

衛生福利部國民健康署「罕見疾病個案通報審查基準機制」(送審資料表)
-特發性或遺傳性肺動脈高壓

[Idiopathic or Heritable pulmonary arterial hypertension, IPAH or HPAH] -

1. 臨床症狀及徵兆之病歷紀錄，需包含病史、個人史及用藥史等(必要)
2. 心臟超音波及心導管檢查正式報告或影本，並具有肺動脈壓的數據(必要)
3. 排除次發性肺動脈高壓相關原因之檢驗報告及佐證資料(必要)
4. 與本疾病相關之正式入院及出院病歷摘要(必要)
5. 如為遺傳性疾病，應檢附詳細家族史與相關基因檢測報告(必要)
6. 其他檢查均需附上正式報告或影本
7. 需附上相關影像資料
8. 其它說明如'out of proportion'肺高壓

項目	填寫部分
病史 (History)	Symptoms : (Duration, Times or Frequency) <input type="checkbox"/> Dyspnea <input type="checkbox"/> Fatigue <input type="checkbox"/> Syncope <input type="checkbox"/> Chest Pain <input type="checkbox"/> Peripheral edema Personal history: _____ Medications: _____
身體檢查 (Physical examination)	<input type="checkbox"/> Right ventricular heave <input type="checkbox"/> Clubbing fingers <input type="checkbox"/> Loud P2 <input type="checkbox"/> Hepatojugular reflux <input type="checkbox"/> Peripheral edema <input type="checkbox"/> Ascites <input type="checkbox"/> High jugular venous pressure <input type="checkbox"/> 心雜音 舒張期 Gr. _____/VI at _____ 收縮期 Gr. _____/VI at _____
胸部 X 光 (Chest X-ray)	<input type="checkbox"/> Enlarged pulmonary artery <input type="checkbox"/> Enlarged right ventricle
心電圖 (ECG)	<input type="checkbox"/> Right axis deviation <input type="checkbox"/> Right ventricular hypertrophy <input type="checkbox"/> Peaked P-wave <input type="checkbox"/> Other _____
肺功能檢查 (Pulmonary function test)	<input type="checkbox"/> Standard spirometry _____ <input type="checkbox"/> DLCO _____ (Diffusing capacity of the lung for carbon monoxide)
經胸心臟超音波 (Transthoracic echocardiography)	Right heart dilatation: _____ 估算的 RVSP: _____ mmHg, TR severity: _____, PR severity: _____

項目	填寫部分
實驗室數據 (Laboratory data)	<input type="checkbox"/> Complete blood count, aPTT, PT <input type="checkbox"/> LFT : AST_____, ALT_____, Bilirubin_____ total protein_____, Albumin_____, others_____ <input type="checkbox"/> BUN_____, Creatinine_____, Na_____, K_____ <input type="checkbox"/> Arterial blood gas: paO ₂ :_____ paCO ₂ :_____ <input type="checkbox"/> Thyroid function test: TSH:_____ free-T4____ or [TSH:_____ T4:_____ T3:_____] <input type="checkbox"/> HIV Ab: <input type="checkbox"/> Positive(+) <input type="checkbox"/> Negative(-)
肺部通氣灌流掃描 (Lung perfusion & ventilation scan)	<input type="checkbox"/> Negative(-) <input type="checkbox"/> Positive(+):_____
胸部電腦斷層 (Chest CT or Chest HRCT)	<input type="checkbox"/> Negative(-) <input type="checkbox"/> Positive(+):_____
經食道心臟超音波 (Transesophageal echocardiography)	<input type="checkbox"/> Negative(-) <input type="checkbox"/> Positive(+):_____
腹部超音波 (Abdominal ultrasound)	<input type="checkbox"/> Negative(-) <input type="checkbox"/> Positive(+):_____
自體免疫相關檢驗及會 診免疫風濕科 (Autoimmune profile and rheumatology consult)	<input type="checkbox"/> ANA:_____ <input type="checkbox"/> Anti-Sc170:_____ <input type="checkbox"/> Anti-RNP:_____ <input type="checkbox"/> C3:_____ C4:_____ FR:_____
右心導管及血管反應度 測試(Right Heart catheterization and vasoreactivity test)	PAP(S/D/M) :___/___/___ mmHg RAP :___/___/___ mmHg PAWP* :___/___/___ mmHg C.O. :_____ L/min C.I. :_____ L/min/m ² PVR:_____ Wood units, or PVRI:_____ <input type="checkbox"/> Vasodilator test : <input type="checkbox"/> Negative(-) <input type="checkbox"/> Positive(+):_____
備註(病人為非典型之 表現，不完全符合以上 之診斷標準，但仍診斷 為此疾病之理由)	
其他說明	

Abbreviation:

*Pulmonary arterial wedge pressure (PAWP)或 Pulmonary capillary wedge pressure (PCWP) 或 Left atrial pressure (LAP)

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必須檢附文件

1. 臨床症狀及徵兆之病歷紀錄，需包含病史、個人史及用藥史等
2. 心臟超音波及心導管檢查報告
3. 排除次發性肺動脈高壓相關原因之檢驗報告及佐證資料
4. 與本疾病相關之正式入院及出院病歷摘要
5. 如為遺傳性疾病，應檢附詳細家族史與相關基因檢測報告

1. 病史^a, EKG, ABG 和 Chest PA 支持 PH.
2. 在心臟超音波中估算到 sPAP \geq 30mmHg.

1. Mean PAP(平均肺動脈壓力) \geq 25mmHg.
2. PAWP^b(肺動脈楔壓力) \leq 15mmHg.
3. PVR(肺血管阻力) \geq 3WU

1. ANA $<$ 1:80 (ANA \geq 1:80 時，需會診免疫風濕科醫師，排除是否為免疫疾病引起之肺高壓)
2. Negative in HIV Ab, U3/RNP and Anti-RNP.
3. 證明非 Hyperthyroidism.
4. 證明非 Chronic hemolysis(CBT, aPTT, PT).

1. 證明非先天性心臟疾病/Eisenmenger syndrome, L→R shunt CHD without small defects^c, Post-cardiac surgery nor PVOD by HRCT, CT, MRI or Pulmonary angiography (chose one or more).
2. 證明非 Pulmonary artery embolism (CTEPH) in V/Q lung scan.
3. 證明非慢性肝臟疾病所引起的肺高壓，例如 Portopulmonary hypertension 等.
4. 證明非 Chronic lung disease，例如 COPD or ILD 等.
5. 證明非 PPHN or BPD.
6. 證明非 Perinatal lung maldevelopment.
7. 證明非 Congenital pulmonary abnormality.
8. 證明非 PH with unclear and/or multifactorial mechanisms in Group V.

特發性或遺傳性肺動脈高壓 Idiopathic or Heritable pulmonary arterial hypertension

Abbreviation:

a: 症狀包括 dyspnea, syncope, chest pain, peripheral edema, fatigue.

b: Pulmonary arterial wedge pressure (PAWP) 或 Pulmonary capillary wedge pressure (PCWP) 或 Left atrial pressure (LAP)

c: small defects: VSD $<$ 1 cm, ASD $<$ 2cm, PDA $<$ 0.2cm.

HRCT: high resolution CT. PVOD: pulmonary venous obstructive disease. CTEPH: chronic thromboembolic pulmonary hypertension. COPD: chronic obstructive pulmonary disorder. IPD: idiopathic pulmonary disease. PF: pulmonary fibrosis. PPHN: Persistent pulmonary hypertension of newborn. Group V:; Group V in clinical classification of PH in 2013(表一、肺高血壓分類).