and other functions substances like anthisan, procaine, and quinidine are inhibiting the effect of the local hormones.

Summary

Many substances used medicinally as local anaesthetics, as analgesics, and as spasmolytics have common properties, among which is included the ability to act like quinidine on the heart. Antihistamine compounds also belong to this large group of drugs. The side-effects they produce are therefore to be expected. They can also be used for other purposes—as local anaesthetics, as quinidine substitutes for fibrillation, as quinine substitutes for myotonia, for travel sickness, and to relieve pain. Similarly, local anaesthetics such as procaine, or analgesics such as pethidine, have an antihistamine action which is considerable.

The basis of these common properties is that the substances which possess them depress the effects of acetylcholine, histamine, and adrenaline; these substances control activity locally in many tissues.

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CONGENITAL HEART DISEASE*

A REVIEW OF ITS CLINICAL ASPECTS IN THE LIGHT OF EXPERIENCE GAINED BY MEANS OF MODERN TECHNIQUES

BY

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[WITH SPECIAL PLATE]

PART II

Atrial Septal Defect

There were 35 cases of A.S.D.—12 male and 23 female. Their ages ranged from 5 to 61, eight being under 14, six being adolescent, and 21 adult. There were no symptoms in 20, and only slight breathlessness or difficulty in keeping up with others in 10. Of the remaining five, two had Lutembacher's syndrome and were in congestive failure; one had severe hypertension resulting from polycystic kidneys, and two had considerable limitation of cardiac reserve. Only one had haemoptysis.

The physical signs included a small or normal pulse, a normal or high normal jugular venous pressure without exaggeration of the *a* wave, a tapping cardiac impulse, a lifting right ventricular outflow tract, visible or palpable pulsation of the pulmonary artery, a pulmonary systolic murmur with or without thrill, a pulmonary diastolic murmur, and wide splitting of the second heart sound at the pulmonary area without accentuation of the pulmonary element.

A thrusting hyperdynamic cardiac impulse, as described by Roesler (1934), simulating the left ventricular thrust of patent ductus or V.S.D., was felt in five cases and was attributed to a grossly overfilled right ventricle forming the apex beat of the heart. A lifting right ventricular outflow tract or palpable right ventricular conus could be appreciated in the third and fourth intercostal spaces to the left of the sternum in most cases, and explained the precordial bulge of the chest wall that might be seen in this situation. Visible or palpable pulsation of the pulmonary artery in the second left space was also common; but neither this nor the palpable conus was well documented. Precise figures for their incidence are therefore not given.

Although the systolic murmur was classed as pulmonary, being usually best heard in the second space, it was maximal in the third space in six instances. It was accompanied by a thrill, usually rather faint, in nine. In four cases there was a loud mitral systolic murmur and in eight a mitral diastolic murmur; but only two of these had other evidence of Lutembacher's syndrome. Nevertheless, all mitral murmurs in A.S.D. were attributed to mitral valve disease, presumably rheumatic.

Functional pulmonary incompetence occurred in half the cases. In respect of the size of the shunt there was no difference between those with a Graham Steell murmur and those without.

Noticeable splitting of the second heart sound was characteristic and was heard in practically all cases. The split was usually distinctly wider in A.S.D. than in normal controls, but there was some overlap between the two. The intensity of the pulmonary element was commonly normal, not accentuated. The wide split was attributed to right bundle-branch block or to delay in the emptying-time of a grossly overfilled right ventricle.

The electrocardiogram showed the pattern of partial or well-developed right bundle-branch block in all but two cases, and the S wave in Lead V_1 was never conspicuous (under 5 mm.). The secondary R wave in Leads V_1 or V_2 was rarely very tall. The only case that showed a really high R wave had a considerable degree of pulmonary hypertension.

Skiagrams revealed right ventricular enlargement, dilatation of the pulmonary artery, and pulmonary plethora in varying degree in all cases, as described by Roesler (1934) and by Bedford, Papp, and Parkinson (1941). Skiagrams in the antero-posterior view were often difficult to distinguish from those of V.S.D. or patent ductus; but in the second oblique position the chamber responsible for the enlargement could usually be recognized.

Cardiac catheterization was undertaken in 25 cases (Table IV). The mean pulmonary artery pressure was

TABLE IV.—*Atrial Septal Defect*

Case No.	Age	Mean Pul. Art. Pressure (mm. Hg)	O_2 Saturation			Ratio of Pul. to Systemic Flow	Pul. Resistance $\left(\frac{\text{P.A. Pressure}}{\text{Pul. Flow}}\right)$
			S.V.C.	R.A.	P.A. R.V.		
1 H	16	13	70	83	85	2·5:1	1·1d
3 1	24	18	68	80	81	2·5:1	1·4
4 9	36	12	58	80	81	2·7:1	0·9d
5 11	16	8	63	82	81	2·5:1	0·6d
6 24	51	16	59	81	82	3:1	1·0d
8 44	14	13	62	—	76	1·8:1	1·5
10 47	6	10	64	78	83	2·5:1	0·8d
11 63	12	17	67	76	78	1·6:1	2·0
12 78	35	6	71	81	82	1·8:1	0·7d
13 79	12	64	52	70	68	1·6:1	8·0*
14 95	18	13	62	85	85	3·2:1	0·9d
15 97	16	14	68	86	90	4:1	0·7d
16 109	14	7	71	81	89	3·5:1	0·4d
17 111	5	10	69	85	85	3:1	0·7d
18 118	29	6	65	88	90	4·5:1	0·3d
19 148	53	12	67	82	82	2·3:1	1·0d
20 149	5	13	69	86	87	4:1	0·7d
21 150	30	6	71	88	88	3·2:1	0·4d
23 159	21	18	63	81	84	3·3:1	1·4
25 162	18	13	73	82	85	2:1	1·3

* Increased pulmonary resistance. d = Diminished pulmonary resistance.
(No attempt to enter the pulmonary artery was made in 5 cases; these have been omitted from the Table).

*Conclusion of the St. Cyres Lecture delivered on June 13 at the Royal Society of Medicine under the auspices of the National Heart Hospital. Part I appeared in last week's issue.

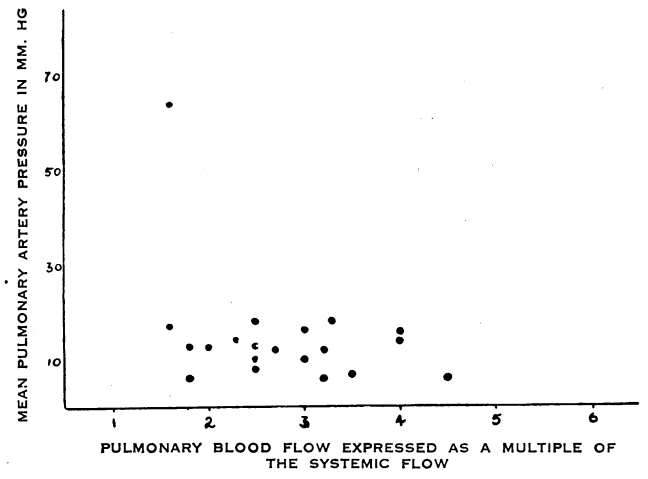

FIG. D.—Graph showing the relationship of the pulmonary blood flow to the pulmonary artery pressure in A.S.D.

usually normal or nearly so, and the peripheral resistance in the lungs was usually diminished (Fig. D). This explains why the right auricular pressure does not rise, why the clinical and electrocardiographic signs point to right ventricular dilatation rather than hypertrophy, and why the pulmonary element of the second heart sound is not accentuated.

The pulmonary blood flow was two to three times the systemic flow in 10—that is, about 10 to 15 litres a minute. In five cases it was a little less, and in eight cases more.

Reversal of the shunt was not demonstrated in any case in this series. But the stage was set for such a development in two patients. One of these had considerable pulmonary hypertension for no known reason, and one had slight pulmonary hypertension associated with severe emphysema. Two other cases, not included in this study, came to necropsy with central cyanosis and lowered arterial oxygen saturation. Both had reversed interatrial shunt resulting from the effects of pulmonary hypertension associated with emphysema in one case and with cystic disease of the lungs in the other. It is believed that reversal of the shunt in A.S.D. is rare in uncomplicated cases, even when the heart fails; it is suggested that an increased peripheral resistance in the lungs is more likely to be responsible, and that this is an accidental association, not even an indirect consequence of the shunt. Such pulmonary hypertension may operate not only by raising the right auricular pressure but also by lowering the left (see under Eisenmenger's syndrome).

Differential Diagnosis

Pulmonary hypertension, especially when complicated by pulmonary incompetence, is too often confused with A.S.D., and with too little reason. There are many points of difference. In pulmonary hypertension the jugular venous pressure tends to be raised and the *a* wave may be conspicuous; the second heart sound is split, but not widely so, and the pulmonary element is grossly accentuated; the electrocardiogram usually shows a prominent P pulmonale and tall R waves in Lead V_1 without widening or much notching; skiagrams do not show pulmonary plethora, but merely dilatation of the pulmonary artery and its two main branches. Again, cyanosis always favours cor pulmonale, and, even if a reversed interatrial shunt is responsible for it, cor pulmonale is still likely to be present, as explained above.

The distinction between A.S.D. and V.S.D. has already been discussed.

Idiopathic dilatation of the pulmonary artery, with pulmonary incompetence causing dilatation of the right ventricle and the electrocardiographic pattern of right bundle-branch block, may cause real difficulty; but pulmonary plethora is absent.

Simple mild pulmonary stenosis with dilatation of the pulmonary artery, normal but well-filled pulmonary vessels, and noticeable splitting of the second heart sound is often confused with A.S.D. But these mild cases are usually associated with a normal left ventricular cardiac impulse and with a normal electrocardiogram. Moreover, the pulmonary artery is always impalpable.

Finally, a normal heart with clockwise rotation (due perhaps to slight scoliosis), unusual prominence of the pulmonary artery, a functional pulmonary systolic murmur, noticeable splitting of the second heart sound, a rather tapping type of cardiac impulse (left ventricle displaced posteriorly), a secondary R wave in Lead V_1, and well-filled pulmonary vessels may resemble mild A.S.D. very closely. The clinical diagnosis depends chiefly on the degree of the changes enumerated. There are very few cases of A.S.D. that are mild enough not to show something beyond the range of normal variation.

Pulmonary Stenosis

No form of congenital heart disease has caused so much confusion and muddled thinking as pulmonary stenosis. First, apart from Fallot's tetralogy, it was considered rare (Abbott, 1932; Taussig, 1947); more recently it has been realized that it is not so rare (Currens, Kinney, and White, 1945; Allanby and Campbell, 1949), and in fact it is common. Secondly, the presence or absence of cyanosis, whether it is peripheral or central, and what determines it still need clarification. Thirdly, certain titles used in modern papers to describe various forms of pulmonary stenosis, while obviously acceptable at their face value, are not acceptable when used to define recognizable clinical entities: thus Selzer *et al.* (1949) used the title "the syndrome of pulmonary stenosis with patent foramen ovale" to describe cyanosed cases of pulmonary stenosis with reversed interatrial shunt; and Allanby and Campbell (1949) used the title "congenital pulmonary stenosis with closed ventricular septum" to describe, apparently, any form of pulmonary stenosis that was not Fallot's tetralogy. As is shown later, neither of these descriptive titles covers the facts that they are meant to convey. Fourthly, no satisfactory description of the clinical features of pulmonary stenosis has yet been published.

There were 61 cases of pulmonary stenosis in this series (excluding pulmonary atresia), of which 23 could be classed as "simple," 5 as pulmonary stenosis with reversed interatrial shunt, and 33 as Fallot's tetralogy. Several different types of "simple" stenosis were encountered, depending on: (1) the state of the pulmonary artery—whether aneurysmal, dilated, normal, or small; (2) whether the stenosis was valvular or subvalvular; (3) whether the stenosis was mild or severe; (4) whether the ventricular septum was closed or patent; and (5) whether the auricular septum was closed or patent.

These variables are not academic, but have a profound practical influence on the clinical features of the condition. To avoid too much repetition the classical picture of severe valvular stenosis with closed septa will be described first, and then the effect of each of the variations on this picture can be discussed.

Simple Pulmonary Valvular Stenosis (Severe) with Closed Septa

Five cases were studied. They were characterized by many constant features.

1. Cyanosis, when present, was peripheral. When the cardiac output was low, cyanosis might be increased by compensatory polycythaemia, and the facies was sometimes bloated.

2. Breathlessness developed in childhood or adolescence, and was progressive. Angina pectoris and syncope occurred in one.

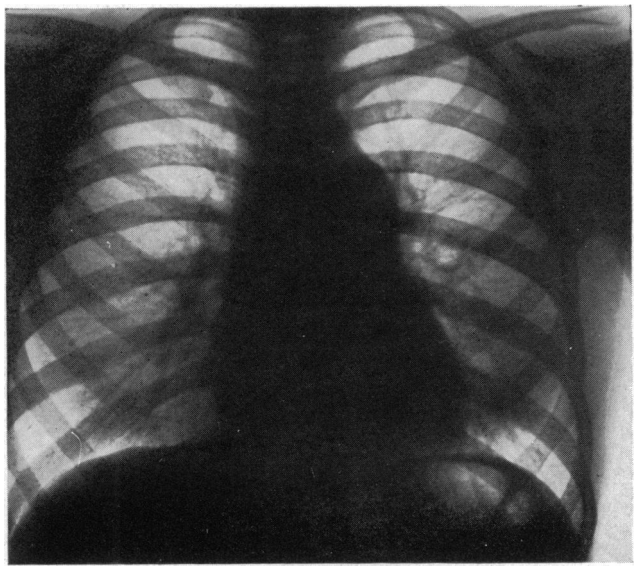

FIG. 5.—Skiagram in a case of moderately severe pulmonary stenosis.

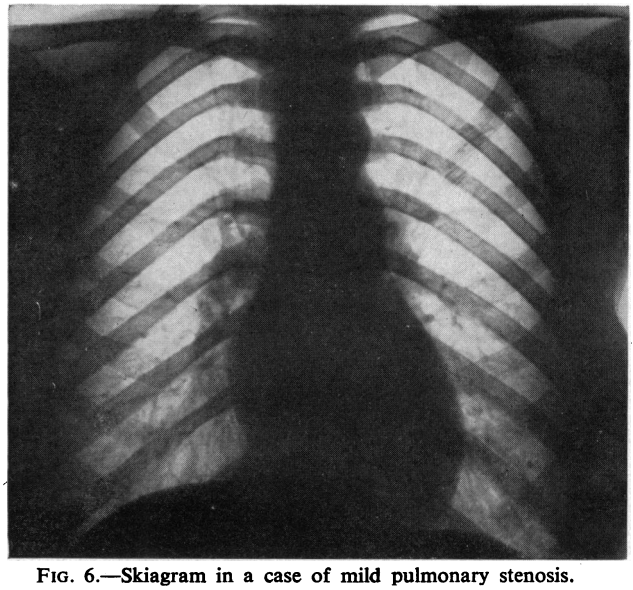

FIG. 6.—Skiagram in a case of mild pulmonary stenosis.

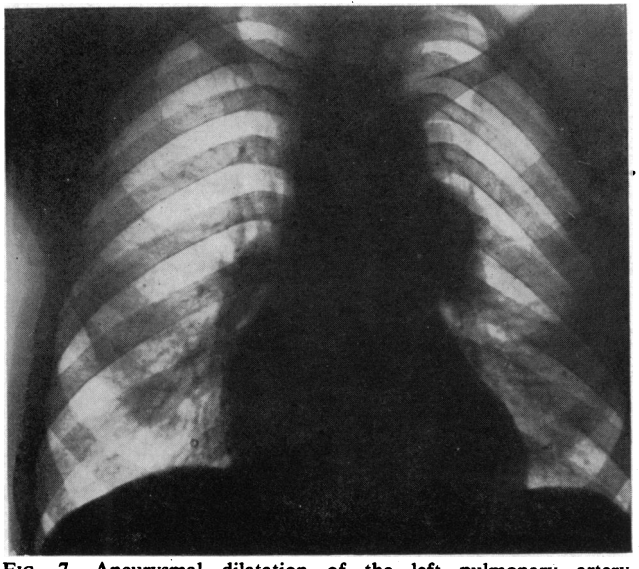

FIG. 7.—Aneurysmal dilatation of the left pulmonary artery associated with pulmonary stenosis.

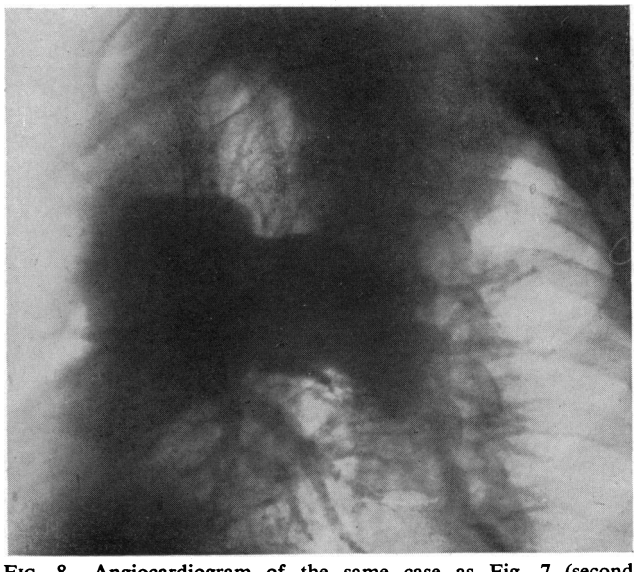

FIG. 8.—Angiocardiogram of the same case as Fig. 7 (second oblique position).

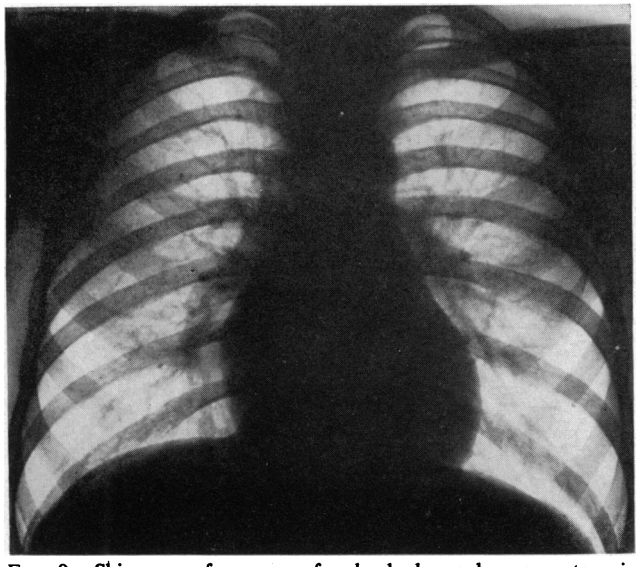

FIG. 9.—Skiagram of a case of subvalvular pulmonary stenosis showing no dilatation of the pulmonary artery.

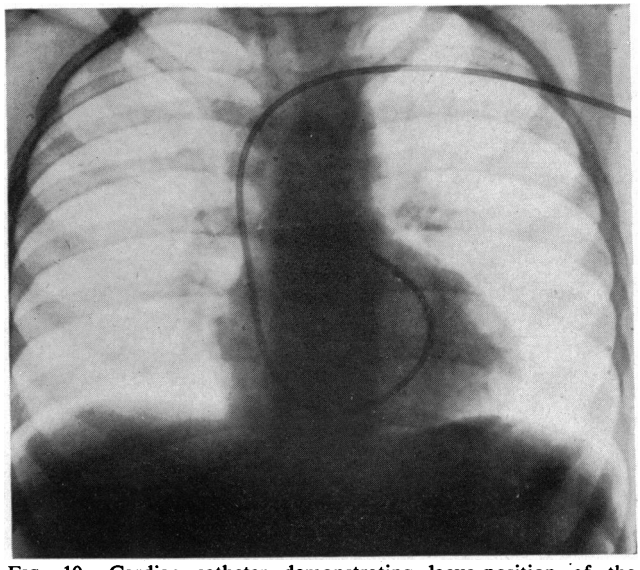

FIG. 10.—Cardiac catheter demonstrating laevo-position of the pulmonary artery in a case of Fallot's tetralogy.

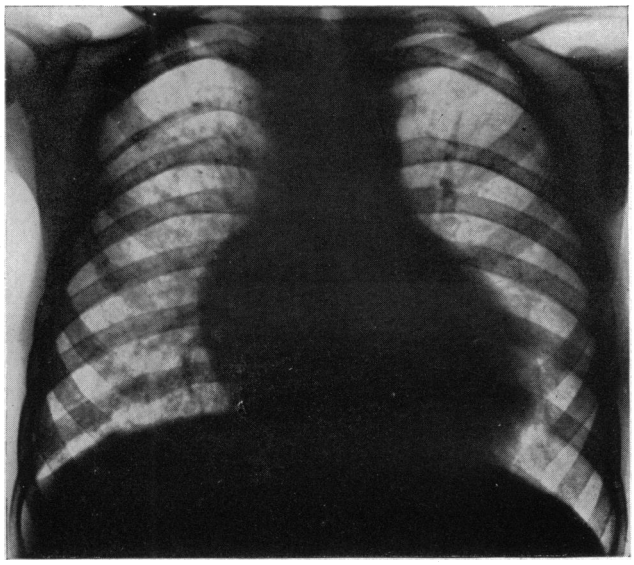

FIG. 11.—Tricuspid atresia with left ventricular enlargement and bronchial vessels supplying the lungs.

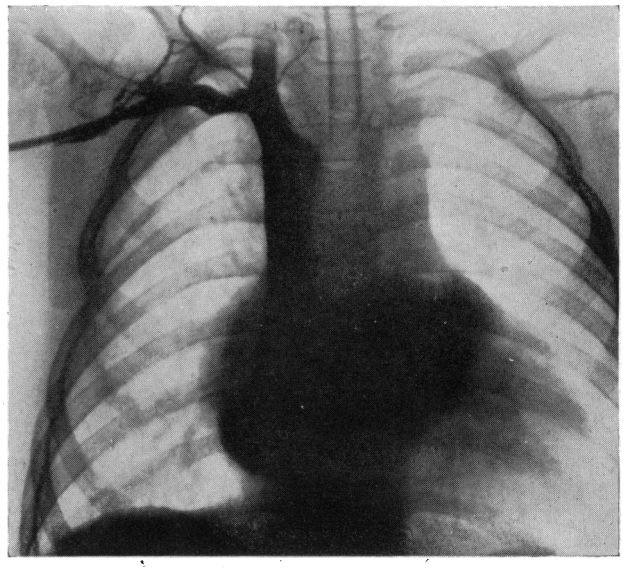

FIG. 12.—Angiocardiogram showing immediate filling of the left auricle in a case of tricuspid atresia (note L.A. appendix).

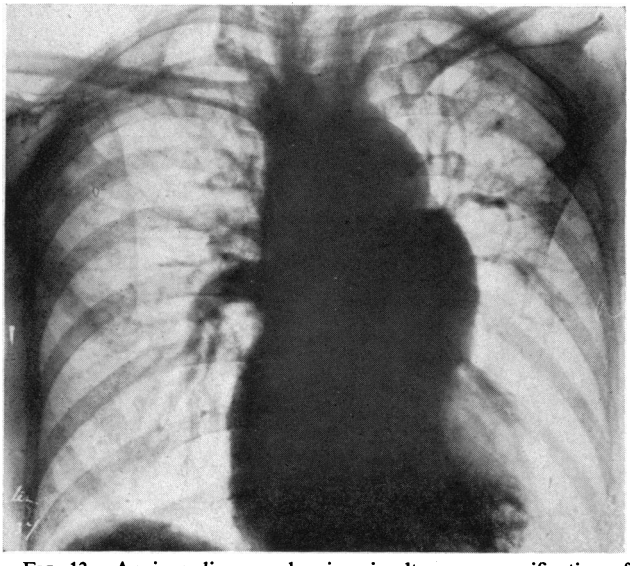

FIG. 13.—Angiocardiogram showing simultaneous opacification of the aorta and pulmonary artery in a case of pulmonary hypertension with reversed aorto-pulmonary shunt due to patent ductus.

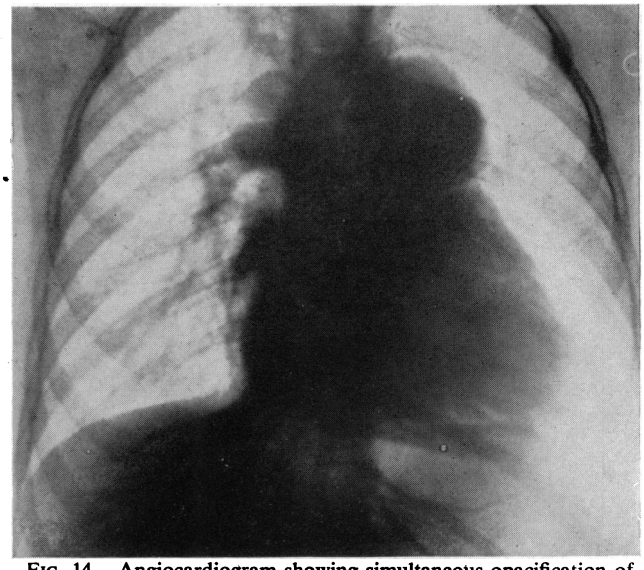

FIG. 14.—Angiocardiogram showing simultaneous opacification of the aorta and pulmonary artery in a case of true Eisenmenger's complex.

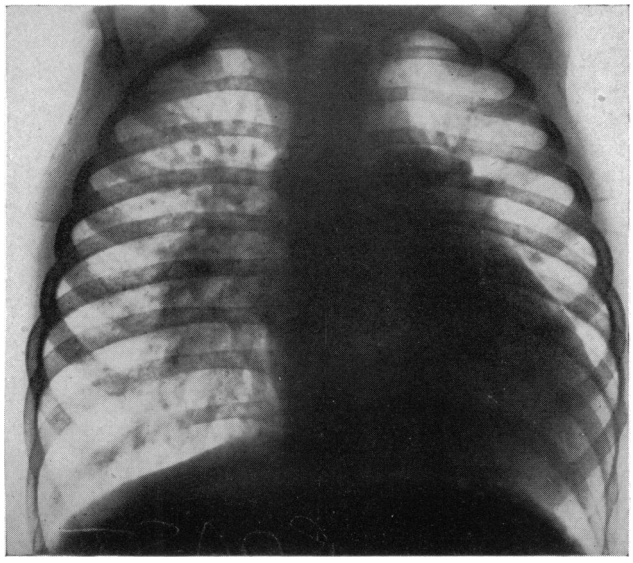

FIG. 15.—Skiagram showing pulmonary plethora in a case of transposition of the great vessels.

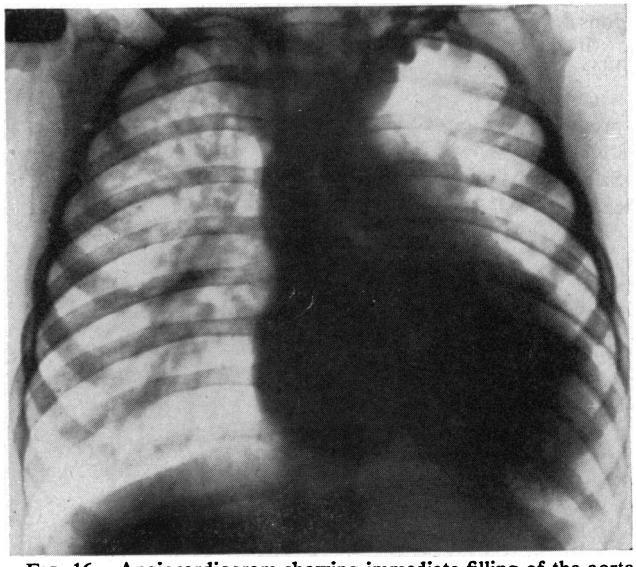

FIG. 16.—Angiocardiogram showing immediate filling of the aorta from the right ventricle in a case of transposition of the great vessels.

Death from congestive failure is expected in all of these before long, unless pulmonary valvulotomy saves them.

3. The pulse was small or on the small side of normal.

4. The jugular venous pressure was usually raised, and in at least two cases giant *a* waves were seen in the neck. This presystolic venous pulse was most conspicuous, towered above the *c* and *v* waves, was abrupt and collapsing in quality, so that it came to be called a "venous Corrigan," and was transmitted to the liver.

5. The cardiac impulse was tapping, the right ventricular outflow tract lifting, and the pulmonary artery pulse invisible and impalpable.

6. All had a high systolic murmur and thrill maximal in the second or second and third left spaces.

7. The second heart sound was invariably single at the pulmonary area.

8. The electrocardiogram showed prominent sharp P waves and strong right ventricular preponderance without right bundle-branch block. The R waves were very tall and the T waves were sharply inverted in right ventricular surface leads or their equivalents (Fig. E). The U waves were also inverted in these leads in some of the cases.

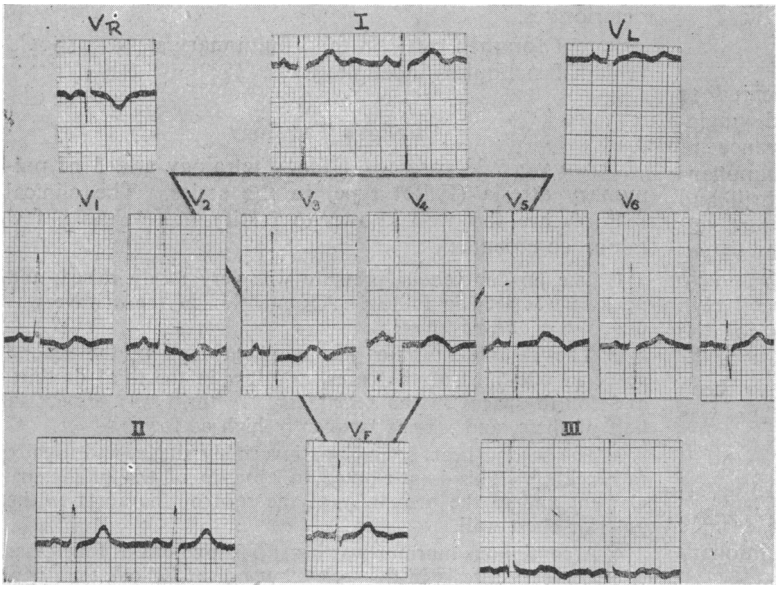

FIG. E.—Electrocardiogram in a case of severe pulmonary stenosis showing almost monophasic R waves, inverted T waves, and inverted U waves in right ventricular surface leads.

9. Skiagrams showed enlargement of the right ventricle, often considerable, and dilatation of the pulmonary artery. The peripheral vascular shadows were normal or rather light (Plate, Fig. 5).

Cardiac catheterization revealed an extremely high mean pressure in the right ventricle (over 35 mm. Hg), and a low pressure in the pulmonary artery (under 8 mm. Hg). Samples from the pulmonary artery, right ventricle, right auricle, and superior vena cava were similar.

Penetration of the pulmonary artery may be dangerous in this condition, for if the catheter blocks the pulmonary valve there is no other way for the blood to escape.

Severe Pulmonary Stenosis with Reversed Interatrial Shunt

Five cases were studied in this series. Three were cyanosed from birth. This is inevitable in the most severe cases, because the low left auricular pressure and raised right auricular pressure must keep the foramen ovale open. But cyanosis certainly increased in childhood or adolescence, and was markedly accentuated by effort. Angina pectoris and syncope occurred in one case. The onset of cyanosis may have been delayed in the other two, as described by Allanby and Campbell (1949). Such cases are presumably not quite so severe at birth but tighten up later. When this occurs reversed interatrial shunt can develop only if the foramen ovale happens to be patent—that is, in about a quarter of the cases.

The physical signs were similar to those of severe stenosis without shunt, with the addition of cyanosis, clubbing, and polycythaemia.

Skiagrams always showed diminished pulmonary vascular markings beyond the dilated pulmonary artery.

Cardiac catheterization (in three cases) revealed extremely high mean pressures in the right ventricle (over 45 mm. Hg) and low pressures in the pulmonary artery (below 5 mm. Hg). Pulmonary venous samples were normal; samples from the left auricle, left ventricle, and femoral artery were similar, and contained a large proportion of venous blood shunted from the right auricle. In a typical case the systemic blood flow was 4.3 litres a minute, and the pulmonary flow 1.4 litres a minute; the shunt was therefore 2.9 litres a minute.

Angiocardiography shows immediate filling of both auricles and simultaneous opacification of both ventricles and of the aorta and pulmonary artery.

Differential Diagnosis.—Severe pulmonary stenosis without cyanosis is difficult to confuse with any other condition. Pulmonary stenosis with reversed interatrial shunt may be distinguished from Fallot's tetralogy by the history, by greater cardiac enlargement, and by the presence of a dilated pulmonary artery. A low murmur or absence of a thrill favours Fallot's tetralogy. Doubt may be resolved by means of cardiac catheterization or angiocardiography.

Treatment—Pulmonary valvulotomy (Brock, 1948) is the treatment of choice for all severe cases of pulmonary stenosis with or without reversed interatrial shunt.

Mild Valvular Stenosis with Closed Septa

Seven cases were investigated, and these differed radically in nearly all respects from the severe type. (1) None had peripheral cyanosis; indeed, they all looked healthy. (ii) There were no symptoms; one patient was an athlete. (iii) The pulse was normal. (iv) The venous pressure was normal, and large *a* waves were not seen. (v) The cardiac impulse was usually a normal gentle left ventricular thrust, and there was little evidence of right ventricular enlargement. (vi) All had a high pulmonary systolic thrill and murmur. (vii) The second heart sound was usually split in the normal fashion, the pulmonary element being soft or of average intensity. (viii) The electrocardiogram showed normal P waves and virtually normal QRS complexes, although the R wave in Lead V_1 was a little high in three of them. (ix) Skiagrams showed no cardiac enlargement, a dilated pulmonary artery, and normal peripheral vascular markings (Fig. 6).

Cardiac catheterization revealed a normal mean pulmonary artery pressure (over 8 mm. Hg) and moderate elevation of the right ventricular pressure (15–30 mm. Hg). Samples from the pulmonary artery were similar to those from the right ventricle, right auricle, and S.V.C., and were normally unsaturated.

Similar mild cases have been described by Mannheimer *et al.* (1949), at Stockholm, and by Dexter *et al.* (1950).

The differential diagnosis is from the normal heart with prominent pulmonary artery and functional pulmonary systolic murmur, from mild atrial septal defect, and from the "maladie de Roger."

No treatment is required.

Pulmonary Stenosis with Aneurysmal Dilatation of the Pulmonary Artery

Only one such case was seen. The clinical features were those of mild pulmonary stenosis, and the *x*-ray appearances (Fig. 7) were a surprise. The mean pressure in the pulmonary artery was 11 mm. Hg, in the right ventricle 22 mm. Hg. Angiocardiography confirmed the nature of the hilar shadows (Fig. 8).

Idiopathic dilatation of the pulmonary artery without pulmonary stenosis (Greene *et al.*, 1949) may be complicated by pulmonary incompetence, enlargement of the right ventricle, and right bundle-branch block. Two such cases were encountered in this series. In both, the pulmonary artery and right ventricular pressures were normal.

Subvalvular Stenosis

In two cases simple pulmonary stenosis appeared to be subvalvular on cardiac catheterization and angiocardiography. Both were characterized by absence of the common post-stenotic dilatation of the pulmonary artery (Fig. 9) and by a relatively low systolic thrill and murmur which were readily appreciated in the third left space. Otherwise the findings were the same as for valvular stenosis. They happened to be moderately severe (P.A.P. 11 and 8; R.V.P. 44 and 31 mm. Hg respectively). When mild, these cases may easily be mistaken for the "maladie de Roger," or for subaortic stenosis.

Since completing this series a third case has been seen, in which a separate infundibular chamber was demonstrated.

Pulmonary Stenosis with V.S.D. (or A.S.D.)

Four such cases were revealed as a result of cardiac catheterization. Two had been diagnosed as pulmonary stenosis, and two as "maladie de Roger."

The stenosis was valvular in all four. It was severe in one (P.A.P. 5; R.V.P. 37 mm. Hg), moderate in two (P.A.P. 17 and 32; R.V.P. 40 and 40 mm. Hg respectively), and mild in one (P.A.P. 10; R.V.P. 16). Despite the stenosis the pulmonary blood flow was at least twice the systemic flow in two of the cases.

Clinical diagnosis may be puzzling, for the signs of pulmonary stenosis and V.S.D. may be combined. As a rule, however, one or the other dominates the picture and determines the course. The association is important for several reasons: (1) it shows that the notation "pulmonary stenosis with closed ventricular septum" cannot be used clinically to distinguish "simple" pulmonary stenosis from Fallot's tetralogy, because the presence or absence of V.S.D. may be impossible to determine clinically, and may not interfere with the clinical features or course of the pulmonary stenosis; (2) it offers further evidence of the clinical confusion between mild pulmonary stenosis and the "maladie de Roger"; and (3) it is probable that similar cases occur in which the stenosis is very tight—reversal of the shunt might then occur and the case would come to resemble Fallot's tetralogy (but without a riding aorta).

Since completing the series a similar type of case was investigated, but the left-to-right shunt proved to be interatrial and resulted from A.S.D. The stenosis was mild (P.A.P. 8; R.V.P. 31; infundibulum 18 mm. Hg). Left auricular samples were 93% saturated, and the pulmonary blood flow was twice the systemic flow. Considerable pulmonary plethora was present radiologically.

Pulmonary Stenosis with Patent Foramen Ovale

That this is not synonymous with pulmonary stenosis with reversed interatrial shunt is obvious, and if there is no cyanosis the presence of patent foramen ovale is unsuspected. Unlike pulmonary stenosis with A.S.D., there is no left-to-right interatrial shunt.

One such acyanotic case was discovered in this series. This was that of a girl of 14 without symptoms and with typical signs of moderate pulmonary valve stenosis. The catheter slipped through a patent foramen ovale, and left auricular samples were 96% saturated with oxygen. The mean right auricular pressure was 4 cm. of saline, the left 5.5 cm. The pulmonary artery pressure was 8 mm. Hg, and the R.V. pressure was 32 mm. She is clearly liable to develop a reversed interatrial shunt if the situation deteriorates.

Patent foramen ovale in mild pulmonary stenosis should remain functionless indefinitely.

Fallot's Tetralogy

There were 33 cases of Fallot's tetralogy and 3 of pulmonary atresia (Fallot type) in the series. The clinical features are too well known to justify more than a few minor comments:

1. The jugular venous pressure was very rarely raised, and no case was seen in failure. Accentuation of the *a* wave was uncommon and never great.

2. The systolic murmur was maximal high in the second space in one-third of the cases, and low in the third or third and fourth spaces in the remainder. A thrill was present in half of them, and was as frequently high as low.

3. The second heart sound was invariably single, and usually rather loud. This is attributed to absence of the pulmonary element and to the proximity of the root of the aorta to the anterior chest wall.

4. A continuous murmur was heard in three out of four cases of pulmonary atresia in the series, but not in Fallot's tetralogy. Necropsy in three of these cases showed that the murmur depended upon a broncho-pulmonary anastomosis at a fairly high arterial level, not upon a patent ductus. Since these figures were completed two more cyanotic cases with continuous murmurs have been examined and diagnosed as atresia partly because of these murmurs. Cardiac catheterization was not, therefore, undertaken, and angiocardiography proved that both diagnoses were correct. The murmur was left-sided in two of these cases, mainly right-sided in two, and bilateral in one. Clinically it is impossible to distinguish pulmonary atresia from a persistent truncus arteriosus unless well-formed pulmonary arteries arise from the root of the aorta, or unless the latter is obviously much enlarged. Angiocardiograms are also similar in all other respects.

5. Cardiac catheterization was carried out in 21 cases of Fallot's tetralogy. The mean pulmonary artery pressure was under 8 mm. Hg (−1 to +7) in all but one of 15 cases in which that vessel was entered. The right ventricular pressure ranged from 22 to 55 mm. Hg, and was commonly around 40. Samples from the P.A., R.V., R.A., and S.V.C. were usually very unsaturated and were similar in all but two cases: in those two the pulmonary artery samples were less unsaturated. No explanation for this can yet be offered.

6. The catheter was passed through a patent foramen ovale in two cases. It was interesting to be able to demonstrate the absence of reversed interatrial shunt in both instances, the left auricular pressure being higher than the right, and left auricular

samples being normally saturated with oxygen ; and this despite mean right ventricular pressures of 44 and 37 mm. Hg and a pulmonary artery pressure of 6 mm. in one of them. It seems that the right ventricle is able to accommodate itself to work against the systemic blood pressure without difficulty, and does not suffer from the extremely high pressure generated on effort in cases of pure pulmonary stenosis. This also suggests that reversed interatrial shunt depends more upon elevation of the right auricular pressure than upon the lowering of the left.

7. Both during cardiac catheterization and on inspecting the angiocardiograms, laevo-position of the pulmonary artery was often noticed. This was usually more convincing than dextro-position of the aorta (Fig. 10).

Tricuspid Atresia

This is the only cyanotic form of congenital heart disease with a diminished pulmonary blood flow associated with left ventricular enlargement and a rudimentary right ventricle. The cardiac impulse, *x*-ray appearances (Fig. 11), and electrocardiogram (Fig. F) usually make

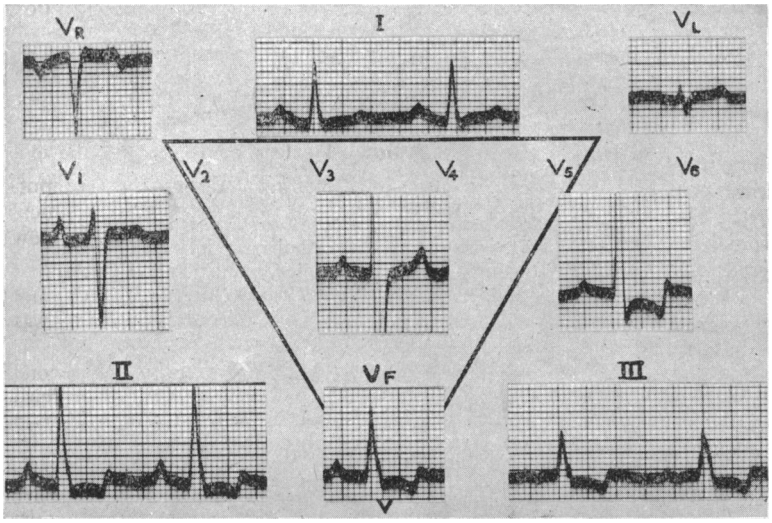

FIG. F.—Electrocardiogram of a case of tricuspid atresia showing the "P mitrale," and left ventricular preponderance (vertical heart).

the diagnosis easy. If not, angiocardiography provides convincing evidence (Fig. 12).

Cardiac catheterization is unsatisfactory, because failure to enter the right ventricle may be due to technical difficulty and cannot be attributed to tricuspid atresia with any surety. However, a reversed interatrial shunt can be demonstrated, and under the clinical circumstances this confirms the diagnosis.

Eisenmenger's Complex

The majority of cases in which a diagnosis of Eisenmenger's complex was suggested turned out otherwise ; only three were finally accepted after full physiological and angiocardiographic studies, and subsequent necropsy disproved one of them.

The Eisenmenger's syndrome consists of central cyanosis, tapping cardiac impulse, lifting conus, visible and palpable pulsation of the pulmonary artery, a systolic murmur and thrill in the third left space, normal splitting of the second heart sound with accentuation of the pulmonary element, an electrocardiogram showing right ventricular preponderance, and *x*-ray appearances characterized by right ventricular enlargement, dilatation of the pulmonary artery and its two main branches, and poorly marked peripheral vascular shadows.

Cardiac catheterization reveals a raised pressure in the pulmonary artery and right ventricle, a similar degree of unsaturation in samples from the pulmonary artery, right ventricle, right auricle, and S.V.C., and normally saturated samples from the pulmonary veins and left auricle (if obtained through a patent foramen ovale). The arterial oxygen saturation is reduced.

Angiocardiography shows simultaneous opacification of the aorta and pulmonary artery from the right ventricle, no dye entering the left auricle initially.

Differential Diagnosis

Clinically, advanced idiopathic pulmonary hypertension may closely resemble Eisenmenger's syndrome at any age, differences being largely a matter of degree. Thus there is usually less cyanosis, and the small pulse and cold extremities suggest that some of it is peripheral ; breathlessness is more severe, and there may be angina or syncope ; the jugular venous pressure is higher and the *a* wave more conspicuous ; there is no thrill at the base, and if there is a murmur it is unimpressive ; the electrocardiogram nearly always shows a conspicuous P pulmonale and stronger right ventricular preponderance ; and the heart looks larger radiologically. Physiological studies reveal higher pressures in the pulmonary artery and right ventricle, and pulmonary venous samples (if they can be obtained) show the same degree of unsaturation as arterial samples. No shunt can be seen in the angiocardiogram.

Idiopathic pulmonary hypertension with reversed interatrial, interventricular, or aorto-pulmonary shunt is the usual explanation for most cases of "proved" Eisenmenger's syndrome. The clinical features resemble those of pure idiopathic pulmonary hypertension as described above ; but cyanosis and clubbing are more conspicuous, and the angiocardiogram shows simultaneous opacification of the aorta and pulmonary artery.

The absence of a thrill and murmur favours reversed interatrial or aorto-pulmonary shunt, and both cardiac catheterization and angiocardiography should demonstrate an interatrial shunt ; but reversed interventricular shunt is clinically Eisenmenger's syndrome itself. At necropsy, however, the aorta is not overriding.

One of the accepted cases of Eisenmenger's syndrome included in this series (Fig. 13) proved to be pulmonary hypertension with reversed aorto-pulmonary shunt through a patent ductus. This case is being reported in detail by Dr. Maurice Campbell.

The question is constantly being asked whether there is really any such thing as Eisenmenger's complex ; whether it is possible to be sure of an overriding aorta at necropsy when the pulmonary outflow tract is normal. One case in the series seems to provide a positive answer to this question.

This was that of a girl aged 7, with considerable effort dyspnoea, squatting, moderate cyanosis and clubbing, slight polycythaemia, a normal rather than small arterial pulse, a normal venous pressure and pulse, a tapping cardiac impulse, lifting conus and visible pulmonary artery pulsation, a systolic murmur and thrill in the third left space, and a normally well-split second heart sound with accentuation of the pulmonary element. Her electrocardiogram showed normal P waves and slight to moderate right ventricular preponderance. Skiagrams showed no obvious enlargement of the heart, but the pulmonary artery was dilated and the peripheral vascular shadows diminished. On cardiac catheterization the pressure was 39 mm.

Hg in the pulmonary artery, 19 in the right ventricle, and +2 in the right auricle, samples from these positions being the same (66–69% saturated). The arterial sample was 71% saturated. Angiocardiography showed simultaneous opacification of the aorta and pulmonary artery without early filling of the left auricle (Fig. 14).

It is clear in this case that the pressures are not high enough to warrant a diagnosis of pulmonary hypertension with reversed interventricular or aorto-pulmonary shunt, and an overriding aorta must be postulated.

Transposition

Two patients with transposition were investigated, girls aged 3 and 12. The clinical diagnosis is usually easy, and depends on the recognition of pulmonary plethora in an obviously cyanotic case (Fig. 15). The rest of the findings closely resemble those of Eisenmenger's complex.

The catheter was passed down the aorta as well as into the pulmonary artery in both cases studied. The mean right ventricular pressure was of course high, 31 and 41 mm. Hg; the pulmonary artery pressure 11 and 64 respectively (aortic 74 and 69). Samples from the pulmonary artery were considerably more saturated with oxygen than samples from the aorta, and this proved the diagnosis. Venous blood entered the lungs from the right ventricle via a ventricular septal defect in both cases, and pulmonary venous blood escaped from the semi-closed pulmonary circuit via an atrial septal defect in the first case and presumably via bronchial and azygos veins in the second. The pulmonary blood flow was greatly increased, being three times the systemic flow in both patients. Angiocardiography also confirmed the diagnosis (Fig. 16).

The elder girl with atrial septal defect was less cyanosed and less breathless than the other; indeed, she was remarkably well. The creation of an artificial atrial septal defect or of a pulmonary-azygos venous anastomosis would seem to be justified.

Classification

In the light of these findings a new clinical classification of congenital heart disease is offered (Table V).

TABLE V.—*Classification of Congenital Heart Disease*

No Shunt

General	Left-sided	Right-sided
Dextrocardia Idiopathic hypertrophy Von Gierke's disease Heart-block Familial cardiomegaly	Coarctation of the aorta Right-sided aortic arch Complete or incomplete aortic rings Bicuspid aortic valve (or supernumerary cusps) Aortic or subaortic stenosis Left coronary artery arising from pulmonary artery	Idiopathic dilatation of the pulmonary artery Simple pulmonary stenosis (with or without patent F.O.). Ebstein's disease

With Shunt

Acyanotic Left-to-right Shunt (Pulmonary Plethora)	Cyanotic Right-to-left Shunt
Left ventricular enlargement: Patent ductus Ventricular septal defect (with or without mild pulmonary stenosis) Perforated aortic sinus into P.A. or R.V. (or R.A.) *Right ventricular enlargement:* Atrial septal defect (with or without mild pulmonary stenosis). Anomalous pulmonary veins joining S.V.C. or R.A.	*Diminished pulmonary blood flow:* Low P.A. pressure: 1. Left ventricular enlargement: Tricuspid atresia 2. Right ventricular hypertrophy: Fallot's tetralogy Pulmonary atresia (Fallot type) Persistent truncus Pulmonary stenosis with reversed interatrial shunt High P.A. pressure: Eisenmenger's complex Pulmonary hypertension with reversed aorto-pulmonary, interventricular, or interatrial shunt *Pulmonary plethora:* Transposition

Summary and Conclusions

A fresh account of congenital heart disease is presented in the light of a series of 200 cases proved by means of cardiac catheterization, angiocardiography, thoracotomy, or necropsy.

The technical difficulties of cardiac catheterization are discussed.

The clinical incidence of the more common or well-recognized anomalies is given.

The physical signs of congenital heart disease are reviewed and revalued. Particular attention has been paid to the jugular pulse, the cardiac impulse, the right ventricular outflow tract, functional mitral and pulmonary diastolic murmurs, and the second heart sound.

New facts concerning most forms of congenital heart disease are presented.

The left-to-right shunts with pulmonary plethora usually have a normal or low pulmonary resistance. When the pulmonary resistance is high the shunt is diminished and is in danger of being reversed; an acquired Eisenmenger's syndrome results. This pulmonary hypertension does not depend on the duration of the shunt, but on some inherent predetermined factor, and operates early.

It is suggested that the term "maladie de Roger" should be limited in its meaning, and should be used only to describe uncommon cases of V.S.D. characterized by nothing more than a Roger thrill and murmur. The majority of cases of V.S.D. present a very different picture.

Many types of pulmonary stenosis are described, the chief of which are: (1) severe pulmonary stenosis (usually valvular) with closed septa; (2) severe pulmonary stenosis (usually valvular) with reversed interatrial (or interventricular) shunt; (3) mild pulmonary stenosis (valvular or infundibular) with or without functionless patent foramen ovale; and (4) pulmonary stenosis with left-to-right shunt through an associated V.S.D. or A.S.D. Pulmonary stenosis with late reversed interventricular shunt was not encountered with certainty, but must occur; it would closely resemble Fallot's tetralogy, but there would be no overriding aorta.

Eisenmenger's complex should be diagnosed under appropriate clinical circumstances only when the pulmonary artery pressure is not high enough to cause reversed aorto-pulmonary, interventricular, or interatrial shunt, through a patent ductus, V.S.D., or A.S.D. Pulmonary hypertension with reversed aorto-pulmonary, interventricular, or interatrial shunt might be called acquired Eisenmenger's syndrome, because it is clinically indistinguishable from Eisenmenger's complex.

A new classification of congenital heart disease is offered.

I wish to thank my colleagues on the staff of the National Heart Hospital for allowing me to investigate some of their cases and to make use of material so obtained. I also wish to thank Mr. Latham, our senior technician, for carrying out the blood-gas analysis.

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