中文題目：以肺高壓表現的下腔靜脈型心房中隔缺損併部分肺靜脈回流異常一案例報告

英文題目：A rare case of inferior vena cava type of sinus venosus atrial septal defect with partial anomalous pulmonary venous return, presenting as symptomatic pulmonary hypertension

作　　者：陳彥維[1],朱俊源[1,2,3], 黃建偉[4], 李文賢[1,2,3], 許栢超[1,2] 邱正安[1,2], 蘇河名[1,2], 林宗憲[1,2], 李智雄[1,2], 顏學偉[1,2], 溫文才[1,2], 賴文德[1,2], 許勝雄[1,2]

服務單位：

1. 高雄醫學大學附設中和紀念醫院內科部
2. 高雄醫學大學附設中和紀念醫院心臟內科
3. 高雄醫學大學醫學院臨床醫學研究所
4. 高雄醫學大學附設中和紀念醫院心臟血管外科

Introduction: Inferior vena cava (IVC) type of sinus venosus atrial septal defect (ASD), or inferior sinus venosus defect (inferior SVDs) is a rare condition, compared with other types of ASD. Some was reported with partial anomalous pulmonary venous return (PAPVR). We presented a case, presenting as pulmonary hypertension and received surgical repairment.

Case presentation: A 37-year-old woman without specific past medical history had intermittent mild chest tightness and dizziness for 1-2 years. She denied shortness of breath, syncope, palpitation or limb edema before. She came to our outpatient department for help.

At our hospital, electrocardiogram showed right axis deviation with incomplete right bundle branch block and few ventricular premature complexes. Transthoracic echocardiography (TTE) showed moderate tricuspid regurgitation (TR), pulmonary hypertension with estimated right ventricular systolic pressure (RVSP) 56mmHg. Suspected ASD with left to right shunt was found. Thus, we suggested transesophageal echocardiography (TEE) and right heart catheterization. TEE showed suspected inferior vena cava type of sinus venosus atrial septal defect with left to right shunting through. Cardiac computed tomography (CT) showed a large atrial septal defect, measured about 50.8x25.4mm, Area:9.94cm2. Also, right ventricular hypertrophy was found. Pulmonary artery catheterization demonstrated significant oxygen set up (oxygen saturation of superior vena cava: 63%, right atrium: 95.8%, inferior vena cava: 81.3%), and increase mean pulmonary artery pressure (mPAP) 25mmHg. Coronary catheterization showed no specific stenosis. We then consulted cardiovascular surgeon for surgical evaluation. Due to symptomatic pulmonary hypertension and right heart hypertrophy, surgical repair was suggested. After well preparation, we performed right atriotomy. PAPVR with right inferior pulmonary vein to right atrium was found. We performed autologous pericardial patch repair of PAPVR and ASD. The patient tolerated the operation and had an uneventful recovery.

Discussion: ASD accounts for about 15-20% of all congenital cardiac anomalies. Sinus venous ASD accounts for only 5-10% of ASD. An anomalous connection involved one or more pulmonary veins is presented in most patients with sinus venosus ASD. TEE is used for diagnosis for sinus venous ASD and cardiac CT and MRI is rising recently. Unlike secundum ASD, percutaneous intervention is hard to approach and surgical treatment is often need. We presented a rare case of

IVC type of sinus venosus ASD with PAPVR. This case initially was symptomatic due to pulmonary hypertension and received surgical treatment successfully.

Conclusion: We presented a case of IVC type of sinus venosus ASD with PAPVR. This type of ASD is rare and surgery treatment is often need.