**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)  □ Dyspnea  □ Fatigue  □ Syncope  □ Chest Pain  □ Peripheral edema  Personal history:\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_  Medications:\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_ |
| 身體檢查  (Physical examination) | □ Right ventricular heave □ Clubbing fingers  □ Loud P2 □ Hepatojugular reflux  □ Peripheral edema □ Ascites  □ High jugular venous pressure  □ 心雜音  舒張期Gr. /VI at  收縮期Gr. /VI at |
| 胸部X光  (Chest X-ray) | □ Enlarged pulmonary artery  □ Enlarged right ventricle |
| 心電圖  (ECG) | □ Right axis deviation  □ Right ventricular hypertrophy  □ Peaked P-wave  □ Other |
| 肺功能檢查  (Pulmonary function test) | □ Standard spirometry  □ 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: |
| 實驗室數據  (Laboratory data) | □ Complete blood count, aPTT, PT  □ LFT : AST , ALT , Bilirubin  total protein , Albumin , others  □ BUN , Creatinine , Na , K  □ Arterial blood gas:  paO2: paCO2:  □ Thyroid function test:  TSH: free-T4 or [TSH: T4: T3: ]  □ HIV Ab: □ Positive(+) □ Negative(-) |
| 肺部通氣灌流掃描  (Lung perfusion & ventilation scan) | □ Negative(-) □Positive(+): |
| 胸部電腦斷層  (Chest CT or Chest HRCT) | □ Negative(-) □Positive(+): |
| 經食道心臟超音波(Transesophageal echocardiography) | □ Negative(-) □Positive(+): |
| 腹部超音波 (Abdominal ultrasound) | □ Negative(-) □Positive(+): |
| 自體免疫相關檢驗及會診免疫風濕科  (Autoimmune profile and rheumatology consult) | □ ANA:  □ Anti-Sc170:  □ Anti-RNP:  □ 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/m2  PVR: Wood units, or PVRI:  □ Vasodilator test :  □ Negative(-) □ 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)

**衛生福利部國民健康署「罕見疾病個案通報審查基準機制」(審查基準表)**

**-特發性或遺傳性肺動脈高壓  
[Idiopathic or Heritable pulmonary arterial hypertension, IPAH or HPAH] -**

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
6. □病史a, EKG, ABG和Chest PA支持PH.
7. □在心臟超音波中估算到sPAP≧30mmHg.
8. □Mean PAP(平均肺動脈壓力) ≧25mmHg.
9. □PAWPb(肺動脈楔壓力)≦15mmHg.
10. □PVR(肺血管阻力)≧3WU
11. □ANA<1:80 (ANA≧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)
12. □Negative in HIV Ab, U3/RNP and Anti-RNP.
13. □證明非Hyperthyroidism.
14. □證明非Chronic hemolysis(CBT, aPTT, PT).
15. □證明非先天性心臟疾病/Eisenmenger syndrome, L→R shunt CHD without small defectsc, Post-cardiac surgery nor PVOD by HRCT, CT, MRI or Pulmonary angiography (chose one or more).
16. □證明非Pulmonary artery embolism (CTEPH) in V/Q lung scan.
17. □證明非慢性肝臟疾病所引起的肺高壓，例如Portopulmonary hypertension等.
18. □證明非Chronic lung disease，例如 COPD or ILD等.
19. □證明非PPHN or BPD.
20. □證明非Perinatal lung maldevelopment.
21. □證明非Congenital pulmonary abnormality.
22. □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).