The Diagnosis of Aortic Septal Defect by Retrograde Aortography

Report of a Case

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This rare congenital malformation is a round or oval opening between the ascending aorta and the main pulmonary artery above the semilunar valves. It is practically impossible to differentiate this malformation from a patent ductus arteriosus clinically because both malformations may present the same physical, fluoroscopic, roentgen and electrocardiographic findings. Even angiocardiography and cardiac catheterization do not differentiate these congenital malformations. The authors have studied 2 patients with aortic septal defects. One was operated at another clinic for a suspected patent ductus arteriosus, and the other case presented here was diagnosed by retrograde aortography.

The Diagnosis of congenital aortic septal defect is one of the most difficult in the field of congenital malformations of the heart. Fortunately, it is one of the rarer malformations. A thorough review of the literature has revealed 24 reported cases.

During the past four years, we have encountered 2 cases of aortic septal defect. One, a 9 year old white male who presented findings of a patent ductus arteriosus, was operated upon elsewhere and no ductus was found. We believe that the case presented below is the first instance of an aortic septal defect diagnosed preoperatively by means of retrograde aortography, which was first introduced for visualization of the aorta and its branches, by Castellanos and Pereiras.

REPORT OF CASE

M. P. was first seen at age of 3½ months on March 8, 1949 because of fever, rapid respirations and cough of several days duration. A diagnosis of bronchopneumonia was made. Heart examination revealed the apex in the fifth left intercostal space, just outside the midclavicular line. There was a continuous murmur over the pulmonary area, which was transmitted towards the apex. The systolic murmur could also be heard over the second and third right intercostal spaces. Blood pressure was 90/15 in the upper extremities. Femoral pulsation was normal.

Fluoroscopic and roentgen-ray examinations performed after the bronchopneumonia had cleared, revealed a moderate enlargement of the transverse diameter of the heart with a straightening of the pulmonary area and with a definite hilar dance (fig. 1). The left oblique view (fig. 2) showed a moderate enlargement of the inflow tract of the left ventricle. An electrocardiogram (fig. 3) showed a left axis shift in the standard leads, a horizontally shaped heart in aVL and aVF and no evidences of heart strain in the precordial leads. A stethogram showed a continuous murmur in pulmonary area. (The stethogram was taken and read by Dr. A. Luisada.)

An angiocardiogram in the left oblique view showed reopacification of the pulmonary artery after the filling of the aorta; although these findings were compatible with the diagnosis of a patent ductus arteriosus, the presence of a systolic and diastolic murmur at such a young age (3½ months), and the recent experience with a 9 year old child who failed to show a patent ductus at operation, led us to believe that this patient might have an aortic septal defect. Since the patient was too young to have catheterization performed, and since catheterization does not differentiate aortic septal defect from patent ductus arteriosus, retrograde aortography was done. Five cc. of a 70 per cent Diodrast solution was injected into the left axillary artery and several films were taken. Figure 4 shows the dye entering the pulmonary artery from the aorta just above the semilunar valves. There is no evidence of a patent ductus arteriosus. A film, taken one second later, shows the entire heart filled with Diodrast.
DIAGNOSIS OF AORTIC SEPTAL DEFECT

Because of these findings, an operation was not advised and the patient is being kept under observation.

DISCUSSION

A thorough review of the literature reveals 24 reported cases. As far as we could ascertain,

no previously reported case was diagnosed clinically; the diagnoses were made either at autopsy or following surgery. Cotton and Hektoen reported the first case in the American literature in 1899 and 1900 and summarized the nine previously reported cases. Since then 9 single cases have been reported. Gibson, Potts and Langewisch recently reported 4 cases in children, all of whom had

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**Fig. 1.** Moderate enlargement of the transverse diameter of the heart in the posteroanterior view. Straightening of pulmonary area. Good vascular pulmonary markings.

**Fig. 2.** Left anterior oblique view. Moderate enlargement of the inflow tract of the left ventricle.

**Fig. 3.** Standard leads show a left axis deviation (important since patient was only 3½ months of age). Leads aVL and aVF show a horizontal position of the heart and the precordial leads show no evidences of heart strain.
findings suggestive of patent ductus arteriosus, and in none was a patent ductus found at operation. Two of their cases had been catheterized previously and one had an angiocardiographic study done. The blood obtained from the pulmonary artery showed a significantly higher oxygen saturation than the blood from the right ventricle, and the angiocardiogram showed a refilling of the pulmonary artery after the dye entered the aorta; both of these findings were considered strongly suggestive of a patent ductus arteriosus and yet at operation none was found.

Anatomically, there exists a communication between the aorta and pulmonary artery a short distance above the semilunar valves; the communication is usually round or oval in outline and is 10 to 12 mm. in diameter. It is not in the location of the ductus arteriosus and is not to be considered as a short ductus, neither is it an aneurysm of the right sinus of Valsalva, which occasionally ruptures into the pulmonary artery, right auricle or right ventricle.

The hemodynamics of this rare lesion are very similar to that of a patent ductus arteriosus (fig. 5). Just as in the latter condition, blood is shunted from the aorta into the pulmonary artery and there is, therefore, no cyanosis. Systolic and diastolic murmurs may be heard in the pulmonary area and may be transmitted downwards. Although one may expect the continuous murmur to be located at a slightly lower level than that of a patent ductus arteriosus, and the diastolic murmur either may be missing or may not have the rumbling quality so characteristic of the murmur of patent ductus arteriosus, it has not been possible so far to differentiate the two; the character, location and duration of the murmur in the patent ductus arteriosus is subject to considerable variations. A wide pulse pressure may be present in both malformations. Fluoroscopic and x-ray examinations may reveal a prominent convexity in the pulmonary area and a hilar dance in either case. Angiocardiog-

Fig. 4. Angiocardiogram obtained by retrograde aortography. Dye is seen entering the pulmonary artery from the aorta just above the semilunar valves. Note the semilunar valves, coronary artery and no evidences of a patent ductus arteriosus.

Fig. 5. Schematic drawing illustrating the reported locations of aortic septal defects. These sites are either just above the semilunar valves or at a higher level in the ascending aorta. The location of patent ductus arteriosus is also shown in order to emphasis the fact that the sites of the two anomalies differ.
raphy does not differentiate these two conditions because Sussman's sign (the localized bulging of the descending aorta in the region of the ductus just beyond the isthmus) is not always present and is furthermore not pathognomonic; and the indirect sign of reopacification of the pulmonary artery after the filling of the aorta may be present in either malformation. Even catheterization of the heart does not differentiate one anomaly from the other because the higher oxygen content of the blood obtained from the pulmonary artery as compared with that from the right ventricle occurs in either malformation. Differentiation of an aortic septal defect from a patent ductus arteriosus, is, however, of paramount importance because an aortic septal defect is usually not amenable to surgery. Patent ductus arteriosus can, of course, be easily corrected. As far as we know there is only one instance of successful surgical treatment of aortic septal defect.

We believe that so far ours is the only reported case proved to have an aortic septal defect by retrograde aortography. As far as we know this appears to be the only method that may differentiate an aortic septal defect from a patent ductus arteriosus. Dexter recently stated that, "venous catheterization is of little or no diagnostic value in anomalies of the aortic arch, but on one occasion aortic septal defect has been demonstrated by passage of the catheter from the pulmonary artery into the aorta, but in other suspected cases this could not be accomplished." Details on this case were not given by the author.

SUMMARY

A case of an aortic septal defect in a 3½ month old child is presented. The diagnosis was made by retrograde aortography.

REFERENCES

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