THE PULMONARY AND SYSTEMIC CIRCULATIONS IN CONGENITAL HEART DISEASE

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The application of cardiac catheterization to the study of the human circulation (Forssmann, 1929; Cournand and Ranges, 1941) provided a new technique for obtaining information about the pulmonary circulation and also for the study of the physiological effects of congenital heart disease (Bing *et al.*, 1947*a*; Dexter *et al.*, 1947*b*; Cournand *et al.*, 1949). Extensive studies have been made in which the pulmonary circulation in congenital heart disease has been investigated and in particular the pulmonary blood flow and peripheral resistance have been evaluated (for instance Bing *et al.*, 1947*b*.; Handelsman *et al.*, 1948; Hickam, 1949; Wood, 1950). It has long been known, however, for the systemic circulation that the elasticity of the arterial walls in relation to their *Windkessel* (pressure reservoir) function is of great importance as well as the peripheral resistance (Frank, 1899 and 1926; Broemser and Ranke, 1930; Wezler and Böger, 1939). Cournand (1947) has recently pointed out that the same consideration applies to the pulmonary circulation but so far no quantitative estimations of the " elasticity resistance " in the pulmonary circulation have been presented.

Cardiac catheterization used for the investigation of congenital heart disease yields data that may be used for the study of the systemic circulation as well as the pulmonary, so that it is possible to compare the effect of the cardiac abnormality on the two circulations although hitherto attention has usually been concentrated on the pulmonary circulation.

Formulae that give mathematical expression to the arterial *Windkessel* function are available and some are applicable to the data obtained by catheterization studies in normal subjects or in those with many forms of acquired or congenital heart disease. These or other formulae applied to the analysis of the changes occurring in the two circulations are capable of yielding valuable results enabling these changes to be studied with greater precision. From cardiac catheterization studies made in patients with suspected or proven congenital heart disease we have been able to make such an analysis in a variety of conditions and have confirmed the importance of the elasticity resistance in the pulmonary arterial system and of the interrelationship of the various components that determine the arterial blood pressure.

METHODS

The data used have been obtained by the catheterization technique described by Holling and Zak (1950). Resting oxygen consumptions have been measured in a Benedict spirometer on the fasting patients immediately before catheterization; blood samples have normally been taken from each of the superior vena cava, inferior vena cava, right atrium, right ventricle, and pulmonary artery, from other chambers or vessels if entered and from a systemic artery by puncture; blood oxygen contents have been estimated by the Haldane apparatus (Douglas and Priestley, 1948). Blood pressure records were obtained by a modified

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Hamilton manometer (Hamilton *et al.*, 1934) or by the Southern Instruments blood pressure recorder * using a Kelvin-Hughes pen recorder, both instruments having a resonant frequency of over 15 c.p.s. with a No. 8 cardiac catheter attached. Zero pressure levels have been taken approximating to the patient's mid-axillary line. Patients, if not anæsthetized, were sedated with phenobarbitone, grains 1–2 according to age and size. Anæsthesia, when used, was induced by rectal thiopentone supplemented by intravenous thiopentone if required, care being taken as far as possible to avoid undue fluctuations in the patient's condition.

The blood flows were calculated according to the Fick principle. Whenever possible the pulmonary arterial oxygen content has been used for mixed venous blood, but where left to right shunts were present samples proximal to the site of the shunt have had to be used. Where normal, the systemic arterial oxygen saturation has been substituted for the pulmonary venous oxygen saturation in calculating the pulmonary flow; where reduced on account of a right to left shunt a value of 96 per cent has been assumed for the pulmonary venous oxygen saturation. Where it was reduced and a pulmonary factor was suspected the arterial oxygen saturation while breathing oxygen has been determined; if this was then found to be normal the resting arterial oxygen saturation has been substituted for the pulmonary venous saturation.

All patients submitted to cardiac catheterization in whom pressures were measured by the Hamilton or S.I. manometer were considered, but strict criteria were applied in the selection of patients for analysis. The majority were necessarily rejected as unsuitable on various grounds; thus, all patients with unknown, possibly raised pulmonary venous pressure, those with shunts into the pulmonary artery or with suspected pulmonary regurgitation (for reasons given later), all patients in whom the pulmonary artery was not entered so that no pressure record was obtained, and all those in whom the record obtained was not sufficiently clear to indicate the incisura marking the division between systole and diastole (mostly those with pulmonary stenosis) and all patients where for any reason the results seemed doubtful or unreliable have been excluded. Finally, 24 patients were selected as suitable for study.

The following symbols have been used to define the various characteristics of the pulmonary or systemic circulations, co-existing in time, as obtained from the pressure curves and cardiac output data:

p_s = Systolic blood pressure in mm. Hg.
p_d = Diastolic blood pressure in mm. Hg.
P_p = Pulse pressure ($=p_s-p_d$) in mm. Hg.
p_m = Integrated mean pressure in mm. Hg.
V_m = Minute volume of pulmonary or systemic blood flow in cm.^{3}/min.
W = Peripheral flow resistance in dynes. sec./cm.^{5}
E' = Effective volume elasticity coefficient of the arterial system (=" elasticity resistance ") in dynes./cm.^{5}
The quotient E'/W, which is without dimensions.
S, D, and $S+D$ = The systolic, diastolic, and total pulse times respectively in σ ($=10^{-3}$ sec.).

In obtaining the data from the pressure curves the mean values obtained throughout a respiratory cycle have been used. The systemic arterial blood pressures were measured by a sphygmomanometer in the earlier investigations and by arterial puncture (brachial or femoral) in all the later.

Applying Poisseuille's law (analogous to Ohm's law and the basis of the so-called Aperia's formula) to the circulatory conditions in the pulmonary and systemic circuits, the total flow resistance is given by the equation,

$$W=(p_m-p_v)/c \quad \ldots\ldots\ldots\ldots\ldots \quad (1)$$

where p_m is the mean arterial pressure, p_v the mean venous pressure in dynes./cm.^{2}, and c the mean blood flow per second ($V_m/60$) in cm.^{3}/sec. In the systemic circulation, in normal conditions, the mean venous pressure does not exceed about 3–6 mm. Hg so that compared with the magnitude of the arterial pressure it may be neglected (Wezler and Böger, 1939) and p_m may be substituted for (p_m-p_v) in the above equation. Where the mean venous pressure exceeds 6 mm. Hg this approximation is not permissible and in some of our cases it has been necessary on this account to use the right ventricular diastolic pressure as a measure of the mean venous pressure, as pressures in the systemic veins have not been recorded. For the pulmonary circulation, in some cases of atrial septal defect or anomalous pulmonary venous drainage it is possible to make direct measurements of the mean venous pressure (Cournand *et al.*, 1947; Hickam, 1949), and in others the diastolic pressure in the left atrium can be determined or the right atrial diastolic pressure may be substituted as

* An instrument operating on the electrical capacitance change principle manufactured by Southern Instruments, Ltd., Camberley, Surrey.

an approximation; where, however, there is no such abnormality it is necessary to assume that the mean pulmonary venous pressure approximates to zero. In the presence of any condition, notably mitral stenosis, where the pulmonary venous pressure may be abnormally high such an approximation is not of course permissible.

The term W is expressed in absolute units in dynes. seconds/cm.^{5}, and represents the total effective resistance of the small arteries, arterioles, and capillaries. To obtain W in these units it is necessary to multiply the pressures measured in mm. Hg by the factor 1332 (representing the product of the sp. gr. of mercury and the acceleration due to gravity).

The mean pulmonary arterial blood pressure has been determined by integration of the pulse curve, as has been done by Cournand and his co-workers (Riley *et al.*, 1948) and by Hickam (1949). The basis of this method is illustrated in Fig. 1, and is that described by Wezler and Böger (1939), who used it to analyse

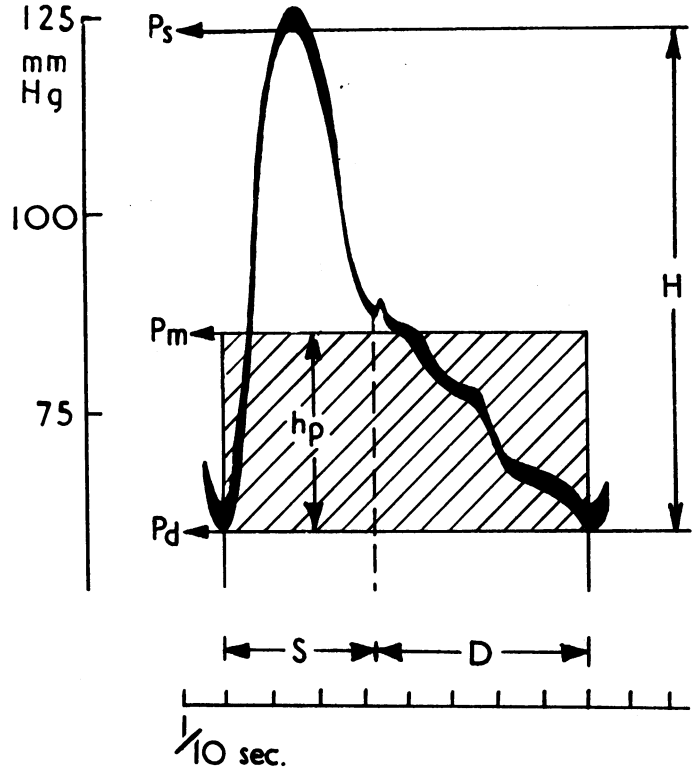

FIG. 1.—Diagram illustrating the method used for obtaining the mean arterial blood pressure by integration of the pulse curve. For full explanation see text.

systemic arterial pulse curves. The area of the pulse curve is measured by planimetry and the value so obtained is divided by the length $S+D$ to gain the value h_p, the height of the shaded rectangle the area of which represents the area measured. The mean pressure of the pulse curve is then obtained from the relation $h_p \times (p_s - p_d)/H$, where H is the height of the pulse curve and $(p_s - p_d)$ is the pulse pressure (i.e. the amplitude of the pulse curve in mm. Hg). The mean blood pressure in mm. Hg is the sum of this value and the diastolic pressure.

The quotient h_p/H gives the fraction of the pulse pressure that equals the mean pressure of the pulse curve. From values obtained by the above method in a series of pulse tracings we have found this quotient for the pulmonary arterial pulse curves to have a value of 0·398 with a standard deviation $\pm 0{\cdot}08$ (or expressed as a percentage about 40% $\pm 8\%$). It will be seen that the coefficient of variation is rather high so it is clearly preferable, for the pulmonary artery, to perform planimetry in each case rather than to use this value to calculate the mean pressure from the pulse pressure.

In many of our cases systemic arterial pulse curves were not available and in them the mean systemic pressures have been calculated from the sphygmomanometric figures, using the value of 42 per cent for the above quotient as found by Wezler and Böger (1939) from a large series of subclavian arterial pulse curves.

Otto Frank (1899) and Wezler (1938) in their theory of blood pressure, derived from consideration of a *Windkessel* model, have given an equation, applicable to steady conditions, relating the diastolic pressure to the various components determining it. These components are the mean blood flow per second (c), the effective volume elasticity coefficient (E') of the arterial system, the peripheral flow resistance (W) and the systolic and diastolic times.

$$p_d = \frac{cW(1-e^{S\times E'/W})}{1-e^{(S+D)\times E'/W}} \quad \ldots\ldots\ldots\ldots\ldots (2)^*$$

This equation is very difficult to solve for E', but Broemser and Ranke (1930) have obtained an approximation based upon the same considerations,

$$p_d = \frac{P_p \,.\, W}{E' \,.\, D} \quad \ldots\ldots\ldots\ldots\ldots\ldots (3)$$

In this equation it is possible to substitute for W, using equation (1), the expression $(p_m-p_v)/c$, and as $c=V_m/60$ the last equation can be written,

$$E' = \frac{(p_m-p_v)\times(p_s-p_d)\times 60}{p_d\times D\times V_m} \quad \ldots\ldots\ldots\ldots\ldots (4)$$

It is possible to use this equation to obtain values for E' by the use of pressure measurements and estimations of the blood flows obtained by independent methods such as are used in cardiac catheterization. The value of E', like W, is expressed in absolute units of the C.G.S. system; in calculating it, therefore, the pressures must be expressed in dynes./cm.^{2} and the diastolic time in seconds.

These formulae are based upon considerations of normal circulatory dynamics beyond the semilunar valves and are not therefore applicable in cases where there is either incompetence of these valves or an additional flow of blood through some abnormal channel into the arterial system. It is for this reason that we have had to exclude from analysis many patients in whom there was evidence or suspicion of pulmonary regurgitation or of a shunt through a patent ductus arteriosus or similar channel into the pulmonary artery.

Normal Values

In most cases of congenital heart disease there are abnormal features present affecting the pulmonary circulation. In order to analyse and assess the significance of the changes in the various factors upon which the pulmonary arterial pressure depends, it is necessary to have some knowledge of their normal values. It is known from the observations made by Riley *et al.* (1948), Stead *et al.* (1947) and Hickam and Cargill (1948) that large changes can occur in the volume of the pulmonary flow in various circumstances with only slight changes of pulmonary arterial pressure, which implies that the other factors controlling this pressure are themselves subject to large variations. Normal values must therefore be related to standard conditions and the resting or basal state is used throughout our study.

Adequate data concerning these normal values for the pulmonary circulation are not yet available and figures for the pulmonary blood flow and arterial pressure in normal subjects have been obtained only in a comparatively small number of persons, nearly all of whom have been adults. It is known from studies of the systemic circulation (Wezler, 1942) that the values of the factors involved vary with the age of the subject, and knowledge of the values in children is particularly scant. In order to assess the significance of our observations in congenital heart disease we had first to obtain some approximations for the normal values related to age. The figures we have adopted are given in Table I; values for intermediate ages have been obtained by interpolation.

Systolic and diastolic pressures in normal subjects over twenty years of age have been reported by Cournand (1947), Werkö (1947), Dexter *et al.* (1947*a*) and Lagerlöf and Werkö (1948). The figures we have accepted for subjects of twenty years and over represent the average of these, and using our observation on the relationship of the mean pressure of the pulse curve to the pulse

* e is the base of the natural or Napierian logarithm.

TABLE I

NORMAL VALUES FOR THE PULMONARY CIRCULATION RELATED TO THE AGE OF THE SUBJECT

Age in Years	5	10	20 and over
Systolic pressure (P_s) mm. Hg		23	
Diastolic pressure (P_d) mm. Hg		8	
Pulse pressure (P_p) mm. Hg		15	
Mean pressure (P_m) mm. Hg		14	
Pulse rate per minute	96	82	74
Diastolic time (D) σ	390	460	500
Pulmonary blood flow (V_m) cm.^{3}/min.	3000	4000	6000
Peripheral resistance (W) $\frac{\text{Dynes-sec.}}{\text{cm.}^5}$	400	300	200
Elasticity resistance (E') $\frac{\text{Dynes}}{\text{cm.}^5}$	1800	1200	700
$\frac{\text{Elasticity resistance}}{\text{Peripheral resistance}}\left(\frac{E'}{W}\right)$	4·5	4·0	3·5

pressure (i.e. mean pressure of pulse curve=40% of pulse pressure) we have calculated the value for the mean pressure given. As no published figures are available for children it has been necessary to assume the same values for all ages.

Cardiac outputs in normal resting adult subjects have been estimated using cardiac catheterization techniques by Cournand (1945), Cournand *et al.* (1945), McMichael and Sharpey-Schafer (1944), Stead *et al.* (1945), Werkö (1947) and Lagerlöf and Werkö (1948) in a total of over eighty subjects. The average value obtained from these figures is 6 l./min. which is 30 per cent higher than the average value for normal adults obtained by Wezler and Böger (1939) using physical methods. The latter have obtained average values for subjects aged 5 and 10 years and we have added 30 per cent to these to obtain comparable approximations for the normal outputs in children. As some justification for this procedure it may be noted that the values so obtained agree with the average values we have found in children for the systemic blood flow.

Using these basic figures we have calculated a normal value for the pulmonary peripheral resistance in adults=200 dynes. sec./cm^5. Lagerlöf and Werkö (1948) have recorded systemic intra-arterial pressures in adults by arterial puncture and using their average figures (130/80 mm.) we have calculated the corresponding value for the systemic peripheral resistance=1400 dynes. sec./cm^5. In normal subjects the systemic and pulmonary blood flows are equal so these values are comparable and it will be seen that the systemic and pulmonary peripheral resistances are in the relation 7 : 1.

In order to calculate the elasticity resistance (E') by the method used here it is necessary to know the diastolic time. Lagerlöf and Werkö (1948) give 74 as the resting pulse rate in their normal adults during catheterization; from this figure, using the relation $S/S+D=0{\cdot}375$ (an average value obtained from our pulse tracings in 21 subjects), we have calculated an approximate normal diastolic time (D)=500σ. Using this a normal value for the elasticity resistance (E') in the pulmonary arterial system a figure of 700 dynes/cm.^{5} has been obtained. Correspondingly, again using Lagerlöf and Werkö's figure for the systemic arterial pressure, the normal value obtained for the elasticity resistance in the systemic arterial system is 1700 dynes/cm.^{5} By their physical methods in normal adults Wezler and Böger (1939) obtained a value for this factor of 1800 dynes./cm.^{5} which agrees well with our figure. The systemic and pulmonary elasticity resistances are in the relation 2·4 : 1. It

follows from the different relations between the peripheral and elasticity resistances in the systemic and pulmonary circulations that the quotient E'/W (system.)=1·2 and E'/W (pulm.)=3·5.

Using the pulse rates given by Wezler (1942) for children of 5 and 10 years as a basis for calculating the diastolic time as above we have obtained the values given in Table I for the above-mentioned factors at those ages. For the systemic circulations we have used the blood pressure figures for children given by Fleisch (1927) as a basis for our calculations.

It is appreciated that many of these values are rather artificial but so long as the necessary observations on normal children are not available some such approximations are inevitable. When direct estimations have been obtained it will obviously be necessary to substitute these. Meanwhile the departures from normal that we shall consider are in most cases so large that even considerable errors in the normal values adopted would not affect the validity of our conclusions. In all cases we have elected, in accordance with the views of Rossier *et al.* (1950), to present the absolute values rather than to attempt a relation to body surface.

Results in Normal Hearts

All our patients have been studied on account of suspected congenital heart disease so that we are able to present results from only one subject where the findings seemed to indicate that the heart was normal.

Case 1. H. T. (No. P268), 16, m. Suspicion of congenital heart disease was raised on a soft systolic murmur in the pulmonary area, with a normal pulmonary second sound, found after a mass radiograph was said to show an abnormal shape of heart with a prominent pulmonary conus. There were no symptoms and no disability. Investigations here showed that the heart was displaced to the left by a sternal depression; the cardiogram showed right axis deviation with normal chest leads; cardiac catheterization did not reveal any evidence of intracardiac abnormality but the cardiac output at rest was increased with a normal oxygen consumption.

The values calculated for the pulmonary and systemic hæmodynamics are given in Fig. 2. In this and all subsequent similar figures the pulmonary circulation is represented on the left and the systemic on the right; the height of the columns represents the values of the various factors in terms of the percentage of the normal values for the age of the subject. In this figure absolute values are given above the appropriate column in each case. The cardiac output is increased (+65%), but in both circulations the peripheral and elasticity resistances are diminished so that the pressures are all within normal limits. Both the resistances in each circulation are equally affected so that the quotient E'/W is within normal limits in both. This case serves as an example of the alteration of the resistances in the normal subject, already referred to, which results in the accommodation of a change in the cardiac output without changes of the blood pressures, shown here to be occurring similarly in both circulations.

Results in Left to Right Shunts

We have been able to analyse the findings in eight patients with left to right shunts leading to an increased pulmonary blood flow. The results are given in Table II. There are two patients with atrial septal defect, two with anomalous pulmonary veins, and four with ventricular septal defect, one of these last possibly having an atrial septal defect also.

Atrial Septal Defect

Case 2. B. W. (No. O626), 7, m, came under observation for symptoms associated with a paroxysmal arrhythmia (? atrial flutter). His exercise tolerance was scarcely limited. Systolic and faint diastolic murmurs were heard at the apex; the pulmonary second sound was duplicated and increased. Radioscopy showed moderate cardiac enlargement due to the right ventricle with enlarged, pulsatile pulmonary arteries. The cardiogram showed no axis deviation in the standard leads but a notched R wave in V1. Cardiac catheterization demonstrated a shunt into the right atrium, proved due to an atrial septal defect by passage of the catheter into the left atrium.

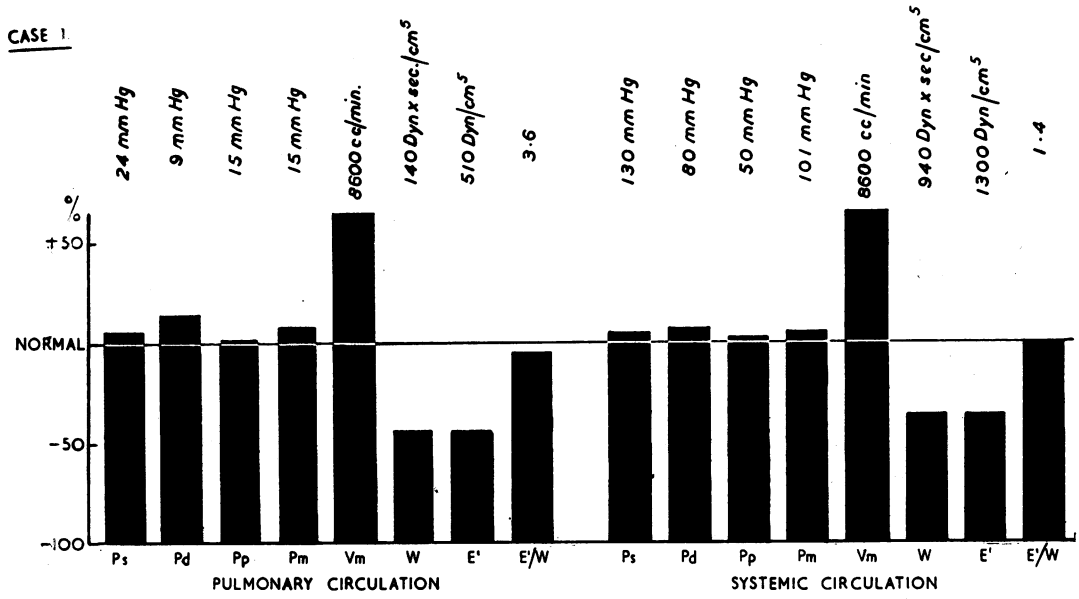

FIG. 2.—Diagrammatic representation of the hæmodynamic analysis in a patient with a normal heart. In this and subsequent similar diagrams the height of each column represents the observed value of the factor indicated by the symbol at its foot (for list of symbols see page 226) in terms of its percentage change from normal; in this case the absolute figures are also given above each column. The pulmonary circulation is shown on the left and the systemic on the right. Similar changes are present in both circulations; the minute volume (*Vm*) is increased but the peripheral (*W*) and elasticity (*E'*) resistances are so reduced that the blood pressures are within normal limits. Case 1.

In this patient the shunt is 7·3 l./min., leading to a pulmonary blood flow of 11·3 l./min.: the right atrial pressure is within normal limits, but the right ventricular systolic pressure is slightly elevated. An interesting feature is the small systolic pressure drop on passing into the pulmonary artery from the right ventricle (34 mm. dropping to 21 mm. Hg), which may be due to a " relative stenosis " produced by the dilatation of the pulmonary artery (Burchell *et al.*, 1950). Taking into account the patient's age the pulmonary arterial pressures are probably within normal limits although below our " normal " values; this, despite the increased blood flow, results from the extremely low peripheral resistance (*W* (pulm.)=60 units which is −83% of normal and the lowest value we have found in our series). The elasticity resistance is also greatly reduced (E'(pulm.)= 500 units or −68% of normal) so that the pulse pressure is not increased despite the high stroke volume (110 cm.³). The quotient E'/W (pulmonary) is high. In the systemic circulation the blood flow, pressures, and peripheral resistance are essentially normal with the elasticity resistance only slightly increased.

Case 3. J. R. (No. O291), 7, m., had negligible disability but examination had revealed a cardiac abnormality. There was a soft systolic murmur maximal at the apex; a diastolic murmur was not certainly heard. The pulmonary second sound was split but not loud. Radioscopy showed moderate cardiac enlargement mostly due to the right ventricle with enlargement of the pulmonary arteries and some pulsation near the hila of the lungs. The cardiogram showed some right ventricular preponderance (R.V.P.) with a notched R in V1. Cardiac catheterization revealed a shunt into the right atrium.

The systemic flow in this patient could not be satisfactorily estimated owing to a large difference between the oxygen contents of the two venæ cavæ and the impossibility of obtaining a true mixed venous blood sample in the presence of the shunt into the right atrium, so data are only available for the pulmonary circulation in which the blood flow is 12 l./min. As in the previous patient the right atrial pressure is normal, the right ventricular systolic slightly elevated and there is a small pressure gradient between the right ventricle and the pulmonary artery (37–30 mm. Hg systolic). The mean

pulmonary arterial pressure is normal, but the pulse pressure (25 mm. Hg or +67% of normal) is increased with a slight rise of the systolic and a considerable reduction of the diastolic pressure. These changes are accounted for by the low peripheral resistance and normal elasticity resistance, the quotient E'/W (pulmonary) being very high. The high pulse pressure reflects the normal elasticity resistance occurring with the large stroke volume of 128 cm.[3]

Anomalous Pulmonary Venous Drainage

Case 4. W. B. (No. O392), 12, m. Congenital heart disease was diagnosed at 6 years. Investigation had revealed some bronchiectatic changes in the left lower lobe, presumed the sequel of acute respiratory illness in infancy. There had been dyspnœa on exertion since early childhood. Clinical examination revealed dextrocardia, a systolic murmur and thrill over the præcordium (no diastolic murmur) and a loud pulmonary second sound. Radioscopy showed dextrocardia with situs inversus, no ventricular enlargement, but pulmonary arteries enlarged with moderate pulsation. Cardiac catheterization demonstrated a shunt into the venous atrium, and pulmonary veins were intubated entering this directly from the left lung.

The pulmonary blood flow in this patient is of the same order as in the preceeding two cases but here, in contrast, the pressures are increased. The systolic, diastolic, mean, and pulse pressures are all about double the normal, whereas the peripheral and elasticity resistances and the quotient E'/W (pulmonary) are not significantly changed from normal, so that the pressure changes are effected by the increased blood flow only. The systemic circulation is essentially normal in all respects.

Case 5. M.G. (No. H288), 8, f., had been listless as an infant and subsequently had some limitation of exercise tolerance. Congenital heart disease was diagnosed at 5 years. There was a loud systolic murmur and thrill at the apex and in the pulmonary area where the second sound was duplicated and loud. Radioscopy revealed enlargement of the right ventricle and also probably of the right atrium, with enlarged, pulsatile pulmonary arteries. The cardiogram showed R.V.P. Cardiac catheterization demonstrated a shunt into the right atrium and pulmonary veins draining into or near the lower end of the superior vena cava were intubated

Allowing for the age difference, the increase of the pulmonary flow in this patient relative to the normal value is similar to that in the last, but the mean pressure is much higher (p_m=+465% of normal) due to the high peripheral resistance (+106% of normal). Proportionately the diastolic pressure is more increased than the systolic so that the pulse pressure is less changed. This reflects the low elasticity resistance (−31% of normal), the significance of which is all the greater in the presence of the high pressure. The relation of this to the enlargement of the pulmonary artery will be considered later. Again the systemic circulation shows no significant changes from normal, apart from a rather high elasticity resistance.

Ventricular Septal Defect

Case 6. R. F. (No. O142), 12, m., had no disability and congenital heart disease was diagnosed on examination at 4 years. There was a systolic thrill and murmur in the pulmonary area with a diastolic murmur at the apex, and a loud pulmonary second sound. Radioscopy showed both ventricles to be enlarged with pulmonary arteries also enlarged and showing pulsation to the periphery of the lung fields. The cardiogram showed no definite preponderance. Cardiac catheterization demonstrated a shunt into the right ventricle.

The very large pulmonary flow is here associated with high values for the pulmonary systolic, diastolic, and mean pressures, these being due to the volume of the flow as the peripheral resistance is on the low side of normal. The greatly increased pulse pressure (+220%) similarly results from the high stroke volume as the elasticity resistance also is low. These relationships are well shown in Fig. 3A. In the systemic circulation the blood flow is normal and the other features are on the low side of normal.

Case 7. D. O'D. (No. O634), 11, f. Congenital heart disease had been diagnosed at 2 years on routine examination but later there was some dyspnœa on exertion. There were systolic and diastolic murmurs at the apex and the pulmonary second sound was greatly increased. Radioscopy showed cardiac enlargement

due to the right ventricle, great enlargement of the main pulmonary artery, a hilar dance, and pulsation to the periphery of the lung fields. The cardiogram showed no definite preponderance. Cardiac catheterization demonstrated a shunt into the right ventricle.

The pulmonary flow is even larger than in the last patient, being the largest we have found: the general pattern of the changes is the same except that the elasticity resistance is more nearly normal. Again the systemic circulation is essentially normal in all respects.

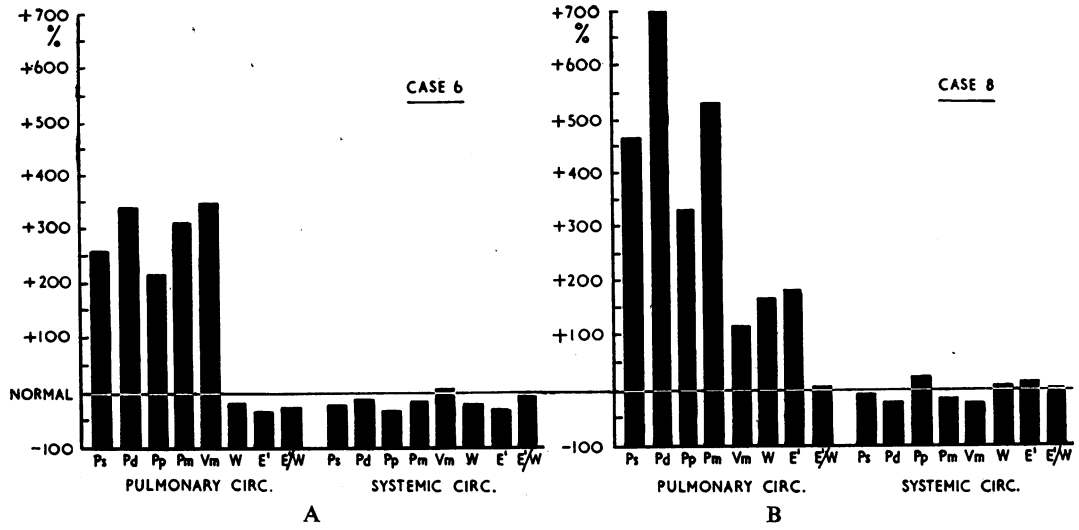

FIG. 3.—(A) Ventricular Septal Defect. The diagram illustrates the pattern of the hæmodynamic changes and shows that the pulmonary hypertension arises entirely from the increased blood flow (*Vm*), the resistances being reduced (Case 6). (B) Ventricular Septal Defect. In contrast to Case 6 the diagram shows that in this patient the pulmonary hypertension is due to an increase of the resistances (*W* and *E'*) as well as to an increased blood flow (Case 8). The systemic circulation is relatively unaffected in both cases.

Case 8. V. H. (No. O467), 48, f., had had recurrent attacks of bronchitis from childhood with some winter asthmatic attacks; dyspnœa on exertion had increased during the last three years. There was slight cyanosis; systolic and diastolic murmurs were widely heard over the præcordium with an increased pulmonary second sound. Radioscopy showed cardiac enlargement chiefly due to the right ventricle; the pulmonary arteries were greatly enlarged with considerable pulsation. The cardiogram showed some R.V.P. Cardiac catheterization demonstrated a shunt into the right ventricle; the arterial oxygen saturation at rest was 87 but rose to 100 per cent on breathing oxygen.

This patient shows a great increase in the pulmonary arterial pressures, the diastolic being relatively the most affected; these changes are the result of several factors as the blood flow, peripheral and elasticity resistances are all increased. The changes in the two resistances are of the same order so that the quotient E'/W (pulmonary) is normal. The systemic circulation yields normal figures. These relationships are illustrated in Fig. 3B.

Case 9. D. H. (No. O163), 3, m., was unwell from infancy having a persistent cough and dyspnœa on effort, although this seems to be decreasing as he grows. A systolic thrill was palpable all over the præcordium with systolic and diastolic murmurs best heard at the apex. Radioscopy showed considerable cardiac enlargement and enlarged pulmonary arteries with pulsation to the periphery of the lung fields. Cardiac catheterization demonstrated a moderately large shunt into the right ventricle and the oxygen content differences were suggestive but not decisive of a small shunt into the right atrium.

The pattern of the changes in the pulmonary circulation closely resembles that in Case 4 in type and degree, the increases in all the pressures being solely a reflection of the high blood flow. The systemic circulation is essentially normal although the elasticity resistance is slightly increased.

Summary of Findings in Left to Right Shunts

Surveying all the patients of this group three classes may be distinguished. All have increased pulmonary blood flows but some have normal pulmonary arterial pressures with low peripheral resistances. The rest have increased pressures but are further divisible into two types, those where the hypertension is purely an effect of the large flow and the peripheral resistance is low or normal, (exemplified by Case 6, Fig. 3A) and those where the hypertension is augmented by an increase in the resistance (exemplified by Case 8, Fig. 3B). The latter are distinguished by the relatively greater rise in the diastolic pressure.

Some of these patients have no disability but where this is present it is not associated with the largest pulmonary flows; study of the findings soon reveals that the best correlation is between disability and the magnitude of the pulmonary peripheral resistance; thus the two patients with the greatest disability (Cases 5 and 8) have peripheral resistances +106 and +170 per cent respectively whereas their flows are only +142 and +117 per cent respectively; on the other hand, for example, Case 6 who has no disability has a peripheral resistance −14 per cent but a flow +348 per cent.

The difference between the changes in the pulmonary and systemic circulations is well shown in Fig. 3 and Table II. The altered conditions of the pulmonary circulation are established independently of any change in the systemic and we shall show that this applies in other forms of congenital heart disease.

Results in Cases with Pulmonary Hypertension

Eisenmenger's Complex

Whatever the merits or demerits of arguments for considering Eisenmenger's complex to be a distinct anatomical entity (Selzer, 1949) we consider that it is a useful label for what appears to be a physiological entity. The criteria we have adopted for putting two of our patients into this group have been the demonstration of pulmonary arterial hypertension, with evidence of a right to left shunt confirmed by failure to obtain full arterial oxygen saturation on breathing oxygen. The findings in these patients are given in Table II.

Case 10. D. C. (No. P036), 44, f., was found to have congenital heart disease on examination at the age of 6 months but cyanosis was not apparent until her school years at the earliest. Disability was slight until her late twenties since which time dyspnœa and substernal pain had steadily become increasingly severe limiting factors. A diastolic thrill and murmur with a short systolic murmur were found to the left of the sternum. Radioscopy showed cardiac enlargement mainly due to the left ventricle with great dilatation of the main pulmonary arteries; pulsation was easily seen in these. Cardiac catheterization showed the criteria stated above.

The changes in the pulmonary circulation are illustrated in Fig. 4A. It will be seen that the greatest change in the pressures affects the diastolic value, a reflection of the great increase in the peripheral resistance (+800% of normal). The pulse pressure is also greatly increased although here, in contrast to the previous group, the stroke volume is small (38 cm.^{3}); this is accounted for by the very high elasticity resistance (+729% of normal). The quotient E'/W (pulmonary) is therefore normal. The systemic circulation has a normal flow and mean pressure, but the elasticity resistance and therefore the pulse pressure are rather low.

Case 11. D. L. (No. O625), 33, f., had about a year before admission a sudden onset of dyspnœa on exertion which steadily increased; cyanosis became apparent about three months later. There were no signs of cardiac failure. A systolic thrill with systolic and diastolic murmurs to the left of the sternum were present; the pulmonary second sound was increased and duplicated. Radioscopy showed cardiac enlargement mainly due to the left ventricle with pulsation in the large pulmonary arteries. Cardiac catheterization showed the same criteria.

The changes in the circulation are seen in Fig. 4B and the resemblance of the pulmonary circulatory pattern to that in the previous patient is obvious. The systemic pressures are normal although here the flow is slightly reduced and the resistances are raised.

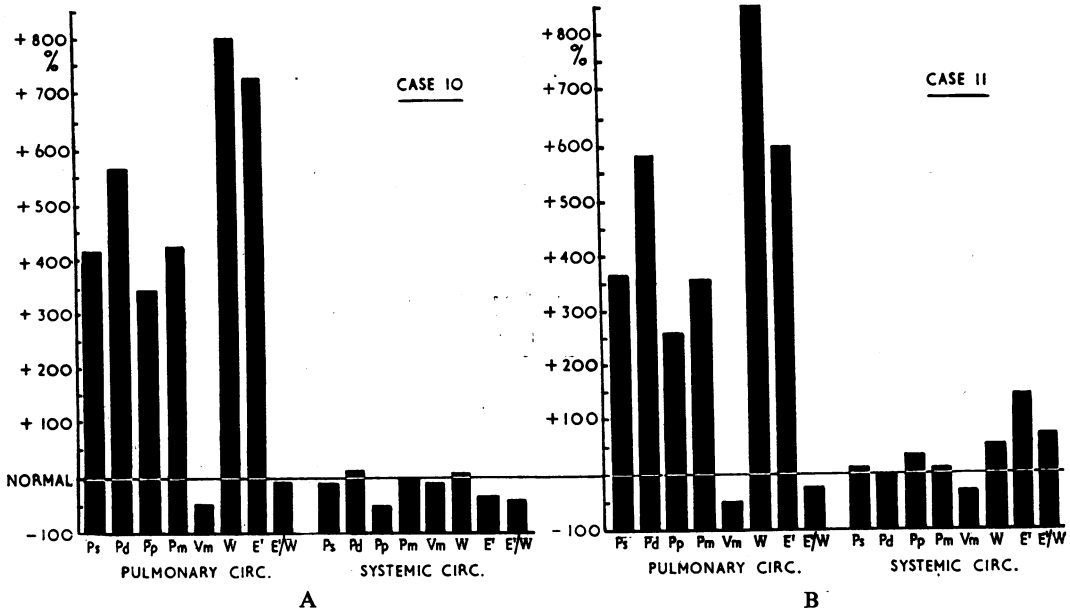

FIG. 4.—(A) Eisenmenger's Complex. The pattern of the changes in the pulmonary circulation shows clearly how the hypertension is due entirely to the increased resistances (*W* and *E'*) while the blood flow is greatly reduced. The normal mean systemic arterial pressure (*Pm*) and blood flow (*Vm*) are in marked contrast (Case 10). (B) Eisenmenger's Complex. The pulmonary pattern in this patient shows a striking resemblance to that in Fig. 4A. In the systemic circulation the reduced blood flow has been compensated for by changes in the resistances so that the pressures are normal (Case 11).

These patients show a remarkable resemblance to each other but stand in marked contrast with the previous group in that the pulmonary blood flows are reduced; the mechanism producing hypertension here is solely the increase in the resistances. In keeping with the previous group the high peripheral resistances are associated with greater disability and again the brunt of the physiological disturbance is borne by the pulmonary circulation.

Transposition of the Great Vessels

In only one patient, with partial transposition and a ventricular septal defect, were full data available for analysis. We have elected to present the results at this point for, as will be seen, the hæmodynamic changes lie intermediate between those of the preceding two groups.

Case 12. H. K. (No. O109), 13, m., had been cyanosed from birth and had suffered moderate disability due to dyspnœa on exertion. There was a loud systolic and a rumbling diastolic murmur to the left of the sternum; the pulmonary second sound was greatly increased. Radioscopy showed some cardiac enlargement mostly affecting the right ventricle; the pulmonary arteries were enlarged with a hilar dance. The cardiogram showed R.V.P. During cardiac catheterization both the aorta and pulmonary artery were entered, the oxygen saturation of blood in the latter being found the higher.

The pattern of the changes is shown in Fig. 5A. Like the two patients above the greatest change in the pulmonary arterial pressures affects the diastolic value (+563% of normal) and correspondingly the chief factor in producing the hypertension is the high peripheral resistance. The elasticity resistance also is increased but proportionately less so; so that E'/W (pulmonary) is low. Unlike the two patients above, but resembling the group with left to right shunts, there is an increase of the pulmonary blood flow, so that all the three possible factors are here involved in producing the hypertension. In common with both groups the increased pulmonary peripheral resistance is associated with some disability and the systemic circulation shows only minor changes from normal.

TABLE II

DATA AND RESULTS OF ANALYSIS IN CASES 2 TO 12

Case No.	Age and Sex	Diagnosis	Disability *	Shunt (cm.³ min.)	Pulse/ min.	Right atrial pressure (mm. Hg)	Right ventricular pressure (mm. Hg)	Pulmonary arterial circulation							Systemic arterial circulation						
								p_s (mm. Hg)	p_d (mm. Hg)	p_m (mm. Hg)	V_m (cm.³/min.)	W (Dynes. sec./cm.⁵)	E' (Dynes./cm.⁵)	E'/W	p_s (mm. Hg)	p_d (mm. Hg)	p_m (mm. Hg)	V_m (cm.³/min.)	W (Dynes. sec./cm.⁵)	E' (Dynes./cm.⁵)	E'/W
LEFT TO RIGHT SHUNTS																					
2	7 M	A.S.D.	O	7300	102	6/0	34/1	21 −8%	6 −25%	12 −14%	11,300 +232%	60 −83%	500 −68%	8·3	116 +15%	72 +6%	91 +11%	4000 +18%	1800 −7%	3600 +61%	2·0
3	7 M	A.S.D.	O	—	94	2/0	37/0	30 +30%	5 −38%	15 +7%	12,000 +253%	100 −72%	1500 −4%	15·0	—	—	—	—	—	—	—
4	19 M	P.V.A.†	I–	8000	82	—	55/0	50 +118%	15 +88%	30 +114%	14,500 +142%	170 −15%	800 +14%	4·8	115 −4%	65 −19%	86 −14%	6500 +8%	1100 −21%	1700 ±0%	1·6
5	8 F	P.V.A.†	II–	5700	125	18/6	100/13	97 +322%	65 +712%	79 +465%	8700 +142%	700 +106%	1100 −31%	1·6	105 −1%	65 −7%	82 −4%	3000 −17%	2200 +16%	4300 +92%	2·0
6	12 M	V.S.D.	O	15,100	84	12/8	78/8	83 +261%	35 +338%	58 +314%	19,700 +348%	240 −14%	740 −33%	3·1	95 −17%	65 −11%	78 −13%	4600 +5%	1360 −17%	1440 −27%	1·1
7	11 F	V.S.D.	O–I	19,200	105	5/0	90/5	93 +304%	40 +400%	63 +350%	23,600 +462%	200 −31%	1000 −13%	5·0	110 ±0%	75 +4%	90 +2%	4400 +5%	1600 −6%	1300 −32%	0·8
8	48 F	V.S.D.	? II	8700	96	10/0	124/2	130 +465%	64 +700%	89 +535%	13,200 +117%	540 +170%	2000 +186%	3·8	120 −8%	60 −25%	85 −15%	4500 −25%	1500 +7%	2000 +18%	1·3
9	3 M	V.S.D. ?+A.S.D.	O	5800	117	6/0	53/4	58 +152%	26 +225%	41 +193%	8000 +207%	410 −7%	2000 ±0%	4·9	90 +5%	60 +7%	73 +6%	2200 −15%	2700 +27%	5200 +66%	1·9
EISENMENGER'S COMPLEX																					
10	44 F		III	2100 (R to L)	88	8/2	118/0	120 +421%	53 +562%	74 +428	3300 −45%	1800 +800%	5800 +729%	3·2	115 −12%	90 +13%	101 +1%	5400 −10%	1500 +7%	1100 −35%	0·7
11	33 F		III	1200 (R to L)	94	22/6	110/16	108 +370%	54 +575%	71 +407	3000 −50%	1900 +850%	4900 +600%	2·6	145 +12%	80 ±0%	107 +7%	4200 −30%	2100 +50%	4200 +147%	2·0
TRANSPOSITION OF GREAT VESSELS																					
12	13 M		II	700 (overall) (L to R)	108	5/0	93/0 L.V. 100/0	94 +308%	53 +563%	74 +428%	6200 +35%	960 +256%	2100 +100%	2·2	100 −15%	68 −8%	83 −11%	5500 +11%	1200 −25%	1900 −5%	1·6

* Disability classified as described by Campbell, 1948.

P.V.A. = Pulmonary venous anomaly.

TABLE III

DATA AND RESULTS OF ANALYSIS IN CASES 13 TO 24

Case No.	Age and Sex	Diagnosis	Disability *	Arterial† oxygen saturation (%)	Pulse/ min.	Right atrial pressure (mm. Hg)	Right ventricular pressure (mm. Hg)	Pulmonary arterial circulation							Systemic arterial circulation						
								p_s (mm. Hg)	p_d (mm. Hg)	p_m (mm. Hg)	V_m (cm.³/min.)	W (Dynes. sec./cm.⁵)	E' (Dynes./cm.⁵)	E'/W	p_s (mm. Hg)	p_d (mm. Hg)	p_m (mm. Hg)	V_m (cm.³/min.)	W (Dynes. sec./cm.⁵)	E' (Dynes./cm.⁵)	E'/W
PRIMARY PULMONARY HYPERTENSION																					
13	25 M		Variable	96·6	68	12/3	100/10	102	45	67	4200	1300	2600	2·0	105	55	76	4200	1500	2100	1·4
				—				+643%	+462%	+378%	−30%	+550%	+272%		−19%	−46%	−25%	−30%	+7%	+24%	
14	19 F		III	72·6 (98·5)	126	10/3	85/10	88 +282%	56 +600%	68 +385%	4900 −16%	1100 +471%	2000 +167%	1·8	110 −8%	70 −13%	87 −10%	4900 −16%	1400 +5%	2700 +100%	1·9
15	22 M		III–IV ✠	90·2 (100)	91	26/12	125/17	124 +440%	76 +850%	94 +570%	1900 −68%	4000 +1900%	6800 +872%	1·7	120 ±0%	90 +13%	103 +6%	1900 −68%	4400 +214%	4100 +141%	0·9
PULMONARY STENOSIS (P.S.)																					
16	13 M	P.S. with L to R shunt	O–I	98·6	79	13/5	117/8	27 +17%	14 +75%	19 +36%	12,700 +176%	120 −56%	240 −77%	2·0	110 −8%	70 −5%	87 −6%	4800 +4%	1500 −6%	1800 −14%	1·2
17	23 M	Simple P.S.	I–	99·0	67	6/0	114/0	16 −30%	4 −50%	6 −57%	5400 −10%	90 −55%	470 −33%	5·2	130 ±0%	86 +8%	105 +4%	5400 −10%	1600 +14%	1400 −18%	0·9
18	22 F	„	O	98·2	91	5/0	60/4	21 −9%	8 ±0%	13 −7%	8700 +45%	120 −40%	470 −33%	3·9	130 ±0%	70 −13%	95 −6%	8700 +45%	900 −36%	1800 +6%	2·0
19	12 M	„	I–	97·8	120	12/5	180/0	17 −27%	7 −13%	10 −29%	4000 −9%	200 −28%	910 −17%	4·5	110 −4%	60 −18%	81 −10%	4000 −9%	1600 −2%	4300 +118%	2·7
20	15 F	„	O–I	95·2	78	13/5	83/0	25 +9%	11 +38%	16 +14%	6200 +24%	210 −16%	550 −42%	2·6	125 +2%	75 ±0%	96 +1%	6200 +24%	1200 −21%	1700 −13%	1·4
21	18 M	„	I(+)	98·5	92	16/3	150/8	50 +118%	13 +63%	27 +93%	5000 −11%	440 +100%	2600 +225%	6·0	125 +1%	70 −7%	93 −3%	5000 −11%	1500 +10%	2500 +37%	1·7
22	9 F	„	III	92·2	153	19/7	163/13	31 +35%	8 ±0%	17 +18%	2100 −45%	660 +113%	9300 +607%	14·1	110 +4%	70 ±0%	87 −1%	2300 −40%	2800 +57%	7800 +245%	2·8
23	16 M	Fallot's tetralogy	I(−)	86·0	100	9/6	125/6	34 +48%	8 ±0%	18 +29%	3700 −29%	390 +63%	4000 +345%	10·4	120 −4%	80 +7%	97 +1%	5700 +10%	1300 −14%	2000 ±0%	1·5
24	14 F	„	III–IV	87·1	103	13/6	132/11 Inf. ch. 54/8	27 +17%	13 +63%	18 +29%	4900 +2%	490 +89%	860 −14%	1·8	115 −7%	90 +22%	101 +6%	7400 +54%	1030 −35%	770 −66%	0·7

* Disability classified as described by Campbell, 1948.

† Figures in parenthesis indicate value whilst breathing oxygen.

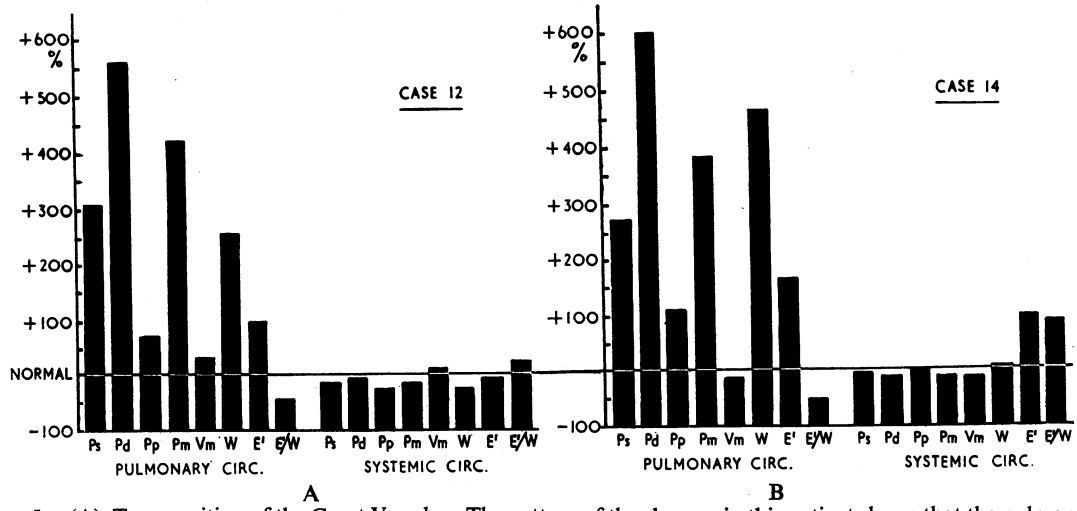

FIG. 5.—(A) Transposition of the Great Vessels. The pattern of the changes in this patient shows that the pulmonary hypertension is chiefly due to an increase of the peripheral resistance (*W*) but that the high blood flow (*Vm*) and elasticity resistance (*E'*) are additional factors. The essentially normal systemic pattern stands in marked contrast to the pulmonary (Case 12). (B) Primary Pulmonary Hypertension. The essential change in the pulmonary circulation is seen to be the hypertension associated with the increased peripheral resistance (*W*). Apart from the increased elasticity resistance the systemic circulatory pattern is essentially normal (Case 14).

Primary Pulmonary Hypertension

In three patients we have found pulmonary arterial hypertension where there was no left to right shunt and where the normal arterial oxygen saturation either ordinarily or whilst breathing oxygen excluded a right to left shunt. One of these (Case 15), reported fully elsewhere (Soothill, 1951), had no evidence of any pulmonary disease to account for these changes and would appear to be an example of primary pulmonary hypertension (Brinton, 1950) or idiopathic right ventricular hypertrophy (De Navasquez *et al.*, 1940). Another (Case 13) had a complex story of illness of obscure nature not obviously involving the lungs but showed some discreet mottling of unusual character in the lungs on X-ray; the pulmonary disease seemed insufficient to account for the hæmodynamic changes observed and may well have been coincidental. The third (Case 14) had more obvious pulmonary disease but the general picture again suggested that the circulatory changes were not entirely accounted for by the lung changes; while it is appreciated that cor pulmonale cannot be excluded we have included it here as an example of pulmonary hypertension without a cardiac malformation. The findings in these patients are given in Table III.

Case 13. D. K., 25, m., came under medical observation at the age of 20 years while in India when he was suspected of having ankylosing spondylitis and was also treated for tropical eosinophilia. At 22 years he first complained of dyspnœa and within a year first noticed œdema. His symptoms and his disability were variable, generally responding to treatment with rest in bed but relapsing on resumption of activity. When referred here there was an apical systolic murmur and an increased pulmonary second sound. Radioscopy showed great enlargement of the pulmonary arteries but with some pulsation only in the main branches; both lung fields showed scattered, sharply defined, probably calcified, miliary opacities. The cardiogram showed R.V.P. Cardiac catheterization revealed pulmonary hypertension but no evidence of any intracardiac abnormality.

The mean pulmonary arterial pressure is greatly increased (67 mm. Hg) with the systolic somewhat more affected than the diastolic, unlike the other cases in this group, so that the pulse pressure is considerably increased (57 mm.). These changes exist with a low minute volume, but a stroke volume (62 cm.^{3}) not much changed from normal because of a rather slow pulse rate. Although both the resistances are increased the chief change is in the peripheral resistance, so that the quotient E'/W (pulmonary) is low. The systemic circulation shows some hypotension, a reduced blood flow being associated with normal resistances.

Case 14. M. L., 19, f., had suffered from bronchitis and asthma from early childhood but at an early age had already shown cyanosis and dyspnœa on exertion and had had several attacks of right-sided heart failure before being investigated. No murmurs were heard and the pulmonary second sound was thought to be normal. Radioscopy showed moderate cardiac enlargement due to the right ventricle with great enlargement of both pulmonary arteries but pulsation only just seen in the main vessels. The cardiogram showed R.V.P. Cardiac catheterization revealed pulmonary hypertension and no evidence of a left to right shunt; the arterial oxygen saturation while breathing oxygen was normal.

The pattern of changes in the circulation is shown in Fig. 5B where will be seen the great increase of the pulmonary diastolic pressure (56 mm. Hg) reflecting the high peripheral resistance (+471% of normal), the blood flow being rather low. The elasticity resistance is less affected (+167% of normal), so that again the quotient E'/W (pulmonary) is low. The changes in the systemic circulation are similar to, but less than, those in the last patient, and the pressures are probably within normal limits.

Case 15. V. B., 22, m. (Soothill, 1951), was well until a year before his investigation when he began to complain of dyspnœa on exertion which increased rapidly and was followed nine months later by congestive failure. After treatment he improved and cardiac catheterization was performed but he died a few months later in a second attack of cardiac failure. No murmur was heard but the pulmonary second sound was increased and duplicated. Radioscopy showed considerable cardiac enlargement due to the right ventricle and large pulmonary arteries confined, as was the pulsation, to the more proximal branches. The cardiogram showed R.V.P. with S–T depression and inversion of T waves from V1–V6. Cardiac catheterization yielded results similar to those in the preceding patient. Post mortem, the thoracic viscera were examined by Dr. de Navasquez who reported that the findings, apart from some secondary changes in the larger pulmonary vessels, were those of idiopathic right ventricular hypertrophy (De Navasquez *et al.*, 1940).

The changes in the pulmonary circulation are the same as in the previous case but here are seen in extreme form, the great hypertension existing with a much reduced cardiac output (−68% of normal) so that the peripheral resistance is +1900 per cent of normal, the highest value found in our series. In the systemic circulation the low cardiac output is compensated for by increases in the resistances so that the pressures are nearly normal.

The three patients in this group have in common great pulmonary hypertension with reduced pulmonary blood flows, the major factor in the production of the hypertension being the great increase of the peripheral resistance. Two of them had evidence of associated parenchymatous lung changes in the form of central cyanosis abolished on breathing oxygen. There is some resemblance between this group and Eisenmenger's complex group, but here the elasticity resistance is relatively less increased and some at least of the increase observed is almost certainly secondary to the stretching of the vessel walls by the hypertension. The predominant change here is the increased peripheral resistance. This is associated in all the cases with disability and in Case 15 the observation of an extremely high resistance was shortly followed by death. In all three the cardiac output was reduced and this has led to some changes in the systemic circulation, but these are largely directed to maintaining normal pressures and are all trivial compared with the pulmonary changes.

Results in Pulmonary Stenosis

In nine patients with pulmonary stenosis the pulmonary arterial curves and other results were satisfactory for analysis. Six were examples of uncomplicated pulmonary valvular stenosis, i.e. patients in whom there was no evidence of a shunt at any site; only two of our many cases of Fallot's tetralogy yielded suitable curves. The remaining patient (reported in detail elsewhere, Deuchar and Zak, 1951) was an example of the combination of pulmonary stenosis with an increased pulmonary blood flow. The findings in these patients are given in Table III.

Pulmonary Stenosis with a Left to Right Shunt

Case 16. D. A. (No. P259), 13, m., was diagnosed as having congenital heart disease at 8 months but was very little disabled. There was a systolic thrill and murmur. On radioscopy the lung fields did not appear light but there was no abnormal pulsation. The cardiogram showed R.V.P. Cardiac catheterization

demonstrated pulmonary stenosis and also a left to right shunt (8·6 l./min.) which was thought probably to be due to anomalous pulmonary venous drainage into the right atrium.

The pattern of the changes in the circulation is shown in Fig. 6A, and apart from the pressure drop across the pulmonary valve (represented by the shaded column showing the right ventricular systolic pressure) it will be seen that it most closely resembles in form that shown in Fig. 3A illustrating a patient with a left to right shunt through a ventricular septal defect, although here the changes are smaller. The pressures in the pulmonary artery are slightly raised and this is due solely to the increased blood flow as the resistances are much reduced, especially so the elasticity resistance which is only 240 units (−77% of normal), giving a low value for the quotient E'/W (pulmonary). The systemic circulation is essentially normal in all respects.

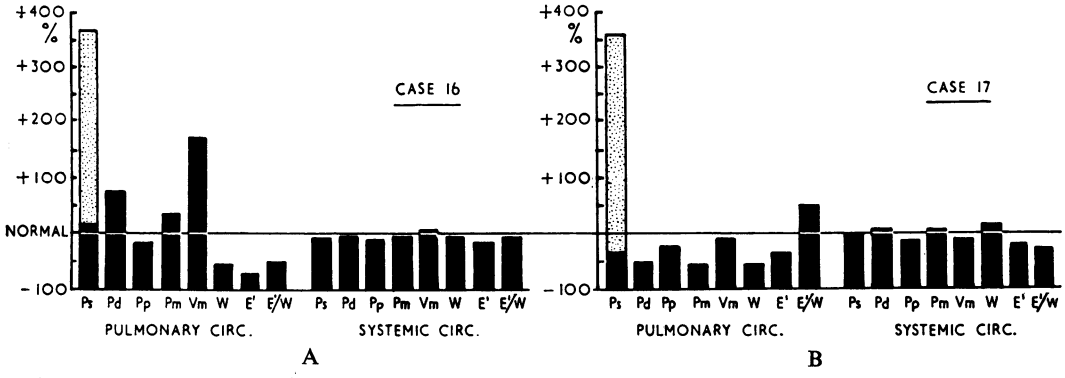

FIG. 6.—In this and the succeeding diagrams the dotted column above the pulmonary *Ps* column represents the right ventricular systolic pressure in terms of its change from normal; it gives some indication of the severity of the stenosis. (A) Pulmonary Stenosis with a left to right shunt. The increased pulmonary blood flow is here responsible for the high normal pulmonary arterial pressure despite the great reduction in the resistances. The systemic circulation is essentially normal. Comparison with Fig. 3A shows that the total pattern of the changes is the same, though less in degree in this case (Case 16). (B) Uncomplicated Pulmonary Stenosis. The pulmonary circulatory pattern shows how a normal blood flow is associated with low pressures and low resistances in this patient; again the systemic pattern is virtually normal (Case 17).

Uncomplicated Pulmonary Stenosis

Case 17. R. R. (No. P273), 23, m., had no symptoms until 14 years of age; dyspnœa on exertion was then noticed and had since increased slightly but when investigated he was still able to work as a farm labourer. There was a systolic thrill and murmur in the pulmonary area with a diminished pulmonary second sound. Radioscopy showed moderate cardiac enlargement due to the right ventricle and gross dilatation of the pulmonary trunk with pulsation visible in the dilated portion but not beyond. The cardiogram showed R.V.P. with T inversion from V1–V4. Cardiac catheterization demonstrated pulmonary stenosis with no evidence of any shunt.

In this patient there is a normal blood flow but definite pulmonary hypotension due to the low resistances, of which the peripheral resistance is the more reduced so that the quotient E'/W (pulmonary) is greater than normal. This pattern of changes is shown in Fig. 6B. The systemic circulation is again essentially normal.

Case 18. I. P. (No. H302), 22, f., was diagnosed in childhood as having congenital heart disease but had no disability even after an illness which seems to have been bacterial endocarditis. There was a systolic thrill and murmur in the pulmonary area. Radioscopy showed no general enlargement of the heart but a little fullness of the right ventricle with slightly dilated pulmonary arteries and some pulsation there. The cardiogram showed no definite evidence of ventricular preponderance. Cardiac catheterization revealed a mild degree of pulmonary stenosis with no evidence of a shunt.

The cardiac output is slightly increased in this patient but in both circulations the pressures are within normal limits as there is a compensatory reduction in the peripheral resistances. The rather

large stroke volume (95 cm.^{3}) is associated in the two circulations, however, with different changes in the pulse pressure; this is slightly reduced in the pulmonary but increased in the systemic reflecting correspondingly slight, contrasted changes in the elasticity resistances. The pattern in the pulmonary circulation closely resembles that illustrated in our normal case (Fig. 2).

Case 19. C. A. (No. P247), 12, m., had been known to have congenital heart disease for many years and complained of some tiredness on exertion but had little disability. There was a systolic thrill and murmur to the left of the sternum with a diminished pulmonary second sound. Radioscopy showed a normal size heart but gross dilatation of the left pulmonary artery. The cardiogram showed R.V.P. with T inversion from V1–V5. Cardiac catheterization demonstrated a severe degree of pulmonary valvular stenosis (p_s in the R.V.=180 mm. Hg, the highest we have recorded) with no shunt.

The cardiac output is within normal limits and the pulmonary circulatory pattern exactly resembles Case 17 above in type though the changes are slightly smaller. In the systemic circulation the only deviation from normal is a slight increase in the pulse pressure associated with increase of the elasticity resistance. It is interesting that a repeat study after pulmonary valvulotomy showed a considerable reduction of the right ventricular pressure but that in all respects the pulmonary and systemic circulation figures were unchanged.

Case 20. P. N. (No. O511), 15, f., first noticed some dyspnœa on exertion at the age of 10 years; this had not changed and she suffered little disability. There was a systolic thrill and murmur to the left of the sternum with a diminished pulmonary second sound. Radioscopy showed considerable enlargement of the right ventricle and prominence of the main pulmonary artery, its branches being small. The cardiogram was normal in the standard leads but the chest leads showed R.V.P. Cardiac catheterization revealed a moderate degree of pulmonary stenosis with no evidence of a shunt.

The cardiac output and the pulmonary arterial pressure although higher than the " normal " values are probably within normal limits. The peripheral resistance is on the low side of normal, but the elasticity resistance is somewhat more reduced giving a low value for the quotient E'/W (pulmonary). The systemic circulation is normal.

Case 21. J. F. (No. P206), 16, m., was diagnosed as having congenital heart disease at the age of 3 years when the murmur was heard but had no disability until about 8–10 years, when he got tired easily but was able to walk two miles. There was a systolic murmur and thrill to the left of the sternum with diminution of the pulmonary second sound. Radioscopy showed some enlargement of the right ventricle with pulmonary arteries within normal limits. The cardiogram showed R.V.P. Cardiac catheterization revealed severe pulmonary stenosis with no shunt. Two years later at the age of 18 years there was some suggestion of increasing disability and he had an attack of unconsciousness. There was no change in the physical signs. Cardiac catheterization was repeated (Table III) and showed only some change in the pulmonary arterial pressure.

At both catheterizations a degree of pulmonary hypertension was found to accompany the stenosis; the pressure tracing from the first catheterization was not satisfactory for analysis. The cardiac output, the mean pulmonary arterial pressure, and the peripheral resistance were essentially the same on each occasion, but the first time the diastolic pressure was relatively more elevated than the systolic, whereas the second time the reverse is the case, due to the increased elasticity resistance on this occasion. The pattern of changes seen at the second catheterization is illustrated in Fig. 7A. The systemic circulation was essentially normal on both occasions.

Case 22. C. H. (No. O468), 9, f., was found to have a murmur at six years but the time of onset of her disability was uncertain; dyspnœa on exertion had gradually increased and was moderately severe at the time of investigation. She also suffered from a chronic cough. There was a systolic thrill and murmur associated with a diminished second sound in the pulmonary area; rhonchi were heard in both lungs. Radioscopy showed considerable cardiac enlargement mainly due to the right ventricle; the pulmonary conus area was prominent but the branches of the pulmonary artery were small. The cardiogram showed R.V.P. Cardiac catheterization revealed a severe degree of pulmonary stenosis; the arterial oxygen was only just below normal and this was thought to reflect her pulmonary disease rather than to indicate a shunt.

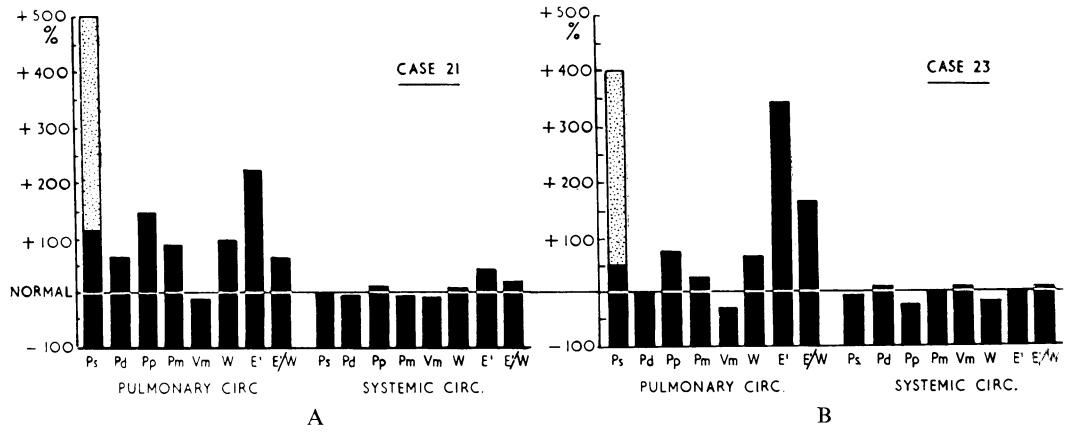

FIG. 7.—(A) Uncomplicated Pulmonary Stenosis. The pulmonary pattern in this patient contrasts with that in Fig. 6B; here the normal blood flow is associated with pulmonary hypertension due to an increase of the resistances, of which the elasticity resistance (E') is seen to be the more important (Case 21). (B) Fallot's Tetralogy. The pulmonary pattern here shows changes similar to those in Fig. 7A but with a reduced blood flow due to the right to left shunt and with the elasticity resistance change even more prominent (Case 23). In both cases the systemic patterns are essentially normal.

In the pulmonary circulation the mean arterial pressure is virtually normal with slight widening of the pulse pressure, but this is associated with a low blood flow so there is some elevation of the peripheral resistance (+113% of normal) and considerable increase of the elasticity resistance (+604% of normal), giving a high value for the quotient E'/W (pulmonary). The low blood flow and normal pressure in the systemic circulation are associated with similar but lesser changes in the resistances.

Fallot's Tetralogy

Case 23. D. S. (No. O168), 16, m., had been cyanosed from infancy and had dyspnœa on exertion but was not greatly disabled. There was a systolic murmur to the left of the sternum. Radioscopy showed a sabot-shaped heart (Fig. 8) with enlargement of the right ventricle and light lung fields. The cardiogram showed R.V.P. Cardiac catheterization findings were in keeping with Fallot's tetralogy, the right to left shunt being 3 l./min.

The pulmonary arterial diastolic pressure is normal but the systolic is raised, so the pulse pressure is increased despite a small stroke volume (38 cm.^{3}). This reflects a considerable increase of the elasticity resistance (+345%). The normal diastolic and slightly raised mean pressure with the low blood flow are associated with a slightly raised peripheral resistance (+63%). The quotient E'/W (pulmonary) is high. The systemic circulation is normal in all respects. These patterns are illustrated in Fig. 7B.

Case 24. D. J. (No. P142), 14, f., had cyanosis and some dyspnœa on exertion from infancy but had not been greatly disabled until 12 years old when, following what seemed a respiratory illness, she began to get cyanotic attacks and suffered considerable decrease in her exercise tolerance. There was a systolic thrill and murmur to the left of the sternum with diminution of the pulmonary second sound. Radioscopy showed some enlargement of the right ventricle with nothing remarkable about the pulmonary arteries. The cardiogram showed R.V.P. Cardiac catheterization findings were in keeping with Fallot's tetralogy (right to left shunt=2·5 l./min.) with a double pressure change between the right ventricle and the pulmonary artery suggesting the presence of both infundibular and valvular stenosis (confirmed at operation).

The pulmonary blood flow is normal and there is some increase of the pulmonary arterial pressure, chiefly affecting the diastolic level, so the peripheral resistance is increased (+89%). The elasticity resistance is slightly diminished and the quotient E'/W (pulmonary) is low. There is a normal mean systemic arterial pressure but a low pulse pressure associated with some increase of the blood flow: accordingly both the resistances are reduced, the elasticity the more so.

Summary of Cases with Pulmonary Stenosis

In this group the only feature common to all cases is a pulmonary stenosis, ranging in degree as can be seen from the right ventricular pressures in Table III, from moderate to severe. It is clear that there is no typical circulatory pattern associated with this owing to the large number of other variables in these patients. Despite the small number in this series and the complexity of the inter-relationships involved it seems to us that some correlations are suggested by our findings. The first case is unusual in having a left to right shunt associated with the stenosis but apart from that does not differ in any fundamental way from the rest of the group in whom the blood flows range from slightly raised to considerably diminished; so there seems no reason for not including this patient in our consideration of the group as a whole.

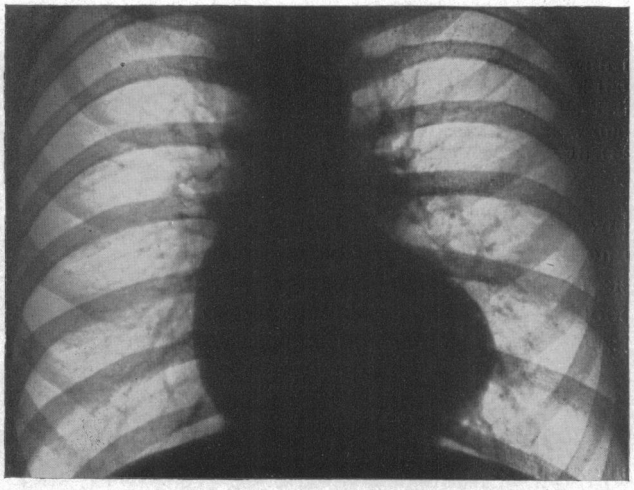

FIG. 8.—Fallot's Tetralogy. Plain radiograph of the chest showing the sabot-shaped heart and small pulmonary arteries associated with a high elasticity resistance (Table III and Fig. 7B). Case 23.

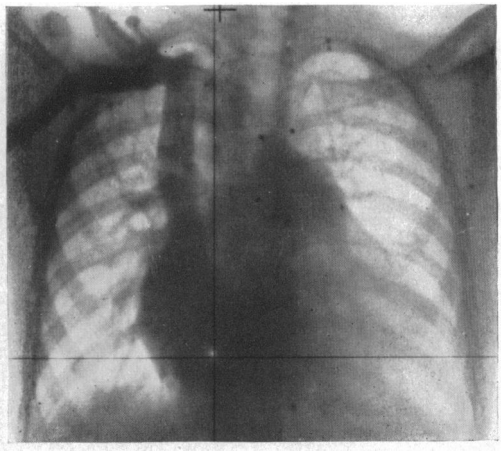

FIG. 9.—Pulmonary Stenosis with left to right shunt. Angiocardiograph showing the large pulmonary arteries associated with a low elasticity resistance (Table III and Fig. 6A). Case 16.

The pulmonary arterial pressures in the group range from the moderately increased (Case 21) to the considerably diminished (Case 17). In those with a high pressure, this is due in Case 16 to a high blood flow with low resistances, in Case 23 predominantly to high elasticity resistance, and in Case 24 to a high peripheral resistance; Cases 21 and 23 show some increase of both resistances. In all the other patients the resistances are low.

There is a range of pulmonary elasticity resistances from −77 per cent (Case 16) to +607 per cent (Case 22) of normal and comparing these values with the size of the pulmonary arteries as judged on the radiographs, radioscopy and the angiocardiographs we have observed that there is some correlation between dilatation of the pulmonary artery and a low elasticity resistance and vice versa. Thus in Case 16 the large pulmonary arteries shown on angiocardiography (Fig. 9) are associated with the lowest elasticity resistance; in Case 17 dilatation is again found with a low elasticity. Conversely in Case 23 (Fig. 8) the typical sabot shape due to small pulmonary arteries is associated with a high elasticity resistance. In Case 22, where the elasticity resistance was the highest in this group, although there is some prominence in the pulmonary conus area on the plain radiograph, the angiocardiograph shows that much of this is due to the ventricular enlargement and that, the pulmonary arteries are small, especially in the lung fields.

There seems to be no simple relation between the degree of stenosis and the disability (e.g. Case 19 appears to be one of the most severe stenoses but his disability is slight) which is scarcely sur-

prising in view of the other independent factors that, prima facie, would be expected to affect disability. In Table III, within the limits imposed by separating the different diagnostic groups, the patients with pulmonary stenosis are arranged in order from above down according to the magnitude (in percentage of normal) of the pulmonary peripheral resistance. Thus all cases from 16 to 20 inclusive have low peripheral resistances and it can be seen that all these patients have little or no disability; the remainder have increased peripheral resistances and the disabilities here are greater. With the close interdependence of blood flow and peripheral resistance it is not surprising that a somewhat similar correlation exists in this group between low blood flow and disability but we shall discuss later reasons for believing that the resistance is the more important factor.

The patients in this group, like those in the others, show predominantly normal patterns in the systemic circulation, independent of the type of pattern found in the pulmonary circulation. Even more striking is the close adherence of the mean systemic arterial pressures to the normal values; the range observed is only from -10 to $+6$ per cent of normal.

Discussion

To appreciate the significance of these observations it is necessary to understand how the various components interact to determine the actual blood pressure. Our knowledge in this field derives from the work of Frank (1899 and 1926) and his followers, e.g. Broemser and Ranke (1930), Broemser (1939), and Wezler and Böger (1939), in their physical and mathematical analysis of the mechanical constitution of the arterial system and the theory of blood pressure. The conceptions of the blood flow and the peripheral flow resistance and their interrelationships are well known and need no further description here. The role of the effective volume elasticity coefficient and its relationship to the *Windkessel* function of the main vessels is less known. The term itself is defined by the expression $E' = \triangle p / \triangle V$, where $\triangle p$ is the change of pressure inside the vessel resulting from changing its volume content by $\triangle V$. Its value is also given by the expression $E' = \kappa / V$, where κ is a measure of the elasticity of the vessel wall and V the volume content of the vessel (Frank, 1899 and 1926). It will be seen that E' is more than a simple expression of the elasticity of the vessel wall involving, as it does, also a consideration of the size of the vessel. We are here speaking of elasticity in its true physical sense, i.e. the greater the elasticity of the wall, as expressed by κ, the less the distensibility of the vessel so that the effective volume elasticity coefficient is a measure of the resistance of the vessel to distension. Hence our description of it as the " elasticity resistance."

The interrelationships of these components are naturally complex and have been discussed in detail by Wezler (1943) but briefly they can be summarized as follows. (1) An increase of the minute volume by itself results in an increase of diastolic and systolic pressures; the effect on the pulse pressure will depend upon the heart rate as this affects the stroke volume. (2) An increase of the peripheral resistance alone leads to a rise in the diastolic and systolic pressures but predominantly affects the former thereby reducing the pulse pressure. (3) An increase of the elasticity resistance alone leads to a fall in the diastolic pressure and an increase in the systolic thereby widening the pulse pressure, as has been demonstrated directly in heart–lung preparations (Knebel, 1941*a* and *b*).

All these statements presuppose a change of one of the components only but in practice any combination of changes may occur producing correspondingly modified alterations of the blood pressure. It is not possible, therefore, by a simple consideration of the pressures recorded to judge the changes, and the values of the components must be calculated in each case.

It has been possible hitherto to study these changes in the systemic circulation by means of physical methods based on the velocity of the pulse waves in the main arterial vessels and these methods have been applied extensively in normal persons and patients with systemic hypertension or a variety of circulatory disturbances (for full references see Wezler, 1943). The application of formulae derived by Frank (1926) and by Wezler and Böger (1939) and simplified by Broemser and Ranke (1930) to the data obtained during cardiac catheterization now gives us a way of studying these changes simultaneously in both the systemic and pulmonary circulations.

It is known from the work on the systemic circulation just referred to that the normal values for these components vary with age. At present factual data are only available for calculation of the normal pulmonary values in adults, so, as described, we have had to construct from a variety of sources some approximations for the values of childhood. From these calculations it is possible to give some quantitative expression to the well known differences between the two circulations (Cournand, 1947 and 1950). Normally, the minute volume of the two circulations is the same; it follows that the pressure differences between them must arise from differences of resistances. From our figures it can be seen that the greater difference is in the peripheral resistance (which for the systemic circulation is 7 times as large as for the pulmonary in adults), the difference in the elasticity resistances being only about one-third of this. This is shown in the important quotient E'/W with a value 3·5 for the pulmonary circulation as opposed to 1·2 for the systemic in normal adults. These resistances are not, however, fixed quantities and we are discussing values observed under resting or basal conditions; this is well illustrated by the figures obtained in our normal case (Fig. 2) where the high cardiac output has been accommodated in both circulations by equal changes in the two resistances so that the pressures are maintained within normal limits.

In our patients we have observed a wide range of variations from the normal circulatory patterns and many have shown pulmonary hypertension. In the patients with left to right shunts we have observed examples of this hypertension due solely to high minute volume (Fig. 3A) and others due partly to an increase of the resistances (Fig. 3B), but in all the increased flow is the chief factor. Other members of this group must not be overlooked who, like our normal case above, have accommodated the increased flow by reducing the resistances and so maintained normal pressures. By contrast the two patients with Eisenmenger's complex have pulmonary hypertension with small minute volumes and therefore due solely to changes in the resistances (Fig. 4A and B). The group with primary pulmonary hypertension similarly have hypertension due to an increase of resistances although here the elasticity is less affected (Fig. 5B). Our case of transposition of the great vessels (Fig. 5A) occupies a somewhat intermediate position. We have also observed slight pulmonary hypertension in a few patients with pulmonary stenosis and in these the responsible components are again the resistances, but from Fig. 7 and the values in Table III for Case 22 it is clear that the elasticity resistance is of greater importance here. Pulmonary hypertension in these patients may therefore result from a considerable variety of mechanisms.

In the pulmonary stenotic group we have already drawn attention to the inverse relation between the size of the pulmonary artery and the observed elasticity resistance; thus dilated pulmonary arteries have been observed in cases with very low values and, conversely, small arteries with high values. This important observed relationship agrees with the relationship expected from the definition of the elasticity resistance which, as already noted, is concerned with the vessel size in such a way that, other things being equal, the larger the vessel the less the elasticity resistance. The importance of the elasticity resistance in relation to the pulmonary arterial pressure in this group has just been noted, hence the importance of the vessel size. The qualification " other things being equal " in the statement concerning elasticity resistance and vessel size is important. If the vessel be distended by a high pressure the size will be large but the elasticity resistance may be large also owing to the increase of the factor κ when the vessel wall is stretched to near its elastic limit. This accounts for the significance we have attached in some patients to relatively low elasticity resistances occurring with considerable hypertension, and also for the absence of the simpler relation between elasticity resistance and vessel size in many of the earlier cases with pulmonary hypertension, although it commonly holds in the first group when hypertension is slight or absent.

The elasticity resistance must clearly play an important role also in the matter of pulsation of the pulmonary vessels as seen on radioscopy. We have endeavoured to investigate this in some selected cases that have already been discussed elsewhere by Campbell (1951), adding a few others from the pulmonary stenotic group for contrast. Campbell has concluded that increased pulmonary blood flow is the important factor and that raised arterial pressure although having some effect in producing visible pulsation is less important.

It is possible from our data to analyse the relationship more exactly. During systole, part of the volume of blood ejected from the ventricle flows directly into the peripheral circulatory bed, the rest serving to dilate the arterial tree from its diastolic volume; during diastole this latter portion is also driven into the periphery by the elastic recoil of the arterial wall; this is the essence of the *Windkessel* function of the arterial system. If the volume $\triangle V$ in the expression $E'=\triangle p/\triangle V$ is the volume just mentioned that dilates the arteries, then $\triangle p$ is the pulse pressure, available from the pressure records. E' is calculable and therefore it is possible to obtain $\triangle V$. This volume is a measure of the " storing capacity " of the *Windkessel* function of the arteries and is spoken of as the pulse volume, as opposed to the stroke volume of which it forms a part. The stroke volume (Vs) being also known it is possible to determine from the simple function $\triangle V/Vs$ the portion of it which is stored in the arterial tree in the way described. This value is related closely to the quotient E'/W and we shall discuss this elsewhere (Deuchar and Knebel, 1952).

The values we have obtained in 17 patients are given in Table IV together with the size of the pulmonary artery and the degree of pulsation seen on radioscopy as reported by Dr. Campbell.

The two examples of atrial septal defect (Cases 2 and 3) provide an instructive contrast; both have large and nearly equal stroke volumes yet in the former there was striking pulsation whereas in the latter it was seen only in the larger pulmonary vessels. From the table it can be seen that this is associated with a much larger pulse volume and " stored portion " in the former than in the latter ($\triangle V/Vs=0{\cdot}36$ and $0{\cdot}17$, respectively). This suggests that a large pulse volume or " stored portion " may be in some patients an important determining factor in producing visible pulsation. Further study of the table shows that all the patients in the anomalous pulmonary venous drainage and ventricular septal defect group have large pulse volumes and stored portions ($\triangle V/Vs$ ranging from $0{\cdot}30$ to $0{\cdot}56$) associated with considerable pulsation. (Case 9 has a large pulse volume with $Vs=21$ cm.^{3} as the age of the patient was only 3 years). In the pulmonary stenotic group there was one patient (Case 18) who showed some pulsation in the large pulmonary vessels. This patient stands in striking contrast to the other two with pulmonary stenosis in respect especially of the pulse volume, which is several times greater in the case with pulsation than in the others (Table IV); interestingly the pulse pressure in this patient was lower than in the others ($P_p=13$ and 26 or 23 mm. Hg, respectively).

Less easy to explain on the basis of the pulse volume is the pulsation seen in the Eisenmenger, transposition, or primary pulmonary hypertension groups for although the pulsation is generally somewhat less obvious it is none the less present with pulse volumes that are on the average considerably less than those we have so far considered. In all these cases, however, on account of the high peripheral resistance they have, the " stored portion " is high. It may be that this being so the rate of dilatation of the arteries in systole is greater and therefore more obvious on radioscopy although the amplitude may not be so great. It would be interesting to investigate this by means of electrokymographic studies.

It seems, therefore, that in many cases a large pulse volume is the chief factor in producing visible pulsation, as can be readily understood, but even when this is small a high " stored portion " appears by some less obvious mechanism to be important; as already noted a high " stored portion " often occurs when the peripheral resistance and hence the arterial pressure is high, so that this latter mechanism is probably the one concerned in patients showing visible pulsation in whom there is hypertension without an increased blood flow.

In all the diagnostic groups we have considered there are some patients who are disabled to a greater or lesser degree but the reasons why one patient should be disabled and another not are by no means clear. It is easy to visualize the mechanical effect of a pulmonary stenosis restricting the blood flow and producing disability, but, from our cases it is clear that the disability is not related to the severity of the stenosis alone (see Case 19 in Table III). The cause of disability in other groups such as those with left to right shunts is less apparent. We have noticed in the course of this study, and have already commented upon, a relationship that seems to exist between an increased pulmonary peripheral resistance and the presence of disability in all of these groups, even those with

TABLE IV

PULMONARY CIRCULATORY HAEMODYNAMICS AND APPEARANCES OF THE PULMONARY ARTERIES ON RADIOSCOPY *

Case No.	Age	Elasticity Resistance E' Dynes/Cm.^{5}	Peripheral Resistance W Dynes. sec./cm.^{5}	E'/W	Pulse Pressure (mm. Hg)	Stroke Volume Vs (cm.^{3})	Pulse Volume ΔV (cm.^{3})	Stored Portion $\Delta V/Vs$	Size of Right P.A.	Pulsation			
										Right P.A.	Middle Third	Peripheral Third	Hilar Dance
ATRIAL SEPTAL DEFECT													
2	7	500	60	8·3	15	110	40	0·364	++	++	++	(+)	(+)
3	7	1500	100	15·0	25	128	22	0·172	++	++	(+)	−	−
ANOMALOUS PULMONARY VENOUS DRAINAGE													
4	19	800	170	4·8	35	177	58	0·328	++	++	+	(+)	−
5	8	1100	700	1·6	32	69	39	0·566	++	++	+	+	−
VENTRICULAR SEPTAL DEFECT													
6	12	740	240	3·1	48	234	87	0·372	++	++	++	+	+
7	11	1000	200	5·0	53	236	71	0·301	++	++	+	+	+
8	48	2000	540	3·8	66	138	44	0·319	+(+)	+++	++	+	+
9	3	2000	410	4·9	32	69	21	0·304	++(+)	++	++	++	−
EISENMENGER'S COMPLEX													
10	44	5800	1800	3·2	67	38	15	0·395	++	++	++	+	+
11	33	4900	1900	2·5	54	32	15	0·468	++	+	−	−	−
TRANSPOSITION OF THE GREAT VESSELS													
12	13	2100	960	2·2	41	58	26	0·448	+++	++	+	(+)	−
PRIMARY PULMONARY HYPERTENSION													
13	25	2600	1300	2·0	57	62	29	0·468	+++	+	(+)	−	−
14	19	2000	1100	1·8	32	39	21	0·538	++	−	−	−	−
15	22	6800	4000	1·7	48	21	9	0·428	++(+)	++	+	−	−
PULMONARY STENOSIS													
18	22	470	120	3·9	13	95	37	0·390	+(+)	+	(+)	−	−
22	9	9300	660	14·1	23	14	3	0·214	−	−	−	−	−
23	16	4000	390	10·4	26	37	9	0·243	−	−	−	−	−

* The data relating to radioscopic appearances were kindly supplied by Dr. Maurice Campbell.

pulmonary stenosis. With these it is difficult to dissociate the possible influence of the pulmonary blood flow and of the peripheral resistance as these tend to be inversely related and prima facie in this group a reduction of pulmonary blood flow seems a likely cause of disability. In the group of left to right shunts, however, the position is somewhat different as here the blood flow is increased in all cases, but disability here also correlated best with high values of the peripheral resistance, and in the other groups the high peripheral resistance is probably the determining factor in producing a low blood flow.

We would suggest that a high peripheral resistance may reflect a loss of flexibility in the circula-

tory adjustments, which we have already seen (vide Case 1) are normally able to accommodate changes in blood flow such as are required on exercise, and that such loss of flexibility is an essential mechanism by which disability may arise whatever the anatomical abnormality present. Although the correlation we have observed has involved the peripheral resistance, this is probably only because it plays the predominant role in the regulating mechanisms and elasticity resistance changes may also affect the flexibility of adjustment. Our series has not, however, been sufficiently large to allow of investigation of this point which, in the light of our observations regarding the relation between vessel size and elasticity resistance, may be of importance in understanding the significance of post-stenotic dilatation of the pulmonary arteries in pulmonary stenosis.

Another feature that we regard as of fundamental physiological importance has been noted to be common to all groups and that is the difference between the changes observed in the two circulations. The nature of these differences has already been stressed in each group and is well shown in the figures illustrating the circulatory dynamics. In all cases the systemic circulation is essentially normal or any adjustments found are directed towards maintaining normality of arterial pressure especially and also of blood flow where possible. In contrast the pulmonary circulation may suffer large changes from normal and these can sometimes be interpreted as being necessary, in view of the cardiac abnormality present, to assist the maintenance of normal conditions within the systemic circulation. Our findings, therefore, would suggest that the regulatory mechanisms of the systemic circulation are predominant and that such regulation as exists in the pulmonary circulation is subordinate to the needs of the systemic.

SUMMARY

A method has been devised, using data obtained by cardiac catheterization studies and formulæ relating to the *Windkessel* (pressure reservoir) function of the arterial system, that enables the elasticity resistance as well as the peripheral resistance to be calculated for both systemic and pulmonary circulations.

Sets of normal values according to age have been calculated for both circulations. These figures give a quantitative expression to the differences between the systemic and pulmonary circulations.

The data obtained from cardiac catheterization studies on 24 subjects with congenital heart disease have been used and the findings are presented in groups according to the anatomical diagnosis; the patterns of the circulatory changes in these groups have been illustrated and described.

Pulmonary hypertension has been observed in many patients and the different mechanisms of its production have been discussed with reference to the observed findings. It has been shown that in congenital heart disease it may result from a variety of circulatory changes requiring full analysis for their differentiation.

In cases with pulmonary stenosis a relation has been observed between vessel size and the elasticity resistance in the pulmonary arterial system which is in agreement with the theoretically predicted relation and affects the circulatory dynamics.

The relation between the elasticity resistance and the occurrence of visible pulsation in the pulmonary arteries on radioscopy has been examined, and it has been shown that these are probably connected by the influence of the former on the pulse volume or "stored portion" of the stroke volume in conditions either of increased flow or high pressure.

A relationship has been observed in all groups between a high pulmonary peripheral resistance and disability; the significance of this has been discussed briefly.

In all groups the circulatory patterns in the two circulations have shown striking differences; the systemic circulation is always adjusted towards normal whatever the changes in the pulmonary circulation.

We wish to express our great indebtedness and thanks to Dr. Maurice Campbell for his encouragement and assistance in producing this paper and also for permission to report details of several of his patients. We also wish to thank Mr. R. C. Brock for the use of the case records of some of his patients and Dr. G. A. Zak for the catheterization results in Cases 8, 20 and 21. We are greatly indebted also to the Department of Medical Photography and Illustration, Guy's Hospital, for preparing the figures for publication.

REFERENCES

Bing, R. J., Vandam, L. D., and Gray, F. D., Jr. (1947*a*). *Bull. Johns Hopkins Hosp.*, **80**, 107 and 121.
——, ——, ——, (1947*b*). *Bull. Johns Hopkins Hosp.*, **80**, 323.
Brinton, W. D. (1950). *Brit. Heart J.*, **12**, 305.
Broemser, P. (1939). *Verh. dtsch. Ges. Kreislaufforsch.*, 289.
——, and Ranke, O. F. (1930). *Z. Biol.*, **90**, 467.
Burchell, H. B., and Wood, E. H. (1950). *Proc. Staff Meet. Mayo Clin.*, **25**, 41.
Campbell, M. (1948). *Guy's Hosp. Rep.*, **97**, 1.
——, (1951). *Brit. Heart J.*, **13**, 438.
Cournand, A. (1945). *Fed. Proc.*, **4**, 207.
—— (1947. *Bull. New York Acad. Med.*, **23**, 27.
—— (1950). *Circulation*, **2**, 641.
——, and Ranges, H. A. (1941). *Proc. Soc. Exper. Biol. Med.*, **46**, 462.
——, Baldwin, J., and Himmelstein, A. (1949). *Cardiac Catheterization in Congenital Heart Disease.* New York: Commonwealth Fund.
——, Motley, H. L., Himelstein, A., Dresdale, D., and Baldwin, J. (1947). *Amer. J. Phys.*, **150**, 267.
——, Riley, R. L., Breed, E. S., Baldwin, E. de F., and Richards, D. W. (1945). *J. Clin. Invest.*, **24**, 106.
De Navasquez, S. J., Forbes, J. R., and Holling, H. E. (1940). *Brit. Heart J.*, **2**, 177.
Deuchar, D. C., and Knebel, R. (1952). *Z. Ges. exp. Med.* In the press.
——, and Zak, G. A. (1952). *Guy's Hosp. Rep.*, **101**, 1.
Dexter, L., Haynes, F. W., Burwell, C. S., Eppinger, E. C., Sagerson, R. P., and Evans, J. M. (1947*a*). *J. Clin. Invest.*, **26**, 554.
——, ——, ——, ——, Sosman, M. C., and Evans, J. M. (1947*b*). *J. Clin. Invest.*, **26**, 561.
Douglas, C. G., and Priestley, J. G. (1948). *Human Physiology.* 3rd Edition. Oxford: Oxford Univ. Press.
Fleisch, A. (1927). *Handbuch der normalen und pathologischen Physiologie*, **7**, part 2, 1273. Edited by Bethe and Emsden. Berlin: Springer.
Forssman, W. (1929). *Klin. Wschr.*, **8**, 2085 and 2287.
Frank, O (1899). *Z. Biol.*, **37**, 483.
—— (1926). *Z. Biol.*, **85**, 91.
Hamilton, W. F., Brewer, G., and Brotman, I. (1934). *Amer. J. Phys.*, **107**, 427.
Handelsman, J. C., Bing, R. J., Campbell, J. A., and Griswold, H. E. (1948). *Bull. Johns Hopkins Hosp.*, **82**, 615.
Hickam, J. B. (1949). *Amer. Heart J.*, **38**, 801.
——, and Cargill, W. H. (1948). *J. Clin. Invest.*, **27**, 10.
Holling, H. E., and Zak, G. A. (1950). *Brit. Heart J.*, **12**, 153.
Knebel, R. (1941*a* and *b*). *Arch. exp. Path. Pharmak.*, **197**, 485 and 496.
Lagerlöf, H., and Werkö, L. (1948). *Acta. Phys. Scand.*, **16**, 75.
McMichael, J., and Sharpey-Schafer, E. P. (1944). *Brit. Heart J.*, **6**, 33.
Riley, R. L., Himmelstein, A., Motley, H. L., Weiner, H. M., and Cournand, A. (1948). *Amer. J. Phys.*, **152**, 372.
Rossier, P. H., Maier, C., and Volkmann, M. (1950). *Cardiologia*, **15**, 147.
Selzer, A. (1949). *Arch. intern. Med.*, **84**, 798.
Soothill, J. F. (1951). *Guy's Hosp. Rep.*, **100**, 232.
Stead, E. A., Jr. Hickam, J. B., and Warren, J. V. (1947). *Tr. Ass. Amer. Physicians*, **60**, 74.
——, Warren, J. V., Merrill, A. J., and Brannon, E. S. (1945). *J. Clin. Invest.*, **24**, 326.
Werkö, L. (1947). *Acta. Med. Scand. Suppl.*, **193**, 1.
Wezler, K. (1938) *Z. Biol.*, **98**, 438.
—— (1942). *Z. Altersforschung*, **3**, 199, and **4**, 1.
—— (1943). *Wein Arch. Inn. Med.*, **37**, 226.
——, and Böger, A. (1939). *Ergebn. Physiol.*, **41**, 292.
Wood, P. (1950). *Brit. med. J.*, **2**, 639 and 693.