Diagnosis of Congenital Aortic Septal Defects

Description of Two Cases and Special Emphasis on a New Method which Allows an Accurate Diagnosis by Means of Cardiac Catheterization

By H. A. H. D'Heer, M.D. and C. L. C. van Nieuwenhuizen, M.D.

Two cases of aortic septal defect, diagnosed in life by means of cardiac catheterization, are described. An explanation is given of a method, used in these cases, whereby a correct diagnosis could be made before operation. Other methods for diagnosis and differential diagnosis are discussed.

Among congenital cardiac anomalies which are now amenable to cardiac surgery, none is more difficult to diagnose than aortic septal defect. All authors emphasize the difficulty of making a correct diagnosis before operation, and most of the cases which have been described were at first diagnosed as patent ductus, ventricular septal defect, truncus arteriosus or other anomalies. Among 34 cases described in the literature, 17 were diagnosed at necropsy and 12 during operation for patent ductus. On only five cases was the correct diagnosis made before operation: by retrograde aortography in three and by catheterization in two cases.

In our two cases, the diagnosis was made with certainty by the interpretation of the various positions of the catheter during cardiac catheterization.

Since the application of surgery for the relief of these anomalies depends upon correct diagnosis, we will, after a short description of our two cases, review the various methods of making a diagnosis and discuss the differential diagnosis.

Case Reports

Case 1. H. D., a 7 year old girl, was admitted to our Cardiologic Department on March 13, 1953 with the diagnosis of possible atypical patent ductus. Cardiac anomaly was first recognized when she was 3 months old.

Her complaints were fatigue, exertional dyspnea and palpitation. There was no cyanosis and she did not squat. There were also complaints of coughing and a history of frequent bronchitis. On physical examination, we found a child with a reddish color, but no cyanosis, and no clubbing of the fingers was present. The blood pressure was 105/70 mm. Hg. A harsh systolic murmur was present in the second to the fourth intercostal space along the left sternal border, beneath the manubrium and in the second and third right intercostal spaces. The second pulmonic sound was extremely loud.

The hemoglobin was 78 per cent, and the red blood cell count was 3,820,000. The electrocardiogram showed evidences of left ventricular hypertrophy with strain. On x-ray examination there was no hilar dance, but an enormous enlargement of the pulmonary artery and great pulmonary engorge ment was evident. The heart was enlarged to the right and to the left.

Catheterization was performed for the first time on March 17, 1953. At this time it was possible to pass the catheter into the aorta (fig. 1). The pressures were: aorta 100/55 mm. Hg, mean 72; pulmonary artery 100/65, mean 77; right ventricle 90/0, mean 45 mm. Hg. There were no differences between the pressure curves of the aorta and pulmonary artery. The blood samples showed a slight bidirectional shunt between aorta and pulmonary artery.

In considering the pathway taken by the catheter, it was evident that it did not enter the aorta through a patent ductus, because its position was too far median, and it was lying in the ascending aorta, the tip not reaching the site in the arch at which the ductus arteriosus enters the aorta.

There remained, therefore, two possible routes that the catheter may have followed: an aortic septal defect or a high ventricular septal defect. To secure more information we decided to perform a second catheterization, and to take as many blood samples as possible in the pulmonary artery and in the right ventricle. At the second catherizer-
Fig. 1. Case 1. Catheter is in the aorta, having reached it via right ventricle and pulmonary artery.

Fig. 2. Case 1. Catheter, via right ventricle and pulmonary artery, passed through an aortic septal defect into the ascending aorta toward the aortic valves.

Fig. 3. Case 1. Superposition of the two catheter positions in figures 1 and 2. The defect must be situated in the aortic septum at a certain distance above the aortic valves.

Fig. 4. Case 2. Catheter is in the aorta, having reached this vessel via right ventricle and pulmonary artery.

Fig. 5. Case 2. Pressure recording while withdrawing the catheter from aorta into pulmonary artery and pushing it into a “capillary.” Note, there is no ventricular pressure pattern between aorta and pulmonary artery pressures, and the two pressures are equal.

...tion (March 26, 1953), however, it was possible again to guide the catheter into the aorta, but on this occasion downward into the ascending aorta toward the aortic valves rather than upward into the ascending aorta toward the arch (fig. 2). Through these observations, a high ventricular septal defect was excluded, and when the films showing the two catheter positions were superposed, it could be seen that a large defect above the aortic valves must be present (fig. 3).

The child was operated upon on April 24, 1953 by Prof. Dr. A. G. Brom and Dr. A. L. E. Schaepkens van Rijpmst. A large defect with a diameter of 5 cm. between the large pulmonary artery and the aorta above the semilunar valves was found. The defect was partially closed by a large ligature. The postoperative course was uneventful.

Case 2. C. E., a 6 year old girl, was admitted to the hospital on March 29, 1954. Heart disease was observed immediately after birth by the physician. When she was three months old, she became cyanosed for the first time. At the time of admission, she tired very quickly, had dyspnea and cyanosis and squatted. She had also frequently bronchitis. On physical examination acrocyanosis and slight clubbing were observed. The blood pressure was 110/80 mm. Hg. A systolic murmur varying from...
heart, especially the right atrium and the left ventricle. There was pulmonary engorgement, but no hilar dance. The electrocardiogram showed right ventricular hypertrophy.

Heart catheterization was performed on April 4, 1954. The pressure in the pulmonary artery was 110/55, the mean being 85 mm. Hg. Then we were able to put the catheter into the carotid artery (fig. 4). While withdrawing the catheter from the aorta, we could push it into the pulmonary artery “capillary,” without obtaining a ventricular pressure recording between aorta and pulmonary artery (fig. 5).

The possibility of an aortic septal defect was then thought of and a second attempt was made to enter the aorta via the defect; and, by manipulating the catheter, to direct it into the ascending aorta toward the aortic valves. This was accomplished (figs. 6 and 7). Again, superposition of the two x-ray films (fig. 8) demonstrated with absolute certainty the presence of an aortic septal defect situated above the aortic valves.

Comment

On embryologic grounds, aortic septal defect lies close to truncus arteriosus and the difference is only a matter of gradation. Some authors have designated aortic septal defect as partial truncus. Clinically, aortic septal defect resembles patent ductus and, especially, patent ductus with pulmonary hypertension. It is also frequently mistaken for a high ventricular septal defect.

Diagnosis of Aortic Septal Defect

(1) Clinically, this condition is a possibility, and we must have this diagnosis in mind, therefore, in any case with a continuous or systolic and diastolic murmur and right ventricular hypertrophy in the electrocardiogram. On x-ray films, there is an enlarged heart and a very large pulmonary artery.

(2) In cardiac catheterization, the impossibility of catheterizing the aorta when a patent ductus is suspected, always suggests the possibility of an aortic septal defect. When a catheter can be passed into the aorta, the following possibilities exist: ventricular septal defect or overriding aorta, patent ductus, truncus arteriosus and transposition of the great vessels.

The difference between ventricular septal defect and aortic septal defect is very difficult
to find out by location of the catheter in the aorta only. However, it may be possible in aortic septal defect, while withdrawing the catheter out of the aorta, to see that the aortic pressure changes into the lower pressure of the pulmonary artery. When, in aortic septal defect, the pressures in aorta and pulmonary artery are equal, it may be possible to withdraw the catheter out of the aorta and push it immediately into the “capillaries” of the pulmonary artery, without obtaining a ventricular pressure pattern between the aorta and pulmonary artery pressures (fig. 5).

Patent ductus is easier to distinguish from aortic septal defect. When the catheter enters the aorta via a patent ductus, it nearly always goes into the descending aorta, without ascending in the arch. In the occasional instance when it starts toward the arch, almost always it will enter the left carotid artery and not the right. Through an aortic septal defect or an overriding aorta, the catheter follows the arch and in that way enters the descending part, or it ascends in any of the carotid or subclavian arteries.

Differentiation of aortic septal defect and truncus arteriosus is often very difficult. On determining the oxygen content of various blood samples, both anomalies can give exactly the same picture. Only the method described in this paper, by which catheters are made to take two directions after entering the aorta, can give a complete differentiation.

Transposition of the great vessels gives a totally different clinical picture since it is not possible to place a catheter in the pulmonary artery.

While determination of the oxygen content of blood samples, obtained from the pulmonary artery and right ventricle, may aid in differentiating these anomalies and, particularly, in excluding a ventricular septal defect (or an overriding aorta), the only absolutely sure method of differentiation is to pass the catheter into the aorta via the aortic septal defect and photograph it first, after it has been directed toward the arch and again, after it has been directed toward the aortic valves. X-ray films, thus obtained, will show clearly that the defect is situated in the ascending aorta and at a certain distance above the aortic valves (figs. 3 and 8).

(3) The oxygen content of blood of right brachial artery and femoral artery on effort can also give much information. This can be used in cases in which a shunt into the pulmonary artery and a pulmonary hypertension are present. On effort the blood of the femoral artery will be more desaturated than that of the brachial artery in the presence of a patent ductus with reversed shunt. However, when there is an aortic septal defect, the desaturation of the blood in the two arteries will be the same, since the shunt will lie proximal to the mouth of the innominate artery. Nearly the same information can be reached by obtaining simultaneous oxymetric readings from both ears, during effort, by means of a two-ear oxymeter.

(4) Retrograde aortography is also a good method for making a correct diagnosis of aortic septal defect. Since in our cases the diagnosis was confirmed by catheterization, aortography was not necessary, and we have had no experience with this method in the diagnosis of aortic septal defect.

**Summary**

Two cases of aortic septal defect are described. The correct diagnosis could be made only by catheterization of the heart. After the catheter was put into the aorta via the aortic septal defect, it was advanced first in a forward direction toward the aortic arch and carotid artery and then in a backward direction through the ascending aorta toward the aortic valves. By superposition of the x-ray films, showing the two catheter positions, a defect in the ascending aorta, at some distance above the aortic valves, was clearly demonstrated.

Various methods for making a correct diagnosis of aortic septal defect are discussed. It is pointed out that cardiac catheterization yields the most important diagnostic information. This consists mainly in the findings discussed in the preceding paragraph, but also in the oxygen content of blood samples obtained from different chambers and vessels. When great pulmonary hypertension exists, only the method described in the above paragraph insures a correct diagnosis.
SUMMARIO IN INTERLINGUA

Es describite duo casos de defecto aortico-septal. Le correcte diagnose esseva possibile solmente per catheterisation cardiac. Post que le catheter esseva inserite in le aorta via le defecto aortico-septal, illo esseva movite (1) in avante verso le arco aortic e le arteria carotide e (2) in retro a transverso le aorta ascendente in le direction del valvulas aortic. Per superimponer le pelliculas roentgenographic del duo positiones del catheter, illo esseva possimile de monstrare clarmente le presentia de un defecto in le aorta ascendente alique supra le valvulas aortic.

Es discutite varie methodos pro le correcte diagnose de defectos aortico-septal. Nos signalia que catheterisation cardiac provide le plus importante information diagnostic. Isto consiste principalmente in le supra-discutite constatationes sed etiam in datos in re le contento de oxygeno in specimens de sanguine obtenite ab varie camera e vasos. In casos de grande hypertension pulmonar, solmente le supra-describite metodo (vide paragrapho 1) assecura un correcte diagnose.

REFERENCES

Diagnosis of Congenital Aortic Septal Defects: Description of Two Cases and Special Emphasis on a New Method which Allows an Accurate Diagnosis by Means of Cardiac Catheterization

H. A. H. D'HEER and C. L. C. VAN NIEUWENHUIZEN

doi: 10.1161/01.CIR.13.1.58
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1956 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/13/1/58

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/