Supplement

Table S1: TF composition per topic

1. **Definitions:** *Torbicki, Lang, Pepke-Zaba, Dorfmuller, Madani + Corkery*
2. **Diagnosis:** *Gopalan, Hoeper, Ogo, Meyer, Jais + Meszaros*
3. **Epidemiology:** *Sitbon, Matsubara, Klok, Jansa, Dorfmuller, D'Armini*
4. **Acute PE:** *Klok, Torbicki, Hoeper, Vonk Noordegraaf, Gopalan + Meszaros*
5. **Pathophysiology:** *Lang, Humbert, Vonk Noordegraaf, Gopalan, Dorfmuller, Fadel*
6. **PEA:** *Jenkins, D'Armini, Fadel, Madani, Brenot, Ghofrani, Kim*
7. **BPA:** *Kim, Ogo, Lang, Brenot, Meyer, Matsubara, Galie, Madani*
8. **Medical treatment:** *Humbert, Ogo, Hoeper, Sitbon, Ghofrani, Galie, Pepke-Zaba, Fadel*
9. **Multimodality:** *Jais, Brenot, Mayer, Matsubara, Torbicki, Jansa, D'Armini, Jenkins + Corkery*
10. **Rehabilitation in CTEPH:** *Vonk Noordegraaf, Gruenig, Klok, Jais, Jansa + Corkery*
11. **Global research lines:** *Pepke-Zaba, Humbert, Sitbon, Ghofrani, Galie, Kim, Gopalan, Jenkins + Meszaros*

Table S2: Search strategies for each topic/question

	Topic	Database searched	Search terms
1	Definition	MEDLINE (via PubMed) without date limits	chronic thromboembolic pulmonary hypertension AND definition Chronic Thromboembolic Disease (CTED) AND definition
2	Diagnosis	MEDLINE (via PubMed), EMBASE (via Ovid), and Cochrane Library (Wiley) Years Covered: 1980-2019	Chronic Thromboembolic Pulmonary Hypertension OR CTEPH AND imaging Chronic Thromboembolic Disease OR CTED AND imaging
3	Epidemiology	MEDLINE (via PubMed) without date limits	- First of all: “chronic thromboembolic pulmonary hypertension” OR “CTEPH” AND “Epidemiology” - Then: “Frequency”, “surveillance”, “occurrence”, “incidence”, “prevalence”, “pulmonary embolism”, “venous thromboembolism”, “perfusion defect”, “lung scan”, “anticoagulation”, “oral anticoagulant”, “vitamin K antagonist”, “Non-Vitamin K antagonist oral anticoagulants”
3	PE to CTEPH	MEDLINE (via PubMed) without date limits	For Table S7: Publication range from 1990-2019 (the 1st paper from 1992 by Bill Auger for description of CTEPH findings was baseline). MeSH Terms: pulmonary embolism; thromboembolism; hypertension, pulmonary; tomography, x-ray computed. Subheading: diagnosis For table S8: ("risk factors"[MeSH Terms] OR ("risk"[All Fields] AND "factors"[All Fields]) OR "risk factors"[All Fields]) AND "chronic thromboembolic pulmonary hypertension"[All Fields] For table X3: ("diagnosis"[Subheading] OR "diagnosis"[All Fields] OR "diagnosis"[MeSH Terms]) AND "chronic thromboembolic pulmonary hypertension"[All Fields]
5	Pathophysiology	MEDLINE (via PubMed) without date limits	chronic thromboembolic pulmonary hypertension, pulmonary arterial hypertension, bronchial arteries, systemic vessels, microvascular, anastomoses, shunt, pathology, histology, small vessel disease, right ventricle, maladaption, wave reflection patterns, CTPA, MRI, ECG gating, blood volume maps
6	PEA	MEDLINE (via PubMed) , EMBASE (via Ovid), and Cochrane Library (Wiley) until Sept 2019	CTED OR CTEPH OR chronic thromboembolic disease OR chronic thromboembolic pulmonary hypertension AND pulmonary endarterectomy OR pulmonary thromboendarterectomy AND surg OR thorax surgery
7	BPA	MEDLINE (via PubMed) without date limits	“balloon pulmonary angioplasty” AND “percutaneous transluminal pulmonary angioplasty”; Balloon pulmonary angioplasty: 2363 citations Percutaneous transluminal pulmonary angioplasty: 458 citations
8	Medical therapy	MEDLINE (via PubMed)	chronic thromboembolic pulmonary hypertension AND medical treatment

			chronic thromboembolic pulmonary hypertension AND pulmonary vasodilators chronic thromboembolic pulmonary hypertension AND anticoagulation chronic thromboembolic pulmonary hypertension AND riociguat chronic thromboembolic pulmonary hypertension AND guanylase cyclase stimulators chronic thromboembolic pulmonary hypertension AND phosphodiesterase type 5 inhibitors chronic thromboembolic pulmonary hypertension AND prostacyclin chronic thromboembolic pulmonary hypertension AND prostacyclin receptor agonist
9	Multimodality	MEDLINE (via PubMed) without date limits	multimodality[MeSH Terms]) OR multimodality[Text Word]) OR multimodal[MeSH Terms]) OR multimodal[Text Word]) OR combination[MeSH Terms]) OR combination[Text Word]) OR hybrid[MeSH Terms]) OR hybrid[Text Word]) OR additional[MeSH Terms]) OR additional[Text Word]) OR sequential[MeSH Terms]) OR sequential[Text Word]) OR combined[MeSH Terms]) OR combined[Text Word]) OR bridge[MeSH Terms]) OR bridge[Text Word]) OR bridging[MeSH Terms]) OR bridging[Text Word]) OR therapy[MeSH Terms]) OR therapy[Text Word]) OR angioplasty[MeSH Terms]) OR angioplasty[Text Word]) OR endarterectomy[MeSH Terms]) OR endarterectomy[Text Word]) OR thromboendarterectomy[MeSH Terms]) OR thromboendarterectomy[Text Word]) AND chronic thromboembolic pulmonary hypertension[MeSH Terms]) OR chronic thromboembolic pulmonary hypertension[Text Word]
10	Rehabilitation	MEDLINE (via PubMed) without date limits	(("Cardiac Rehabilitation"[Mesh] OR "cardiopulmonary rehabilitation"[tw] OR "cardiovascular rehabilitation"[tw] OR "cardiac rehabilitation"[tw] OR "pulmonary rehabilitation"[tw] OR "Rehabilitation"[Mesh] OR "rehabilitation"[Subheading] OR "rehabilitation"[tw] OR rehabilit*[tw] OR "Early Ambulation"[mesh] OR "Exercise Therapy"[mesh] OR "Endurance Training"[mesh] OR "Motion Therapy, Continuous Passive"[mesh] OR "Muscle Stretching Exercises"[mesh] OR "Plyometric Exercise"[mesh] OR "Resistance Training"[mesh] OR "Early Ambulation"[tw] OR "Exercise Therapy"[tw] OR "Endurance Training"[tw] OR "Continuous Passive Motion Therapy"[tw] OR "Muscle Stretching Exercises"[tw] OR "Plyometric Exercise"[tw] OR "Resistance Training"[tw] OR "Muscle Stretching Exercise"[tw] OR "Plyometric Exercises"[tw]) AND ("chronic thromboembolic"[tw] OR "chronic thromboembolic disease"[tw] OR "chronic thromboembolism"[tw] OR "chronic thrombo embolic pulmonary"[tw] OR "Pulmonary Embolism"[Mesh] OR "Pulmonary Embolism"[tw] OR "Pulmonary Embolisms"[tw] OR "Pulmonary Embolism"[tw] OR "Pulmonary Embolisms"[tw] OR "Pulmonary Thromboembolism"[tw] OR "Pulmonary Thromboembolisms"[tw] OR "Pulmonary Thrombo-embolism"[tw]))
11	Research	MEDLINE (via PubMed), EMBASE (via Ovid)	*"CLINICAL TRIALS AS TOPIC"/ AND ((CTEPH).ti,ab OR exp *"HYPERTENSION, PULMONARY"/ OR ("chronic thromboembolic pulmonary hypertension").ti,ab)) [DT 2009-2019] ((CTEPH).ti,ab OR exp *"HYPERTENSION, PULMONARY"/ OR ("chronic thromboembolic pulmonary hypertension").ti,ab) AND exp *"DRUG THERAPY, COMBINATION"/21 exp REGISTRIES/ AND ((CTEPH).ti,ab OR ("chronic thromboembolic pulmonary hypertension").ti,ab) 32

			(((CTEPH).ti,ab OR exp *"HYPERTENSION, PULMONARY"/ OR ("chronic thromboembolic pulmonary hypertension").ti,ab) AND(("biobank" OR "biorepository" OR "biological samples").ti,ab ORexp "DATABASES, GENETIC"/ OR exp "BIOLOGICAL SPECIMENBANKS"/ OR exp PHENOTYPE/)) [DT 2009-2019] [Humans] 147

Table S3: Hemodynamic characteristics in cohorts of CTEPD patients without PH at rest

	Taboada 2014 [1]	Van Kan 2016 [2]	Held 2016 [3]	Guth 2018 [4]	Claeys 2019 [5]	Swietlik 2019 [6]
n	42	14	10	12	14	34
age	49 ± 16	48 ± 16	62 ± 14	51 (34-65)	47 ± 16	53±17
mPAP, mmHg	21 (5)	19 ± 3	20.5 (4.3)	20 ± 3	20.5 (17.5 – 23.3)	20 (18–22)
PVR, dyn.s.cm-5	164 (104)	147 ± 34	244 (151)	3.4 (2.8-4.4)	154 ± 48.9	1.9 (1.4–2.4)
PQ slope, mmHg.min.L-1	-	2.7 ± 1.2	-	3.6 ± 1.0	3.48 (2.24 – 4.36)	1.6 (1.1–3.4)

Mean ± SD, median (IQR)

Table S4: Survey on VQ SPECT, DECT and MR for CTEPH

Commentor	Does your institution offer SPECT VQ routinely	Does your institution offer the possibility of extracting Planar images from SPECT data	would you as clinicians like to have access to these planar images as add-on to SPECT to be made available on your PACS system
DG (UK)	Yes	Yes	Yes
JP-Z (UK)	Yes	Yes	Yes
EM (Germany)	No Patients referred for PEA or BPA usually had a SPECT VQ before admission	No	Yes
FK (Netherlands)	No	No	Yes
EG (Germany)	Yes	Yes	Yes
MMH (Germany)	Yes	Yes	Yes
NK (USA)	Yes	Yes	Yes
PJ (Czech Republic)	No	N/A	Not filled in
PB (France)	Yes	Yes	Yes
TO (Japan)	Yes	No	Yes
XJ (France)	Yes	Yes	Yes
IL (Austria)	Yes	Yes	Yes
AT (Poland)	Yes (done in a contracted separate nuclear medicine laboratory)	Yes	Yes
MD (Belgium)	Yes	Yes	Yes
AD (Italy)	Yes	Yes	Yes

15 respondents.
12/15 institutions do VQ SPECT. Of these, the clinicians working in the 11 of the 12 that do VQ SPECT would like planar reprojections from SPECT to be made available for review.

Commentor	Is the CT scanner in your institution capable of doing dual energy (DECT)?	If yes to above, do you do DECT as routine rather than conventional CTPA	Is Cardiac MR routinely done as a part of PH/CTEPH workup in your institution?	If Yes to above, which of the following do you use MR for? 1. MRA (+/- perfusion) 2. Rotating MIP 3. RV function baseline 4. RV function post-Rx
DG (UK)	Yes	Yes	Yes	ALL
JP-Z (UK)	Yes	Yes	Yes	ALL
EM (Germany)	Yes	No We use DECT for additional information	No only in selected cases for RV assessment	N/A
FK (Netherlands)	No	N/A	No	N/A
EG (Germany)	Yes	Yes	Yes	3,4
MMH (Germany)	Yes	Not filled-in	Yes Regularly done, but not as part of clinical routine but as research tool (Change MRI study)	
NK (USA)	Yes	No. Only On request	No. Only on request	
PJ (Czech Republic)	No	N/A	No	
PB (France)	Yes	Yes	No	Angio DSA used regularly and occasionally cone beam CT during BPA sessions
TO (Japan)	Yes	Yes	Yes	3,4
XJ (France)	No	N/A	No	
IL (Austria)	Yes	No	Yes	N/A
AT (Poland)	No	N/A	No	N/A
MD (Belgium)	Yes	Yes	No	
AD (Italy)	Yes	Yes	Yes	3,4

15 respondents.
DECT: 11/15 have access but only 7 are routinely doing DECT over standard CTPA. 2/15 do DECT on request
MRI: 7/15 routinely have access to MRI and 2/15 perform MRI on request. Out of the 7 institution that had routine access to MRI, only 2 are using all components of MR (assessment of pulmonary vasculature and evaluation of RV function). Three centres use MRI only for RV function evaluation at baseline & post treatment. One institution does MRI routinely as it is part of a study protocol.

Table S5: What makes a high-quality CTPA?

Image quality determines the diagnostic value of a CTPA but is a complex entity for which there is no single objective measure. Current generation of CT systems of 64 slices and higher are consistently capable of producing high spatial resolution images that are of high quality but the CTPA protocols vary according to the available hardware as well as technical and patient related factors such as body habitus, motion and cardiac function. It is beyond the scope of this paper to address all aspects of CTPA protocols, but the fundamentals are outlined below.

The basic principle is to acquire a movement-free volumetric dataset of the thorax during peak enhancement of the pulmonary circulation with reduced venous contamination. High temporal resolution to ensure fast coverage of the thorax using the shortest acquisition time can be achieved by increasing detector size and anatomic coverage per rotation. This is of particular importance in dyspneic patients to avoid breathing-related artefacts. Caudocranial direction of acquisition will also provide the best chance of motion free imaging of the lower lobes. ECG gating can diminish motion artifacts arising from cardiac pulsation and result in sharper outlines of the vessel contour and can be facilitated without increasing the breath-hold time, an important consideration in pulmonary hypertension patients but is not routinely implemented in standard CTPA protocols.

High injections rate (eg, 4-5 mL/sec) and iodine concentrations of 350 or 370 mgI/mL will provide improved enhancement that is necessary to distinguish thrombus from intraluminal contrast material. The injection duration is approximately equal to the sum of the scan duration and the delay time. As the standard CTPA protocol is optimised for pulmonary arterial opacification, a longer delay from the injection of contrast medium to image acquisition is necessary for depicting bronchial collaterals. The volume of contrast medium is individually tailored for each patient taking in to account the patient size, cardiac and renal function. A saline chaser at the same injection rate can help in minimising streak artifacts by improving the washout of contrast medium from the superior vena cava and innominate veins. Prior to the acquisition, patients are also instructed to avoid inadvertent Valsalva manoeuvre as it can cause sudden influx of un-opacified blood from the inferior vena cava.

Pulmonary artery attenuations of 300 to 350 HU (ie, 250–300 HU net contrast enhancement) is considered as the preferred level of CTPA contrast enhancement {Bae:2010er}. The theoretic minimum attenuation of pulmonary artery required to detect acute and chronic emboli is computed as 211 HU {Wittram:2007fh}. Thinnest possible collimation will improve detection in the subsegmental level.

Images are reconstructed at minimum of 1mm slice thickness using a soft tissue kernel in the axial plane. An elemental advantage of the CT volumetric datasets is the possibility to perform multiplanar reformats through the longitudinal axis of a vessel. This can be of immense benefit in the interrogation of the complex vascular anatomy and aid in the differentiation of factors that mimic pulmonary thromboembolism. It is also mandatory to adjust the display window widths and levels (soft tissue window 400 /30 to 40 HU; pulmonary parenchyma 1500/ -800 to -600 HU; pulmonary embolism–specific setting 700/100 HU) in order to improve the conspicuity of the abnormalities. Maximum Intensity Projection (MIP) is a widely used rendering tool that involves projecting the highest attenuation voxels on every view throughout the volume onto a 2D image. MIP's can be adjusted to variable thickness and is very useful for creation of a vascular map for display. [7, 8]

Table S6: CTPA features of acute PE and CTEPH [9]

	Acute PE	**CTEPH**
Pulmonary vasculature	- Dilatation of main pulmonary artery (in severe PE) - Central or eccentric filling defect surrounded by contrast material forming acute angles with the arterial wall ["polo mint" when orthogonal or "railway track" sign when parallel to long axis of the vessel] - Distension of completely occluded vessel - Normal calibre segmental vessels	- Asymmetric enlargement of central pulmonary arteries - Eccentric thrombus (may or may not be calcified) forming obtuse angles with the vessel wall - Intimal thickening/irregularities - Reduced calibre of occluded vessel with absent flow distal to the total obstruction - Small segmental and subsegmental vessels with abrupt cut-offs - Intravascular webs and bands - Stenosis with or without post-stenotic dilatation - Tortuous vessels - Atherosclerotic calcification of vessel wall
Systemic vasculature		- Enlarged bronchial and non-bronchial collaterals
Cardiovascular	- Right ventricular dilatation (in severe PE) - Retrograde flow of contrast material into distended azygos vein, inferior vena cava and intrahepatic veins (in severe PE)	- Right ventricular dilatation and hypertrophy - Retrograde flow of contrast material into distended azygos vein, inferior vena cava and intrahepatic veins
Lung parenchyma	- Wedge-shaped peripheral areas of hyper-attenuation - Opacities with central ground glass and a rim of consolidation ["reverse-halo" sign]	- Mosaic attenuation - Focal ground glass opacities - Parenchymal bands - Subpleural scars - Cavitating infarcts - Pleural thickening - Cylindrical bronchial airway dilatation - Air trapping

Table S7: Known risk factors for CTEPH compared to patients with IPAH, and their prevalence in patients with PE

	Reported Odds Ratio CTEPH versus IPAH (95%CI) [10]	**Estimated prevalence among patients with acute pulmonary embolism** [11]
Thyroid replacement therapy	6.1 (2.7–15)	5-7%
Malignancy	3.8 (1.5–10)	10-25%
Recurrent venous thromboembolism	15 (5.4–43)	20-30%
(Sub)-massive pulmonary embolism	13 (not provided)	30-60%
Anti-phospholipid antibodies	4.2 (1.6–12)	2-5%
Factor VIII >230 IU/dl	2.5 (not provided)	10-30%
Non-O blood group	2.1 (1.2–3.9)	60-80%
Ventriculoatrial shunt or (infected) pacemaker	76 (7.7–10,350)	<1%
Splenectomy	18 (1.6–2.4)	1-2%

Table S8: Overview and accuracy of diagnostic tests that provide clues to the presence/absence of CTEPH, as derived from studies in patients with a history of acute PE and/or suspected CTEPH

Diagnostic test	Sensitivity	Specificity	Remarks
"Leiden CTEPH prediction score" [11] [12]	85-91%	75%	- Score based on the following factors: Unprovoked PE (+6 points), hypothyreoidism (+3 points), symptom onset >2 weeks before PE diagnosis (+3 points), RV dysfunction at index CTPA (+2 points), diabetes mellitus (-3 points) and thrombolytic/surgical therapy (-3 points); a total score >6 points denotes patients at high risk of CTEPH. - Not developed to serve as single diagnostic test but to identify 25% of patients with highest pre-test probability.
Cardiopulmonary exercise testing (CPET) [9, 13]	83%	92%	- Typically appears as ineffective ventilation with elevated alveolar–capillary gradients of oxygen and carbon dioxide; o Elevated slope of minute ventilation (V'_E)/carbon dioxide output (V'_{CO2}) ratio. o Elevated ventilator equivalents for oxygen (EQ_{O2}) and carbon dioxide (EQ_{CO2}). o Low and decreasing end-tidal carbon dioxide tension (P_{ETCO2}), elevated alveolar–arterial oxygen tension gradient ($P_{A–aO2}$) and elevated arterial–end-tidal carbon dioxide gradient ($P_{A–ETCO2}$).
BNP/NT-proBNP [14, 15]	82%	70%	- Applied threshold was age and sex normal value as determined by the manufacturer.

ECG [16, 17] [14]	77%	89%	- Generally, ECG signs of right heart strain include P-pulmonale, right bundle branch block, T-wave abnormalities in the chest leads and right-axis deviation. - Specifically, an ECG is indicative of CTEPH in case of either 1) rSR' or rSr' pattern in lead V1, 2) R:S >1 in lead V1 with R >0.5 mV, or 3) QRS axis >90^{o}.
ECG and BNP/NT-proBNP combined ('CTEPH rule-out criteria') [11, 12] [14]	91-96%	65%	- Not prospectively evaluated in management study (trial ongoing NCT02555137, results expected in 2020).
Echo [9, 18-20]	70-100%	72-89%	- Evaluations include estimating peak velocity of tricuspid valve regurgitation, calculation of atrioventricular pressure gradients and detection of indirect signs of pulmonary hypertension such as right atrial and right ventricular dilatation, reduced right ventricular contractility and Doppler flow abnormalities in the right ventricular outflow tract. - Conclusions derived from an echocardiographic examination should aim to assign a level of probability of pulmonary hypertension.
V/Q scintigraphy (planar) [9, 18, 21, 22]	90-100%	51-100%	- In CTEPH, at least one but more commonly several, segmental or larger mismatched perfusion defects are present. - Small peripheral unmatched and non-segmental defects in perfusion can also be present in patients with PAH. - Larger unmatched perfusion defects may also be seen in other pulmonary vascular disease such as PVOD.
V/Q SPECT [23]	97%	81%	- Data for V/Q SPECT in CTEPH are still sparse.
CTPA [24]	76%	96%	- Specific CTPA characteristics of CTEPH are summarized in **Table S7**.

			- Sensitivity was 99% and specificity was 97%, when only high quality CT scans including DECT were included.
Dual-energy CT (DECT) [9]	97%	86%	- Data for V/Q SPECT in CTEPH are still sparse. - The calculation of iodine distribution in the lung parenchym offers qualitative and quantitative insights into pulmonary haemodynamics (correlates with mPAP and PVR measurements on right heart catheterisation) and helps differentiation between small airway disease from pulmonary artery vascular disease.
Contrast-enhanced/perfusion MRI [9, 23, 25]	83-100%	81-99%	- MRA is superior to DSA in depicting the precise proximal beginning of the thromboembolic material, with accuracy lower at the subsegmental level - MRI scanning will also allow for accurate assessment of the RV function and morphology

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