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in case of functional impairment of the lung due to excessive pleural thickening or to confirm the diagnosis of asbestosis by open biopsy in difficult cases. The outlook for workers exposed to asbestos has improved greatly since the advent of better dust control systems; greater awareness of the condition; early removal of the affected worker from the job; better health care facilities for the workers; and the use of antibiotics and ancillary measures for improved tracheobronchial toilet. Patients now often live long enough to demonstrate the possible carcinogenic or cocarcinogenic effect of asbestos fibers.

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Hypoplasia of Right Ventricle and Tricuspid Valve in Three Siblings*

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A case of hypoplasia of the right ventricle and tricuspid valve is reported in the third affected sibling of a family. The condition, which has a strong familial tendency, usually is responsible for death in infancy. Exceptional patients live to adult life. The functional problem is that of inflow obstruction into the right ventricle. A transatrial right-to-left shunt is characteristic and results in cyanosis. Diminished right ventricular forces in the electrocardiogram represent an important point in differential diagnosis among patients with cyanotic congenital heart disease.

A strong familial tendency1-2 is noted in congenital isolated hypoplasia of the right ventricle and tricuspid valve.

This report describes the case of the third sibling in a family to be afflicted with this entity. The cases of the first and second siblings of the family concerned were reported earlier by Raghib and associates3 and by Davachi and co-workers,4 respectively. A brief summary of the cases of the two siblings reported will be given before a more detailed description of the present and third case.

CASE REPORTS

Case 1 (Reported by Raghib and associates3)

A male infant, first born of the family, was first examined at the age of two days because of cyanosis. There were no cardiac murmurs and no signs of congestive heart failure. The electrocardiogram showed a QRS electrical axis of +100 degrees with a precordial pattern indicating either left ventricular hypertrophy or hypoplasia of the right ventricle.

A thoracic roentgenogram revealed that the heart was slightly enlarged and the pulmonary vascular markings appeared to be decreased. A venous angiocardiogram revealed

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a massive right-to-left shunt at the atrial level with opacification of the right ventricle, thereby excluding tricuspid atresia. In the neonatal period, a diagnosis of pulmonary valvular atresia was then made but, at operation, no abnormality of the pulmonary valve was found and no corrective procedure was done. In the interval between the time of the operation and death at the age of three months, the infant manifested progressively increasing cyanosis and dyspnea.

At necropsy, both the right ventricle and tricuspid valve were hypoplastic, while the pulmonary valve was normal. A patent foramen ovale was present.

**Clinical Observations**

This male infant, being the third consecutive sibling in the family, was born approximately three years after the second child (case 2). No abnormalities during pregnancy had been noted. Cyanosis was noted shortly after birth and this increased in degree within 24 hours. When admitted at the age of 36 hours, the physical examination showed a vigorous, cyanotic, male infant. The lungs were clear. The heart was not enlarged. The first cardiac sound appeared normal and the second sound was single. A grade II/VI, systolic, ejection-type murmur was present along the upper left sternal border. The liver was palpable 2 cm below the right costal margin.

Thoracic roentgenograms revealed a normal-sized heart with diminished pulmonary vascular markings (Fig 1). The electrocardiogram, being similar to that of cases 1 and 2, showed a QRS axis of +60 degrees with a precordial lead pattern of absence of right ventricular forces (Fig 2).

Angiocardiography showed a hypoplastic right ventricle with a normal pulmonary valve (Fig 3). A right-to-left shunt was present at the atrial level and a left-to-right shunt through a patent ductus arteriosus.

When the infant was two days old, a surgical anastomosis between the right pulmonary artery and the ascending aorta (Waterston's procedure) was performed. This resulted in diminution of the cyanosis but chronic cardiac failure persisted in spite of digitalization.

The patient was readmitted at the age of three months with cyanosis, tachypnea and tachycardia and subcostal and inter-
costal retraction. Examination showed the lungs to be clear but the cardiac size was enlarged. A soft, continuous murmur was present over the back. A gallop rhythm was present. Treatment consisted of increasing the dosage of digitalis and administration of diuretics and oxygen.

Cardiac catheterization and angiography were repeated. These studies showed the right ventricular pressure to be 32/0-13 (mm Hg). The right atrial mean pressure was 9 mm Hg with a and v waves being 19 and 8 mm Hg, respectively. Pressures in the left atrium were as follows: mean 13, a 16, v 20.

The angiocardiograms again revealed hypoplasia of the right ventricle and a right-to-left shunt at the atrial level. The aorticopulmonary anastomosis appeared to be functioning poorly. A balloon atrial septostomy (Hashkind's procedure) was performed but the general condition of the child did not improve. One week later, a Blalock-Taussig anastomosis was carried out, following which the child experienced extreme respiratory difficulties and died 36 hours after the latter operation at the age of 14 weeks.

 Necropsy Observations

The main pathologic findings were restricted to the heart. The left ventricular chamber was large, in contrast to the presence of a small right ventricle (Fig. 4). The great vessels were normally related. Signs were evident of the previously performed Waterston's procedure and the recently done left-sided Blalock-Taussig anastomosis. The diameters of the pulmonary trunk and of each main pulmonary artery were 8 and 5 mm, respectively.

Marked degrees of hypoplasia of the right ventricle and the tricuspid valve were apparent. The tricuspid orifice, which measured 12 mm in diameter, was guarded by a hypoplastic tricuspid valve. The tricuspid leaflets and their basal attachments were otherwise normal (Fig. 4b and c).

The right ventricular sinus (inflow) portion was hypoplastic (Fig. 4c). The distance between the tricuspid valvar ring and the apex of the right ventricle was 15 mm. The distance between the right ventricular apex and the pulmonary valve was 20 mm. The crista supraventricularis was normally positioned. The infundibular portion of the right ventricle, lying cranial to the aforementioned crista supraventricularis, was normal in aspect (Fig. 4d). The distance between the crista and the pulmonary semilunar cusps was 8 mm.

In addition to the papillary muscle of the conus and a posterior and an anterior papillary muscle, an anomalous papillary muscle was located between the latter two. The anomalous muscle had chordal connections to the septal half of the posterior tricuspid leaflet.

The right ventricular endocardium was slightly thickened by gray, fibrous tissue. Each of the three pulmonary cusps was normally formed and delicate. The orifice of the valve measured 8 mm in diameter.

The right atrium was normally formed, though its wall was markedly hypertrophic. The chamber was dilated. The floor of the fossa ovalis, which was 1 mm thick, showed a tear 8 mm in length representing the previously performed atrial septostomy.

The cardiac chambers on the left side of the heart were dilated and hypertrophied but otherwise normal. There were no valvular anomalies in the left side of the heart.

Grossly, the lungs showed no abnormalities. Histologic study of the pulmonary vessels showed medial hypertrophy with a slight degree of arterialization of pulmonary veins. The muscular pulmonary arteries were slightly thin-walled.

**Comment**

The congenital anomaly herein reported, that of hypoplasia of the right ventricle and tricuspid valve, is
relatively uncommon. As far as we are aware, about 14 cases have been reported in the English language literature, including the new case (case 3) presented in this report.

Of these, eight of the subjects belonged to families in which more than one sibling was affected. Medda and associates' cases involved two siblings of one family and Sackner and co-workers, three of another. In the family concerned in this report, three siblings were involved. Thus, while the condition may appear as an isolated one in a given family, there is a suggestion of a strong familial tendency. It is of interest that both sexes are affected and with approximately equal frequency, even in the familial cases.

The basic hemodynamic situation is one of inflow obstruction into the right ventricle. In the presence of this abnormality, a right-to-left shunt through the foramen ovale is classical and cyanosis is a common consequence.

The occurrence of cyanosis and other features has, in some instances, suggested a false diagnosis of pulmonary valvular stenosis with a right-to-left shunt.

On roentgenograms, neither the configuration of the heart nor the diminished vascularity of the lungs is diagnostic.

The electrocardiogram is of importance, since it clearly reveals that the right ventricle contributes little to the QRS electrical forces. As such, the electrocardiographic findings rule out the tetralogy of Fallot as the underlying malformation. The correct diagnosis of isolated hypoplasia of the right ventricle and tricuspid valve may be achieved by catheterization studies and angiography, as was done in the current case. A familial history of this condition is of considerable aid in the diagnosis in

FIGURE 4. Gross specimen of heart. a. External view from in front showing normally related great arteries (Ao = aorta; PT = pulmonary trunk). The left ventricle (LV) is large, in contrast to the small size of the right ventricle (RV). b. Interior of right atrium (RA) showing dilatation of the chamber and hypertrophy of the wall. There is a tear (arrow) in the floor of the fossa ovalis caused by balloon atrial septostomy. The tricuspid orifice (TO) is narrow. c. Interior view of right atrium (RA) and sinus part (RVS) of right ventricle. The tricuspid valve is hypoplastic but its leaflets and their basal attachments are otherwise normal. An anomalous papillary muscle (P) arises from the anteroseptal wall and has chordal connections to the septal half of the posterior leaflet. d. Interior of right ventricle, pulmonary valve and pulmonary trunk (PT). In contrast to the usual size of the inflow part of the right ventricle (RVS), the infundibulum (RVI) is of normal size.

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an affected sibling.

Of about 14 cases of the condition reported in the English literature, it is common that death occurs in infancy. Exceptional cases have however, been reported.

Popper and associates\(^1\) reported an isolated case of hypoplasia of the right ventricle and tricuspid valve occurring in a 39-year-old man. Saccner and associates\(^2\) reported the condition to occur in three siblings with the respective ages of 13, 22 and 39 years. In an isolated and unreported case referred to us by Drs. Ann B. Catts and Coolidge S. Wakai, the patient was 39 years old at the time of death.

In hypoplasia of the tricuspid valve and right ventricle, the principles of treatment are similar to those of other congenital conditions causing obstruction to flow into the right ventricle, such as tricuspid atresia or pulmonary atria with hypoplasia of the right ventricle and intact ventricular septum.

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Localized Unilateral Pulmonary Edema: An Unusual Presentation of Left Heart Failure*

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A patient with mitral insufficiency caused by infarction of the anterior papillary muscle, presenting with localized unilateral pulmonary edema, is reported. On the basis of previous reports it appears that unilateral presentation of acute pulmonary edema, although unusual, is not so rare as is generally believed. The exact underlying mechanism is not clear but various hypotheses put forward to explain the unilateral distribution include the effect of gravity and posture, variations in the pulmonary venous pressure and some disturbance of the neurogenic control of capillary size and permeability. We believe that a wider recognition of the unilateral presentation of acute pulmonary edema is important to avoid unnecessary delay in diagnosis and proper management.

Unilateral pulmonary edema presents an interesting and confusing diagnostic problem. Lack of awareness of the fact that pulmonary edema can be unilateral may lead to mistakes in diagnosis and important delay in treatment of the condition. The usual and more widely recognized form of roentgenologic presentation of pulmonary edema is one of bilaterally symmetrical opacity occupying the central zones of the lungs with a bibasilar predominance. Although a few case reports of unilateral or segmental pulmonary edema have appeared in the literature, the existence of such an entity is not commonly appreciated.

In this report we describe a patient with an unusual presentation of left heart failure, in that unilateral pulmonary edema, due to mitral insufficiency, was seen radiographically to be localized largely to the right upper lobe.

Case Report

A 62-year-old white man was admitted to Barnes Hospital on January 12, 1969. He was well until December 1, 1968, when he developed pain in the upper abdomen and left lower chest. The pain usually followed a meal and was partially relieved by antacids. An electrocardiogram showed ST segment abnormality suggestive of anterolateral myocardial injury. Examination by his private physician showed marked tachycardia and an apical pansystolic murmur which had not been heard before. The chest roentgenogram revealed an in-

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