Cardiac Catheterization
in Congenital Heart Diseases*

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Cardiac catheterization was done for the first time 25 years ago by Forssman, who performed the maneuver on himself. However, this procedure was employed in the study and diagnosis of heart disease only after Courand and Ranges demonstrated the value and safety of the method in 1941. The method was subsequently completed by the addition of several collateral techniques. The most useful application of cardiac catheterization is in the diagnosis and study of congenital heart diseases.

Method and Technique

The patient should be admitted to the hospital one day before cardiac catheterization. If any evidence of infection or heart failure is found or if there are arrhythmias, the test is postponed until recovery. The patient is again kept under observation in the hospital for 24 hours following the test. Antibiotics are given before and after the procedure. Except for this, no other premedication is required for adults. Anesthesia or sedatives are sometimes needed in children, even though it is common experience that an intelligent and gentle nurse can obtain more cooperation than a large dose of sedatives.

A 100 cm. long radio-opaque catheter is introduced through the median basilic vein of the left or right arm (saphenous vein in children under 5 years of age) which had been previously exposing it by a small incision through the skin, under local anesthesia. Then it can be advanced gently under fluoroscopic control, through the innominate vein, superior vena cava, right atrium, right ventricle, and pulmonary artery. The catheter can subsequently be advanced within the pulmonary artery until a small branch is blocked. Thus, pressures from the venous side of the pulmonary circulation can be measured (so called pulmonary capillary pressure.) Then, while the catheter is pulled back, pressure readings and blood samples are taken in rapid succession from the right, left and main pulmonary artery, from the right ventricle and right atrium, from the inferior and superior vena cava and from any unusual structure which may be entered. Simultaneously with blood samples from the catheter, arterial blood samples from an indwelling needle introduced into the brachial or femoral artery, may be obtained during the procedure.

Measurement of oxygen consumption while blood samples are simultaneously obtained from the pulmonary artery and a systemic artery, makes possible the determination of cardiac output per minute through

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the use of Fick's principle. The following formula is then employed.  

$$1. \text{ Cardiac output (ml. per min.)} = \frac{O^2 \text{ intake (ml. per min.)}}{O^2 \text{ content of arterial blood (vol. per cent)} - O^2 \text{ content of mixed venous blood (vol. per cent)}} \times 100$$

The same principle has been widely applied for the calculation of left-to-right shunts (septal defects or patent ductus arteriosus). It must be pointed out that there are many causes of errors in the measure of this shunt, so that the data sometimes differ from the results of autopsy or surgery.

Three different kinds of data are obtained during the procedure by observing:

(a) position of catheter;
(b) pressure tracings; and
(c) oxygen content of blood samples.

The position of the catheter cannot be determined with certainty by fluoroscopy alone. In particular, it is impossible to determine on which side of a valve the catheter tip lies. As each cardiac chamber or vessel has characteristic pressure patterns, pressure tracings are helpful in order to ascertain the location of the tip of the catheter, whenever a blood sample is obtained.

An abnormal position of the catheter, when its location is precisely ascertained, is the most important information which can be obtained by cardiac catheterization; the catheter may be passed through or into a patent ductus arteriosus, an aortic septal defect, a ventricular septal defect, an overriding or transposed aorta, an atrial septal defect, an anomalous pulmonary vein or a left superior vena cava.

Cardiovascular pressures may be measured with reasonable accuracy by means of a strain gage, an electromanometer, or an optical manometer connected with the catheter. However, the motion of the catheter by the heart beat may cause a severe distortion of the pressure pulses, thus somewhat invalidating an otherwise promising aspect of this study.

The following figures of pressure can be considered normal from a survey of the literature:

<table>
<thead>
<tr>
<th>Pressure Source</th>
<th>Systolic (mm. Hg.)</th>
<th>Diastolic (mm. Hg.)</th>
<th>Mean (mm. Hg.)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right atrium</td>
<td></td>
<td></td>
<td>from -2 to +5</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>from 17 to 31.5</td>
<td>from -0.5 to +7 (end of diastole)</td>
<td></td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>from 11 to 29</td>
<td>from 4 to 13</td>
<td>from 8 to 19</td>
</tr>
<tr>
<td>Pulmonary</td>
<td></td>
<td></td>
<td>from 5 to 13</td>
</tr>
</tbody>
</table>

No significant gradient of systolic pressure exists normally between right ventricle and pulmonary artery.* On the other hand, the systolic pressure of the right atrium is about the same as the right ventricular

*It has been stated that a dilatation of the pulmonary artery may cause a moderate drop of the systolic pressure creating a gradient of 15 to 20 mm. Hg.
diastolic pressure. It is called, therefore, right atrial filling pressure.

Each organ consumes a different amount of oxygen. The blood which comes from the kidney, for instance, is rich in oxygen. The blood from the liver is poor in oxygen while that from the coronary sinus is the poorest of all. The venous blood of the right atrium is therefore a mixture of various venous bloods having different oxygen saturations. When the blood coming from a collateral vein enters the inferior vena cava, it does not mix well and has a tendency to form laminar flow. This may persist through the atrium and even within the right ventricle, so that the pulmonary arterial blood is often the only truly mixed blood. On account of this, the oxygen content of blood samples varies from chamber to chamber and even within the same chamber. Variations as great as 2.3° have been found in a couple of samples taken from the right atrium without moving the catheter.

The following differences in oxygen content are generally accepted as representing the maximum limits of normal. Superior vena cava and inferior vena cava usually show a difference of from 0 to 2 vol. per cent. Therefore, a gradient of at least 2 volumes per cent between the blood of the vena cava and that of the right atrium is necessary in order to determine the existence of a left-to-right shunt in the right atrium. However, if the samples of the two cavae are approximately the same and there is a difference in oxygen content between them and the right atrium of 1.5, this may be considered as indicative of left-to-right shunt, especially if there is clinical evidence supporting this diagnosis. Similarly, a difference greater than 0.9 vol. per cent between right ventricle and right atrium indicates either a left-to-right shunt between the ventricles or regurgitation of highly oxygenated blood from the pulmonary artery. On the other hand, the existence of a step-up in oxygen content in the pulmonary artery should not be considered as definite proof of the presence of patent ductus arteriosus because oxygenated blood from the left ventricle may be shunted directly across the defect and the pulmonary valve to the pulmonary artery in cases of high ventricular septal defect.

Failure to catheterize the pulmonary artery cannot be considered as evidence of pulmonic stenosis or atresia; unsuccessful attempts to pass the catheter through a clinically suspected septal defect or ductus arteriosus does not rule out its presence.

The pressures in the left side of the heart are normally higher than in the right side. Whenever there is an abnormal communication between the two sides, a left-to-right shunt occurs. Later on, pulmonary hypertension may develop. It usually causes an increase of pressure in the right ventricle and right atrium which may overcome the physiological difference and reverse the shunt causing cyanosis. If there is pulmonic stenosis associated with a septal defect, the pressure in the right heart is high since birth, and cyanosis may be present shortly afterwards. The same is true when the aorta is dextroposed.
Clinical Conditions Requiring Catheterization

Partial anomalous venous connection.—This malformation consists of the draining of one or more pulmonary veins into the right side of the heart. The most common sites are the superior vena cava, the right atrium, and the left innominate vein. When the pulmonary veins are draining into the superior vena cava or innominate vein, diagnosis through catheterization can be made either by entering the pulmonary vein from the systemic vein or by finding a high oxygen content of the blood samples taken in this vessel in comparison with those of the inferior vena cava. However, when the anomalous pulmonary vein drains into the right atrium, it is impossible to differentiate this condition from an atrial septal defect: both conditions show a step-up in oxygen content in the right atrium in comparison with that taken in the superior and inferior vena cavae. Even when the pulmonary vein is catheterized, this is not a definite proof that it is draining into the right atrium because it is impossible to tell whether the catheter passed from the right atrium into the anomalous pulmonary vein or through an atrial septal defect and the left atrium into the pulmonary vein.

Dye curves obtained by injecting Evans Blue T-1824 into the right pulmonary artery do not help either because it has been proved that there is a preferential drainage of the right lung into the right atrium in the majority of cases of atrial septal defect. Moreover, anomalous pulmonary veins and atrial septal defect may be associated creating an even greater diagnostic problem.

Atrial septal defect.—Atrial septal defect is one of the most common congenital heart anomalies and an accurate diagnosis becomes of the greatest importance because of its possible surgical closure. Usually the pressure of the left atrium exceeds that of the right by at least one or two mm. Hg and the blood flows from left to right. Since the left atrial blood is fully saturated with oxygen, there is no cyanosis. The amount of blood shunted across the defect depends upon the size of the artery. This blood flows through the right atrium, right ventricle, pulmonary artery, pulmonary veins and then returns to the right atrium again. This causes some dilatation of the right atrium, right ventricle, and pulmonary arteries. Although an atrial septal defect can be diagnosed clinically with relative accuracy, its definite diagnosis can be established only by catheterization. Cardiac catheterization shows an increased saturation of the blood of the right atrium in comparison with that of the inferior and superior venae cavae. The catheter may be passed through the septal defect. However, this is not a definite proof of a clinically significant defect because passing of the catheter may be accomplished also through a foramen ovale which is patent only from right to left being closed by a valve. This exists in about 25 per cent of normal cases.

Catheterization studies are also of value in the evaluation of the pressure in the right ventricle and pulmonary artery since some patients develop early pulmonary hypertension and such finding may affect the decision regarding surgical closure of the atrial septal defect.
Increase in oxygen content of the right atrium may also be due to:
—rupture of an aortic aneurysm into the right atrium;
—aorta-cava fistula;
—ventricular septal defect plus tricuspid insufficiency;
—A.V. communis with shunt from left ventricle to right atrium; or
—atrial defect (septum primum) with mitral insufficiency.

**Ventricular septal defect.**—Ventricular septal defect is a less benign malformation than previously thought. Many patients die in early infancy\(^1\) due to heart failure. The defect is usually located in the basal portion of the septum (pars membranacea). Defects of the muscular septum are less common and are considered more benign; it is possible that the defect is reduced in size during ventricular systole,\(^1\) thus causing a smaller shunt. Pulmonary hypertension occurs more frequently when the defect involves the pars membranacea. The difference in pressure between the left and right ventricles causes a shunt from left to right. The amount of blood passing through the defect depends not only upon the size of the defect but also upon the resistance offered by the pulmonary circulation; blood shunted across the defect increases the pulmonic flow and the return to the left heart. Therefore, evidence of enlargement of the left ventricle, right ventricle, pulmonary arteries and left atrium is encountered. The murmur caused by this malformation is usually harsh and loud and is mostly located in the third left interspace. However, a similar type of murmur is also found in atrial septal defects and even in cases with atypical ductus.

Right heart catheterization may furnish definite data in the majority of cases. A step-up in oxygen content of more than 1 volume per cent in the right ventricle is usually found in cases of ventricular septal defect. The association of pulmonary hypertension is more frequent with ventricular than with atrial septal defect. However, it should be noted that an increase of right ventricular oxygen content may also be due to: patent ductus with pulmonic regurgitation; rupture of aortic aneurysm into the right ventricle; or to a streamlined jet of a left-to-right shunt through an atrial septal defect (missed during sampling in the right atrium).

**Patent ductus arteriosus.**—The diagnosis of patent ductus arteriosus presents no problem when a continuous murmur can be heard over the pulmonic area. The absence of a diastolic component during the first few years of life has been admitted by several authors and has been attributed to small gradient of pressure during diastole between aorta and pulmonary artery.\(^1\) Occasionally, cardiac enlargement and heart failure may occur early in life and a prompt diagnosis may be life saving. Cardiac catheterization may establish a definite diagnosis if the catheter is passed through the ductus into the aorta. On the other hand, failure to catheterize the ductus does not rule out its presence. Arterialization of the blood of the pulmonary artery cannot be considered as a definite proof of patency of the ductus because arterialized blood of the left ventricle may be directly shunted across a ventricle septal defect into the pulmonary artery without admixture of that of the right ventricle. In such cases aortography
is necessary for the differential diagnosis.

*Pure pulmonic stenosis.*—Pulmonic stenosis without ventricular or atrial septal defect is a relatively common congenital malformation. The stenosis may be valvular, infundibular, or both. The valvular stenosis is caused by a fusion of the cusps which form a diaphragm-like or conical obstruction with a small central orifice of 2 to 4 mm. in diameter, offering great resistance to the flow of blood from the right ventricle to the pulmonary artery. As a result, the right ventricular work increases tremendously and the pulmonary flow is reduced; pressure increases in the right ventricle and decreases in the pulmonary artery, thus creating a “gradient.” The pressure of the right atrium is also elevated, even without right ventricular failure. If the foramen ovale is closed, no shunt is possible. Therefore, if there is cyanosis, it is a “peripheral” cyanosis resulting from slow circulation and low cardiac output. Arterial saturation is normal.

A definite diagnosis can be made clinically in the typical cases. However, catheterization is useful, not only for evaluation of the abnormal physiology pre- and post-operatively but also as a way to ascertain the type of stenosis. This differentiation is important since the type of operation depends on the type of obstruction. When there is an infundibular stenosis, a zone of intermediate pressure may be registered while the catheter is withdrawn through the outflow tract of the right ventricle. On the other hand, failure to detect an “intermediate zone of pressure” does not necessarily exclude the existence of infundibular stenosis.

There is a group of patients who clinically show almost no disability, a systolic murmur and split second pulmonic sound, slight evidence of hypertrophy or right bundle branch block, and normal vascularization of the lung fields upon fluoroscopy. The differential diagnosis in this group can be made only by catheterization. Considering that a small “gradient” of pressure between the right ventricle and pulmonary artery has been found in cases of dilatation of pulmonary artery or increased pulmonic flow, there has been discussion about the minimum gradient of pressure necessary for diagnosing pulmonic stenosis. There is general agreement that a minimum of 20 mm. is required in order to establish such a diagnosis.

Since there are cases of pulmonic stenosis with marked elevation of the right ventricular pressure who show slight or no disability, determination of pressure by cardiac catheterization is of great value for the evaluation of the patient for surgery. Even when the patient is symptom-free, if the pressure is higher than 80 mm. of Hg., surgery is advisable, while a pressure higher than 100 mm. Hg. is definite indication for surgery.

The blood samples usually reveal no significant change in oxygen content in the various chambers of the heart. Occasionally, when the opening of the valve is very small and the catheter stays in the pulmonary artery for a long time (care should be taken to avoid this), the oxygen content of the pulmonary artery may become low as a result of the obstruction produced by the catheter and of the diminished cardiac output.
Pulmonary stenosis with patent foramen ovale.—Right atrial pressure is usually high in cases of pure pulmonic stenosis even in the absence of right heart failure. If the foramen ovale is patent, mixed venous blood is shunted from the right to the left atrium. This blood then mixes with the fully saturated blood coming from the lungs and passes to the ventricle and aorta. Therefore cyanosis may be present at birth.

Except for arterial unsaturation of the blood, the data of catheterization are similar to those of pulmonic stenosis with closed septa. The typical cases offer no diagnostic difficulty. Whenever the differential diagnosis with a Tetralogy of Fallot is difficult, catheterization should be done because a Blalock-Taussig operation would be detrimental. A definite diagnosis can be made through cardiac catheterization when the pressure of the right ventricle is much higher than that of the aorta. Then ventricular septal defect and dextroposed aorta can be excluded. When the pressure in the right ventricle and aorta is at about the same level, ventricle-to-face circulation times determined by fluorescein in children, and ether and Evans Blue in adults, also help in ruling out an over-riding aorta.

Tetralogy of Fallot.—This is the most frequent cyanotic malformation of the heart. As well known, it consists of pulmonic stenosis, dextroposition of the aorta, and high ventricular septal defect; right ventricular hypertrophy is a necessary consequence. Infundibular stenosis is the usual finding in Tetralogy of Fallot; however, cases with valvular stenosis are not uncommon.

In typical cases, the aorta overrides the septum by 40 per cent. On account of the pulmonic stenosis, there is an increased resistance for the blood to reach the pulmonary artery and, as a result, blood is shunted from the right ventricle into the aorta causing some unsaturation of the arterial blood. The amount of the shunt will depend on the severity of the stenosis and the degree of overriding.

In the majority of cases the diagnosis can be made clinically. Atypical cases need catheterization; however, sometimes angiocardiography is even more helpful. Catheterization usually shows a normal or low pressure in the pulmonary artery and a high pressure in the right ventricle. The right ventricular pressure should be about the same level as the aortic pressure. Some degree of left-to-right shunt may be demonstrated by a step up in oxygen content in the right ventricle. Overriding of the aorta is confirmed if the catheter is passed into it. Unfortunately the aorta and pulmonary artery are seldom entered.

A definite diagnosis can be made when low pulmonic pressure and high right ventricular pressure are found; the aorta is catheterized; right ventricular and aortic pressure are at about the same level.

When the aorta is not catheterized, no step-up in the right ventricle is found, and right ventricular pressure is at the same level as the systemic, the ventricle-to-face circulation time is of help for the evaluation of a right-to-left shunt from the right ventricle to the aorta. A short right ventricle-to-face circulation time confirms the diagnosis of Tetralogy of Fallot, while prolonged circulation time would be against it. However,
normal circulation time does not rule out Tetralogy of Fallot.\textsuperscript{3, 20} Selective angiocardiography\textsuperscript{22} with the catheter in the right ventricle is the ideal method for the evaluation of these cases. It makes possible to visualize the infundibulum, the pulmonary artery and the pulmonary valve, besides showing the overriding aorta. It cannot be a substitute for catheterization because it fails to give information about the hemodynamic changes.

Tricuspid Atresia.—The patient may survive in spite of this condition if there is an atrial septal defect. Blood coming from the cavae and the coronary sinus passes through the atrial defect to the left atrium where it mixes with fully saturated blood coming from the pulmonary veins and then goes to the left ventricle. From this chamber it reaches the systemic circulation through the aorta and also reaches the pulmonary circulation by one of the following routes:

(a) through a ventricular septal defect, a small right ventricle and the pulmonary artery.
(b) through a pulmonary artery arising from the left ventricle;
(c) through patency of the ductus or collateral circulation.

Two basic data are needed in cases of tricuspid atresia:

(1) Pulmonary circulation—The amount of blood reaching the lungs may be reasonably estimated by fluoroscopic and x-ray examination in the majority of the cases, but sometimes it is difficult to determine the need for an anastomosis by clinical means. Catheterization does not help much in getting this information. As is expected the catheter fails to enter the right ventricle. It passes into the left atrium and thence may be passed to the pulmonary veins or left ventricle. The position of the pulmonary artery is such that it is almost impossible to intubate this vessel. The blood that reaches the lungs comes from the left ventricle but it may not be mixed well enough for even an approximated estimation of pulmonary flow. Angiocardiography may be more useful than cardiac catheterization.

(2) Size of the atrial septal defect—When the defect is small, the right atrium has to work against a greater resistance in order to deliver all the blood received from the cavae. As a result, it hypertrophies and dilates. Liver pulsations may be an indication that the septal defect is small and have been observed even in one case proved at autopsy to have a single atrium.\textsuperscript{23} The magnitude of the P waves of the ECG may be of value in the evaluation of the right atrial hypertrophy, which in turn would be inversely proportional to the size of the defect. No definite conclusions can be reached at present. Angiocardiography usually gives valuable information about the size of the defect.

Taussig-Bing Syndrome.\textsuperscript{24} In this condition, the aorta arises completely from the right ventricle while the pulmonary artery, usually very large, overrides the septum. There also is a ventricular septal defect. Through this defect, some arterial blood from the left ventricle reaches the aorta while
venous blood from the right ventricle reaches the pulmonary artery. Catheterization demonstrates that the blood of the pulmonary artery has a higher oxygen concentration than that of the aorta. The pressures of both vessels are at about the same level. When the defect is small, the patient dies early in life. Attempts to increase the mixture of venous and arterial blood by creating an atrial septal defect have been made in some cases with good results. Catheterization is of help in the evaluation of these cases, prior to surgery, in order to find out if an atrial septal defect already exists, and after surgery in determining the hemodynamic changes due to the operation. In complete transposition of the great vessels, catheterization of the pulmonary artery is more difficult and failure to accomplish this would not permit definite conclusions.

Eisenmenger complex.—This malformation consists of a ventricular septal defect associated with some degree of over-riding of the aorta and right ventricular hypertrophy. Since resistance of the pulmonary vascular bed is less than the systemic, shunting is predominantly left-to-right, at least in an early stage. However, pulmonary vascular changes may develop early in life and, as a result, pulmonary vascular resistance increases and may even become higher than the systemic. Then, a predominant right-to-left shunt occurs. Therefore, the time of appearance of cyanosis is in relationship with the degree of pulmonary resistance. The mild cases of Eisenmenger may be confused with moderate pulmonic stenosis, ventricular septal defect, or even Tetralogy of Fallot. The severe cases may be confused with the Taussig-Bing syndrome.

Cardiac catheterization contributes to the differential diagnosis in the first group of cases by showing:

(a) equal systolic pressure in the right ventricle and pulmonary artery;
(b) low saturation of blood in the femoral artery.

In the second group, catheterization makes possible the demonstration of either a small step-up in oxygen content in the pulmonary artery or none at all. In the Taussig-Bing syndrome, it would show a higher oxygen content in the pulmonary than in the systemic artery, because the pulmonary artery arises from the left ventricle.

Ebstein’s Disease.—This anomaly consists of a downward displacement of part of the tricuspid valve and is usually associated with an atrial septal defect. Due to this association, cyanosis is frequently present. Although difficult, a clinical diagnosis may be possible. In some cases, differential diagnosis with a pure pulmonic stenosis can be made only by cardiac catheterization. If the pulmonary artery is entered, the systolic pressure of this vessel is similar to that of the right ventricle. Tricuspid insufficiency, when present, would result in high pressure in the right atrium with little gradient between this cavity and the right ventricle. On withdrawal of the catheter from the ventricle, a change from the ventricular pattern of pressure to an atrial pattern may occur near the apex while no further change takes place at the region of the tricuspid valve (displacement of the tricuspid leaflets).
CONCLUSIONS

Cardiac catheterization is a valuable tool for diagnosis and evaluation of congenital malformations of the heart. Many physical and roentgenological signs have been clarified by this method. However, there is no substitution for a good history and physical examination. The decision to submit a patient to cardiac catheterization should be made only after complete clinical evaluation of the patient. It should be pointed out that certain risks are involved in this procedure. Therefore, only a trained team can undertake this procedure with success.

RESUMEN

La cateterización cardiaca es un procedimiento valioso para determinar y valorar las malformaciones del corazón. Muchos signos físicos y roentgenológicos se han aclarado por este método. Sin embargo, no hay substituto para una buena historia y un buen examen físico. La decisión para someter un enfermo a la cateterización, debe hacerse sólo después de un estudio clínico completo. Debe señalarse que hay ciertos riesgos en el proceder. Por tanto sólo el personal bien preparado puede emprender el método satisfactoriamente.

REFERENCES

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