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Aspects of congenital heart disease as elucidated by cardiac catheterization

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ASPECTS OF CONGENITAL HEART DISEASE AS ELUCIDATED BY CARDIAC CATHETERIZATION

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INTRODUCTION

It seems to be a common misconception among many members of our society that the scientist working in his laboratory is doing so only because the result of his endeavor may be of practical benefit to mankind. Thus the atomic scientist theorized about the atom in order to afford us with an atomic bomb and Fleming concerned himself with the findings on his agar plate because he could see its far reaching effect on mankind.

Now it is apparent to most individuals engaged in research that they are seeking fundamental truths apart from their effect on the technicological or socio-political progress of our society. Therefore it is true in a sense that the scientist does reside in an ivory tower insofar as his mind and energies are free to meditate, to theorize, to experiment and to give vent to his insatiable curiosity concerning the "whys" of our universe.

So it was that Werner Forssmann (25-26) did not realize the vast clinical application of cardiac catheterization when he threaded a small tube into his own basilic vein in 1929.

HISTORICAL CONSIDERATIONS

Catheterization of the heart by the intravenous route was first performed in Germany, by Forssmann (25-26), who was interested in the rapid injection of drugs into the heart. The first catheterization was performed by Forssmann on himself. In his first attempt, Forssmann, aided by another physician, introduced 35 cm. of a number 4 ureteral catheter into an arm vein. Because, as he writes, his colleqgue considered continuation of the test too dangerous, it was interrupted although, "I felt perfeetly fine." After another week Forssmann made another attept, alone. This time he was able to introduee the catheter into the right auricle and verified the position of the catheter in a mirror which was held by a nurse in front of the flueroscopie screen; "I observed the progress of the catheter in a mirror which was held by a nurse in front of the fluoroscopic screen." Forssmann mentioned then that he had no untorward sensation and that he considered the procedure safe. There is no doubt that his should be the credit for the first successful catheterization of the heart.

Klein (40) werking in Prague, used catheterization of the right side of the heart for the first time to determine the cardiac output of man by means of the Fick priniciple. Klein performed 18 catheterizations, in 8 of which the heart was intubated; in some instances, the catheter also entered the inferior vena cava of the right ventricle. The technic of Klein is almost indentical with that used by most investigators at the present time, and his should be the credit for the first physiologic application of the method.

From 1930-1939 the technique of right heart catheterization has been widely

used in Europe for injecting contrast substance in order to visualize the right chambers of the heart and pulmonary vascular tree.

In 1941, Cournand and Ranges (20) described the introduction of a radiopaque catheter into th right auricle of a human subject and standardized the method for the determination of the cardiac output. Since then, a great number of investigators have used this method for diagnostic and physiologic studies both here and abroad. An accurate determination of the pressures existing in the heart and great vessels of man by means of the catheter was performed by Bloomfield and co-workers (9) and Lenegre and Maurice (46). The recording of pressure curves is now indispensable to cardiac catheterization.

CATHETERIZATION TECHNIQUE

Some of the minute details of catheterization technique will not be dwelled upon in this paper since its primary purpose is not be a laboratory manual for those involved actively in cardiac catheterization. Adequate descriptions of general teshnique may be found in the literature (7, 14, 19, 21, 48, 56, 66, 68).

A general description will be given however in order to give the reader some idea of catheterization procedure.

Under strict asepsis a nick is made in the median vein. There is preliminary infiltration of novocain. A 10 gauge Lindeman type needle inserted and then the catheter is introduced into the vein through the needle. Several types of catheters are in use. One type described by Cournand and Ranges (20)has one eye at the tip of the catheter. Goodale et al. (32) described another type which, in addition to this opening, has two small eyes at each side and a groove connecting the side eyes at the tip. The latter type is apt to cause less obstruction to the catheter by endothelial or endocardial tissue on withdrawal of blood. The end of the catheter is connected to a heparinized saline reservoir to provide constant drip infusion through the catheter (15 mg. of heparin per 1000cc of saline solution). Heparinization of the patient is unnecessary.

Further passage of the catheter is done under fluorscopy.. It is important to move the catheter under fluoroscopic visualization only. Otherwise the position of the catheter tip cannot be ascertained and serious errors in the diagnosis of intracardiac lesions will result. The danger to the patient and physician of repeated x-ray exposure should be kept in mind.

To avoid over exposure, it is advisable to limit screening time five minutes at five milliamperes. The operator should wear a lead apron and protect his hands by a lead shield or a lead apron placed under the arm and along the side of the body of the patient. Despite these precautions, over exposure has been observed in physicians performing repeated catheterizations; therefore routine observation of the differential blood count is essential.

When the tip of the catheter is in the heart, the tubing of the saline drip is discontinued and a large syringe filled with a little saline is adapted to the 3 way stop cock. Two or three ccs of blood are withdrawn into this syringe thus washing the catheter with right heart blood. Then the valve is turned and the blood for analysis collected in a second syringe containing mineral oil as an air seal, 15-20 ccs of blood can be collected in 25 seconds by using only the slightest amount of suction. Following the withdrawal of blood, the saline drip through the catheter is restored. The oxygen content of the blood may be determined with the conventional Van Slyke method (65) or according to the method of Wood (68). This investigator has devised a photoelectric device which determines immediately the oxygen saturation of samples of blood withdrawn from the catheter.

General anesthesia is almost never used. In adults, catheterization is performed without any premedication. In children from 1-10 years of age, a morphine-scopolamine mixture is employed.

Protocol for the determination of cardiac output is obtained by simultaneous sampling of mixed venous blood, femoral arterial blood and collection of expired air by Tissot spirometer.

In the recording of the pressure through the catheter, a measuring instrument should reproduce and record the physiologic events in a true and undistorted manner, and the recording system should have an adequate sensitivity and adequate frequency response.

It has shown that most of the existing manometers for the determining the pressure in the body cavities containing air or liquid suffer from certain defects both clinically and physiologically. This is the result particularly of the relationship between amplitued distortion, phase distortion, and the degree of damping.

In general, two systems for pressure recording are used. The first is based on optical principles. The optical manometer is the prototype of these instruments. A number of other workers (30, 37, 61) constructed similar apparatus. Hamilton's (37) manometer is purely optical, in contrast to most of the other manometers which use mechanical connecting parts. The greatest advantage of Hamilton's optical system is that it permits attainment of a very high modulus of volume elasticity. It is probably still the most accurate tool for the registration of pressures, but its operation requires a certain amount of technical skill, patience, and knowledge. In this respect, however, it does not differ from its electrical counterparts.

The other systems used are based on purely electrical principles. The movements of a manometer membrane are at once transformed into electrical energy which is then transferred to the ultimate recorder. The transformation of the pressure impulse may be affected by (1) changes in electric

resistance of a wire in proportion to its length, (2) a piezo electric effect, (3) induction phenomena, and (4) changes in capacitance. In the latter system, an electric plate condenser is used in which the distance between the plates and thus the capacitance can be changed.

Dry cell batteries are sufficient to energize the transducers while the relatively large electric output of the transducer permits direct operation of many indicating and recording instruments. Lambert and Jones (44) described in detail the properties of resistance wire manometers. They selected a gauge with relatively low sensitivity. They found the gauges to be lineal and stable as long as a constant input was maintained. Upon application of an approximately square wave pressure front, the manometer indicated about 99 per cent of the pressure change within a fraction of a second.

The ideal means of recording pressures would be from a pressure pick-up unit, located at the ti of the catheter directly. Such pressure pick-ups, based on a differential transformer, have been build by Gauer and Gienapp (29). Ellis, Gauer, and Wood (23) applied such a system for the recording of artefact-free pressure tracings from the heart and great vessels of man and animals. Their results indicate that the contour and magnitude of the central pulse can usually be recorded with a nonometric system which possesses a uniform sensitivity of up to 5 cycles per second but only 32 per cent response at 60 cycles per second.

PERTINENT NORMAL AND ABNORMAL PHYSIOLOGIC FINDINGS IN CARDIAC CATHETERIZATION

In order to understand and appreciat the technique of cardiac catheterization and the results obtained therein one must have some basic understanding of cardiac and circulatory physiology.

It is assumed that the basic features of cardiac anatomy are known. The heart, a four chambered organ, has as its primary purpose the action of pumping non-aerated blood to the lungs and aerated blood to the general circulation. The non-aerated or venous blood flow into the right heart; it is then pushed into the lungs where it is aerated. After aeration the oxygenated blood returns to the left heart where it received its final push into the systemic circulation.

Now certain fundamentals of cardiac and hemo-dynamics were known before the era of cardiac catheterization. Graphic representations of pressure changes within the auricle and ventricle were recorded from animals, records of arterial and venous pulses in man were obtained by means of the polygraph, and electrical changes were noted by EKG tracings. Oxygen content and carbon dioxide content of arterial and venous blood were calculated, and cardiac output in man could be measured by means of the indirect Fick Method. How then does the catheterization technique augement our understanding of cardiac physiology, and how does it enter the realm of clinical diagnosis?

The feasibility of threading a tube into the heart means that now we can record directly pressure changes in the heart; determine the oxygen

content and carbon content of the blood within the cardiac chambers. The output of the heart can be calculated directly by means of the Fick (24) Method. These direct measurements are indeed of great import for it enable investigative studies in cardiac dynamics to have practical implications in the tealm of cardiac pathology--particularly as it pertains to cardiac anolomies ameliorated by surgical intervention.

It is necessary however before understand pathological findings to first be familar with the normal. Numerous studies done by Bing (5,7) and Dexter (21) on control subjects serve as a standard of reference. It has been shown that the following are the average pressure values are found in normal subjects: right auricle plus or minus 5 mm. Hg., right ventricle 25/0 mm. Hg., pulmonary artery about 25/0 mm. Hg.

There was some difficulty in establishing a normal range for oxygen content of the blood in the cardiac chambers(carbon dioxide values are not used since it is known that the carbon dioxide values are continually changing in response to slight alterations in respiratory or circulatory equilibrium (21). The reason for this difficulty arise from the fact that the right auricle receives venous blood from three major sourses i.e. superior vena cava, inferior vena cava and coronary sinus, each having a different oxygen content. Fowler (27) has shown that inferior vena cava blood may be 2 Vol. per cent over that in the superior vena cava, whereas blood from the coronary sinus is very low in oxygen content, being usually below 7 Vol. per cent. It is obvious therefore that the venous blood is not mixed in the right auricle. This mixing occurs only after the blood has reached the pulmonary artery. It was found by Dexter (21) and Bing (5,7) in control patients that the greatest increase in the oxygen conetnt of the blood

in the right auricle over that in the superior vena cave is 2 Vol per cent the greatest increase in oxygen content of blood in the right ventricle over that present in any part of the right auricle is 1 Vol. per cent, and the greatest increase in oxygent content of blood in the pulmonary artery over that present in any part of the right ventricle was 0.5 Vol. per cent.

In contrast to the descrepencies in oxygen content of blood obtained from the right auricle and right ventricle there was remarkable uniformity in values obtained from the pulmonary artery. The maximum variation observed hardly exceeded the method for determining oxygen content. It is believed therefore that true mixing of venous blood occurs uniformly in the pulmonary artery in man, and that the pulmonary artery may be used as a source of mixed venous blood in determining cardiac output by the direct Fick Principle.

An early misleading finding was that of a higher than normal oxygen content in the pulmonary artery when the tip of the catheter was as far out as it would go in the pulmonary artery. It took but little reasoning, deduction and further investigation however to wxplain this finding on the grounds that the catheter had occluded the lumen ofthe small artery and that therefore the blood withdrawn from the catheter really came back from the pulmonary capillaries and veins where xoygen content would be that of arterial blood.

To recapitulate briefly at this point--through catheteriation studies we have extablished directly:

1. The normal pressures found in the right heart and pulmonary artery.

2. The average variation in oxygen content of blood in the right heart and pulmonary artery.
3. The oxygen content of mixed venous blood
4. The oxygen content of arterial blood as getten from the pulmonary capillaries.

Now for consideration of cardiac output and the Fick Principle.

In 1870 Fick (24) pointed out that the difference between the concentrations of oxygen or carbon dioxide in arterial and mixed venous blood represents the amount of oxygen taken up or carbon dioxide given up by each unit of blood as it blows through the lungs. If, in addition, the total amount of oxygen absorbed or carbon dioxide eliminated in a given time be known, the amount of blood flowing through the lungs during that period may be calculated by means of the following formula:

$$\frac{\text{O2 intake (ml/min) times 100}}{\text{O2 Content of peripheral blood} - \text{O2 content of Mixed Venous Blood (Vol. \%)}}$$

The blood flow through the lungs obviously represents the output of the right ventricle, since the output of the left ventricle is the same its output is determined as well. With the advent of catheterization mixed venous blood can be obtained and the direct FickMethod can be used.

After having considered pertinent normal results obtained from cardiac catheterization we may now understand more clearly abnormal findings. For example in many cardiac abnormalities there exists a shunt between the right and left heart by using the direct Fick method were are now able to calculate the volume of the shunt.

To elaborate:

The peripheral blood flow may be calculated in the fashion already noted except that the sample of the mixed venous blood must be obtained proximal to the shunt. In cases with patent ductus it may be obtained by multiple samples of blood from the right ventricle, in cases of ventricular septal defect, from the right auricle; and in cases of auricle septal defect from the superior vena cava. Since true mixing becomes progressively poorer in this sequence errors in the application of the Fick principle of blood flow becomes greater.

Determination of pulmonary arterial blood flow necessitates measurement of the oxygen content of the pulmonary artery and of venous blood. Normally, blood in the main branches of the pulmonary artery is completely mixed (Dexter 21). This is also true in most cases of auricle septal defect. In ventricular septal defects and in ductus arteriosus mixing may be less complete. In these cases multiple sampling in the plumonary artery usually yields a representitive value from which the calculation of pulmonary blood flow can be made. The oxygen content of pulmonary venous blood is most easily obtained in individuals with a normal circulation and in those with left to right shunts by determining the oxygen content of the systemic arterial blood since these two are identical. In an individual with right to left shunt, the oxygen content of the pulmonary venous blood can be obtained by measuring the oxygen content of the pulmonary capillary blood. Failing this, a value of 95-98 per cent saturation must be assumed. This admittedly is open to criticism especially when pulmonary disease with imperfect oxygen diffusion exists.

The effective pulmonary blood flow is of special significance for it represents the volume flow of blood which after its return to the right auricle, ultimately reaches the pulmonary capillaries. It has been called the effective pulmonary blood flow since it representsthat component of mixed venous blood which becomes effectively oxygenated in the lung (5). In the absence of a pathologic shunt, no oxygenated blood reaches the pulmonary artery and no venous blood reaches the aorta, so that the effective pulmonary blood flow equals the pulmonary arterial or systemic blood flows. Calculations have shown that in congenital malformations of the heart in which a septal defect is present, the volume flow through the shunt is predominately unidirectional. The only exceptions are complete and partial transposition of the great vessels. In these malformations the unidirectional shunt is incompatible with life. The overall intracardiac shunt represents the difference between the systemic and the pulmonary arterial blood flow. If the systemic flow exceeds the pulmonary arterial blood flow, the shunt is directed from right to left. If, on the other hand, the pulmonary arterial flow exceeds the systemic flow, the intracardiac shunt is directed from left to right. It should be borne in mind that in most instances there is reciprocal admixture through the defect. The actual volume of the left to right current is represented by the difference between the pulmonary arterial effective flow (total right to left shunt). Conversely, the volume of right to left mixing currents is represented by the difference between the systemic flow and the effective pulmonary blood flow (total left to right shunt). In the presence of right to left admixture of blood the volume of total mixed venous blood is decreased. This will result in a fall in the ratio of effective pulmonary flow over systemic flow, with a subsequent decrease pulmonary blood flow and decreased oxygen saturation. The arterial

oxygen saturation is not the result of decreased pulmonary blood flow but rather due to a decrease in the ratio of effective pulmonary blood flow over systemic flow or expressed mor simply, is due to the right to left shunt. The decrease in pulmonary blood flow reduces this ratio. Reduction of pulmonary blood flow alone, for example, in isolated pulmonic stenosis does not result in arterial oxygen unsaturation.

In regard to congenital anomolies as indicated by other abnormal catheterization results; it has been found generally speaking that increased oxygen content in the right heart above the expected normal variation indicated a cardiac shunt of blood from left to right. The same is true as concerns increased pressure recordings within the heart (4,5,7,15,19,21,).

DANGERS OF CARDIAC CATHETERIZATION

Although cardiac catheterization is not as dangerous a procedure as one might expect, it is certainly not innocuous. Some of the complications that may arise are as follows:

1. Cardiac arrhythmias
2. Venous thrombosis
3. Chills
4. Acute attacks of anoxia when the pulmonary artery is completely occluded.
5. Air emboli
6. Venospasm
7. Pulmonary infarction

Cardiac arrhythmias:

Disturbances of rhythm are the most frequent and most important complication to consider. Almost every type of disturbance in cardiac rhythm has been encountered. In the most complete study of electrocardiographic changes during catheterization Goldman et al.(31) observed 50 patients under constant visual electrocardiographic control. Sixty per cent of these showed auricular premature systoles. These were most frequent when the catheter tip appeared to be in contact with the auricular septum and particularly when the catheter was passing through the auricular septal defect. Nodal premature systoles were seen in 78 per cent of the patients. Superventricular tachycardia of auricular or nodal origin occurred in 28 per cent, and auricular flutter in 6 per cent. In 85 per cent there were ventricular premature beats, and in 78 per cent short bursts of ventricular tachycardia developed. Ventricular flutter was seen in 6 per cent of the cases.

Dexter and his group (21) observed 2 patients who developed transcient auricular fibrullation which subsided spontaneoulsy in the course of one-half hour. Johnson and his co-workers (38) reported auricular tachycardia in a patient with an auricular septal defect when the catheter was passed through the opening.

Slight transcient heart block, right bundle-branck blocks and pre-excitation phenomema have been seen by (41, 54, 45).

Ventricular premature beats appear quite frequently. They are produced noticably when the tip of the catheter bouches the wall adjacent to the tricuspid valve. The catheter should never be allowed to curl up in one of the chambers. Ventricular premature beats occur also quite regularly when the tip of the catheter touches the wall of the right ventricle(31).

Micheal and his group (50) reported 113 consecutive venous catheterizations. Abnormal rhythms developed in more than one-third. The tendency was significantly greater in the presence of congenital heart lesions and in patients whose electrocardiographs were abnormal prior to the procedure. These observations are in line with certain cardio-vascular abnormalities which should be considered definite contra-indications to the procedure. Dexter (21) listed them as follows: spontaneous ventricular premature systoles, recent history of paroxysmal ventricle tachycardia, subacute bacterial encocarditis, coronary occlusion within 6 weeks, any manifestation of digitalis toxicity, diffuse myocarditis(e. g. rheumatic) and auricular fibrillation.

Zimdahl (69) does state however that venous catheterization has been performed on fairly ill patients and often seriously ill patients with no serious effects. It has been used for example in congestive failure, pericardial tamponade, acute hemorrhage, shock, anemia and penetrating wound of the chest.

Venous thrombosis:

Venous thrombosis is not frequent following catheterization. Bing(4) has observed it in about 0.5 per cent of all catheterization. In no case has the thrombosis been progressive. Cournand and his associates (18) reported that in more than 260 catheterizations with the catheter left in place continuously for more than 24 hours no unfavorable complications were encountered beyond the formation at times of a mural thrombus near the site of insertion of the catheter in the brachial vein.

The occurrence of local venous thrombosis at the site of introduction of the catheter is not common. Careful handling of the veins and tissues and frequent moistening of the catheter with saline solution as it is withdrawn and inserted forward will help this (4).

Chills:

Chills are primarily due to pyogenic substances probably remaining in the catheter from a previous test. Chills may be avoided by thoroughly rinsing the catheter with sterile water followed by rinsing with a detergent solution. The catheter should be boiled for one-half hour immediately preceding catheterization (7).

Bing also states that he has found it useful to insert a stylet through the catheter, with its tip reaching within 1 and one-half inches of the catheter tip. This makes the catheter more rigid and places the lever point directly at the bend of the catheter.

Anoxia:

Acute attacks of anoxia due to occlusion of the pulmonary artery were observed in Bing's laboratory in two cases of isolated valvular pulmonic stenosis. The attacks resulted probably from a sudden decrease in the cardiac output following occlusion of the pulmonary artery. The resulting fall in cerebral blood flow probably initiated the attacks.

Air emboli:

Air emboli have been observed by Bing (7) in 4 of some 1,800 cases. The neuorlogic symptoms were primarily of pontine origin, but all symptoms disappeared after a period of hours, without residual damage.

Venospasm:

Venospasm may occur when there is difficulty in inserting the catheter into the vein or when the catheter cannot be introduced into the heart on the first attempt, due to anatomic anomalies or the veins (7).

Pulmonary infarction:

Houssay et al. (38) reports pulmonary infarction occurring in 19 patients with severe stenosis or congenital heart failure following wedging of a catheter into a branch of the pulmonary artery for measurement of pulmonary

"capillary" pressure. This complication was reduced when the catheters were sterilized by autoclaving instead of by soaking in cyanide.

It is unlikely that catheterization of the human heart damages the endocardium. Edwards and his group (22) reported that post mortem examination of patients on whom catheterization had previously been performed failed to show any demonstrable lesions of the veins, heart, pulmonary artery or lungs attributable to the passage of the catheter.

CLINICAL APPLICATION OF CARDIAC CATHETERIZATION IN CONGENITAL HEART DISEASE

The precise diagnosis during life of congenital heart lesions has now more than an academic interest. Ligation of patent ductus is a well established method of therapy while surgical procedures for tetrology of Fallot are of great assistance. Even if the presence of a ductus is obvious it may be of value to know whether other lesions are present or absent when considering ligation. Enlargement of the pulmonary artery as a main finding has long presented a difficult differential problem for clinicians and radiologists when one must consider such conditions as: patent interventricular septum, mitral stenosis, pulmonary stenosis, Eisenmenger's complex, patent ductus, cor pulmonale, emphysema, primary pulmonary hypertension in making the differential diagnosis.

Congenital heart disease may be classified on the basis of clinical, physiologic or pathologic findings. Maude Abbott's classification is based on the presence or absence of cyanosis and accordingly she divides congenital heart disease into 3 groups: (1) An acyanotic group (no abnormal communication) (2) A group consisting of individuals with arteriovenous shunts who show terminal reversal of flow; (3) A group consisting of individuals with cyanosis(1). This classification has certain disadvantages. For instance, although septal defects are classified as belonging to group II, cyanosis may never appear. Patent ductus arteriosus, also included in this group, rarely leads to cyanosis. Furthermore, cyanosis does not represent the primary physiologic disturbance but is fundamentally the result of a right to left shunt.

Taussig (61) divides congenital malformations of the heart into two groups: Group I, malformations which deprive the body of an adequate amount of oxygenated blood; Group II, malformations which permit the body to receive an oxygen supply sufficient for growth of the individual. Using this classification, Eisenmenger's complex is brought into Group II, despite the fact that in this malformation the cyanosis may be as severe as that encountered in the tetralogy of Fallot.

A classification on a purely physiologic basis will suffer from the disadvantage that malformations with similar anatomic findings are brought into different physiologic groups. These disadvantages are outweighed by the fact that surgical intervention depends on physiologic rather than on anatomic findings. Thus, group I, outlined below, contains all conditions in which it may be helpful to increase the effective pulmonary blood flow by surgical means. An attempt has been made by Bing (4) to classify cardiac malformations according to the disturbances in the hemodyanamics which result from pathologic malformations.

CLASSIFICATION

I. Pulmonary Flow Less than Systemic Flow. Pulmonary Artery Pressure Usually Decreased

A. Tetralogy of Fallot
B. Pseudotruncus arteriosus
C. Tricuspid atresia
D. Single ventricle
    1. with pulmonary artery arising from a rudimentary outlet chamber
    2. with pulmonic stenosis
E. Transposition of the great vessels with pulmonic stenosis
F. Patent foramen ovale with pulmonic stenosis
G. Ebstein's disease with patent foramen ovale
H. Anomalous venous return
    1. pulmonary arteriovenous fistula
    2. superior vena cava emptying into the left ventricle

II. Pulmonary Flow Greater than Systemic Flow and/or Pulmonary Artery Pressure Normal or Increased

A. Eisenmenger's complex
B. Transposition of the great vessels
    1. complete
    2. partial
C. Isolated septal defect
    1. auricular septal defect
        a. uncomplicated
        b. Lutembacher's disease
    2. ventricular septal defect
D. Aortic atresia with patent ductus arteriosus
E. Single ventricle
    1. with both great vessels arising from the rudimentary outlet chamber
    2. with the aorta arising from the rudimentary outlet chamber
F. Truncus arteriosus
G. Patent ductus arteriosus
H. Anomalous venous return with pulmonary vein emptying into the vena cava or the right auricle

III. Pulmonary Flow Equals the Systemic Flow at Rest and After Exercise

A. Pure pulmonic stenosis
B. Patent foramen ovale as an isolated anomaly
C. Coarctation of the aorta
    1. adult type
    2. infantile type without patent ductus arteriosus
D. Double aortic arch

Group I: Pulmonary Artery Flow Less than Systemic Flow.

Tetralogy of Fallot:

The four characteristics of the tetralogy of Fallot are:

1. A defect of the membranous portion of the interventricular septum.

2. Dextroposition of the aorta which overrides both the right and left ventricles.

3. Pulmonary stenosis. This is essentially a stenosis of the infundibulum of the right ventricle, which is either narrowed throughout, or exists as a "third ventricle," separated from the right ventricle, which lies below it by a constricted opening. Associated with this is a thin-walled, narrow, pulmonary artery, and an abnormal pulmonary valve, which is bicuspid, defective or stenosed. The ductus arteriosus is closed; in extreme cases, pulatresia occurs. In such cases, the ductus remains open and carries blood to the lungs.

4. Right ventricular hypertrophy.

This is the most frequent congenital cardiac malformation with cyanosis and includes about 75 per cent of all cyanotic patients with congenital heart disease.

Results of catheterization:

(1) Movements of the catheter: Usually the catheter may be introduced into the right auricle and the right ventricle without particular difficulty in cases in which there is marked right auricular enlargement. Dexter (21) and Bing (4) as well as Cournand (9) have been able to introduce the catheter into the pulmonary artery through a stenosis in many cases. In rare instances the catheter may pass through the septal defect into the left ventricle. The aorta may also by intubated from the right ventricle.

(2) Calculation of Flow: The flow through the pulmonary artery is below normal whereas the systemic flow shows marked variation above and below normal. A comparison of the systemic and the pulmonary flows show that the former exceed pulmonary artery flow by from 0 to 9.7 liters per minute (4). Consequently, the over-all direction of the shunt is from right to left. There does occur some reciprocal admixture however. This is indicated by the observation that the oxygen content of the right ventricular blood exceeds that of the right auricular blood. These findings indicate that the volume of blood flowing through thesystemic capillaries is particularly high in individuals with severe pulmonic stenosis, large interventricular septal defects, and marked dextro-rotation of the aorta. In patients with a moderate degree of pulmonic stenosis and small septal defects, the systemic flow is approximately normal. The hemodynamics of the systemic flow in the tetralogy of Fallot depends to a large extent on mechanical factors brought about by the cardiac malformation as well as on the resistance of the peripheral arterioles.

(3) Pressures in the Cardiac Chambers and in the Pulmonary Artery: The right auricular pressure may be normal or elevated. Intraventricular pressure curves obtained in pulmonic stenosis reveal that the systolic pressure in the right ventricle is markedly elevated (from 40-110 mm. above normal.) In most instances the intraventricular pressure rises sharply and falls during the ejection phase. The presence of a considerable diastolic right ventricular volume is demonstrated by the observation that in a great number of patients with the tetralogy of Fallot the general level of the ventricular diastolic pressure is elevated above the minimum values which occur early in diastole (Bing, Vandam and Gray 5).

When the catheter is passed into the pulmonary artery through the stenosis a sharp fall in pressure is observed. In some instances the pressure in the pulmonary artery is above normal. Consequently, the pulmonary artery pressure alone is of no diagnostic importance. The pressure gradient between the right ventricle and the pulmonary artery, however, is significant. Small pressure gradients between right ventricular and pulmonary arterial pressure have been recorded without the presence of pulmonic stenosis in individuals with large right to left shunts. Here the increased systolic blood volume leads to the development of a pressure gradient between the right ventricle and pulmonary artery. This pressure gradient between the right ventricle and the pulmonary artery, however, does not exceed 30 or 40 mm. of Hg.(59,60)

(4) The Pulmonary Capillary Flow: In most of the younger individuals with the tetralogy the values obtained for pulmonary capillary flow agree with those determined for pulmonary artery flow. In the majority of the patients, pulmonary capillary flow exceeds pulmonary artery flow, indicating the presence of extensive collateral circulation. Large calculated difference between pulmonary capillary and pulmonary artery flow is also obtained in individuals with pulmonic stenosis and a congenital artificial ductus arteriosus (Bing, Vandam and Gray 5).

Pseudotruncus:

This is a condition in which there exists a high ventricular septal defect with over-riding aorta and right ventricular hypertrophy. In contrast to the tetralogy of Fallot, the pulmonary artery is atretic and the circulation to the lung is via the bronchial and other systemic arteries or through a patent ductus. In thetypical form of truncus a single great vessel of large caliber originates from both the right and the left ventricles and the cir-

culation to the lungs is established by way of the pulmonary arteries which branch directly from thesingle vessel.

In this condition, the physiologic fings are similar to those observed in the tetralogy of Fallot. Various types of truncus arteriosus have been discribed by Soulie et al. (57).

Tricuspid Atresia:

Congenital absence of the thicuspid valve is associated with a nonfunctioning right ventricle which may be rudimentary or even absent, and with an atresic or hypoplastic pulmonary artery. In addition, the foramen ovale is patent, or an interauricular septal defect is present. There may be no auricular septum. The ductus arteriosus usually remains patent, but it may close. The course of venous blood is therefore as follows: vena cava to right auricle, through the septal to left auricle and vnetricle, to aorta and systemic circulation; and from the aorta through a patent ductus to the lungs.

Catheterization may offer valuable information and aid in the diagnosis of this condition. The diagnosis of tricuspid atresia should be suspected it the catheter enters a high pressure area from which blood of a relatively high oxygen content is obtained. On withdrawal to the right auricle, the catheter tip should pass to a low pressure area containing blood of a corresponding high oxygen content. In the abnormality the systemic flow is nearly always elevated. The effective pulmonary artery flow is also reduced. The pressure in the common ventricle is elevated (4).

Single Ventricle with the Pulmonary Artery Arising from a Rudimentary Outlet Chamber:

In this malformation the rudimentary outlet chamber is separated from the

main ventricle by a muscular ridge. It lies in the region normally occupied by the pulmonary conus; pulmonary stenosis may be present. Single ventricle is usually associated with malformation of the great vessesl. For instance, both vessels may arise from the diminutive outlet chamber or they may be transposed. The present discussion, however, is limited to the single ventricle with coexistent pulmonic stenosis and a single ventricle from which the pulmonary artery arises from the rudimentary outlet chamber.

Usually the catheter does not enter the pulmonary artery or the rudimentary outlet chamber. There is a difference in oxygen content between right aurcle blood and blood in the high pressure chamber of more than 3.5 volumes per cent. The over all shunt is from right to left. The differentiation between tricuspid atresia and this malformation is difficult (4). Unless continous pressure recordings are taken, together with frequent blood samples for blood oxygen determination, the left ventricle may easily be mistaken for a common ventricle.

Transposition of the Great Vessels with Pulmonic Stenosis:
In this condition there is an increase in aertic blood flow and a reduction in the pulmonary blood flow and the effectvie pulmonary blood flow. Similar to transposition of the great vessels without pulmonic stenosis, the systemic and lesser circulations exist as separate units, and a unilateral shunt does not exist (62, 64).

Patent foramen ovale with Pulmonic Stenosis:
This malformation has been described in detail by Selzer et al.(52). In catheterization it has been found that the pressure in the right ventricle

is markedly increased over that in the right auricle whereas the pressure in the pulmonary artery is reduced. There is no significant difference in the oxygen content of blood samples obtained from the right auricle and ventricle. The peripheral arterial oxygen saturation is reduced, demonstrating the presence of a right to left shunt. The systemic flow in these patients is usually normal, and the average effective and pulmonary artery flows are reduced. No significant left to right shunt is seen.

Ebstein's Disease with Patent Foramen Ovale:

This is a rare condition in which part of the tricuspid valve is inserted into the wall of the right ventricle below the auriculo-ventricular orifice. In addition, the valve is deformed and its leaflets fused. The result is to separate the right ventricle into a proximal chamber continuous with the right auricle, and a distal outflow chamber.

Because of the abnormal tricuspid valve, it is difficult for blood to pass from the right auricle into the right ventricle, and the right auricle and the proximal portion of the right ventricle dilate greatly. The right auricular pressure rises above the left auricular pressure, and since a patent foramen ovale is usually present, venous blood is shunted into the systemic circulation.(7)

Catheterization studies show an oxygen content in the superior vena cave of 16.1, in the right auricle of 17.5, and in the right ventricle of 17,8 volumes per cent. The flow as calculated per square meter of body surface indicates that there is a large right to left shunt; the effective pulmonary flow being greatly reduced. The arterial oxygen saturation is decreased;

the pressure in the right ventricle is normal. The right ventricular pressure may be redueed in this condition, usually as a result of hypoplasia of the right ventricular muscle.(4).

Anomalous Venous Return:
Pulmonary Arteriovenous Fistula:

Bing and his co-workers (9) reported catheterization results found in 2 cases of pulmonary arteriovenous fistula. It was shown that in both cases the systemic flow was well over that of the pulmonic flow. The pressure in the pulmonary artery was normal, and the arterial oxgyen content was well under normal.

Superior Vena Cava Emptying into the Left Ventricle:

In this malformation unsaturated Blood from an abnormal left superior vena cava enters the left auricle. This results in arterial unsaturation and cyanosis. Stated briefly the criteria for diagnosis of an anolomous superior vena cava emptying into the left auricle may be summarized as follows:

(1) Catheterization of the left auricle thru the anomalous vein.

(2) Failure to detect evidence of an intracardiac shunt.

(3) Arterial unsaturation

Anomalous Venous Return with Pulmonary Vein Emptying into the Vena Cava or the Rigth Auricle:

The following are the catheterization findings in the above anomaly (9).

(1) The oxygen content of right auricle exceeds that of the superior vena cava.

(2) The pulmonary veins may be catheterized directly from the right auricle.

Group II Pulmonary Artery Flow Greater than Systemic Flow:

Eisnmenger's Complex:

This malformation is anatomically identical with the tetralogy of Fallot except that the pulmonic stenosis is absent. Eisenmenger's complex is extremely rare. Among approximately 800 patients with congenital heart disease of the cyanotic type studied in the Physiological Laboratories of the Department of Surgery of the Johns Hopkins Hospital, only 4 had this malformation (6).

Pulmonary hypertension is present and in some individuals, pulmonary artery flow exceeds systemic flow (6). However, the difference between the two rates of volume flow is uaually slight. The intracardiac shunt may be directed toward the left. The vascular resistance ofthe pulmonary bed is increased. As a result, the systolic and diastolic pressure components recorded from the pulmonary artery are elevated. (4, 17, 57).

Transposition ofthe Great Vessels:

Complete Uncorrected Transposition:

In this condition the aorta arises from the ventricle receiving systemic venous blood and the pulmonary artery arises form the ventricle receiving oxygenated blood. Blood pumped out by the left ventricle through the pulmonary artery to the lungs returns via the pulmonary veins to the left auricle. The aorta receives blood from the right ventricle, and the blood from the systemic circulation returns to the right auricle. In complete transposition the aorta arises anteriorly and the pulmonary atery posteriorly. The position of the ventricles, however, is not reversed. If complete transposition if to be compatible with life, other malformations have to exist

in order to make possible an exchange between the greater and the lesser circulation. These additional defects may be in the form of an auricular septal defect or in the form of a patent ductus arteriosus, which would have to conduct blood from the pulmonary artery into the systemic circulation and vice versa.

An example of catheterization findings will be illustrated by the results of 2 cases published by Campbell et at.(15).

(1) All four chambers of the heart were catheterized. (2) The oxygen content of the right auricular blood exceeded that of the superior vena cava by a significant amount. (3) The oxygen contents of left auricular and left ventricular blood were within 1.5 volumes per cent of one another. The oxygen saturation of peripheral arterial blood was 59 per cent. Right ventricular and left ventricular pressures were almost equal. The patient died following operation, and the necropsy findings showed complete transposition of the great vessels, ventricular septal defect, patent foramen ovale, and hypertrophy of the right and left ventricular chambers.

The results obtained from catheterization of the second cases: (1) The oxygen content of the right ventricular blood was notable higher than that of right auricular blood. (2) The aorta was catheterized through the right ventricle and the oxygen content of aortic blood was increased above that of the right ventricle. (3) The pressure in the right ventricle was elevated.

The studies revealed the presence of a ventricular septal defect. The observation that the oxygen content of the aortic blood was higher than that of right ventricular blood indicated the presence of a patent ductus arteriosus shunting blood from the pulmonary artery into the aorta. The postmortem

examination revealed the following findings: transposition of the aorta and the pulmonary artery, patent foramen ovale, patent ductus arteriosus, ventricular septal defect, hypertrophy and dilatation of both ventricles.

An interpretation of the physiologic data obtained in these 2 patients indicates that a diagnosis of transposition of the great vessels cannot be made from catheterization alone. Without advance knowledge of the nature of the malformation, a diagnosis of pulmonic stenosis with a septal defect or a patent ductus rather than of transposition of the great vessels would have been made. from catheterization. Nevertheless, catheterization contributes greatly to the understanding of the hemodynamics of this malformation, it allows accurate estimation of the volume and the location ofthe shunt in each individual patient. This latter question has become increasingly important because of recent promising advances in the surgery of this malformation by Blalock (8), consisting of the creation of an auricular septal defect combined with construction of an artificial ductus. Before such surgical procedure can be undertaken, it is important to ascertain whether or not there already exists an auricular defect and to determine the volume flow of blood that is shunted through this defect. The volume of the shunt in transposition is calculated in a manner different from that used in the tetralogy of Fallot or isolated septal defects. In the latter cases, for instance, the over all intracardiac shunt is unidirectional and may be computed as the difference between the volume of the systemic and the pulmonary blood flows. Equilibrium between the blood bolumes in the pulmonary and systemic circulation is maintained in these cases, since the amount of the shunted blood is returned to that side of the circulation from which it was originally diverted.

In transposition of the great vessels, on the other hand , a unidirectional shunt cannot exist continuously as it would lead to a progressive depletion of the circulating blood volume in either the pulmonary or the systemic circulation. The crucial hemodynamic alteration in transposition of the great vessesl is that blood shunted from the lesser circulation is that blood shunted from the lesser circulation or vice versa cannot return to that side of the heart whence it was diverted, unless there is an equivalent or opposite shunt. One must postulate, therefore, that in transposition of the great vessels, equal volumes of blood are reciprocally shunted from one circulation to the other(15).

Since the effective pulmonary blood flow is the volume of venous blood which, after its return to the right auricle, is eventually aerated in the lung, it must represent the right to left shunt. According to the considerations presented above, an equivalent amount of blood must be shunted from left to right. Thus the effective pulmonary blood flow represents the shunts between the greater and lesser circulations. The purpose of surgicaltreatment in transposition of the great vessels consists of increasing the effective pulmonary blood flow through the creation of a large auricular septal defect (62, 64).

Partial transposition of the Great Vessels:

In this malformation there is transposition of one of the great vessels while the other vessel originates normally. At this point only complete transposition of the aorta with levoposition and overriding of the pulmonary artery will be discussed. The recognition of this malformation is of importance since clinically it resembles Eisenmenger's complex. One case of this malform-

ation has been reported by Bing and Taussig (64). Catheterization revealed the oxygen content of the right ventricular blood to be significantly higher than that of the right auricle, indicating the presence of a ventricular septal defect. The oxygen content of the pulmonary arterial blood exceeded that of right ventricular by a sizable amount. A gradient of the magnitude noted between the oxygen content of right ventricular blood and pulmonary arterial blood could have been the result of a ductus or of a communication between the pulmonary artery and the left ventricle through a high septal defect, with a pulmonary orifice overriding the lower portion of the ventricular septum. Clinical findings, however, rendered the diagnosis of a patent ductus arteriosus unlikely.

The oxygen content of peripheral arterial blood was considerable less than that of pulmonary arterial and right ventricular blood. This finding suggested that the aorta originated from the right ventricle.

The hemodynamics in this malformation is similar to that in complete transposition of the great vessels. In both malformations a unidirectional shunt cannot exist continuously as it would lead to progressive depletion of the circulating blood volume in either the pulmonary or the systemic circulation. The intracardiac shunt is represented by the effective pulmonary blood flow.

Auricular Septal Defect:

The defect in the auricular septum occupies not in frequently a position close to the foramen ovale. In contrast to the latter, however, the opening is not covered by a valve or membrane. The defect is usually large, averageing 2 to 7 cm. in diameter.

Catheterization studies in this congenital anomaly are primarily those of (1) increase in oxygenated blood in the right auricle as compared to the superior vena cava and (2) ability in some cases to introduce the catheter through the defect into the left auricle. Dexter and his co-workers(21) have shown that in control patients the oxygen content of the right auricle has been observed to be as much as 1.9 volumes per cent higher than that of superior vena cava blood. It is apparent that a sizable defect can be easily over looked unless catheter can be placed in the streqm of arterial blood flowing from the left auricle to the right auricle. The auricular pressure is not helpful in diagnosing this condition.

Defect of the auricular septum is frequently associated with pulmonary hypertension(14), but Barber and his co-workers (3) found no pulmonary hypertension in 21 patients with auricular defects who had undergone cardiac catheterization.

Lutembacher's Disease:

Lutembacher's disease presents the findings of an auricular septal defect and mitral stenosis.

Catheterization studies reveal the following information: The oxygen content of the superior vena cava are markedly below that of the right auricle, whereas the oxygen content of the right auricle and pulmonary artery are about equal. The pressure in the right auricular chamber is below that in the pulmonary artery and the systemic flow is greatly below the pulmonary blood flow. The conclusions from the above data are: interauricular septal defect, slight increase in pulmonary arterial pressure, and a large left to right shunt.(9).

The pressure in the pulmonary artery is relatively normal; this is interesting since this finding contrasts with that observed in isolated mitral stenosis. It is apparent that the septal defect acts as an escape valve, reducing the pressure in the left auricle and the pulmonary vascular bed.

Ventricular Septal Defect:

In this malformation there is typically a small perforation in the ventricular septum. The defect is usually near the base of the septum and usually small resulting in minimal shunt and strain on the heart.

Catheterization studies by Handelman (36), Bing (9), Campbell (15), and Griswold (33) have revealed that in this condition the oxygen content of the right ventricle is markedly above that of the right auricle. Pressure changes offer little assistance.

Differential diagnosis between a patent ductus arteriosus and a high ventricular septal defect is sometimes difficult on the bases of catheterization studies alone, since some of the oxygenated blood from the pulmonary artery may regurgitate into the right ventricle, increasing the gradient between the oxygen contents of right auricular and ventricular blood.

Aortic Atresia with Patent Ductus Arteriosus:

At the time this paper was written only 17 cases of aortic atresia had been reported in the literature and no catheterization studies have been reported.

Single Ventricle with Both Vessels Originating from Rudimentary Outlet Chambers:

Single ventricle is always associated with a rudimentary outlet chamber which represents a persistent bulbus cordis. The rest of theright ventricle is usually completely atretic. Both tricuspid and mitral orifices lead into a main chamber. The following malformations are included in this category:

(1) Single ventricle with both vessels originating from a rudimentary outlet chamber.

(2) Single ventricle with the aorta arising from the rudimentary outlet chamber. The aorta arises anterior to the pulmonary artery; hence a transposition is present.

(3) Single ventricle with pulmonic stenosis.

(4) Single ventricle in which the pulmonary artery arises from a rudimentary outlet chamber.

The last two malformations have been discussed in a previous section. Catheterization studies have not been reported in the first malformation.

Single Ventricle with a Rudimentary Outlet Chamber from which the Aorta Arises:

Catheterization study in this malformation revealed (9) the oxygen content of ventricular blood markedly exceeded that of right auricular blood. Futhermore, the oxygen content of the pulmonary arterial blood was noticeably higher than that of right ventricular blood. The pressure in the ventricle was elevated and about equal to that in the pulmonary artery. The results of the catheterization were interpreted as indicating a transposition of the aorta with the pulmonary artery overriding a high ventricular septal defect. The possibility of a single ventricle was also entertained. Necropsy revealed a single large ventricle from which the pulmonary artery

arose and a small anterior rudimentary chamber from which the aorta arose. Both auricles emptied into the large posterior chamber.

Truncus Arteriosus:

In this malformation the pulmonary artery arises from a single vessel which overrides both ventricles. Physiologically this malformation resembles pseudotruncus arteriosus, which was discussed in a previous section. There have been no catheterization on this anomaly.

Patent Ductus:

In this there remains a communication between the greater and lesser circulation because the ductus fails to close after birth.

Catheterization studies have revealed the oxygen content of the pulmonary artery to be greater than that of right ventricular blood. This is due to the presence of pulmonic insufficiency which results in some of theoxygenated blood regurgitating into the right vantricle simulating the presence of a ventricular septal defect. The aortic flow is considerably greater than the systemic flow, the pulmonary capillary flow exceed the right ventricular out flow. Usually the flow through the ductus comprises about 40 per cent of the left ventricular output. Findings have been reported on patients in whom the pulmonary arterial pressure exceeded that in the systemic circulation and the blood through coursed from the pulmonary artery into the aorta. Studies in this condition have been done by the following workers: Bing, Vandam and Gray (4), Cournand, Baldwin and Himmelstein (19), and Levinson et al. (47). The latter believing that there is a specific local elevation in pulmonary systolic and diastolic pressure in patients with patent ductus arteriosus.

III Pulmonary Flow Equals the Systemic Flow at Rest and After Exercise:

Isolated Pulmonary Stenosis and/or Infundibular Stenosis:

Pulmonary stenosis may involve either the pulmonary valve, or theinfundibulum of the right ventricle, or both. When the valve is involved, the stenosis is usually produced by a dome-shaped diaphragm which has a small opening, a few millimeters in diameter in its center. The outlines of the valve leaflets can usually be seen within the diaphragm which has a small opening , a few millimeters in diameter in its center. Occassionally the stenosed leaflets are thickened and deformed, or bicuspid.

Since the development by Brock (13) of pulmonic valvotomy for valvular pulmonic stenosis and the subsequent development of techniques for correction of infundibular stenosis (11, 12) increasing importance has been attached to the preoperative differentiation of these conditions, since the type of operation depends on the type of obstruction found at the pulmonic valve.

During routine cardiac catheterization it is usually possible to enter the pulmonary artery even when a severe degree of pulmonic stenosis is present (21). If this be the case, the diagnosis of pulmonic stenosis can then be made by the demonstration of a high systolic pressure in the right ventricle and a low systolic pressure in the pulmonary artery. In all cases an attempt is also made to ascertain, it possible, the type of stenosis present. Fluoroscopic observation of the tip of the catheter and continuous monitoring of the pressure being transmitted through the catheter allow localization of the region in the cardiac shadow at which the change form the high right ventricular pressure to the low pulmonary arterial pressure occurs. Under ideal circumstances, it can also be determined if this change occurs

abruptly in the region of the pulmonic valve. If the latter can be demonstrated with certainty, it is an indication but not absolute evidence that the stenosis is of the valvular type.

In infundibular stenosis it is frequently possible, by careful manipulation of the catheter during its withdrawal from the pulmonary artery to the right ventricle, to detect an intermediate zone of pressure in the outflow tract of the right ventricle. This finding is characteristic of infundibular pulmonic stenosis and enables a positive diagnosis to be made. If, however, an intermediate zone of pressure is not detected, this does not necessarily exclude the possibility that infundibular stenosis may be present, for even in cases of typical infundibular stenosis, the tip of the catheter may traverse the infundibular zone so rapidly that the intermediate pressure fail to be recorded (42).

General findings in the cases of isolated pulmonic stenosis are as follows: There is not significant change in the oxygen content of blood samples obtained from any chamber of the heart or pulmonary artery; peripheral arterial blood is fully saturated, demonstrating the absence of a right to left shunt. There is no intracardiac shunt; the systemic effective and pulmonary artery flows are equal. There is a marked elevation of the right ventricular and auricular pressure (30). Silber it al. (53) found that patients with this malformation are able to maintain an adequate cardiac output at rest and with ordinary activity.

Coartation of the Aorta:

An infantile and an adult type of coarctation must be distinguished. In the

infantile type there is a diffuse narrowing of the aorta between theorigin of the left subclavian artery and the point of entrance of the ductus. In the adult type there is a localized constriction of the aorta which usually occurs abovethe point of entrance of the ductus arteriosus. In the infantile type of coarctation the constriction usually causes complete interruption of the aorta and prior to the development of collateral vessels, the circulation to the lower extremities is through a patent ductus. In the adult type of coarctation of the aorta, however, the ductus usually undergoes obliteration and the blood from the ascending aorta reaches the descending aorta through collateral vessels.

Adult form of Coarctation of the Aorta:

The most important physiologic diagnostic procedure in coarctation of the aorta consists in simultaneous optical recording of arterial pressures in the upper and lower extremities. The finding of a dampened pressure curve in the lower extremity and an increase in the delay of the pulse wave from arm to leg are suggestive of coarctation(9).

Infantile type of Coarctation:

No physiologic studies on patients with malformation have been reported.

Double Aortic Arch:

Double aortic arch has become of great clinical importance owing to the progress made in the surgery of this condition by Gross (35). The double aortic arch is the result of the persistence of both fourth brachial arterial arches; the arches fuse with the dorsal aorta to form the descending aorta. The resulting anomaly constitutes a vascular ring. Usually a

large right aortic arch passes behind the esophagus and trachea and connects with a small left aortic arch.

Catheterization studies have not been reported in this malformation.

CONCLUSIONS

It has been shown that cardiac catheterization can demonstrate various congenital heart conditions rather specifically due to changes found in oxygen content and pressure recordings obtained in the right heart. But just as it is true generally in medicine, that laboratory tests cannot provide the complete answer to out problems, so it is true of cardiac catheterization which must be interpreted in the light of the history, physicalfindings, X-Ray, and fluoroscopic data.

Some of the inaccuracies and ambiquities of cardiac catheterization must now be elucidated so that one may more intelligently evaluate the results obtained by this method. The fralities may be divided into three categories:

I Inaccurate recording:

The major source of error found in this grouping lies in the recording of pressures within the heart chambers. There is distortion, due primarily to the movements of the catheter; the main pressure distortion resulting from the use of the catheter are caused by the free damped vibration occurring within the catheter. They are started and maintained by the inevitable extrinsic influences to which the catheter is exposed during pressure determinations.

Although this source of error is not great enough to invalidate the data obtained, it nevertheless is factor that must be kept in mind.

II Changes Resulting in the Individual Caused by Cardiac Catheterization:

It must be quite apparent that an individual cannot go though such a procedure without some trepitation. The primary effect of this, physiologically speaking, will be upon the respiratory and heart rate. This factor is not significant if the recordings are taken while the person is on a plateau, for then constant base readings are obtainable.

III Lack of Specificity in the Method Resulting in Possible Inaccurate Interpretation:

It is within this category that we find the greatest pitfalls. Some examples will be cited.

(1) In cases of interauricular and interventricular septal defects the greatest normal variation in oxygen content between the superior vena cava and the right auricle is 2 volumes per cent; between the right auricle and right ventricle is 1 volume per cent. Since these variations may represent shunts of 1 to 2 liters it is apparent that a sizable auricular septal and ventricular septal defect may be overlooked unless the catheter can be placed in the stream of arterial blood flowing from left to right.

(2) As would be expected the tetralogy of Fallot can not be recognized in its entirety by study with catheterization. Right ventricular hypertrophy is perhaps best indicated by electrocardiography; dextro-position of the aorta is largely a morphological diagnosis and is functionally indistinquishable from a ventricular septal defect in studies with venous catheterization.

(3) The differential diagnosis between a patent ductus arteriosus and a

high ventricular septal defect is sometimes difficult on the basis of catheterization studies alone, since some of the oxygenated blood from the pulmonary artery may regurgitate into the right ventricle increasing the gradient between the oxygen content of the right auricular and ventricular blood which is normally found in interventricular septal defects.

(4) It is possible to confuse an interauricular septal defect with the rare anomaly of the pulmonary vein emptying into the right auricle since both of these conditions result in increased oxygen content in the right auricle.

(5) Even with comprehensive studies including cardiac catheterization it is often difficult if not impossible to definitively diagnose pseudotruncus, single ventricle and other extremely rare anomalies.

SUMMARY

In this paper there has been presented a comprehensive review of the literature relating catheterization findings in congenital heart disease. The categories that have been covered are as follows:

1. History of the development of this method.
2. Pertinent normal and abnormal physiologic findings obtained in catheterization.
3. The technique used in cardiac catheterization.
4. The specific application of cardiac catheterization in congenital heart disease.
5. Complications resulting from catheterization.

There has also been a discussion of the reliabilities and shortcomings of this method.

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