A Rare Form of Isolated Interrupted Aortic Arch: the Value of Two-Dimensional Echocardiography in the Precatheterization Evaluation

Julio A. Morera, Vincenzo Celano, J.-Michael A. Roland, Robert L. Gingell, S. Subramanian, F.G. Torres-Aybar, and Daniel R. Pieroni

Departments of Cardiology and Cardiovascular Surgery, Children's Hospital of Buffalo, State University of New York, Buffalo, New York, USA

SUMMARY. Preoperative evaluation of a 12-year-old girl, previously diagnosed as having a coarctation of the aorta, revealed a rare form of isolated interruption of the aortic arch. Two-dimensional echocardiography played an important role in the delineation of the defect and significantly reduced the need for invasive techniques. It helped to anticipate problems in reaching left-sided structures at cardiac catheterization, saving time and unnecessary trauma. This technique also ruled out associated intracardiac anomalies that were not detailed at cardiac catheterization.

KEY WORDS: Interrupted aortic arch — Two-dimensional echocardiography

Isolated interruption of the aortic arch (IAA) is an extremely unusual vascular anomaly [2, 3] believed at one time to be incompatible with life once the ductus arteriosus (DA) closed [7]. The first case of this type was reported in 1964 [6]. Since then, 9 other patients have been reported in the literature [2, 3].

The following report serves a dual purpose. It depicts a form of isolated IAA (type C2) [1] not described previously to our knowledge, in a living patient [2]. More important, it illustrates the key role that two-dimensional echocardiography (2DE) played in the delineation of the lesion. This technique enabled the cardiologist to plan for cardiac catheterization by confirming the clinical diagnosis. Furthermore, it characterized anatomic peculiarities of the anomaly with significant accuracy; it ruled out associated intracardiac defects and avoided a futile retrograde approach to the ascending aorta and left ventricle. It kept invasive techniques to a minimum and information yield to a maximum.

We believe that a thorough 2DE evaluation, including a search of the aortic arch, ought to be performed in any patient evaluated for coarctation of the aorta or suspected of having an IAA.

Case Report

O.R. was a 12-year-old girl referred to the Children's Hospital of Buffalo for evaluation and eventual repair of an atypical coarctation of the aorta. She was noted to have a systolic murmur at birth, and shortly afterward developed congestive heart failure that responded well to medical measures. Pulses were not palpable in any of the extremities. Cardiac catheterization at 18 months of age demonstrated an obstruction of the aorta just distal to the right carotid artery.

The patient developed normally and led a fairly active life. She remained asymptomatic on maintenance doses of digoxin. Physical examination revealed a thin, but well-developed 12-year-old girl without signs of congestive heart failure. There was a prominent left ventricular impulse, which was displaced laterally. Neither peripheral pulses nor cuff blood pressures were obtainable in any of the extremities. The right carotid pulsations were clearly visible; they were vigorous enough to cause noticeable head movement with each beat. There was a palpable thrill and an audible bruit over the right carotid artery. The left carotid pulse was absent. A grade IV/VI systolic ejection murmur was heard along the left sternal border radiating to the left subclau-
lar fossa. A loud $S_4$ was present at the apex. The liver was not enlarged. Neurologic examination failed to show any hypertensive changes.

The electrocardiogram was consistent with left atrial enlargement and left ventricular hypertrophy. A standard erect chest roentgenogram demonstrated cardiomegaly with normal vascularity. Rib notching was not present. There was an abnormal shadow in the right upper parasternal area adjacent to the region of the ascending aorta (Fig. 1).

Two-dimensional echocardiography proved to be quite helpful. The standard suprasternal long-axis view displayed marked dilatation of the ascending aorta. The proximal aortic arch ended abruptly in a blind cul-de-sac after giving off a single "$S$"-shaped vessel, nearly as large as the ascending aorta itself. This vessel passed posteriorly to the right and cephalad. The left descending aorta was normally positioned and a distinct separation from the blind end of the ascending aorta was visualized (Fig. 2).

Other 2DE views disclosed left atrial enlargement and a concentrically hypertrophied, hypercontractile left ventricle. Both atrioventricular and semilunar valves were normal in

Fig. 1. Plain AP chest roentgenogram: arrows point to density in right upper parasternal area, which represents the dilated ascending aorta and right common carotid artery.

Fig. 2. 2DE (standard suprasternal notch projection, long axis): AAO, ascending aorta; RCC, right common carotid artery; DAO, descending aorta (indicated by arrows).