CASE REPORTS

Total Anomalous Pulmonary Venous Connection and Severe Pulmonic Stenosis in a 52-year-old Man

JOHN O. PASTORE, M.D., CARY W. AKINS, M.D., LEONARD M. ZIR, M.D., MORTIMER J. BUCKLEY, M.D., AND ROBERT E. DINSMORE, M.D.

SUMMARY Total anomalous pulmonary venous connection (TAPVC) is a rare cardiac congenital abnormality which usually results in death in infancy or early childhood. We describe a 52-year-old man with TAPVC and severe calcific pulmonic stenosis. Both diagnoses were suspected clinically and from the chest X-ray. Cardiac catheterization was confirmatory, and the patient underwent surgery. His initial postoperative course has been unremarkable. The hemodynamic factors relevant to long survival and relative freedom from symptoms in TAPVC are considered based on findings in this patient.

TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION (TAPVC) is a rare congenital cardiac abnormality which, untreated, usually results in death in infancy. This generally poor prognosis contrasts with that of most patients with partial anomalous pulmonary venous connection. However, an occasional patient with TAPVC survives into adulthood without therapy and even without severe symptoms.

We have recently treated a patient with TAPVC who, because of his age, nearly asymptomatic state, and associated cardiac abnormalities, is of unusual interest.

Case Report

A 52-year-old man was admitted to the Massachusetts General Hospital for the first time on March 15, 1975, for cardiac catheterization. He was the product of a normal, full-term pregnancy and uncomplicated delivery. He was first noted to have a heart murmur at age two years. Because of it, his activity was restricted during his school years by various physicians. The onset of cyanosis was uncertain, but he was first aware of it at age 22 while swimming.

He led a normal life except for refraining from sports and heavy work. He never squatted, was not aware of dyspnea, and did not tire easily. At the age of 47, he underwent right orchiectomy because of infarction of the right testicle. Four months before the present admission, he had an episode of pleurisy which resolved spontaneously. Three months before admission, he noted the sudden onset of aphasia and transitory right-sided weakness. The motor defect resolved completely over several days but moderate expressive aphasia remained.

Because of his obvious cyanosis and abnormal chest X-ray, cardiac catheterization and possible surgery had been proposed in the past but rejected by the patient. However, following his cerebral vascular accident, the patient became more receptive to investigation of his cardiac defect and consented to admission. On physical examination, he was plethoric and cyanotic with clubbing of his fingers and toes. Blood pressure was 106/70/70 mm Hg in the left arm and 110/70/70 mm Hg in the right arm. The pulse rate was 72/min and regular; respirations were 16/min. He was afebrile.

The jugular venous pulsations were estimated to reveal a central venous pressure of 5 cm H2O. There appeared to be an increased A wave in the jugular venous pulse tracing. The carotid pulsations were normal. The chest configuration was unremarkable. The lungs were clear. Cardiac examination revealed a faint, lateralized left ventricular impulse with a mildly sustained right ventricular heave at the left sternal border. The first heart sound was normal; the second heart sound was single and had a snapping quality at the left sternal border. A right ventricular S4 gallop and a loud right ventricular S4 gallop were audible. No click was heard; there was a grade III/IV harsh systolic murmur extending from the first heart sound to the second heart sound. It was loudest at the left second intercostal space at the sternal margin and radiated to the left infraclavicular region; it did not change with Valsalva’s maneuver. No diastolic murmur was heard.

The abdomen was unremarkable. The extremities were negative except for clubbing and cyanosis. All peripheral pulses were normal.

The hemoglobin was 20.3 gm/100 ml; the hematocrit was 63%. The white blood cell count was 9,000 and platelets were 114,000/mm3. Urinalysis was unremarkable. The blood urea nitrogen, creatinine, and serum electrolytes were normal. Arterial blood gases (breathing room air) revealed a pO2 of 47 mm Hg, pCO2 of 29 mm Hg; and pH of 7.36. Cardiac fluoroscopy (fig. 1) revealed enlargement of the left ventricle without evidence of atrial enlargement. The pulmonary arteries were diffusely increased in size with greater increase on the left than on the right. The pulmonic valve was heavily calcified. The aortic arch was left-sided and normal in size. There was a 2 cm in diameter vascular structure arising at the level of the left pulmonary hilum traveling upward and lateral to the aorta on the left. The superior vena cava on the right was dilated. An electrocardiogram (fig. 2) showed sinus rhythm with frequent ven-
tricular premature beats; the rate was 72/min and the frontal plane axis was +135°. There was evidence of biventricular enlargement and right ventricular hypertrophy.

On March 19, 1975, the patient underwent right and left heart catheterization and right ventricular and vertical vein angiography.

At cardiac catheterization, performed from a right median antecubital vein, a catheter was passed into a large left vertical vein draining the anomalous collecting channel. Oxygen saturation in the vertical vein was 95%; simultaneous brachial artery saturation was 86%. Right heart catheterization revealed the pulmonic valve leaflets to be heavily calcified. The pressure in the right ventricular body and infundibulum was 125/15 mm Hg, while the pulmonary artery pressure was 20/5 mm Hg. The right atrial mean pressure was 3 mm Hg. Left heart catheterization was performed by passing the catheter through an apparently large atrial septal defect into the left atrium where the mean pressure was 3 mm Hg. The left ventricular pressure was 125/12 mm Hg.

The systemic cardiac output (Fick) was 7.4 L/min and the pulmonary output was 9.4 L/min. A left-to-right shunt was 5 L/min, and the right-to-left shunt through the atrial septal defect was 3 L/min.

Right ventricular angiography revealed findings typical of valvular pulmonic stenosis (fig. 3) and on levophase outlined total pulmonary venous connection to a common chamber behind the left atrium. The vertical vein drained this common chamber into the right superior vena cava (fig. 4).

At operation, a markedly enlarged left innominate vein was observed to drain into an extremely large superior vena cava. The aorta was somewhat small for a heart of this size. The right ventricle and right atrium were considerably enlarged. There was myocardial fibrosis scattered throughout the wall of the right ventricle, possibly indicative of previous myocardial ischemia. Palpable through the pulmonary artery was an extremely calcified pulmonic valve with a prominent thrill suggesting pulmonic stenosis. The pulmonary veins drained into a common channel behind the pericardium. This trunk then emptied into an anterior left-sided ascending vein which drained into the left innominate vein.

The patient was cannulated and placed on cardiopulmonary bypass. Systemic hypothermia to 28°C was achieved while the heart was bathed in iced Ringer's lactate. The right atrium was opened to reveal an atrial septal defect measuring approximately 3 × 6 cm. Through this defect one could see a normal mitral valve. The apex of the heart was then elevated, the posterior pericardium was opened, and the anomalous venous trunk was isolated with tourniquets. An anastomosis was performed side to side with the common venous channel to the posterior wall of the left atrium. The atrial septal defect was closed with a Dacron patch through the right atrium.

The pulmonary artery was opened to reveal a severely scarred pulmonic valve that was heavily calcified with only a very small central orifice. The valve was excised leaving a pliable anulus. A #23 Hancock stented porcine xenograft aortic valve prosthesis was sewn into position. In order to seat a prosthesis of this size, the anulus had to be divided and expanded with a diamond-shaped gusset of pericardium backed with Teflon felt. The ascending venous channel on the left side was then ligated. The patient came off cardiopulmonary bypass without difficulty and was then restudied on the operating table. Systemic arterial pressure was 105/75, right ventricular pressure was 45 to 55 mm Hg; peak systolic with an end-diastolic pressure of 8, and the pulmonary artery pressure was 25/12. Peak to peak gradient
FIGURE 3. Right ventricular angiogram, lateral view, demonstrating valvular pulmonic stenosis.

varied between 20 and 25 mm Hg across the porcine prosthesis. It was evident that the gradient was at the valve and not in the subvalvular region. In light of the marked reduction in the RV pressure it was felt that a larger prosthesis would not be necessary. The PO$_2$ at the time of coming off bypass was 440 mm Hg torr. The PO$_2$ in the next 24 hours decreased to 250 to 350 mm Hg torr on 100% oxygen. At the time the patient was extubated he maintained a PO$_2$ of 90 mm Hg torr on room air. His entire postoperative course was benign.

**Discussion**

The patient presented is remarkable not merely because of his age before surgical repair of his cardiac defect but also because he had few cardiac symptoms until he suffered a cerebral vascular accident four months prior to his referral to our hospital.

There has been considerable speculation on the factors relevant to long survival in patients with TAPVC. It seems clear that patients with this defect who come to medical attention in infancy because of cyanosis or congestive failure or both are at increased risk of early death, with or without surgery.

Several series of patients with TAPVC reported over the last two decades contain relatively small numbers of patients who survived into adulthood and even middle age. In general, these favored patients tend to have large atrial septal defects, allowing for a more adequate systemic flow. In addition, the longer surviving patients tend to have normal to near normal pulmonary artery pressures and are thus protected from the irreversible pulmonary vascular changes which herald early demise in less fortunate individuals with TAPVC.

There are, however, exceptions to the above generalization. Singh et al. have described two adult patients over 40 years of age with TAPVC, both treated successfully surgically. One of the two, a 46-year-old man, presented with intolerance for heavy exertion as his only symptom. At cardiac catheterization he had no pulmonic stenosis and a pulmonary artery pressure of 63/32 mm Hg. His atrial septal defect, measured at surgery, was 4 cm in diameter. Their other patient, previously the oldest reported survivor with TAPVC, was a 50-year-old woman with considerable dyspnea. At catheterization her pulmonary artery pressure was 38/16 mm Hg. The atrial septal defect was "large" at operation.

While there are exceptions, it seems likely that factors delaying the onset of obliterative pulmonary hypertension tend to retard the development and severity of symptoms in patients with TAPVC. The long survival of our patient with a markedly severe gradient across the pulmonic valve would support the contention of Gott et al. that "the dampening effect on the greatly increased pulmonary flow by the stenotic pulmonary valve is of some value in retarding the onset of damaging pulmonary vascular changes." Thus, the optimal situation for long survival with this lesion is coexistence of a large atrial septal defect for maximal systemic flow and a low pulmonary artery pressure for preservation of lung vasculature.

We advised surgery in our patient despite his relatively benign presurgical course for several reasons. The preservation of pulmonary normotension had been purchased at the expense of hemodynamic derangement proximal to the stenotic pulmonary valve. The right ventricle was shown at cardiac catheterization to be generating systemic pressures. On close questioning he did give a history of mildly increasing easy fatigue over the year prior to admission.

FIGURE 4. Levophase of pulmonary angiogram demonstrating anomalous pulmonary vein (PV) draining into right superior vena cava (SVC). RPV = right pulmonary vein.
However, the case for operation rested on the desirability of preventing further emboli to the brain.

Surgical Considerations

The repair of this form of total anomalous pulmonary venous connection which falls into the supracardiac type as described in the classification by Darling et al.\(^1\) was performed via a retrocardiac anastomosis as described by Cooley et al.\(^2\) In our patient the atrial septal defect was not large enough to allow the performance of this anastomosis through the right atrium. The closure of the atrial septal defect was performed in the standard fashion for a large atrial septal defect of the adult type, that is, utilizing a prosthetic patch. Since there was no hope of conservative reconstruction of the totally calcified pulmonary valve and because there was evidence of prior right ventricular damage as manifested by the fibrosis scattered throughout the wall of the right ventricle, it was felt that pulmonic valve replacement was indicated. Furthermore, laboratory evaluation by Austen et al.\(^3\) has shown that the acute induction of pulmonary valvular insufficiency in animals could lead to prolonged right ventricular failure. Ross and Somerville\(^4\) have reported that in patients who have gradual deterioration of the fascial artery reconstruction of the right ventricular outflow tract, subsequent pulmonic regurgitation is poorly tolerated. In our patient we were concerned that the very small pulmonary anulus would require a very large patch and might result in high grade pulmonic insufficiency. In light of the recent success with porcine prostheses in multiple sites, especially in conduits for the reconstruction of the right ventricular outflow tract, it was decided that a valve replacement would be done in this patient to prevent severe pulmonic insufficiency. A conduit was not selected because the patient’s very large right ventricle would have made the placement of a conduit difficult. The use of a prosthetic fitting into the normal anular area with only a moderate enlargement of the area with a gusset made the placement of the valve anatomically acceptable and hemodynamically proper. A Hancock stented porcine xenograft aortic valve was chosen because of its good central flow characteristics and the probable need for only short-term anticoagulation.

References

Total anomalous pulmonary venous connection and severe pulmonic stenosis in a 52-year-old man.
J O Pastore, C W Akins, L M Zir, M J Buckley and R E Dinsmore

doi: 10.1161/01.CIR.55.1.206

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/55/1/206