CARDIAC CATHETERIZATION IN THE DIAGNOSIS OF CONGENITAL HEART DISEASE*

LEWIS DEXTER

Assistant Professor of Medicine, Harvard Medical School and Senior Associate in Medicine, Peter Bent Brigham Hospital, Boston

UNTIL recently, congenital heart disease has been largely of academic interest to physicians. Recent developments in cardiac surgery have followed closely upon improvements in thoracic surgery and anesthesia. As new operations on or around the heart have been devised during the last decade, the onus of a correct pre-operative diagnosis has fallen onto the shoulders of the internist. Before the advent of angiocardiography and cardiac catheterization. a correct diagnosis of many congenital heart lesions had to await autopsy examination. Inability to be sure of the correct diagnosis during life made for slow progress in our knowledge of these disorders. Increasing familiarity with the various congenital heart malformations correctly diagnosed during life now makes it possible to study a given abnormality in a detailed manner so that by the usual approach of history, physical examination, fluoroscopy of the heart, and electrocardiogram, a correct diagnosis can usually be made without resorting to more complicated procedures.

In acquired heart disease, it is usually stated that 60 per cent of all information to be obtained is derived from a careful history, 25 per cent from physical examination and 15 per cent from laboratory procedures (x-ray and electrocardiogram). In congenital heart disease, just the opposite is true. About 15 per cent of the information comes from history, 25 per cent from physical examination, and 60 per cent from laboratory procedures. The history may date from birth or childhood, tells us the extent of physical incapacity, may tell us if the patient has been cyanotic. A history of cyanosis may be quite misleading, however, because after one or two physicians have made inquiries on this point,

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the answer becomes almost uniformly positive, whether or not cyanosis really exists. Physical examination reveals the presence of a murmur which has obviously not been acquired and must therefore be congenital, but unlike many acquired murmurs is not diagnostic except in the case of patent ductus arteriosus. Fluoroscopy of the heart reveals abnormalities of contour and the size of some of the chambers. I should like to emphasize, however, with all apologies to any radiologists who may be present, that in our experience x-ray and fluoroscopy are not reliable in evaluating the size of the right auricle, *are* quite reliable as regards the left auricle, and are good in recognizing ventricular enlargement but are not always correct in identifying which ventricle is enlarged. We have found the electrocardiogram to be the more reliable in identifying which ventricle is hypertrophied. Further information to be derived from the x-ray is the size of the pulmonary artery and the prominence of the pulmonary vascular markings at the hilar regions of the lung. These markings are regularly increased when pressure in or flow of blood through the pulmonary artery is increased and, with one exception to be discussed, are of normal or decreased prominence when pressure and flow are normal or decreased. If, with evaluation by these methods, the diagnosis is still uncertain, angiocardiography, which will be discussed by Dr. Morgan, and cardiac catheterization[1] will usually reveal the important lesions present.

Before one can make a correct morphological diagnosis, two principles must be recognized. The physician must first have some idea of the differential diagnostic possibilities and secondly, in the case of congenital heart disease, some concept of the physiological disturbances created by the lesion, since the abnormal flows and pressures produce changes in the contour and size of the heart and pulmonary vessels from which logical deductions concerning the underlying lesions can usually be made.

Under the age of, say, five, congenital heart disease is extremely complicated. It is in this age group that the greatest challenge exists. During this period, the serious lesions and the multiple lesions are lethal. The simpler lesions compatible with life are prone *not* to have distinctive patterns. Above the age of about five, most of the patients with seriously malformed hearts have expired, leaving a relatively few major types with which we, as physicians, are confronted. The majority fall into the following groups: simple pulmonic stenosis, tetralogy of

Fallot, Eisenmenger's complex, atrial septal defect, ventricular septal defect, and patent ductus arteriosus. Less frequently seen are tricuspid atresia, anomalous pulmonary veins emptying into the right atrium, levoposition of the pulmonary artery, transposition of the great vessels, aortic septal defect, and a variety of other lesions. Although coarctation of the aorta is fairly common, it will not be discussed here because cardiac catheterization does not serve any useful diagnostic purpose.

In the discussion that follows, emphasis will be placed on the physiological changes that occur and their influence on the x-ray and electrocardiogram. Only by this approach have we been able to improve our clinical recognition of the various disorders and thus to abandon the use of the cardiac catheter in many cases. It is now reserved for those cases which are not distinctive.

Technique of cardiac catheterization: The cardiac catheter is a modified ureteral catheter with a curved tip. It is sufficiently stiff so that rotation of one end turns the curved tip at the other end. Due to its radiopacity, it may be guided to the desired position under fluoroscopic observation. The median basilic vein is the usual site of introduction. In infants, the jugular or the femoral vein has been used. In the older age group, novocaine is used for anesthesia. In the younger age group, a general anesthetic must be employed. The vein is exposed and the catheter introduced. It is guided fluoroscopically into the subclavian vein, superior vena cava, right auricle, right ventricle, and pulmonary artery. If it is allowed to obstruct the pulmonary artery, it is in direct connection with the pulmonary capillary bed so that blood samples so obtained are arterial instead of venous and the pressure so measured is within a millimeter or so of the true pulmonary capillary pressure.[2] Pressures can be recorded through the catheter and blood samples withdrawn and analyzed for their content of oxygen. In congenital heart disease, left-to-right shunts through defects in the cardiac septa will be characterized by the appearance of arterial (highly oxygenated) blood in the corresponding chamber in the right side of the heart.

Normal variations: Figure 1 shows characteristic pressure tracings in a normal patient. It will be observed that except that the pressure in the pulmonary artery is quite low, the contour is the same as in a systemic artery with a systolic rise, a dicrotic notch due to closure of the pulmonary valve, and a fall to the diastolic level just before the next ventricular systole. The systolic pressure in the right ventricle is iden-

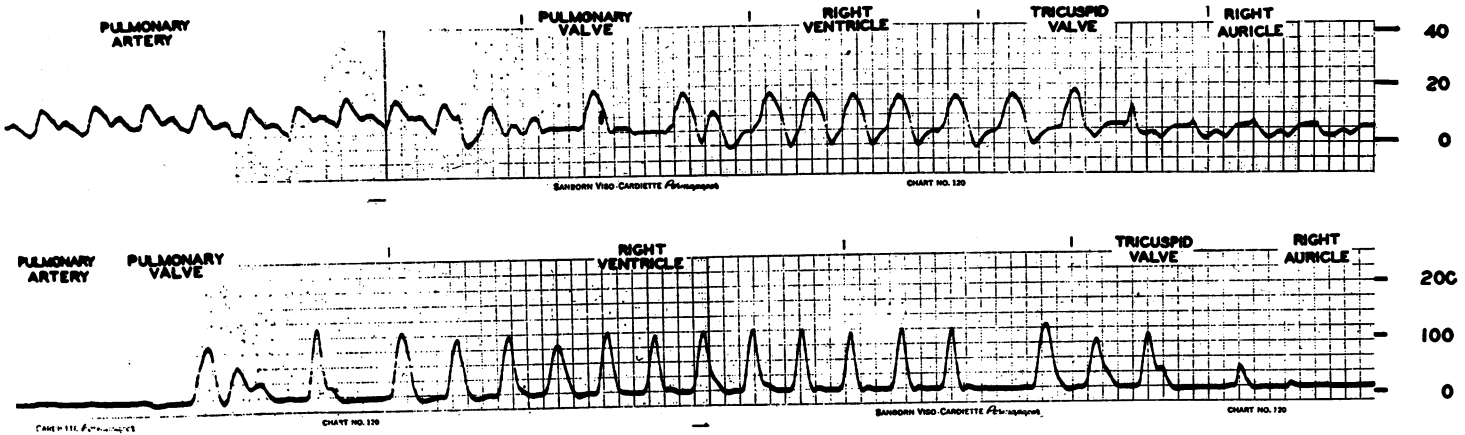

Figure 1—Pressure tracings obtained through a catheter during continuous withdrawal of the catheter from pulmonary artery through right ventricle and into right auricle.

Upper curve: Normal patient. Note that the systolic pressure in the right ventricle is the same as that in the pulmonary artery.

Lower curve: Patient with pulmonic stenosis. Note that the systolic pressure in the right ventricle is higher than that in the pulmonary artery.

tical with that in the pulmonary artery. The diastolic pressure in the ventricle is the same as the pressure at the same time in the right auricle.

Blood samples withdrawn from the different chambers in the lesser circulation normally vary somewhat in their oxygen content. Blood entering the right auricle comes from the superior vena cava, inferior vena cava, and coronary sinus. The amount of oxygen in these different venous bloods varies greatly so that a considerable variation in oxygen content is encountered in different parts of the right auricle. Less variation occurs in the right ventricle and by the time blood has reached the pulmonary artery, the oxygen content is remarkably uniform. Variations in the oxygen content in the various chambers have been described in detail.[3] Because of these normal variations, two or more samples of blood must be withdrawn from each chamber in order to recognize with any degree of certainty an abnormality when it exists.

Pulmonic Stenosis: The commonest example of pulmonic stenosis is the tetralogy of Fallot. Simple pulmonic stenosis without other accompanying defect has always been considered a rarity. It has turned out, however, to be one of the common congenital cardiac lesions[4,5,6] and has usually been diagnosed clinically as ventricular septal defect.

A. *Simple pulmonic stenosis* is characterized clinically by the absence of cyanosis, a systolic murmur of varying intensity in the 2nd, 3rd, or 4th left intercostal space, and right ventricular hypertrophy or right bundle branch block by electrocardiogram. X-ray and fluoroscopy of the heart may or may not show right ventricular enlargement, but in almost all cases there is *prominence of the pulmonary artery accompanied by normal or decreased pulmonary vascular markings* because pressures and blood flow through the lung are normal or reduced. The cause of the enlarged pulmonary artery has been discussed for over one hundred years and no reasonable explanation has yet been given. The combination of the murmur, electrocardiographic and x-ray findings are practically pathognomonic. By cardiac catheterization the diagnosis is made by pressure recording alone. Instead of finding identical systolic pressures in the pulmonary artery and right ventricle, this pressure is higher in the right ventricle than in the pulmonary artery, indicating an obstruction between the two, i.e., pulmonic stenosis (Figure 1).

B. *The tetralogy of Fallot* consists of pulmonic stenosis accompanied by a ventricular septal defect, an aorta which straddles both ventricles (dextroposition of the aorta) and right ventricular hypertrophy. Blood

enters the right auricle and goes to the right ventricle. Due to the obstruction to outflow through the pulmonary valve, there is a diminished blood flow into the pulmonary artery and lung, and a shunting of blood into the systemic circuit producing cyanosis. Since the right ventricle is in direct connection with the aorta, the systolic pressures in the right ventricle and in the aorta are identical. The path taken by the cardiac catheter may be from right ventricle through the stenotic pulmonary valve and into the pulmonary artery. Under these circumstances, pulmonic stenosis will be recognized by the higher systolic pressure in the right ventricle than in the pulmonary artery, dextroposition by identical systolic pressures in the right ventricle and brachial artery, and a right to left shunt by arterial oxygen unsaturation. In some cases, the catheter cannot be introduced into the pulmonary artery. Instead, it passes from the right ventricle directly into the aorta. Under these circumstances, dextroposition of the aorta is recognized by finding identical systolic pressures in the aorta and right ventricle, a right-to-left shunt by arterial oxygen unsaturation, but pulmonic stenosis cannot be demonstrated. One must depend on the appearance of the pulmonary artery and pulmonary vascular markings on the x-ray film. In the tetralogy of Fallot, the pulmonary artery and hilar vascular markings are classically diminished whereas if pulmonic stenosis is not present (Eisenmenger's complex) these are both greatly accentuated. The salient findings in a patient in whom the cardiac catheter was introduced into the pulmonary artery as well as into the aorta were as follows:

Source of Blood	*Pressures mm.Hg*	*Oxygen Content cc/l*
Aorta	105/73	204 (69 per cent)
Pulmonary Artery	7	150
Right Ventricle	105/7	—

Eisenmenger's complex: This condition has the same anatomical defects as the tetralogy of Fallot with the exception that pulmonic stenosis is absent. The septal defect varies in size and as it approaches complete absence, this condition merges imperceptibly with the anatomical condition known as cor triloculare biatriatum, i.e.. two atria and a common ventricle. Although usually the two ventricles are anatomically partially separated by a septum, physiologically they are

common chambers with identical pressures because of the over-riding aorta. Cyanosis develops late, there is a pulmonic systolic murmur, the electrocardiogram shows right ventricular hypertrophy, and the x-ray reveals an enlarged right ventricle, large pulmonary artery, and prominent pulmonary vascular markings. The systolic pressures in the aorta (or brachial artery) and right ventricle are identical (because of aortic dextroposition) as are those in the right ventricle and pulmonary artery (because there is no pulmonic stenosis). Actually there is a "stenosis" in the arterioles of the lung. The pulmonary "capillary" pressure is normal, the pulmonary arterial pressure is aortic in its magnitude, and the pulmonary arteriolar resistance approximates that in the systemic circuit, i.e., it may be 15 to 40 times the normal pulmonary resistance. This high resistance produces an effective "stenosis" as in the tetralogy of Fallot, but it is located in the pulmonary arterioles instead of at the pulmonary valve. The net result is minimal shunting of blood either from left-to-right or right-to-left. The late right-to-left shunt that occurs in these individuals is probably due to a pathological increase in the pulmonary resistance which eventually becomes higher than that in the systemic circuit. Curiously enough, history, physical examination, electrocardiogram, x-ray, and fluoroscopy cannot usually differentiate with any degree of certainty this condition from atrial septal defect. Venous catheterization is diagnostic in Eisenmenger's complex by finding identical systolic pressures in the brachial artery, right ventricle, and pulmonary artery. The following is an example:

Site	*Pressure mm.Hg*
Pulmonary Artery	117/56
Brachial Artery	116/72
Right Ventricle	117/1
Pulmonary "Capillary"	12

Atrial septal defect: Defects may occur in the septum separating the two atria. Whether this defect is a patent foramen ovale or persistent ostium primum or secundum is not revealed by the venous catheter. In any event, there is a large shunt of blood from left atrium to right atrium and from here blood passes through the right ventricle, pulmonary artery, lung, and back again to the left atrium. These indi-

viduals have the largest shunts of blood that we have encountered. Late in the course of the disorder, changes occur in the pulmonary vasculature so that resistance to flow through the arterioles of the lungs becomes high and pressure in the pulmonary artery becomes greatly elevated. The right ventricle, the pulmonary artery, and the pulmonary vascular markings attain great prominence by x-ray. These findings coupled with a similar history, physical examination, and electrocardiogram make its differentiation from Eisenmenger's complex difficult. When the cardiac catheter is introduced into the right auricle, it may sometimes be introduced through the defect into the left atrium and even out into a pulmonary vein. The latter experience makes it difficult to differentiate atrial septal defect from an anomalous pulmonary vein emptying into the right auricle. Arterialized (highly oxygenated) blood is found in the right auricle as compared with the superior vena cava as shown by the following example:

Source of Blood	*Oxygen Content cc/liter*
Superior Vena Cava	159
Right Auricle	203
Right Ventricle	209
Pulmonary Artery	209

Ventricular Septal Defect (Roger's disease): Pathological reports have usually shown ventricular septal defect to be much less common than the other congenital abnormalities under discussion. This has likewise been our own experience. These individuals are not cyanotic and they have a pulmonic systolic murmur which is not distinguishable from that of pulmonic stenosis, atrial septal defect, and Eisenmenger's complex. Both ventricles are called upon to perform increased work so that by x-ray there is prone to be a non-specific cardiac enlargement, and the electrocardiogram shows little or no preponderance of either ventricle. Another condition which may produce similar findings and lead to an erroneous diagnosis is pulmonic stenosis with an associated aortic septal defect, i.e., a communication between the aorta and pulmonary artery just beyond the sites of the aortic and pulmonary valves. Cardiac catheterization reveals a significantly higher oxygen content of blood in the right ventricle than in the right atrium:

Source of Blood	*Oxygen Content cc/liter*
Right Atrium	125
Right Ventricle	156
Pulmonary Artery	156

Patent Ductus Arteriosus: Little need be said concerning this disorder, since it is assumed that all are familiar with the typical machinery murmur. On cardiac catheterization, oxygenated blood is found in the pulmonary artery as compared with values found in the right ventricle:

Source of Blood	*Oxygen Content cc/liter*
Pulmonary Artery	137
Right Ventricle	103
Right Auricle	102

Summary and Conclusions

In conclusion, cardiac catheterization in the diagnosis of congenital heart disease might at present be evaluated as follows:

1. It is diagnostic in the tetralogy of Fallot, Eisenmenger's complex, and uncomplicated pulmonic stenosis.

2. Although patent ductus arteriosus, ventricular septal defect, and atrial septal defect may usually be detected with ease by cardiac catheterization, small defects are easily overlooked. Much depends on the skill of the catheter-passer in inserting the catheter through the defect or in placing the tip of the catheter in the stream of arterial blood that is being shunted from left to right.

3. Cardiac catheterization gives confusing or inadequate data for interpretation when there are multiple defects such as in the case of the coexistence of atrial and ventricular septal defects together with a patent ductus arteriosus. Another example is that of tricuspid atresia with its accompanying atrial septal defect and ventricular septal defect or patent ductus arteriosus. In this case, the only abnormality revealed is atrial septal defect.

4. Small defects in the septa are easily missed. The size and precise location of a defect in a given septum, which is of crying interest to

the cardiac surgeon, is not revealed except in a very general way by cardiac catheterization. It is not possible to detect aberrant coronary arteries, bicuspid pulmonic valves, or anatomical abnormalities confined to the left side of the heart.

5. It should be remembered that this is a physiological method from which certain structural abnormalities can justifiably be deduced, just as angiocardiography, which Dr. Morgan is to discuss, is essentially an anatomical method from which many physiological abnormalities may be deduced. There are limits beyond which interpretation of the findings by cardiac catheterization is not justified. In order to arrive at a correct diagnosis, interpretation should be coupled with all of the other clinical and laboratory data that are available.

REFERENCES

1. Cournand, A. and Ranges, H. A. Catheterization of the right auricle in man, *Proc. Soc. Exper. Biol. & Med.*, 1941, *46*:462.
2. Hellems, H. K., Haynes, F. W. and Dexter, L. Pulmonary capillary pressure in man, *J. Applied Physiol.*, 1949, *2*:24.
3. Dexter, L., Haynes, F. W., Burwell, C. S., Eppinger, E. C., Sagerson, R. P. and Evans, J. M. Studies of congenital heart disease; pressure and oxygen content of blood in the right auricle, right ventricle, and pulmonary artery in control patients, with observations on the oxygen saturation and source of pulmonary "capillary" blood, *J. Clin. Investigation*, 1947, *26*:554.
4. Greene, D.G., Baldwin, E. deF., Baldwin, J. S., Himmelstein, A., Roh, C. E. and Cournand, A. Pure congenital pulmonary stenosis and idiopathic congenital dilatation of the pulmonary artery, *Am. J. Med.*, 1949, *6*:24.
5. Pollack, A. A., Taylor, B. E., Odel, H. M. and Burchell, H. B. Pulmonary stenosis without septal defect, *Proc. Staff Meet., Mayo Clin.*, 1948, *23*:516.
6. Dow, J. W., Levine, H. D., Elkin, M., Haynes, F. W., Hellems, H. K., Whittenberger, J. W., Ferris, B. G., Goodale, W. T., Harvey, W. P., Eppinger, E. C. and Dexter, L. Studies of congenital heart disease; uncomplicated pulmonic stenosis, *Circulation, in press.*