surgery mimicking a left atrial mass at both echocardiography and cardiac angiography. A pericardial defect presenting as a left atrial mass at echocardiography has been reported in a patient, prompting surgical exploration of the left atrium.  

This case suggests that the findings, though impressive, from echocardiography and cardiac angiography may, at times, be misleading in the diagnosis of intra-atrial masses, in the presence of a loculated pericardial effusion.

ACKNOWLEDGMENTS: We are indebted to Mrs. Barbara Steele for her help in writing this manuscript.

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Subaortic Stenosis by Solitary Rhabdomyoma*

Successful Excision in an Infant Following 2D Echocardiogram and Doppler Diagnosis

Ennio De Dominicis, M.D.; Alessandro Frigiola, M.D.;
Gaetano Thiene, M.D.; Lorenzo Menicanti, M.D.;
Loredana Bozola, M.D.; and Giuseppe Finocchi, M.D.

A seven-month-old girl had subaortic stenosis caused by a single intracavitary rhabdomyoma unassociated with tuberous sclerosis. Diagnosis was formulated on the basis of two-dimensional echocardiography and Doppler technique findings alone, and surgery was successful.

(Chest 1989; 95:470-71)

Subaortic stenosis in infancy is usually due to a fibromuscular tunnel, septal hypertrophy, or a posteriorly deviated infundibular septum with ventricular septal defect; tumors, such as cardiac fibroma and intracavitary rhabdomyoma, are a rare substrate. Theoretically, noninvasive diagnosis by two-dimensional echocardiography should be precise enough to program surgery directly without further angiocardioanographic studies. We report the case of a seven-month-old girl in whom an obstructive subaortic rhabdomyoma was detected by two-dimensional echocardiography and the Doppler technique and was successfully excised.

CASE REPORT

A seven-month-old girl was referred for evaluation of a systolic murmur, which had been previously heard at birth. The child was the product of an uneventful, full-term pregnancy and normal delivery and weighed 3,730 g at birth.

A grade 4/6 pansystolic murmur with a crescendo-decrescendo pattern was audible in the second right intercostal space, and ECG showed tall R waves in the left precordial leads. A chest x-ray film showed a normal cardiac shadow. The child was otherwise completely asymptomatic; extracardiac malformations and tuberosclerosis were absent.

Two-dimensional echocardiography revealed the presence of a round mass, 8 mm in diameter, along the left ventricular outflow tract just below the aortic valve, but well distinct from the aortic

Figure 1. Preoperative echocardiogram. Parasternal long axis view (upper) shows mass obstructing left ventricular outflow tract located just beneath the aortic valve. Subcostal view (lower) shows left ventricular outflow tract and mass, which is attached to basal portion of anterolateral wall. LA, left atrium; LV, left ventricle; RV, right ventricle; AO, aorta.

*From the Divisions of Cardiology, Pediatric Surgery, and Pathology, the Civil Hospital, Vicenza, and the Institute of Pathological Anatomy, University of Padua Medical School, Padua, Italy. Supported by the National Council for Research, Target Project "Perinatal Pathology and its Sequelae," Rome.
Reprint requests: Dr. Thiene, Istituto di Anatomia Patologica, Via Gabelli 61, Padova, Italy 35121
semilunar leaflets (Fig 1). Continuous-wave Doppler technique indicated an aortic velocity of about 3.8 m/s, and the estimated instantaneous pressure gradient was about 55 mm Hg (Fig 2).

Surgery was performed without prior catheterization, using extracorporeal circulation with moderate hypothermia. The aorta was cross-clamped, and cardioplegia was induced. The aorta was opened, and a sessile white mass, which appeared implanted in the anterolateral muscle band, could be seen through the valve; the mass was exposed by means of a stay-stitch, and then easily excised. The aorta was then sutured, and bypass was discontinued without any problems. The child's postoperative course was uneventful: echocardiography showed perfect patency of the left ventricular outflow tract, and Doppler technique indicated a velocity of 1.6 m/s across the aortic valve (Fig 2).

Histologic examination of the surgical specimen showed the typical features of rhabdomyoma, consisting of spider cells and myofilament bundles with distinct cross-striations (Fig 3).

At eight months after surgery, the child was completely well, and results of her physical examination were normal.

DISCUSSION

Rhabdomyoma, the most common primary tumor of the heart in the pediatric age group, occurs most commonly in infants. Usually intramural, it presents as multiple, variously sized masses in 90 percent of cases and is associated with tuberous sclerosis in approximately one third of patients.6 Blood flow obstruction is a common finding both in intramural tumors, due to intracavitary extension, and in completely intracavitary tumors,7-12 in the latter, however, the surgical anatomy is much more favorable.

Successful removal of a rhabdomyoma obstructing the left ventricular outflow tract has been described in the literature,7-9,11-13 also following ultrasound diagnosis.8,9,11,13 Interestingly enough, in the patient described by Corno et al,13 surgery was performed on the basis of only two-dimensional echocardiographic findings, and the first diagnosis was made during pregnancy.

Several features make our case noteworthy. The apparently solitary, intracavitary mass was strategically located in the subaortic region, a rare position.8,11,12 Diagnosis was formulated on the basis of two-dimensional echocardiography associated with Doppler technique, which permitted not only visualization of the exact tumor size and location but also estimation of the transaortic gradient. Our patient did not have tuberous sclerosis, justifying the risk of surgery, which was performed in infancy without catheterization.

Since some degree of uncertainty persists regarding the presence of other pinpoint rhabdomyomas elsewhere in the heart, this child currently undergoes periodic follow-up with two-dimensional echocardiography.

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FIGURE 2 (left). Preoperative continuous-wave Doppler from an apical position. Maximum velocity across the aortic valve is almost 3.8 m/s, which corresponds to a gradient of about 55 mm Hg. (Right). Postoperative continuous-wave Doppler tracing shows normal velocity across aortic valve.

FIGURE 3. Classic histologic pattern with "spider cells," some of which show distinct cross-striations (arrow).
Effect of Temporary External Stabilization on Ventilator Weaning after Sternal Resection*  

J. Ali, M.D.; B. Harding, M.D., M.Ch.; and R. deNiord, M.D.

The role of mechanical fixation of the chest wall in the treatment of flail chest remains controversial. We report a case of flail chest resulting from major sternal resection. The application of a temporary external stabilization device improved pulmonary mechanics by decreasing the respiratory rate from 36/min to 10/min while increasing tidal volume and vital capacity from 140 ± 85 ml and 195 ± 90 ml, respectively, to 450 ± 110 ml and 905 ± 310 ml, respectively. The improvement with the temporary device facilitated weaning from mechanical ventilation. We recommend consideration of this technique in selected cases of flail chest resulting from major chest wall resection.

(Chest 1989; 95:472-73)

Wide excision remains the main therapy for tumors of the chest wall.1 Formal thoracotomy closure is adequate therapy with defects under the scapula; however, large sternal defects present a greater therapeutic challenge.2,3 We report a recent case of sternal resection requiring mechanical ventilation and the application of a temporary external device that facilitated weaning from the ventilator.

CASE REPORT

A 28-year-old Saudi man presented to King Khalid Hospital in Tabuk with a lower sternal mass in February 1985. Biopsies suggested an enchondroma, and resection of the lower third of the sternum followed in March 1985. Although no blood gas levels were reported at that time, the patient did not require mechanical ventilation after surgery and was discharged five days later.

*From King Faisal Specialist Hospital and Research Centre, Riyadh, Saudi Arabia, and the University of Toronto, Toronto, Canada.
Reprint requests: Dr. Ali, H 1915, 2065 Bayview Avenue, Toronto, Ontario, Canada M4N 3M5

FIGURE 1. Chest wall defect produced after excision with Marlex mesh coverage.

In November 1987, the patient presented with a one-year history of an upper sternal mass which had increased to a size involving the entire upper third of the sternum. The findings from metastatic workup, including bone scan and computerized tomographic scan of the head, chest, and abdomen, were negative. In December 1987, at left thoracotomy the tumor was found free of major vascular structures, and a wide excision was conducted, including the sternal and clavicular heads of the sternocleidomastoid muscle, the intercostal muscles, and ribs, as well as the medial third of both clavicles. The resulting defect was closed by approximating skin and subcutaneous tissue over Marlex mesh sutured into the bony margins (Fig 1). This wound was associated with significant paradoxical movement of the anterior chest wall. The specimen was examined pathologically, and the diagnosis was revised to a more malignant dedifferentiated chondrosarcoma.

After surgery, it was immediately evident that the patient's condition could not be managed without mechanical ventilation. Initial arterial blood gas levels with the patient on the respirator (fractional concentration of oxygen in the inspired gas [FiO2] of 0.6, rate of 10/min, and tidal volume [Vt] of 800 ml) were as follows: arterial oxygen pressure (PaO2), 246 mm Hg; arterial carbon dioxide tension (PaCO2), 41 mm Hg; and pH 7.34. With spontaneous ventilation the respiratory rate was 35/min, Vt was 100 ml, and tidal capacity (VC) was 280 ml. On a SIMV rate of 12/min and FiO2 of 0.3 and Vt of 700 ml, blood gas levels were as follows: PaO2, 130 mm Hg; PaCO2, 39 mm Hg; and pH 7.44. On this setting the patient was completely controlled, not exceeding the SIMV rate of 12/min. Over the next three days, unsuccessful attempts were made to decrease the SIMV rate to 4/min. Immediate flailing occurred.

FIGURE 2. External acrylic stabilization device over sternal defect.