fibrous ridge (Fig 2). The two orifices were of unequal size; the smaller one was anterior to and to the left of the bigger one (Fig 2 and 3). The valve leaflets were inserted into four groups of very hypertrophic papillary muscles inside the ventricle (Fig 3). The two anterior leaflets fused together on the midline and showed fibrous continuity with the noncoronary aortic cusp. The aortic valve was posterior to and to the right of the aortic pulmonary valve. The main ventricular chamber had the smooth septal surface characteristic of a left ventricle. This cavity was in communication with an anterior and right-sided outlet chamber via two ventricular septal defects, one posterior and restrictive and the other anterior and of the malalignment type (Fig 3). The histologic findings in the lungs showed multibaculum bronchopneumonia, with no signs of pulmonary vascular disease.

**DISCUSSION**

The case herein reported can be regarded as tricuspid atresia with a double-outlet left atrium. To the best of our knowledge, such an entity has never been reported before. Duplicated and accessory atrioventricular orifices were first described as an occasional finding at autopsy in otherwise normal hearts. Various etiologies have, therefore, been considered of interest merely from a developmental point of view. Other authors then described cases of double mitral orifice associated with other congenital cardiac defects. Such a malformation may, therefore, be of relevant importance from a surgical standpoint. The double-orifice mitral valve, in fact, may or may not have a normal function. In our case, no clinical and angiographic signs of atrioventricular valvular incompetence were present.

As far as terminology is concerned, it seems reasonable to distinguish between double and accessory orifice, previously used as interchangeable terms. Indeed, duplication of a valve implies a congenital process leading to the abnormal development of two structures, more or less normal; instead, an accessory orifice does not necessarily mean a congenital malformation but may occur also as a consequence of acquired disease (for instance, endocarditis).

In our case, we believe that a true duplication of the mitral valve took place. Thus, it can be considered a true double-outlet left atrium. Why the primitive mitral orifice subdivided into two openings is not known. The most accepted theory is an anomalous development of the endocardial cushions. So, far, little importance has been granted to the ventricular architecture, which in all described cases, as in other congenital mitral disease, is rather abnormal. In our patient the ventricular architecture was, to say the least, very bizarre (Fig 3). Thus, left ventricular modeling may be the cause of the duplication of the mitral valve and possibly of other congenital mitral valvular abnormalities.

An alternative hypothesis, suggested by the association with tricuspid atresia, could be the failure of the primitive atrioventricular canal to migrate to the right. If this were true, the two atrioventricular orifices should be regarded as a mitral and a tricuspid valve. Such a theory seems untenable because of the presence of a normal atrial septum primum (Fig 2) and the structure of the two valve orifices, neither showing chordal attachments to the septal surface (Fig 3) or the other anatomic features of the tricuspid valve.

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Idiopathic Pulmonary Hypertension Associated with Nodular Pulmonary Infiltrates and Portal Venous Thrombosis

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IDIOPATHIC PULMONARY HYPERTENSION 111
A case of idiopathic pulmonary hypertension is associated with portal venous thrombosis. The patient’s chest roentgenogram revealed numerous nodular densities which at necropy were found to have been caused by an exuberant fibroelastic reaction around small pulmonary arterioles. This is an example of the unusual association of idiopathic pulmonary hypertension and portal venous thrombosis, and to our knowledge, the first report of roentgenographically visible pulmonary nodules secondary to the pathologic alterations of idiopathic pulmonary hypertension.

Idiopathic pulmonary hypertension (IPH) is an uncommon disorder which is roentgenographically characterized by central pulmonary arterial enlargement, cardiomegaly, and clear lung fields. Although it is associated with extensive pathologic changes in the pulmonary vascular system, these changes have not previously been found to cause abnormalities on the chest roentgenogram. Our patient had the previously reported combination of primary pulmonary hypertension and hepatic venous thrombosis, and is, we believe, the first reported example of the pulmonary arteritis of IPH resulting in roentgenographically visible pulmonary nodules.

CASE REPORT

A 16-year-old girl was transferred to University Hospital with a four-week history of dyspnea on exertion. At seven years of age she had been found to have splenomegaly but remained well until the age of 13 years when she experienced hematemesis. Evaluation at that time included a chest roentgenogram which revealed an enlarged pulmonary artery, slight cardiomegaly, and clear lung fields. Hepatic angiography revealed portal hypertension secondary to portal venous thrombosis, and roentgenograms of the upper gastrointestinal tract demonstrated gastric and esophageal varices. She was diagnosed as having portal hypertension secondary to portal venous thrombosis, and a splenorenal shunt was performed without complication.

The patient remained well until the age of 16 years when she noted over a four-week period increasing dyspnea on exertion. She was admitted to the University of Michigan Medical Center. Physical examination showed a resting respiratory rate of 40 per minute and cyanotic nail beds. Systemic blood pressure was 110/70 mm Hg, and body temperature was 38.6°C. Her breath sounds were normal, and she had no thoracic deformities. The pulmonic component of her second heart sound was increased but no murmurs or extra sounds were heard. The remainder of her physical examination was within normal limits.

Laboratory examination showed a hemoglobin value of 11.6 g/100 ml, a white blood cell count of 28,800/cu mm, arterial Po2 of 60 mm Hg, arterial Pco2 of 32 mm Hg, arterial pH of 7.51, and an arterial oxygen saturation of 93.5 percent. A chest roentgenogram revealed enlargement of the pulmonary outflow tract, slight cardiomegaly, and diffuse nodular pulmonary infiltrates (Fig 1 and 2). An ECG showed sinus tachycardia, right axis deviation, and right ventricular hypertrophy. The urine contained many bacteria and leukocytes, and culture of the urine grew Escherichia coli.

The patient was treated for her urinary tract infection with ampicillin and rapidly defervesced. Two days after admission, she was undergoing a series of chest roentgenograms with barium in the esophagus when she became extremely short of breath. She was returned to her room and shortly thereafter became bradycardic and died.

NECROPSY

The left pleural cavity contained 300 ml of sanguinous fluid and the right contained 50 ml of serosanguinous fluid.
though the pathogenesis of these vascular alterations has not been firmly established, Wagenvoort and Wagenvoort\(^4\) have suggested that they occur secondary to intense pulmonary vasoconstriction.

Our patient had all of the pulmonary vascular changes mentioned above, and in addition, had an exuberant fibroblastic reaction around numerous pulmonary arteries. This kind of fibroblastic reaction has not been previously reported in patients with IPH and was probably a focal reaction to her necrotizing pulmonary arteritis. Since no other pathologic changes were found in the lungs, we believe that the nodular pattern of densities on this patient’s chest roentgenogram resulted from these 1 to 3 mm areas of fibroblastic reaction around the pulmonary arteries. Although it has been demonstrated that isolated nodular densities 3 mm in diameter are at the lower limit of size required to cause roentgenographically visible densities, multiple nodular densities as small as 1 mm in diameter may be seen as superimposition shadows.\(^6\) It is likely that most, if not all, of the nodular densities seen on our patient’s chest roentgenogram represent such superimposition shadows.

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Restrictive Cardiomyopathy in Pseudoxanthoma Elasticum*

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A unique case of severe and diffuse endocardial fibroelastosis covering the entire left ventricle and associated with pseudoxanthoma elasticum is presented. The clinical picture was that of an apparently primary restrictive cardiomyopathy. Specific histologic lesions could be recognized in the endocardium.

Pseudoxanthoma elasticum (PXE) heart disease has not been clearly characterized since autopsy reports are extremely rare and hemodynamic data are not available. Two categories of cardiac involvement have been

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