RIGHT VENTRICLE-DEPENDENT CORONARY CIRCULATION IN PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM: A CASE REPORT

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Pulmonary atresia with intact ventricular septum (PAIVS) is a morphologically heterogeneous lesion and accounts for 1–3% of critically ill infants with congenital heart disease. Numerous surgical approaches have been attempted with varying degrees of success, but the mortality rate is still high in most series. The optimal surgical procedure depends on the size and morphology of the tricuspid valve and right ventricle and the presence or absence of right ventricle-dependent coronary circulation. Therefore, it is pivotal to define the precise morphologic and hemodynamic characteristics, especially coronary artery anatomy. In this report, we describe a full-term female neonate with cyanosis soon after birth. Two-dimensional and color Doppler echocardiography corroborated the diagnosis of PAIVS and showed a small right ventricle. Cardiac catheterization indicated PAIVS and further revealed right ventricle-dependent coronary circulation. A systemic-to-pulmonary artery shunt was constructed with a positive immediate result.

Key Words: pulmonary atresia with intact ventricular septum, right ventricle-dependent coronary circulation
(*Kaohsiung J Med Sci* 2005;21:236–40)

Pulmonary atresia with intact ventricular septum (PAIVS) is a rare complex of congenital heart diseases that is characterized by striking morphologic diversity in the degree of right ventricular hypoplasia and infundibular pathoanatomy and the occurrence of right ventricle–coronary artery connections. These variations are the major determinants of treatment strategy and outcome in patients with PAIVS. Numerous surgical approaches have been attempted with varying degrees of success. Better understanding of the anatomy and hemodynamics of the lesion has resulted in improved survival after palliative and definitive surgery [1,2]. This report details a full-term newborn with PAIVS and right ventricle-dependent coronary circulation (RVDCC). We also review the literature on the investigation and management of PAIVS.

Received: December 23, 2004 Accepted: March 10, 2005
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CASE PRESENTATION

The patient was referred to our neonatal intensive care unit 5 hours after birth because of cyanosis and heart murmur. She was a full-term neonate with a birth body weight of 2,420 g and was born via vaginal delivery after an uneventful pregnancy.

Physical examination revealed moderate cyanosis with pulse oximetry of around 80–85% and a grade I–II/VI continuous murmur over the left upper sternal border. Three hours after admission, cyanosis was abruptly aggravated with oxygen saturation down to 40–50%. She was intubated and supplied with 100% oxygen. Prostaglandin E_1 (PGE_1)

infusion was begun under the impression of ductus-dependent cyanotic heart disease.

The 12-lead electrocardiogram showed a normal sinus rhythm with a heart rate of 160 bpm, a frontal QRS axis of +5°, right atrial enlargement, and left ventricular hypertrophy. Chest radiograph showed mild cardiomegaly with a cardiothoracic ratio of 62% and decreased pulmonary vascularity. Two-dimensional color Doppler echocardiography showed a thickened, immobile, and atretic pulmonary valve without antegrade blood flow. Pulmonary blood flow was supplied via a patent ductus arteriosus (Figure 1A). The ventricular septum was intact and the right ventricle was small and hypertrophied. The tricuspid valve annulus measured 10.5 mm, corresponding to a *Z* score of –2.5 (Figure 1B). An interatrial right-to-left shunt was also noted.

Hemodynamic investigation using cardiac catheterization revealed an elevated right ventricular pressure of 57 mmHg and left ventricular systolic pressure of 67 mmHg. Biplane right ventricle angiography showed a small right ventricle and the absence of antegrade flow to the pulmonary trunk. Contrast medium filling the small right ventricle passed into the ventricle-coronary sinusoid with extensive ramifications (Figure 2). The patient received continuous PGE_1 infusion. After vital signs stabilized, a right systemic-to-pulmonary artery shunt (modified Blalock-Taussig shunt) was constructed using a 3.5-mm Gortex conduit through median sternotomy on the 10th day. The postoperative course was uneventful.

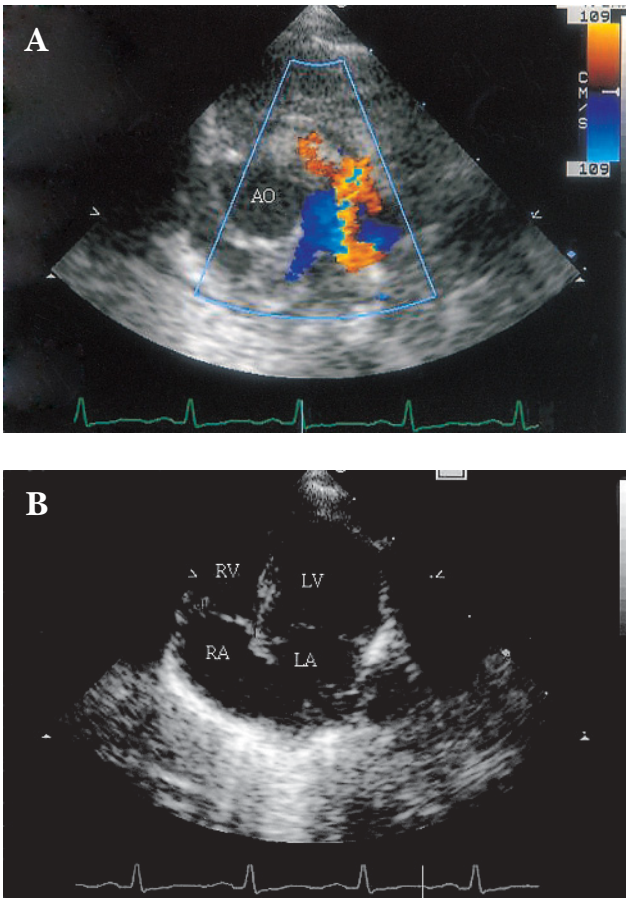

Figure 1. *(A) Two-dimensional echocardiogram parasternal short-axis view showing lack of antegrade flow across the atretic pulmonary valve. A patent ductus arteriosus with left-to-right shunt is also evident. (B) Apical four-chamber view demonstrates a very small right ventricular cavity and a tricuspid valve annulus measuring 10.5 mm. AO = aorta; LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.*

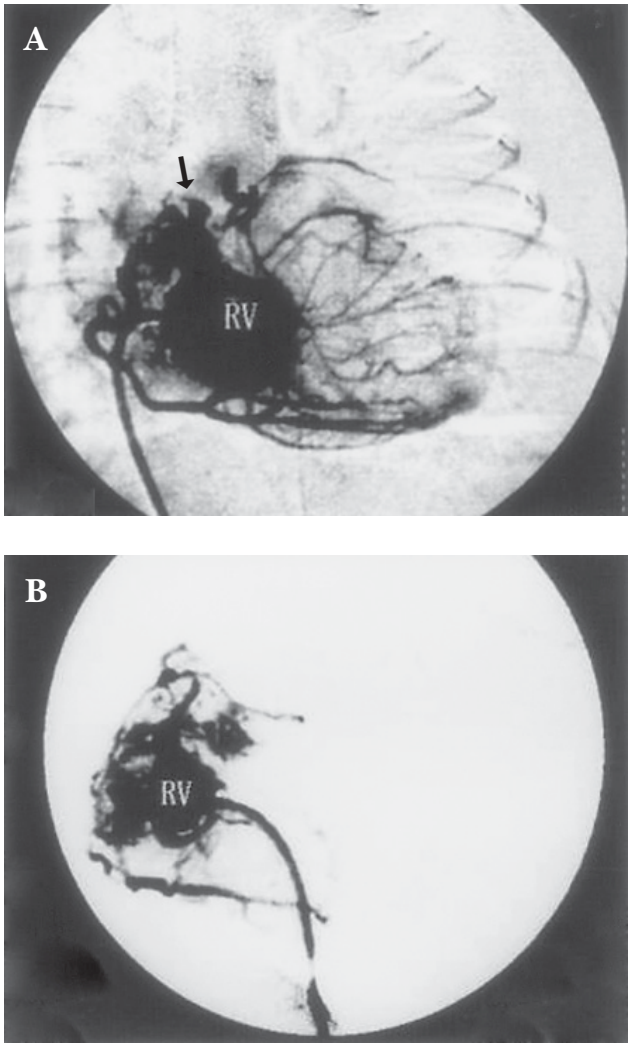

Figure 2. *Biplane right ventriculogram in (A) frontal and (B) lateral views demonstrate atretic pulmonary valve (arrow), intact ventricular septum, a very hypoplastic right ventricle (RV), and extensive right ventriculocoronary communication.*

At the outpatient clinic follow-up at 4 months of age, the patient had oxygen saturation of between 80% and 85% with room air. She was reported to be a good feeder and had steady body weight gain. Echocardiography at this visit revealed patent right Blalock-Taussig shunt and normal left ventricular systolic function.

Discussion

PAIVS is a rare disease with a reported incidence of 4–7 per 100,000 live births. Death *in utero* may occur, with an overall mortality in live-born children of greater than 50% [3]. According to the flow-related theory of cardiac development, a relatively simple primary defect that occurs during morphogenesis of the developing heart may lead to altered pressure and flow patterns, which gradually induces secondary hypoplasia or maldevelopment of major cardiac structures. In PAIVS, many of the important morphologic abnormalities develop secondary to the primary lesion of pulmonary valve atresia.

Coronary artery abnormalities and right ventricular myocardial sinusoids are common in PAIVS and have profound effects on surgical management and outcome. An important concept in management is RVDCC, which has been generally defined as right ventricle to coronary artery fistula associated with obstruction of more than one major coronary artery. In this situation, retrograde coronary blood flow from the hypertensive right occurring during systole mediated by the ventriculocoronary connections may be necessary for myocardial perfusion. Data have shown that the poor prognosis in neonates with PAIVS is generally related to either the presence of a small right ventricle and RVDCC or to florid tricuspid regurgitation and low right ventricular pressure [4].

Identification of a ventriculocoronary connection is one of the weakest areas of echocardiography and is possible only in rare cases with large communication. A report from the Congenital Heart Surgeons Study shows that a negative *Z* score correlates with the presence of ventriculocoronary connections [4]. Recently, Satou et al found that a *Z* score of –2.5 or less may be a good echocardiographic predictor of coronary artery pathology [5], leading researchers to question the need for preoperative diagnostic coronary angiography in patients with tricuspid valve annulus within a normal range. However, most authors still suggest routine preoperative cardiac catheterization for detailed coronary artery anatomy, which is required for accurate diagnosis of RVDCC.

Surgical treatment for PAIVS is complex because of the wide spectrum of malformations within this entity and the presence or absence of RVDCC. Most institutes use a two-stage approach to optimize growth of the right ventricle and tricuspid valve. In patients with mild-to-moderate right ventricle hypoplasia, patent infundibulum, and absence of RVDCC, right ventricle decompression has been advocated as the initial palliative procedure to provide adequate pulmonary blood flow and promote growth of the right ventricle. Right ventricle decompression can be achieved surgically or by interventional transcatheter perforation of the atretic pulmonary valve, either alone or combined with a systemic-to-pulmonary shunt [6]. Recently, radiofrequency energy has been applied to perforate the atretic pulmonary valve in patients with mild-to-moderate right ventricle hypoplasia, which has the advantage of causing less myocardial insult. Initial results have been encouraging [7,8]. Selection of the definitive procedure depends on the development of the right ventricle and the tricuspid valve. Biventricular repair is possible and an ideal definitive procedure in patients with adequate right ventricle and tricuspid valve size. Another alternative method is "one-and-a-half repair" for patients with a right ventricle that is functional but unable to support the entire pulmonary blood flow [9]. In patients with severely hypoplastic right ventricles, a modified Fontan procedure is considered definitive. However, the criteria for determining the best procedure remain unclear. Yoshimura et al have established a management protocol based on quantitative assessment of right ventricular morphology with positive results [10].

In the presence of RVDCC, any procedure reducing right ventricular systolic pressure may result in myocardial ischemia, infarction, and even death [1]. Therefore, patients with RVDCC should undergo systemic-to-pulmonary shunt alone and be placed on a one-ventricle algorithm (modified Fontan procedure), assuming left ventricular function is preserved. Indeed, some patients exhibit globally reduced left ventricular function and should be considered for heart transplantation.

With advanced understanding of coronary artery circulation and the application of one-ventricle repair, survival in patients with PAIVS and RVDCC has improved [11,12]. In the report by Powell et al, all 12 patients with PAIVS and RVDCC initially underwent Blalock-Taussig shunt placement and were then managed toward a modified Fontan procedure. Using this strategy, an 83% 5-year actuarial survival rate was achieved [13].

PAIVS can be diagnosed prenatally, and published data suggest that hypoplasia of the right ventricle and the

tricuspid annulus become more pronounced as pregnancy advances if there is no forward flow through the pulmonary valve [14]. It has been speculated that early valvulotomy in the fetus may be able to alter the prenatal natural history, prevent secondary damage to the fetal heart, and enable better growth of the right ventricle, making it more amenable to biventricular repair. Percutaneous balloon valvuloplasties have been performed recently in fetuses with PAIVS. Following the procedure, there has been significant growth of the tricuspid valve and right ventricle [15]. It seems likely that fetal cardiac intervention may become a useful therapeutic technique for PAIVS.

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存在於肺動脈瓣閉鎖合併完整心室中隔的依賴右心室的冠狀動脈循環 — 病例報告

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肺動脈瓣閉鎖合併完整心室中隔 (**pulmonary atresia with intact ventricular septum**) 具有多樣性的形態表現，此種異常佔了嚴重先天性心臟病的 **1–3%**。僅管外科手術方法日新月異，不過其死亡率仍然偏高。最適當的手術方法取決於右心室及三尖瓣的形態與大小，也要考量是否有依賴右心室的冠狀動脈循環 (**right ventricle-dependent coronary circulation**) 存在。所以，正確的了解病人形態學及血液動力學上的特徵，尤其是冠狀動脈的異常，對於治療方式的選擇是非常重要的。我們報告一位出生後不久即出現發紺的足月產女嬰。心臟超音波檢查顯示肺動脈瓣閉鎖合併完整心室中隔及右心室發育不全。心導管檢查證實心臟含超音波的診斷且更進一步發現依賴右心室的冠狀動脈循環。在詳盡了解病人形態學及血液動力學上的特徵之後，我們選擇為病人進行血管分流術 (**modified Blalock-Taussig shunt**)，初步的治療效果是很令人滿意的。

關鍵詞：肺動脈瓣閉鎖合併完整心室中隔，依賴右心室的冠狀動脈循環

（高雄醫誌 **2005;21:236–40**）

收文日期：**93** 年 **12** 月 **23** 日

接受刊載：**94** 年 **3** 月 **10** 日

通訊作者：吳俊仁醫師

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