endorsement	levermedicinsk
R	assessment for signs of PH is recommended in symptomatic patients with liver disease or portal hypertension and in all candidates for liver transplantation 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 Liver transplantation may be considered in selected patients responding well to	I	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 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 should be considered on an individual basis in patients with PAH associated with portal hypertension,	IIa	Endorsement	levermedicinsk

			normal with PAH			
			therapy			
N			Drugs approved for	ш	Endorsement	
			PAH are not			
			recommended for			
			patients with portal			
			hypertension and			
			unclassified PH (i.e.			
			elevated mPAP, high			
			CO, and a normal			
			PVR)			
Shunt closure in						
patients with						
pulmonary-						
systemic flow ratio >1.5:1						
based on						
calculated						
pulmonary						
vascular						
resistance-						
Recommendati						
on Table 18						
Ν			In patients with ASD,	I	Endorsement	Varetages i DK
			VSD, or PDA and a			hovedsageligt i regi af
			PVR <3 WU, shunt			ACHD subspeciale i
			closure is			dialog med PH teams.
N			recommended	11-	En damante	4
Ν			In patients with ASD, VSD, or PDA and a	IIa	Endorsement	
			VSD, of PDA and a PVR of 3–5 WU,			
			shunt closure should			
			be considered			
N		1	In patients with ASD	IIb	Endorsement	1
			and a PVR >5 WU			
			that declines to <5			
			WU with PAH			
			treatment, shunt			
			closure may be			
			considered			
N			In patients with VSD	IIb	Endorsement	
			or PDA and a PVR >5			
			WU, shunt closure			
			may be considered			
			after careful			
			evaluation in			
N			specialized centres In patients with ASD	III	Endorsement	-
11			and a PVR >5 WU	111	Endorsement	
			despite PAH			
			treatment, shunt			
			closure is not			
			recommended			
Pulmonary						
arterial						
hypertension						
associated with						
adult congenital						
heart disease -						
Recommendati						
on Table 19 N			Dick account is	I	Endorcoment	Varatages ; DV
1N			Risk assessment is recommended for	1	Endorsement	Varetages i DK hovedsageligt i regi af
			patients with			ACHD subspeciale i
			persistent PAH after			dialog med PH teams.
			defect closure			anatos men i il teams.
N			Risk assessment	IIa	Endorsement	1
			should be considered			
			in patients with			
			Eisenmenger			
			syndrome			
R	Bosentan is	I	Bosentan is	I	Endorsement	
	recommended in		recommended in			
	WHO-FC III patients		symptomatic patients			
	with Eisenmenger		with Eisenmenger			
	syndrome		syndrome to improve			
R	The use of	IIb	exercise capacity Supplemental iron	IIa	Endorsement	-
IX.	supplemental iron	110	treatment should be	110	Liuui sement	
	treatment may be		considered in			
	considered in	1		ĺ		
		1	L		I	1

	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	Ilb	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/capillar y involvement – Recommendati on 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 – Recommendati on 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	I	In children with PH, a	I	Endorsement	-
it.	algorithm work-up is		comprehensive	•	Indorsement	
	recommended for		work-up for			
	diagnosis and		confirming diagnosis			
	definition of the		and specific aetiology			
	specific aetiology		is recommended			
	group in paediatric PH patients		(similar to that in adults, but adapted			
	i ii patients		for age)			
N			For confirming PH	I	Endorsement	
			diagnosis, RHC is	-		
			recommended,			
			preferably before			
			initiating any PAH			
N		-	therapy In children with	I	Endorsement	-
11			IPAH/HPAH, acute	1	Endorsement	
			vasoreactivity testing			
			is recommended to			
			detect those who			
			may benefit from			
			calcium channel			
N			blocker therapy			{
N			It is recommended to define a positive	I	Endorsement	
			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			
R	A PAH-specific	I	In children with PAH,	I	Endorsement	
	therapeutic		a therapeutic			
	algorithm is		strategy based on			
	recommended in		risk stratification and			
	paediatric PH		treatment response			
	patients		is recommended, extrapolated from			
			that in adults but			
			adapted for age			
R	Specific paediatric	IIa	It is recommended to	Ι	Endorsement	
	determinants of risk		monitor the			
	should be		treatment response			
	considered		in children with PAH			
			by serially assessing a panel of data			
			derived from clinical			
			assessment,			
			echocardiographic			
			evaluation,			
			biochemical markers,			
			and exercise tolerance tests			
N			Achieving and	IIa	Endorsement	
14			maintaining a low-	11a	Liuoisement	
			risk profile should be			
			considered as an			
			adequate treatment			
			response for children			
			with PAH			4
N			It is recommended to	I	Endorsement	
			screen infants with bronchopulmonary			
			dysplasia for PH			
N		1	In infants with (or at	I	Endorsement	1
			risk of)			
			bronchopulmonary			
			dysplasia and PH,			
			treating lung disease,			
			including hypoxia, aspiration, and			
			structural airway			
1		1		1		
			disease, and			

N			optimizing respiratory support is recommended before initiating PAH therapy 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	IIa	Endorsement	
Pulmonary hypertension associated with left heart disease - Recommendati on Table 22			disease			
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	ΠΡ	Endorsement	

Pulmonary						
hypertension						
associated with						
lung disease						
and/or hypoxia -						
Recommendati on Table 23						
R	Echocardiography is	I	If PH is suspected in	I	Endorsement	
	recommended for the non-invasive		patients with lung disease, it is			
	diagnostic		recommended that			
	assessment of		echocardiography <mark>d</mark> b			
	suspected PH in		e performed and the results interpreted in			
	patients with lung disease		conjunction with			
			ABG, PFTs including			
			DLCO, and CT			
R	Optimal treatment of	I	imaging In patients with lung	I	Endorsement	
	the underlying lung	-	disease and	_		
	disease, including		suspected PH, it is			
	long-term O ₂ therapy in patients with		recommended to optimize treatment			
	chronic hypoxaemia,		of the underlying			
	is recommended in		lung disease and,			
	patients with PH due to lung diseases		where indicated, hypoxaemia, sleep-			
	and and and a dealers		disordered			
			breathing, and/or alveolar			
			hypoventilation			
R	Referral to an expert	I	In patients with lung	I	Endorsement	
	centre is		disease and			
	recommended in patients with		suspected severe PH, or where there is			
	echocardiographic		uncertainty			
	signs of severe PH		regarding the			
	and/or severe right ventricular		treatment of PH, referral to a PH			
	dysfunction		centre is			
	-		recommendede	_		
N			In patients with lung disease and severe	I	Endorsement	
			PH, an individualized			
			approach to			
			treatment is recommended			
N			It is recommended to	I	Endorsement	
			refer eligible patients			
			with lung disease and PH for LTx			
			evaluation			
R	RHC is not	III	In patients with lung	I	Endorsement	
	recommended for suspected PH in		disease and suspected PH, RHC is			
	patients with lung		recommended if the			
	disease, unless		results are expected			
	therapeutic consequences are to		to aid management decisions			
	be expected (e.g. LTx,		uecisions			
	alternative diagnoses					
	such as PAH or CTEPH, and potential					
	enrolment in a					
	clinical trial)					
Ν			Inhaled treprostinil may be considered in	IIb	<mark>Fjernes</mark>	Inhaleret treprostenil er ikke tilgængeligt i
			patients with PH			DK
N			associated with ILD			
Ν			The use of ambrisentan is not	III	Endorsement	
			recommended in			
			patients with PH			
N			associated with IPF The use of riociguat	III	Endorsement	
14			is not recommended			
			in patients with PH			
Chronic			associated with IIP			
thrombo-						

embolic						
pulmonary						
hypertension						
and chronic						
thrombo- embolic						
pulmonary						
disease without						
pulmonary						
hypertension – Recommendati						
on Table 24						
R	Lifelong	I	Lifelong therapeutic	I	Endorsement	
	anticoagulation is recommended in all		doses of anticoagulation are			
	patients with CTEPH		recommended in all			
	×		patients with CTEPH			
N			Antiphospholipid	I	Endorsement	
			syndrome testing is recommended in			
			patients with CTEPH			
N			In patients with	I	Endorsement	
			CTEPH and			
			antiphospholipid syndrome,			
			anticoagulation with			
			VKAs is			
R	It is recommended	I	recommended It is recommended	I	Endorsement	
	that all patients with	1	that all patients with	1	Liluoi semellu	
	CTEPH receive		CTEPH are reviewed			
	assessment of		by a CTEPH team for			
	operability and decisions regarding		the assessment of multimodality			
	other treatment		management			
	strategies made by a		-			
	multidisciplinary					
R	team of experts Surgical PEA in deep	I	PEA is recommended	I	Endorsement	
	hypothermia		as the treatment of			
	circulatory arrest is recommended for		choice for patients with CTEPH and			
	patients with CTEPH		fibrotic obstructions			
	P		within pulmonary			
			arteries accessible by			
R	Interventional BPA	IIb	surgery BPA is recommended	I	Endorsement	
R	may be considered in	110	in patients who are			
	patients who are		technically			
	technically inoperable or carry		inoperable or have residual PH after PEA			
	an unfavourable		and distal			
	risk:benefit ratio for		obstructions			
D	PEA		amenable to BPA			
R	Riociguat is recommended in	I	Riociguat is recommended for	I	Endorsement	Der er i DK gode erfaringer med brug
	symptomatic patients		symptomatic patients			af PDE5i hos denne
	who have been		with inoperable			pt. gruppe
	classified as having persistent/recurrent		CTEPH or persistent/recurrent			
	CTEPH after surgical		PH after PEA			
	treatment or					
	inoperable CTEPH by					
	a CTEPH team including at least one					
	experienced PEA					
	surgeon					
Ν			Long-term follow-up is recommended	I	Endorsement	
			is recommended after PEA and BPA, as			
			well as for patients			
			with CTEPH			
			established on medical therapy			
N			A multimodality	IIa	Endorsement	
			approach should be			
			considered for			
			patients with persistent PH after			
			PEA and for patients			

			with inoperable		
			СТЕРН		
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	lla	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
Ν			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 – Recommendati on 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/assess ment_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	lla	Endorsement

	collaborations with		
	high-volume centres		

а

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.

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 \geq 20 mmHg, Cl <2.0 L/min/m2, SVI <31 mL/m2, and/or PVR \geq 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	В	Endorsement		
The use of PDE5i in patients with HFpEF and isolated post- capillary PH is not recommended	Low	Π	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	Ila	B	Endorsement		

а