there was a concomitant antibody rise. Although the patient survived, no follow-up data were provided. Finally, a case of fatal pneumonia was reported in a middle-aged woman in which adenovirus type 21 was isolated from a stool specimen and intranuclear eosinophilic inclusions were seen in alveolar lining cells. There was a rise in adenovirus CF titer. Interstitial fibrosis and bronchiolitis obliterans were found at autopsy.

Our case was noteworthy in several respects. First, the patient, a previously healthy, civilian adult presented with severe respiratory distress due to adenovirus type 21. The virus was isolated from respiratory secretions, and there was a significant antibody rise. He subsequently had a dramatic response to CPAP and was thereby spared the risk of morbidity and mortality associated with intubation or high concentrations of inspired oxygen. This case and others suggest that CPAP may be particularly suited to severe viral pneumonia. Finally, our patient was free of symptoms and able to tolerate vigorous exercise 15 weeks after discharge. Pulmonary function studies demonstrated resolving, predominantly restrictive, lung disease. The long-term prognosis of adenovirus pneumonia in adults may be more favorable than anticipated based on previous reports of sequelae following adenovirus pneumonia in children and severe influenza pneumonia in adults in whom hypoxia, worsened by exercise, and reduced diffusion capacity have been documented as long as one year after admission.

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Tricuspid Atresia with Double-Outlet Left Atrium*

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Clinical and pathologic findings of an unusual case of cardiac malformation are presented. The main features were those of atresia of the right atrioventricular valve associated with two distinct atrioventricular orifices connecting the morphologically left atrium to the underlying morphologically left ventricle. Other distinguishing characteristics were ostium secundum atrial septal defect, normally related great arteries, with the aorta arising from the main ventricular chamber and the pulmonary artery from the anterior and right-sided outlet chamber, and infundibular and valvular pulmonary atresia.

Duplication of the mitral orifice is a rare congenital cardiac anomaly, often reported as an occasional finding at autopsy. In a few instances, such duplication has been described in association with other congenital cardiac defects such as complete atrioventricular canal, ostium primum atrial septal defect (either isolated or with pulmonary stenosis), bicuspid aortic valve, and coarctation of the aorta.

To the best of our knowledge, duplication of the mitral valve has never been reported in combination with tricuspid atresia. It is the purpose of this report to describe a case with this unusual association.

DEFINITION OF TERMS

In regard to duplication of a valvular orifice the terms double and accessory valvular orifice, have previously been

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TRICUSPID ATRESIA 100
used interchangeably.5,6 Nevertheless, duplication refers to the presence of two complete valvular apparatuses, either in the right or left atrioventricular orifice.1,2 The term accessory valvular orifice is used to refer to the presence of one or more additional openings within the same valvular structure.8 Tricuspid atresia is defined as the absence or imperforation of the tricuspid valve.9

CASE REPORT

A nine-year-old girl was admitted to our hospital for elective cardiac catheterization and reevaluation of her congenital heart disease, for which she had required a Waterston anastomosis at six months of age. On physical examination, she was a rather small, mildly cyanotic girl; both height and weight were under the third percentile. No signs of overt cardiac failure were present. The patient had been receiving digoxin since the time of the Waterston shunt. The pulses were bounding; blood pressure was 115/50 mm Hg. Cardiac examination revealed a hyperactive precordium, a single second heart sound and a blowing continuous murmur all over the chest. The electrocardiogram showed a QRS axis on the frontal plane at about +60° and biatrial and biventricular hypertrophy.

At cardiac catheterization, the ratio of pulmonary to systemic blood flow was greater than 3:1, and pulmonary vascular resistance was low (about 3 units/sq m). The right atrial injection showed a pattern of tricuspid atresia; at the left ventricular injection the subpulmonary infundibulum appeared atretic, with the pulmonary blood supply being entirely dependent on the Waterston anastomosis.

Figure 1. View of right atrium showing blind atrial floor (arrow) and conduit (C) between right atrial appendage and main pulmonary artery. EV, Eustachian valve; and cs, coronary sinus.

Figure 2. Left atrium with two distinct atrioventricular orifices (1 and 2) separated by well-formed fibrous ridge (fr). LAA, Left atrial appendage; and ASD, atrial septal defect, patched surgically.

On the basis of the low value of the pulmonary vascular resistance and the normal pulmonary arterial pressure, the girl was considered a candidate for a Fontan operation.10 The postoperative course was complicated by severe respiratory insufficiency, retention of fluids, and sepsis. She died on the 26th postoperative day.

Postmortem Findings

The most important findings were confined to the heart and lungs. There was viscerotrastr situs solitus with levocardia and d-ventricular loop; surgical repair had been performed by means of a valved conduit between the right atrial appendage and the main pulmonary artery (Fig 1). The heart, when viewed from inside, showed normal systemic and pulmonary venous drainages, tricuspid atresia (Fig 1), and a surgically patched large atrial septal defect, secundum type (Fig 2). The left atrium connected to the underlying morphologically left ventricle by means of two distinct atrioventricular annuli, separated from each other by a well-formed
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 cusp. The aortic valve was posterior to and to the right of the atrioventricular 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 multlobar 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.3-5,8 These anomalies have, therefore, been considered of interest merely from a developmental point of view.6 Other authors then described cases of double mitral orifice associated with other congenital cardiac defects.4,7 Such a malformation may, therefore, be of relevant importance from a surgical standpoint.8,9,10 In this 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.8,8 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).11

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.4,7,8 So far, little importance has been granted to the ventricular architecture, which in all described cases, as in other congenital mitral disease,12 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 leaflets 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.13

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