a smaller reduction in cardiac output. 5,7

In our patient, we successfully reversed the profound hypoxemia with the use of a CPAP mask. We documented in the first three days of hospitalization a progressive decrement in Fio2 requirement, with a marked improvement in arterial oxygen tension along with a trend to normalization of the a/A ratio (Fig 2).

Of all the proposed etiologies for the arterial hypoxemia in pulmonary embolization, our patient's response to CPAP therapy supports the concept of venous admixture occurring possibly secondary to microatelectasis. We further suggest the use of CPAP mask as an adjunctive modality of therapy in the treatment of hypoxemic respiratory failure secondary to pulmonary embolization, provided the necessary guidelines and criteria8 for the selection of these patients are met.

REFERENCES


18 Fleischer F, Hampton AO, Castleman B: Linear shadows in the lung (interlobar pleuritis, atelectasis and healed infarction). Am J Roentgenol 48:610-618, 1941

Left Ventricular Outflow Obstruction Produced by a Pedunculated Fibroma in a Newborn

Clinical, Angiographic, Echocardiographic and Surgical Observations

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An unusual case of a one-day-old infant with significant left ventricular outflow obstruction produced by a pedunculated fibroma is described. The clinical findings were indistinguishable from severe valvar or subvalvar aortic stenosis. The tumor was difficult to detect by echocardiography. It produced an echo-free widening of the left ventricular outflow tract. Left ventricular cine-angiography clearly demonstrated a mobile mass beneath the aortic valve.

Fibromas of the left ventricle are rare cardiac tumors, which are usually located within the anterior wall and/or septum of the left ventricle.1-4 It has been suggested that a fibroma could create inflow or outflow obstruction of either ventricle,1-4 but the actual occurrence of obstruction is rare.4-9 Subaortic obstruction appeared to be present (at autopsy) in three cases,1-9 but hemodynamic or angiographic proof of obstruction has not been described.

The present case documents several unusual or previously undescribed features of a ventricular fibroma: (1) attachment of the tumor to the septum by a peduncle, in contrast to the usual intramural location; (2) its angiographic appearance; (3) its difficult echocardio-

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graphic detection; (4) hemodynamically significant left ventricular outflow obstruction; and (5) surgical removal of the tumor in a neonate.

Case Report

A 3,220 gm full-term female was noted to have symmetrically reduced pulses soon after birth. Systolic blood pressure in both arms by the Doppler technique was 48 mm Hg. Heart rate was 140/min and respiratory rate 40/min. A marked right ventricular lift was palpable. The left ventricular apical impulse was normal; no ejection click was present. A grade 3/6 systolic ejection murmur, which peaked in mid to late systole was loudest at the third left intercostal space. No diastolic murmur or gallop was present. The lungs were clear and the liver not enlarged.

The electrocardiogram was normal for age. A chest x-ray film showed a minimally enlarged heart, with normal pulmonary vascularity and a normally located aortic arch.

A clinical diagnosis of congenital valvular or discrete subvalvular aortic stenosis was made. Cardiac catheterization was performed at age 12 hours. The left ventricle was entered through a patent foramen ovale. The descending aorta was entered through a patent ductus arteriosus. There was a peak systolic gradient of 62 mm Hg between the left ventricular apex and the descending aorta. The systemic index was 3.47 liter/min/m². The pulmonary index was 4.9 liter/min/m² due to a shunt through the patent ductus arteriosus. Pulmonary arterial pressure was 40/23 mm Hg and the left atrial pressure was 10 mm Hg.

A biplane left ventricular cineangiogram revealed a mass in the left ventricular outflow tract, which was mobile and obstructed about 80 percent of the outflow tract. During systole the mass partially protruded through the aortic valve (Fig 1A), while during diastole (Fig 1B) it returned to a subvalvular position. Left ventricular contractility was uniform and the ejection fraction 56 percent.

A diagnosis of a mobile left ventricular tumor producing left ventricular outflow obstruction was made. Since a rhabdomyoma was considered statistically the most likely cell...
type, right ventricular and left atrial cineangiograms were obtained to exclude multicentricity. No additional tumors were found.

Following the catheterization, an echocardiogram was obtained. Even with the knowledge that a mass existed in the left ventricular outflow tract, it was difficult to demonstrate by echocardiography. A group of multiple echoes, as seen with a myxoma or rhabdomyoma, was expected but not found. Instead, a completely echo-free mass during systole was demonstrated directly beneath the aortic valve (Fig 2). The multiple echoes at the subvalvular level during diastole and at the valvular level during systole are probably produced by acoustic interfaces between the top of the tumor and blood. Since the subvalvular mass is wider than the aortic valve, the mass could not project through the valve during systole. No abnormal echoes were recorded lower in the ventricle at the mitral valve level (Fig 3). The motion of the tumor demonstrated echocardiographically is comparable with the motion demonstrated by cineangiography.

At age 34 hours surgery was performed on the infant because of concern about sudden death. Inspection of the external surface of the heart revealed no tumor and no mass could be felt in the wall of either ventricle. Normal aortic valve leaflets were visualized through a transverse aortotomy and the tumor was seen just beneath the valve. A stitch was passed through the tumor and the tumor was delivered through the aortic valve. It was attached by a stalk to the high interventricular septum and a small portion of the undersurface of the noncoronary cusp. The stalk was cut without damage to the septum or valve. No residual tumor could be palpated.

After cardiopulmonary bypass was discontinued the infant gradually became hypotensive and acidotic. Death occurred six hours after completion of the operation. Permission for an autopsy was not granted.

The tumor was firm and about 1 cm long. Microscopically it was covered by endothelial cells and was composed of fibrous tissue and fibroblasts. No myocardial cells or cystic spaces were present.

**Discussion**

Primary cardiac tumors in infants and children are rare. Rhabdomyomas comprise about 70 percent, while fibromas account for about 25 percent. Fibromas are usually large and occupy the anterior wall and/or septum of the left ventricle. The tumor is composed of fibroblasts, dense connective tissue and a small admixture of myocardial cells.

The histologic appearance in the present case was characteristic of a fibroma, but the pedunculated nature of the tumor and its location in the left ventricular outflow tract are unusual. Clinically, the tumor mimicked valvular or discrete subvalvular aortic stenosis because of the symmetrically reduced peripheral pulses and the harsh systolic ejection murmur at the left sternal border.

Left ventricular cineangiography clearly demonstrated the mobile tumor. The angiographic appearance of the tumor is strikingly similar to the pedunculated rhabdomyoma reported by Shafer et al.

The tumor appeared as an echo-free widening of the left ventricular outflow tract during systole. The mass is echo-free because of the homogeneous nature of this tumor and lack of acoustic interfaces within the tumor. This is similar to the echo-free appearance of a cystic cardiac tumor, but contrasts with the typical multiple echoes arising from a myxoma or rhabdomyoma. We were unable to find any previous report of an echo-free solid tumor.

![Figure 3. Echocardiogram at level of mitral valve. On right interventricular septum, mitral valve and posterior left ventricular wall are visualized. Anterior to mitral valve there is no evidence of mass even with critical damping and careful adjustment of gain.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21014/)
A pedunculated or intramural fibroma causing hemodynamically proven left ventricular outflow obstruction has not been described. However, there are three reports of a rhabdomyoma producing subaortic stenosis,11,14,18 and in two of the three cases the tumor was pedunculated.

Surgical excision of the tumor through an aortotomy above the aortic valve was a relatively simple matter. A similar operative approach was employed by Shaber et al14 to remove a subaortic rhabdomyoma. Because of the ease and completeness of removal of the tumor through an aortotomy we would advocate this approach and avoid a ventriculotomy.

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REFERENCES
1 Edwards JE: In Heart Disease in Infants, Children and Adolescents (Moss AJ, Adams FM, eds). City Williams & Wilkins Co, 1968, p 1072
2 Bigelow NH, Klinger S, Wright AW: Primary tumors of the heart in infancy and childhood. Cancer 7:549, 1965
3 Fine G: Primary tumors of the pericardium and heart. Cardiovasc Clinics 208, 1973

Double Ventricular Response to an Extrastimulus in a Patient with Triple Atrioventricular Pathways*

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A patient with the Wolff-Parkinson-White Syndrome was studied through recordings of the intracardiac potentials and programmed atrial stimulation. During programmed atrial stimulation at progressively shorter coupling intervals (A1-A2 intervals), the His deflection was always recorded after the ventricular complex. Thus, at coupling intervals between 295 and 250 msec, there was a double ventricular response, one through the accessory pathway (QRS complex of the Wolff-Parkinson-White morphologic pattern) and the other by the normal atrioventricular pathway (normal QRS complex or with pattern of left bundle-branch block). At a coupling interval of 295 msec, the atrio-His (A-H) interval increased from 200 to 350 msec. This fact and the presence of two distinct A2-H2 intervals are suggestive of the existence of dual atrioventricular pathways, coexisting functionally with a lateral accessory bypass (Kent's bundle).

In spite of having been thoroughly studied through the recordings of the atrioventricular potentials and through atrial stimulation with the technique using extrastimuli, the Wolff-Parkinson-White syndrome (pre-excitation syndrome) can still offer interesting aspects. Thus, a stimulus arising in the atrium or ventricle can be conducted by the accessory pathway or the normal pathway (or both) and produce arrhythmias, which are sometimes difficult to explain. This current report presents the findings in a case of Wolff-Parkinson-White syndrome with a lateral accessory pathway associated with a dual intranodal pathway, with a double ventricular response to a premature atrial depolarization.

CASE REPORT

A 45-year-old man who had diabetes that was controlled by administration of isophane insulin suspension (NPH insulin) had a routine electrocardiogram that showed patterns of the Wolff-Parkinson-White syndrome, type B; he had never had a crisis of tachycardia recorded electrocardiographically. With the patient's consent, he underwent an electrophysiologic study under local anesthesia. The His bundle was recorded by the technique described by Scherlag and associates,1 and the right atrium was scanned by the technique using extrastimuli.2 The electrodes were connected to a multichannel oscilloscope-recorder (Electronics for Medicine DR-12) with surface electrocardiographic leads (leads 2 and V1) and were recorded on photographic paper at a paper speed of 100 mm/sec. The right atrium was paced by a programmed stimulator (designed by Mr. Percival Gomes Neto, engineer, and developed by Indústria de Aparelhos Médicos e Eletrônicos, São Paulo, Brazil); the stimulator

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