

THE DIAGNOSTIC VALUE OF CARDIAC CATHETERIZATION IN ISOLATED PULMONARY STENOSIS AND LARGE INTERVENTRICULAR SEPTAL DEFECTS

BY

E. MANNHEIMER, M.D.

(From the Crown Princess Louisa Children's Hospital, Stockholm, Sweden)

Heart catheterization is a valuable aid in the diagnosis of congenital cardiac malformations of different types. This has been well known since the method was first adopted for clinical use by A. Cournand and his team (Cournand and Ranges, 1941). Bing in Baltimore and Dexter in Boston have both contributed excellent papers on the subject (Bing et al., 1947; Dexter et al., 1947). The monograph 'Cardiac Catheterization in Congenital Heart Disease' by Cournand, Baldwin, and Himmelstein (1949) gives us the results of cardiac catheterization in seventeen cases of congenital malformations of the acyanotic type. Nevertheless blue babies have been so much discussed during recent years that it may be of interest to add something about our experience and methods of catheterization in congenital heart disease without cyanosis.

We carry out catheterization as team work. In the beginning we worked with Lagerlöf and Werkö at St. Eric's Hospital, but since we were able to set up our own unit our team has consisted of Larsson, Möller, Landtman, and myself. Catheters of the Cournand type Nos. 6-9 and an intravenous drip

infusion with 50 mg. of heparin in 1 litre of saline are used. Except in infants and in two or three older children we have not had recourse to general anaesthesia. We believe that in nearly all children

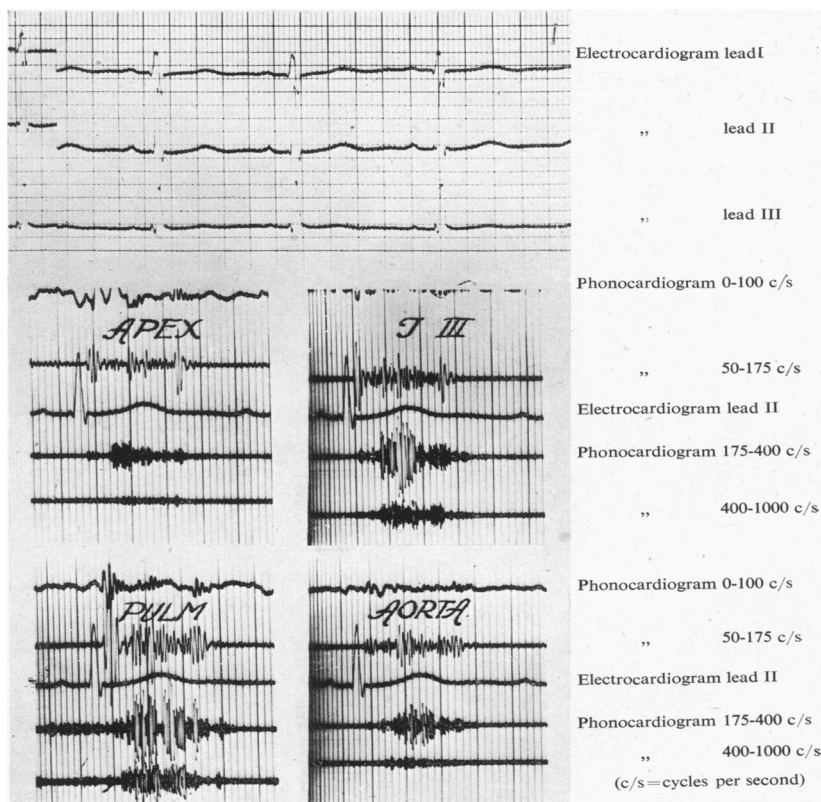


FIG. 1.—Electrocardiograms and phonocardiograms of a four-year-old boy, with isolated pulmonary stenosis.

there is less disturbance without anaesthesia provided the patients are handled in the right way. Blood samples are collected from as many places as the capacity of our laboratory will permit. The Tybjaerg-Hansen pressure apparatus containing a condenser-manometer and an amplifier is used in all cases. It is of great value to have good pressure equipment, and we consider that a water manometer which gives mean values only is not sufficient. Our recording apparatus consists of an Elmquist electrocardiograph permitting the simultaneous recording of six tracings, and an apparatus

Pulmonary Stenosis

The St. Eric's team and ours have seen seven cases of isolated pulmonary stenosis during the last two years. These seven cases differ from the earlier conception of this disease. No cyanosis is present, and the symptoms are so mild that affected persons can live quite a normal life; there is no retardation in growth and only slight limitation of activity. Physical examination reveals a loud, harsh systolic murmur over the pulmonary orifice (fig. 1) and x-ray examination shows a moderately bulging pulmonary arc. The arterial oxygen saturation is

normal (96 per cent. to 98 per cent.) and does not drop more than 4 per cent. to 5 per cent. during exercise, which is normal. In testing arterial oxygen oximeter recordings are helpful, as they make most of the van Slyke determinations superfluous.

It is obvious that in most of the cases the pressure in the right ventricle is pathologically increased (normal value 30 mm. Hg), and in some of the cases to a very high degree. In our opinion this increase in right ventricular pressure above the arterial blood pressure must indicate that no over-riding aorta is present. The highest pressure in the right ventricle was 170/0 and the lowest 58/9. The pressure in the pulmonary artery is always lower than that in the right ventricle, and this fact establishes the diagnosis of a pulmonary stenosis. Withdrawal tracings prove that this

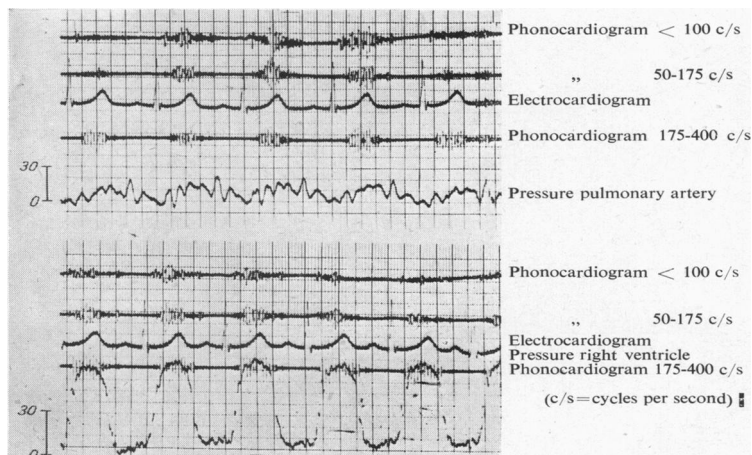


FIG. 2.—Pressure tracings obtained during cardiac catheterization in a case of isolated pulmonary stenosis.

for calibrated phonocardiography (Mannheimer-Stordal type). The oxygen consumption is determined by means of Douglas bags and the gas is analysed in a Haldane apparatus. We use arterial puncture (most often of the femoral artery) in nearly every case in order to obtain values for the arterial oxygen content, arterial oxygen saturation, and oxygen capacity of the blood. By means of these values and the Fick method approximate figures are calculated for the pulmonary and systemic flows and for shunts.

In order to limit myself to those groups of congenital cardiac malformations in which cardiac catheterization has shown new aspects, I have chosen two types, (1) isolated congenital pulmonary stenosis without over-riding aorta, and (2) large interventricular septal defects. Both these groups have some signs in common. First, a loud, harsh systolic murmur immediately leads to the diagnosis of congenital heart disease. Secondly, a more or less pronounced bulging of the pulmonary arc is present in both. In the third place, hypertension in the lesser circulation is a characteristic sign, even if it is not present in all cases.

stenosis is an infundibular one.

Fig. 2 gives pressure tracings from a boy with isolated pulmonary stenosis.

The blood gas analyses give on the whole no significant differences in oxygen contents. In all cases there is a slightly higher oxygen content in the pulmonary artery than in the right ventricle. A patent ductus arteriosus seems, however, rather unlikely as no continuous murmur is present. One case, a 25-year-old woman, was operated on by Crafoord many years ago. The exploratory thoracotomy showed a wide pulmonary artery but no patent ductus arteriosus. The high oxygen content in the pulmonary artery could be due to a small high interventricular septal defect or more probably to random variation.

All seven cases show more or less pronounced post-stenotic dilatation of the pulmonary artery. It is this dilatation that causes the bulging pulmonary arc and also gives rise to the suspicion that these are cases of patent ductus arteriosus. Previously we used to speak of idiopathic pulmonary dilatation. There is no doubt that many of the cases belonging to this group will prove to be cases

of isolated pulmonary stenosis with post-stenotic dilatation.

Interventricular Septal Defects

The new aspects of interventricular septal defects revealed by cardiac catheterization are shown in fig. 3. In all cases the clinical diagnosis was interauricular septal defect, or perhaps a Lutembacher syndrome (atrial septal defect and mitral stenosis), because these signs were observed: retardation in growth; acyanotic, pale, tiny children;

girl. She was a thin, under-developed child and was classed among the morbus coeruleus cases because of slight cyanosis after exercise. X-ray examination showed a picture which suggested an auricular septal defect or a Lutembacher syndrome. The very slight cyanosis and the marked physical under-development pointed the same way. Catheterization, however, revealed a huge ventricular septal defect, with an enormous left to right shunt of about 80 per cent. Thus this heart functioned as a cor triloculare with a common ventricle, the oxygen content of the right ventricle

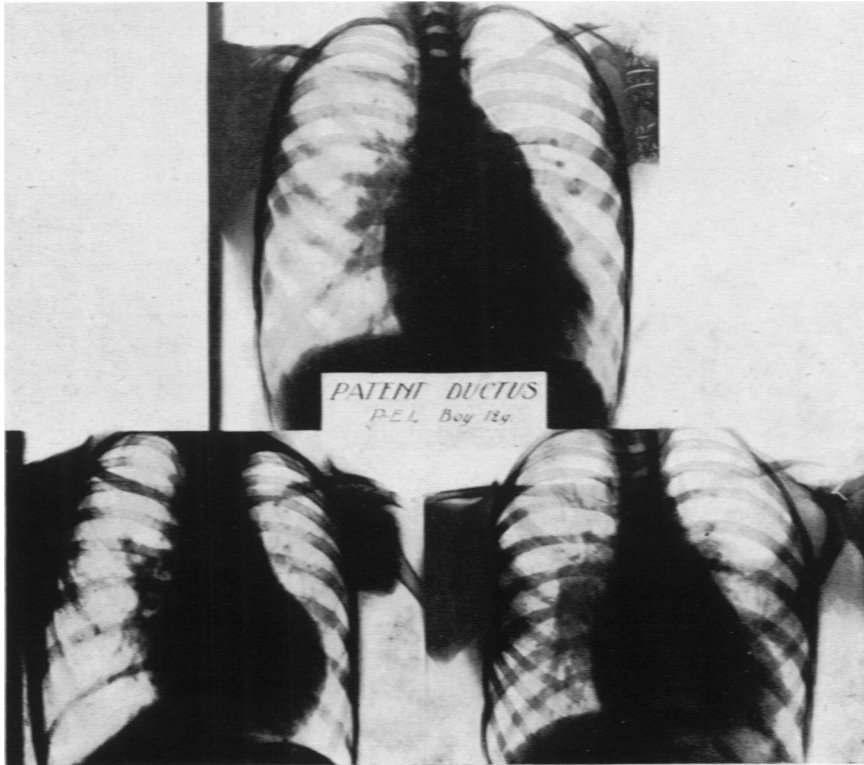


FIG. 3.—Radiographs of one case of patent ductus arteriosus and two of interventricular septal defects; all three showed evidence of increased pulmonary blood flow.

harsh systolic murmur; enlarged heart with marked bulging of the pulmonary arc; and a very pronounced increase in the pulmonary flow showing marked pulsations of the pulmonary artery and hilar dance. In all cases the right auricle was moderately enlarged. After cardiac catheterization was carried out one of these cases (P.E.L.) proved to be a patent ductus with a difference in oxygen content between the pulmonary artery and right ventricle of nearly 4 vol. per cent., thus indicating a left to right extracardiac shunt through the ductus of about 80 per cent.

The second case (R.A.) was that of a 10-year-old

and that of the femoral artery being almost equal. Because of the large left to right shunt, pressure in the right ventricle and pulmonary artery as well as the pulmonary flow were markedly increased. It was of interest that the murmur had its maximal intensity over the pulmonary orifice, although the septal defect was of ventricular origin. The case shows how difficult or even impossible it is to make the correct diagnosis in vivo without catheterization of cases with systolic murmurs and thrills along the left sternal border when there is little or no cyanosis.

Fig. 4 gives the results of cardiac catheterization in this case. Calculating the flows and the shunt

from the differences in oxygen content, the pulmonary flow is five times as large as the systemic flow. In other words, of the amount of blood coming from the lungs and reaching the left heart, 80 per cent. is shunted into the right ventricle and out into the pulmonary artery again. The pressure in the right ventricle and the pulmonary artery is markedly increased and probably about the same as the left ventricle. During systole both ventricles communicate and consequently the systolic pressure will be the same.

In this connexion the third case (B.K.) may be worth mentioning. The clinical picture was the same; the shunt was also very large (75 per cent.) but the pressure in the lesser circulation was normal (Fig. 5). This suggests that the interventricular septal defect did not function during systole. Otherwise the pressures in both ventricles would instantly have been balanced. It is possible that the defect either closed during the systole, perhaps by muscular contraction, or remained anatomically open but no shunt took place (Eek, personal communication).

Summary

Isolated pulmonary stenosis is not a very rare type of congenital heart disease. Patients with this anomaly can live a normal life and the prognosis seems to be remarkably good. The diagnosis is made entirely by cardiac catheterization. Idiopathic pulmonary dilatation as previously described may actually be isolated pulmonary stenosis with post-stenotic dilatation.

Patent ductus arteriosus with enlarged heart and a large shunt, interauricular septal defects, and large interventricular septal defects often cannot be

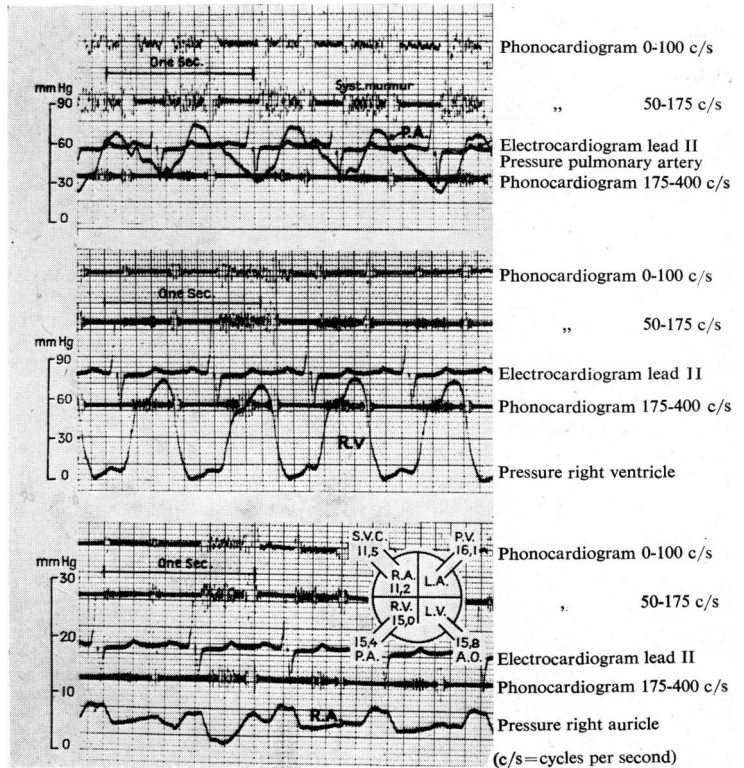


FIG. 4.—Pressure tracing obtained during cardiac catheterization in a case of ventricular septal defect.

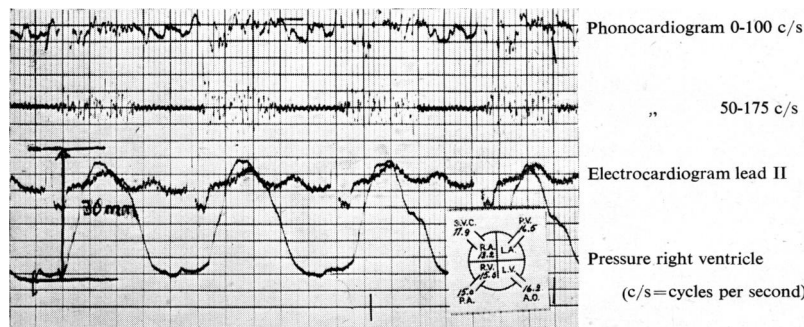


FIG. 5.—Pressure tracing obtained during cardiac catheterization in a case of ventricular septal defect in which probably there was no shunt during the systole.

differentiated clinically. Cardiac catheterization gives the diagnosis in these cases. In many cases with septal defects between the ventricles there is equal pressure in both ventricles. In these the septal defect is undoubtedly open during the systoles. In some other cases, also with large interventricular shunts, the pressure in the right ventricle is normal and much lower than that in the left ventricle. In these cases it is suggested that the defect is anatomically or functionally closed during the systole.

This study would seem to indicate that the classical textbook description of inter-auricular septal defect is not

entirely satisfactory. Such a diagnosis should not be made clinically without a thorough examination, including cardiac catheterization.

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