Abbreviations: LV = left ventricle, RV = right ventricle, TV = tricuspid valve, AV = aortic valve, MV = mitral valve, PA = pulmonary artery, aorta, IABP = intra-aortic balloon pump, ICP = intra-cardiac pressure, PAC = pulmonary artery catheter, CO = cardiac output, PAO = pulmonary artery occlusion, ACE = angiotensin converting enzyme, AV = atrioventricular, NYHA = New York Heart Association, ECG = electrocardiogram, CXR = chest X-ray, SPECT = single photon emission computed tomography, VCD = volume contraction dose, bpm = beats per minute, hemo = hemodynamic

The left circumflex coronary artery showed an area of 50 percent narrowing prior to its bifurcation into the obtuse marginal branch. In addition, there was a significant 70-80 percent stenosis at the origin of the obtuse marginal branch, with no distal circumflex coronary artery visualized. The right coronary artery was a large vessel and was totally occluded at the acute margin, with no distal vessel visualized. The left ventriculogram was still abnormal, showing an akinetic proximal inferior wall, but the apex and anterior wall appeared to contract normally. No further surgical therapy was recommended because the patient was asymptomatic.

At the present time, three and one half years after myocardial infarction and dissection of the right coronary artery, the patient is without cardiac symptoms, and is employed full time as a maintenance worker in a county highway department.

**Discussion**

After reviewing the available autopsy data on the previously reported cases of primary dissecting aneurysm of the coronary artery, Claudon et al. postulated that in the presence of acute myocardial infarction, the following features may suggest a dissecting aneurysm of a coronary artery: 1) relative youth of a patient, since only five of the patients were older than 45 years of age; 2) frequent occurrence in a woman, since 20 of the reported cases were in women; 3) frequently present post partum, since eight of the women were postpartum. Also, they emphasized that “atherosclerosis appears to be highly uncommon” among the patients with primary dissection of a coronary artery.

Findings in our patient did not correlate with these criteria. He was a 56-year-old man with severe three vessel arteriosclerotic coronary artery disease who presented with pericardial effusion secondary to hemopericardium. If this patient had been treated with anticoagulants probably allowed hemopericardium to develop more easily. If this patient had been seen at the present time, most likely the surgical approach would have been ligation of the proximal right coronary artery, with construction of an aortocoronary saphenous vein graft to the distal right coronary artery.

**Reference**


**Congenital Absence of the Pulmonary Valve Associated with Tricuspid Atresia and Intact Ventricular Septum**

Jose Marin-Garcia, M.D.; Juan Roca, M.D.; Leonard C. Blieden, M.B.B. Ch.; Russell V. Lucas, Jr., M.D.; and Jesse E. Edwards, M.D.

A case of congenital absence of the pulmonary valve associated with tricuspid atresia and intact ventricular septum is presented. In this case, the presence of a patent ductus arteriosus and absent pulmonary valve allowed the right ventricle to receive blood from the pulmonary artery. The right ventricle, however, represented a blind pouchlike structure which was not an essential part of the circulation.

In tricuspid atresia, when two ventricles are present, usually the right ventricle is filled through a ventricular septal defect. Uncommonly, the ventricular septum is intact. Under this condition, the pulmonary valve is also atretic and the right ventricle is a non-functioning, slitlike structure in the wall of the left ventricle. We have observed a situation which, to the best of our knowledge, has not previously been reported. In this case, the tricuspid valve was atretic and, although two ventricles were present, these were separated by an intact ventricular septum. An additional unusual feature was the absence of the pulmonary valve, allowing filling of the right ventricle through the pulmonary artery. The right ventricle was an appendage of the pulmonary arterial system, having no hemodynamic significance.

In view of the unusual nature of this condition, we are prompted to place it on record.

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![Congenital Absence of the Pulmonary Valve Associated with Tricuspid Atresia and Intact Ventricular Septum](image)

**Figure 1.** In frontal plane note evidence of right atrial enlargement and biventricular hypertrophy. QRS axis is +80°.

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CONGENITAL ABSENCE OF PULMONARY VALVE

CASE REPORT

Clinical Features

The patient, a male infant, was the product of a normal pregnancy and delivery and weighed 4 kg at birth. At 11 days of age, he was hospitalized because of severe cyanosis and congestive heart failure.

Physical examination revealed cyanosis of the skin and mucus membranes. Cyanosis was equal in the upper and lower extremities. The peripheral pulses were palpable and equal. The cardiac impulse was maximal in the xiphoid area. The first cardiac sound had two components, and the second sound was single. There were no murmurs by auscultation or phonocardiography. The electrocardiogram (Fig 1) showed the QRS axis in the frontal plane to be $+80^\circ$, with right atrial enlargement and biventricular hypertrophy. The thoracic roentgenogram revealed moderate cardiomegaly, with a concavity in the region of the pulmonary artery and a rounded apex. The pulmonary vasculature was decreased. Prior to cardiac catheterization, because of severe metabolic acidosis, the patient was treated with intravenously administered sodium bicarbonate for a 12-hour period.

Catheterization Findings

Cardiac catheterization was performed through the saphenous vein, while the patient received oxygen nasally at...
the rate of 2 liters/min. There was a right-to-left shunt at the atrial level, resulting in decreased systemic arterial saturation (46 percent). The left ventricular pressure was 140/0-15 mm Hg. Injection of contrast material into the right atrium (Fig 2a) revealed this chamber to be dilated. A nonopacified triangular area was seen in the position usually occupied by the inflow portion of the right ventricle. The contrast material passed from the right atrium into the left atrium, thence into the left ventricle (Fig 2b). The latter showed poor excursion and marked hypertrophy of its wall. The aorta, which was dilated, filled from the left ventricle, and the pulmonary artery opacified via a patent ductus arteriosus. A left ventriculogram showed no evidence of a ventricular septal defect. Subsequent to opacification of the pulmonary arteries, the right ventricle was opacified (Fig. 3a, b).

Following the catheterization, an ascending aorta-right pulmonary artery (Waterston) anastomosis was performed, but severe metabolic and respiratory acidosis persisted and death occurred 12 hours after operation.

**Necropsy Findings**

The heart was enlarged, weighing 68 gm (anticipated normal up to 20 gm). The right atrium was dilated and received both the superior and inferior venae cavae. The coronary sinus was normal as was the right atrial appendage. There was no tricuspid orifice or tissue, while a shallow dimple was present at the anticipated location of the tricuspid valve. The only outlet for the right atrium was a widely patent, valvular competent foramen ovale.

The left atrial wall was moderately hypertrophied. The pulmonary veins joined the left atrium, and the mitral valve was normal. The left ventricular wall showed marked hypertrophy, especially of its septal component in the subaortic area (Fig 4a), yielding a picture of hypertrophic muscular subaortic stenosis. No ventricular septal defect was present. The aortic valve was normal and the aorta was dilated, the ascending portion measuring 2.5 cm in diameter. The great vessels were normally related, and the aortic arch was left-sided. A moderate-sized patent ductus arteriosus was present. The pulmonary trunk arose from the right ventricle and was uniform in caliber, measuring 1.5 cm in diameter and branching normally. There was no remnant of pulmonary valvular tissue (Fig 4b). The right ventricle was represented by a pouchlike structure lying basically in an anterior position to the right of the interventricular sulcus. The only communication this chamber had with the circulation was through the pulmonary trunk. This ventricle measured 2 cm in length, while the left ventricle, from apex to aorta, measured 4 cm.

**Comment**

In the case described, the findings are summarized in Figure 5. This shows tricuspid atresia, subaortic stenosis and an intact ventricular septum. The right ventricle was a small, blind, pouch-like structure communicating only with the pulmonary artery. The pulmonary valve was absent, and the pulmonary arterial supply was from the aorta through a patent ductus arteriosus.

Absence of the pulmonary valve is a rare cardiac anomaly. In 1847, Chevers credited Crampton (1830) and Favell (1842) with having been the earliest authors to describe this anomaly.

In each case, associated anomalies were present. In Crampton's, these took the form of what is now known as the tetralogy of Fallot. In Favell's case tricuspid atresia was present but, in contrast to our case in which the ventricular septum was intact, a ventricular septal defect was also present. To our knowledge, the latter case is the only one in which absence of the pulmonary valve in association with tricuspid atresia has been reported. Since then, relatively few cases of congenital absence of the pulmonary valve have been reported. Usually the

FIGURE 4. Gross specimen of heart. (a) Left ventricle (LV) and aorta (A). Hypertrophy of left ventricular wall with particular prominence of ventricular septum in subaortic region. (b) Interior of pouchlike right ventricle (RV) and pulmonary trunk (PT). No pulmonary valvular tissue identifiable.
KARTAGENER'S SYNDROME AND DEAF MUTISM

Figure 5. Diagrammatic portrayal of central circulation indicating that right ventricle filled from pulmonary arterial system; latter, in turn, had received its supply through patent ductus arteriosus. Other features show the characteristic right-to-left shunt of tricuspid atresia.

condition is associated with a ventricular septal defect or tetralogy of Fallot, although in a few cases it has been described as an isolated anomaly or associated with other cardiac anomalies. Absence of the pulmonary valve cusps is easily recognized in some cases; in others, it may be impossible to diagnose clinically and is first identified at necropsy. The classic clinical features are: extreme enlargement of the pulmonary arteries; a single second sound; and a to-and-fro murmur. In the present case, it was not possible to identify the entity clinically because the pulmonary arteries were of normal size, and there was no to-and-fro murmur. These differences might be explained by the fact that the right ventricle was a nonessential appendage of the pulmonary circuit; therefore blood flow across the pulmonary valve was limited.

Since the pulmonary arteries were of adequate size and the patent ductus arteriosus was nonobstructive, how could decreased pulmonary blood flow be explained? It appears that the severe subaortic obstruction led to inadequate cardiac output, decreasing pulmonary and systemic blood flow.

The usual electrocardiographic findings in tricuspid atresia are left axis deviation of the QRS in the frontal plane and left ventricular hypertrophy. Our case was atypical for tricuspid atresia, since the QRS axis was +80° in the frontal plane and biventricular hypertrophy was present. This type of QRS axis has been described in cases of tricuspid atresia associated with transposition of the great vessels or with pulmonary atresia.

Approximately 10 percent of patients with tricuspid atresia have pulmonary atresia, with the right ventricle excluded from the circulation. In the remaining cases of tricuspid atresia, the pulmonary valve is functional and a ventricular septal defect is usually present. In these patients, the right ventricle fills from the left ventricle and empties into the pulmonary trunk. In our case, the right ventricle, acting as an appendage, did not contribute an essential pump for the circulation.

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Kartagener's Syndrome and Deaf-Mutism: An Unusual Association*

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Two of three siblings with deaf mutism in a Negro family had complete Kartagener's syndrome. This association has not been reported before. Kartagener's syndrome is rare in the Negro race.

In 1933, Kartagener emphasized the simultaneous occurrence of situs inversus, bronchiectasis, and sinusitis. This triad is now known as Kartagener's syndrome. The incidence of this combination has been estimated at about 1:40,000 in the general population. To date, over 400 cases have been recorded in the world literature.

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