

## ANOMALOUS PULMONARY AND SYSTEMIC VENOUS DRAINAGE

BY

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In Abbott's (1936) necropsy series of 1,000 cases of congenital heart disease, anomalies of the great veins were found in 13. Of these nine had abnormal major systemic veins and four had anomalous pulmonary veins. This classical series is based on findings before such modern methods as angiocardiology and cardiac catheterization were widely used. Taussig (1947) differentiates cases in which some pulmonary veins drain into the right atrium from those in which all pulmonary veins do so. She regards anomalies of the superior and inferior venae cavae as rare and difficult to diagnose. She records one case in which both superior and inferior venae cavae entered the left atrium, one in which there was no inferior vena cava, and two in which a left-sided vena cava entered the left side of a common atrium. She concludes that anomalies of the superior and inferior venae cavae rarely constitute the sole malformation, and that only the coexisting cardiac malformation permits diagnosis.

With improved diagnostic methods many more examples of abnormal venous drainage are being recognized and reported. A survey of the literature suggests that, for practical purposes, these may be divided into three groups:

- (1) Anomalies involving the systemic veins alone.
- (2) Anomalies involving the pulmonary veins alone.
- (3) Anomalies involving both systemic and pulmonary veins and associated with pulmonary systemic communications.

### ANOMALIES INVOLVING SYSTEMIC VEINS ALONE

Of these the most common is a persistent left superior vena cava. McManus (1941) differentiated two main groups of cases. In group 1 both right and left superior venae cavae persist; this group is further subdivided into (a) those in which the left superior vena cava is connected to the

coronary sinus, either with or without a connexion to the right superior vena cava through a left innominate vein; and (b) those in which the left superior vena cava receives one or more pulmonary veins. In group 2 no right superior vena cava is present. It will be noted that in group 1 (b) arterial blood enters the superior vena cava, but groups 1 (a) and 2 involve systemic veins alone and do not lead to an arterio-venous shunt. A veno-arterial shunt occurs only in those cases, apparently few in number, in which the left superior vena cava enters the left atrium. Examples have been described by McCotter (1916), Rodriguez Diaz, Castellanos, and García Faes (1949), Mankin and Burchell (1953), and Feindt and Hauch (1953). Rarely the inferior vena cava enters the left atrium, producing a veno-arterial shunt (Taussig, 1947; Gardner and Cole, 1955).

A persistent left superior vena cava is most often found as an additional abnormality in patients with some other form of cardiac malformation. Brown (1952) reports a case of aortic coarctation with a single left superior vena cava. Winter (1954) has reviewed the literature and added 30 new cases. In 28 of the 30 cases the left superior vena cava persisted in its entirety, while in two it persisted only in part. Anomalous drainage of pulmonary veins was found in three of these cases: on the other hand a septal defect was present in 25. Other associated anomalies were aortic septal defect, Fallot's tetralogy, truncus arteriosus, coarctation of the aorta, patent ductus arteriosus, and pulmonary stenosis. Campbell and Deuchar (1954) found 46 examples in their series of cases of congenital heart disease. In 18 of these there was partial or complete transposition of the viscera, and 13 of them had either proved or suspected pulmonary stenosis; in four instances the inferior vena cava was absent. In the remaining 28 cases the heart and abdominal viscera were normally placed, but various additional abnormalities were present. In most of their cases the left

superior vena cava joined the coronary sinus to reach the right atrium. They have not seen an example in which it entered a normal left atrium, but have seen it open into a common atrium or into the left half of an atrium with a large septal defect. Anomalous pulmonary venous drainage was noted in one of their cases only. They do not consider the presence of a left superior vena cava to be of much practical importance, but suggest that, if it is suspected, cardiac catheterization is best carried out from the right side, and, if discovered at operation, that ligation should not be performed. On the other hand, Rodriguez Diaz and his co-workers have ligated a left superior vena cava which drained into the left atrium with an excellent result. Difficulties may be encountered with catheterization if the atrium is entered through the coronary sinus from a left superior vena cava (McMichael and Mounsey, 1951).

#### ANOMALIES OF PULMONARY VEINS ALONE

Anomalies may consist of an abnormal number of pulmonary veins or abnormal termination of some or all pulmonary veins (Kugel and Pöschl, 1954). Provided there is no pulmonary-systemic venous communication, the condition appears to be of academic importance only. We know of one patient who was found at pneumonectomy for right-sided bronchiectasis to have a shrunken lung with normal stem bronchus, a hypoplastic pulmonary artery, and no right pulmonary veins (Barclay, 1955).

#### ANOMALIES INVOLVING PULMONARY-SYSTEMIC VENOUS COMMUNICATION

These necessarily involve an arterio-venous shunt which may or may not be associated with a veno-arterial shunt elsewhere. In striking contrast to anomalies which are confined to the systemic or to the pulmonary veins alone, they cause an alteration in the mechanics of the circulation; they may profoundly change the clinical picture and the prognosis of coexisting cardiac malformations; and they often produce a characteristic radiological picture. Anomalous pulmonary venous drainage may be total or partial. In the former case, all the pulmonary veins drain into the right atrium, either directly or via a systemic vein. In partial anomalous pulmonary venous drainage, some pulmonary veins enter the left atrium in the normal manner while others enter a systemic vein or the right atrium.

Brody (1942) found six cases of total anomalous venous drainage recorded in the literature and added a seventh of his own. By 1954 the number

of recorded cases had mounted to 45, to which Keith, Rowe, Vlad, and O'Hanley (1954) added another 14. Yet another case has been added by Arthurton, Gibson, and Woodwark (1954).

Jorgens, LaBree, Adams, and Rigler (1952) first described the radiological features of venous drainage anomalies, demonstrating a persistent left superior vena cava and also drainage of pulmonary veins into the superior vena cava. Snellen and Albers (1952) describe partial pulmonary drainage into the superior vena cava, right atrium, and left innominate vein, and total pulmonary drainage into the superior vena cava, coronary sinus, right atrium, left innominate vein and the portal vein. They draw attention to a "figure of eight" appearance in the postero-anterior radiograph, and stress the frequency of right bundle branch block due to coexisting atrial septal defect. Tori (1952) reports a case in which a right pulmonary vein opened into the superior vena cava, while Ödman (1953) describes a patient with high coarctation of the aorta in whom a persistent left superior vena cava received pulmonary veins and joined the left atrium.

A symposium from the Mayo Clinic on pulmonary-systemic venous connexions includes interesting papers by Edwards (1953), Swan, Burchell, and Wood (1953) and Mankin and Burchell (1953). Edwards emphasizes the important distinction between total and partial pulmonary-systemic venous connexion. In total pulmonary-systemic venous connexion both veno-arterial and arterio-venous shunts are present; the prognosis is poor, most of the patients dying in infancy. In partial pulmonary-systemic venous connexion there is an arterio-venous shunt but not necessarily a veno-arterial shunt; many patients reach adult life. Necropsies were performed at the Mayo Clinic in 135 cases; associated malformations included mitral atresia, Fallot's tetralogy, cor biloculare with subpulmonic stenosis, and atresic common pulmonary veins.

A method of demonstrating the pulmonary drainage pathway by arterial dilution curves is described by Swan and others (1953). The dilution pattern of an indicator (T 1824) is recorded during its initial circulation after injection into the vena cava, pulmonary vein, right pulmonary artery, or left pulmonary artery. Differences in the recorded patterns will demonstrate the presence or absence of anomalous drainage from abnormal venous connexions. In patients who also have an atrial septal defect with a large pulmonary blood flow, it can sometimes be shown that the blood draining from the right lung is shunted preferentially across the defect. By this

method, Swan demonstrated partial drainage of the right lung into the inferior vena cava in one case in which there was no other circulatory defect. In another case cardiac catheterization with arterial dilution curves showed (1) union of the right pulmonary vein with a persistent left superior vena cava, (2) an abnormal communication between coronary sinus and left atrium, and (3) probable presence of an atrial septal defect.

Geraci and Kirklin (1953) discuss surgical treatment by transplantation of the anomalous pulmonary vein into the left atrium; this was carried out by Kirklin (1953) in one case with an uneventful post-operative course and considerable improvement in the patient's dyspnoea and fatigue.

Bruwer (1953) discusses the radiological features. He describes a crescentic sabre-shaped shadow of vascular density running downwards, parallel to or behind the right side of the heart; this no doubt represents a vein draining all or part of the blood from the right lung into the right atrium or inferior vena cava. Other suggestive features are not infrequent. The heart may be shifted towards the right to such an extent that dextrocardia is simulated; this is possibly the result of gross enlargement of the right heart; it is sometimes associated with a smaller volume of the right hemithorax as compared with the left (Dotter, Hardisty, and Steinberg, 1949). The aorta is small, sometimes invisible, possibly in consequence of the diminished volume of blood transmitted through it. The "figure of eight" appearance described by Snellen and Albers (1952) in their cases of total pulmonary-systemic venous connexion also occurs in some cases of partial drainage. The widening of the superior mediastinal shadow on the left is caused by the anomalous left vertical vein draining into the left innominate vein. On the other hand total pulmonary-systemic venous connexion can occur without the "figure of eight" appearance. The pulmonary artery becomes enlarged with total and with partial pulmonary-systemic drainage.

Levinson, Griffith, Cosby, Zinn, Jacobson, Dimitroff, and Oblath (1953) publish 10 cases in adults and children. The diagnosis was made by visualizing a catheter in the anomalous vein, from which pressure curves were venous in type and blood of high oxygen saturation was obtained. In four, the pulmonary-systemic connexion was partial, in two total. Associated anomalies included pulmonary valvular stenosis, a probable tricuspid atresia, and a left superior vena cava entering a sinus venosus in common with the hepatic vein while the right pulmonary vein entered the right atrium. Thurn and Schaeede

(1953) described additional cases but furnished no new information.

Gardner and Oram (1953) have published four cases with a persistent left superior vena cava which was joined by pulmonary veins; they correspond to McManus's group 1 (b). One was verified at operation and one at necropsy. They consider the left-to-right shunt to be the important feature, producing right ventricular enlargement. Cyanosis is absent and on fluoroscopy there is a wide superior mediastinal shadow enveloping the aortic knuckle and pulmonary arc. The differential diagnosis is from an atrial septal defect or from a large ventricular septal defect. The typical superior mediastinal shadow makes the diagnosis very probable, while high oxygen saturation in the left innominate vein confirms it. Surgical correction was attempted in one case, but proved impracticable owing to the small size of the left atrial appendage.

The term "cottage-loaf configuration" was introduced by Whitaker (1954) to describe the appearance in six cases with total pulmonary-systemic venous drainage through a persistent left superior vena cava into the left innominate vein. The clinical features were dyspnoea on exertion, recurring pulmonary infections, poor physical development, slight cyanosis, early finger clubbing, low systemic blood pressure, right ventricular hypertrophy, auscultatory signs of pulmonary hypertension, and prominence of the left side of the chest. In three of his cases there was an atrial septal defect; other anomalies were not recorded. Levin and Borden (1954) report four additional cases of pulmonary venous drainage into the left innominate vein via a left vertical vein; three were proved by cardiac catheterization. One had a patent foramen ovale, one had pulmonary stenosis, and two seemed free from additional defects. Arvidsson (1954) discusses the various routes whereby pulmonary veins may drain into the right auricle. They may enter it directly; they may join the superior vena cava, inferior vena cava, or the coronary sinus; finally they may unite with a left superior vena cava, draining via a left innominate vein into the right superior vena cava. Symptoms arise when a significant volume of blood is recirculated through the lesser circulation; increase of pulmonary blood volume ensues with a corresponding decrease in the systemic blood flow. Wide vessels curving downwards along the cardiac border may suggest the diagnosis, which is confirmed by angiocardiology, catheterization, and blood gas analysis.

Keith and others (1954) review the literature and add 14 cases in children. The main clinical findings were failure to thrive and lack of cyanosis. Murmurs may be absent, but occasionally diastolic murmurs were present; a venous hum was sometimes heard in the pulmonary area. Radiological examination was helpful chiefly in cases where pulmonary veins drained into the left innominate vein; in these circumstances a wide pulsatile arch was seen on either side of the supracardiac area in the anterior mediastinum; together with the remainder of the heart shadow this formed the "figure of eight" noted by earlier authors.

The present paper presents six illustrative cases which have been encountered within a period of six months. In Case 1 the right pulmonary veins entered the right atrium directly; an aorto-pulmonary communication was also present. In Case 2 the pulmonary veins entered a persistent left superior vena cava which joined the coronary sinus and was also connected to the right superior vena cava by a left innominate vein; the patient had aortic coarctation with a patent ductus arteriosus. In Case 3 pulmonary veins drained into a dilated right superior vena cava; whether they entered directly or via a persistent left superior vena cava has not been demonstrated with certainty. Case 4 illustrates an anomaly which we have not found previously described; both venae cavae persisted, the right entering the right atrium while the left entered a normal left atrium which received the pulmonary veins in normal fashion; the ascending aorta was hypoplastic and circulation to the lower limbs was through a widely patent duct; the inferior vena cava and right heart were grossly dilated. Attempted catheterization through the left arm induced spasm of the left superior vena cava with tragic consequences. In Case 5 there was a left superior vena cava draining into the right atrium, associated with a partial transposition of great vessels and patent ductus. Case 6 resembled Case 2 in having a persistent left superior vena cava into which there was pulmonary venous drainage from the left lung. There was no definite evidence, however, of associated patent ductus or aortic coarctation.

CASE 1.—M. B., a burner aged 25 years, married with two children, was known to have a congenital heart lesion shortly after birth. He attended a special school more because of the presence of a murmur than because of its effects on his capacity. He had never been blue. About the age of 12 years he had swollen ankles and was confined to bed for three months. Three times since the age of 16 years he had fainted, and at these times had noticed tingling

in his fingers and toes. He had previously worked as a forester and as a farm labourer without difficulty and could not remember exertional dyspnoea bothering him until recent months, when he had noted slight ankle swelling and breathlessness on heavy exertion. He had lost no appreciable working time as a result of his cardiac condition, but for five years he had had intermittent dyspepsia; in 1951 (age 21 years) he was admitted to hospital on this account and aerophagy was diagnosed. Once more in January, 1955, he reported as an out-patient with digestive symptoms; a barium meal showed no abnormality, but admission was arranged for investigation of his congenital cardiac lesion.

He was a pale man with somewhat slow cerebration, well developed physically. He had no cyanosis or clubbing. Pulses were equal at the wrists and easily palpable in the femoral arteries. The apex beat was just palpable five inches from the mid-sternal line in the sixth left interspace. There was a harsh systolic murmur at the apex, and also a loud systolic murmur at the base, maximal in the third left interspace at the sternal border. There were no palpable thrills. Blood pressure was 135/75 mm. Hg. Other systems were normal. The urine was free from albumin. A blood count was normal apart from a lymphocytosis (haemoglobin 16.2 g. per 100 ml., lymphocytes 5,750 per c.mm.), E.S.R. 1 mm. at one hour.

*Cardiogram.*—There is sinus rhythm 86/min., and right bundle branch block.

*Phonocardiogram.*—At the apex there is a first sound, second sound, and third sound with a faint systolic murmur. At the third costal cartilage at the left sternal border there is a first sound, a split second sound, and a systolic murmur.

*Radiography and Fluoroscopy.*—The heart is globular in shape with slight fullness of the left ventricle (Fig. 1). The aortic window is obliterated. Pulmonary vascular shadows are increased and hilar dance is present.

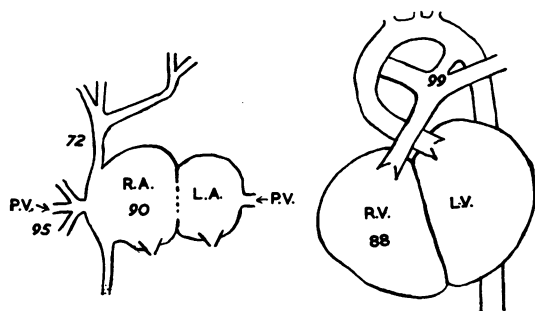
*Cardiac Catheterization.*—Immediately after entering the right atrium, the catheter passed into an anomalous pulmonary vein leading from the right upper lobe (Fig. 2). Later, during withdrawal, it was possible to enter from the right atrium into the pulmonary veins draining the right middle and lower lobes (Figs. 3 and 4). Well-marked hilar pulsation was noticed and the catheter tip showed a marked degree of "to-and-fro" excursion in the right pulmonary artery.

Pressures and oxygen content were as follows:

	Oxygen (Vol. per 100 ml.)	Per- centage Saturation	Pressures (mm. Hg)	
			Dynamic	Mean
Superior vena cava	13.98	72.2	—	—
Pulmonary vein (1)	18.33	94.6	—	—
" (2)	18.50	95.5	—	5
Right atrium	17.51	90.3	8 0	4
" ventricle	17.22	88.8	60 0	20
Pulmonary artery	19.15	98.9	30 8	13
O <sub>2</sub> capacity	19.40	100		



**Conclusion.**—There is anomalous pulmonary venous drainage, the right lung draining into the right auricle and the left lung into the left auricle. A degree of pulmonary stenosis is present. There is also an aorto-pulmonary communication, probably a patent ductus arteriosus but just possibly an aortic septal defect. There may also be an atrial septal defect (in view of right bundle branch block) (Diagram 1).



CASE 1

Figures represent percentage blood oxygen saturation.

**CASE 2.**—M. W., a girl aged 19, a commercial college student, was found to have a cardiac murmur at the age of 1 year. Her only complaint was a tendency to dyspnoea on exertion all her life, rather worse in recent years. She had been living a fairly normal life, however, and played games at school. When breathless, it was noticed that her hands and face became blue, more on the left side of the face and left hand than on the right. Twice she had a slight haemoptysis and once a severe epistaxis. At the age of 15 years she had had a radiograph and cardiogram, but no definite diagnosis was reached. A cardiogram at this time showed probable right ventricular hypertrophy.

She was a well-developed, healthy-looking girl with no cyanosis, no clubbing, and no dyspnoea at rest. The right pulse was stronger than the left. Blood pressure (right) was 115/90 mm. Hg and (left) 90/70 mm. Hg. Femoral pulses were easily palpable. Blood pressure in the legs was 150/90 mm. Hg. The apex beat was diffuse, maximum in the fourth left interspace 3 in. from the midline. Heart sounds were well heard with a loud, coarse, systolic apical murmur and accentuation of the second pulmonic sound. Over a pulsatile dilated vessel above the right clavicle there was a systolic bruit and thrill. No such pulsation was visible over the left carotid.

Blood examination showed a degree of polycythemia with haemoglobin 19.4 g. per 100 ml. and R.B.C. 6,810,000 per c.mm.

**Cardiogram.**—S was prominent in leads 1 and aVL; Q prominent in leads 3 and aVF. Chest leads show evidence of considerable right ventricular hypertrophy and stress.

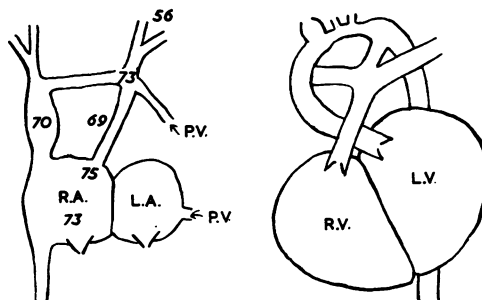
**Phonocardiogram.**—A systolic murmur was heard at the mitral area and was marked over the pulsatile swelling in the right side of the neck. At the second left interspace at the left sternal border there is a

split first sound and a loud second sound without obvious splitting; a systolic murmur occupies the early half of systole.

**Radiography and Fluoroscopy.**—The lung fields are clear. The heart shadow is slightly enlarged with a grossly dilated pulmonary artery. The right hilum pulsates forcibly. The right ventricle is moderately enlarged. The vascular shadow in the right upper mediastinum displaces the trachea and oesophagus to the left. Later it was possible to demonstrate the outline of a coarctation of the aorta and probable low patent ductus in the left anterior oblique view.

**Cardiac Catheterization.**—The catheter passed easily from the right arm into a dilated right atrium, whence it entered a left superior vena cava via the coronary sinus (Figs. 5 and 6). The right ventricle and pulmonary artery were not entered—auricular and venous tracings and pressures were obtained throughout. Oxygen saturations, as shown in Diagram 2, revealed oxygenated blood in both venae cavae and in the right auricle.

**Conclusion.**—This girl had at least four anomalies (Diagram 2): (1) Persistent left superior vena cava draining into a coronary sinus; (2) left pulmonary vein draining into the left superior vena cava with persistent communication between the superior venae cavae; (3) high coarctation of the aorta apparently of some length in the region of origin of the left carotid and left subclavian arteries; (4) a patent ductus arteriosus supplying the lower trunk and limbs under high pressure from the pulmonary artery and probably partly supplying the left carotid and subclavian arteries during exercise, viz., left-sided cyanosis during effort.



CASE 2

**CASE 3.**—M. G., a girl aged 14, the child of healthy parents, had one sister who died from convulsions in early infancy; she, herself, was a full-time child who weighed 6 lb. 5 oz. at birth. She was always undersized and easily tired; she became breathless and rather blue on exertion. She was frequently off school with minor ailments. At the age of 11 she had a mild haemoptysis and a year later she had a more profuse haemoptysis, for which she was treated in a children's hospital.

She was a small child who looked more like 8 than 14 years of age, yet radiographs of the skeleton showed a normal stage of development for her age. There was no clubbing. There was cyanosis of the

*Figs. 1 to 4, Case 1*

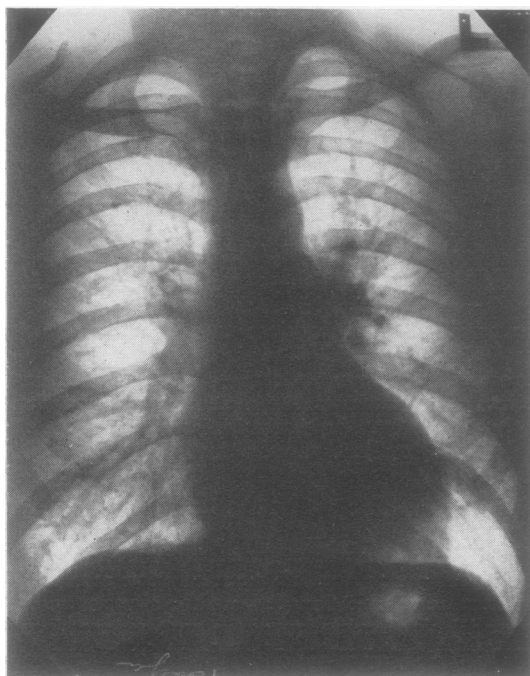


FIG. 1

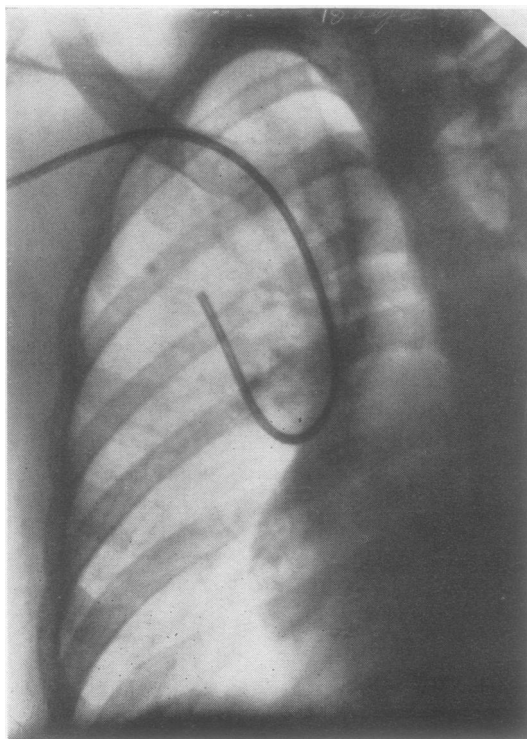


FIG. 2

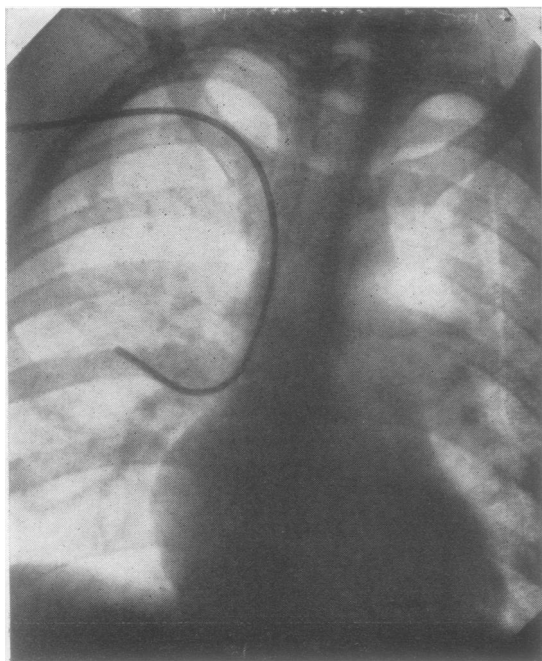


FIG. 3

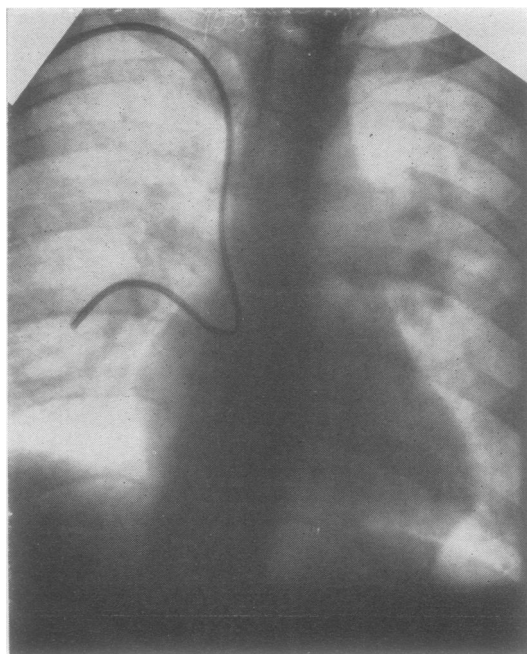


FIG. 4



*Figs. 5 and 6, Case 2*

**FIG. 1**—Postero-anterior radiograph. Prominent pulmonary artery and prominent hilar shadows; slight prominence of the left ventricular contour. Hilar dance noted on fluoroscopy.

**FIG. 2**—The catheter has entered a pulmonary vein draining the right upper lobe from a point at the junction of the superior vena cava with the left atrium. Oxygen saturation 94.6%.

**FIG. 3**—The catheter has entered a pulmonary vein draining the right middle lobe from the same point as in Fig. 2. Oxygen saturation, 95.5%.

**FIG. 4**—The catheter has entered the right atrium and emerged from a point lower down on the right heart border to enter a pulmonary vein draining the right lower lobe.

**FIG. 5**—Postero-anterior radiograph. Note the large pulmonary artery shadow. The double contour shadow in the right upper mediastinum represents a dilated superior vena cava and an aneurysmal innominate artery which displaces the trachea to the left.

**FIG. 6a**—The catheter passes through the right superior vena cava, right atrium, coronary sinus, and left superior vena cava to reach the left jugular vein. Oxygen saturations: left jugular vein, 56%; left superior vena cava, 73%; right superior vena cava, 70%.

**FIG. 6b**—The catheter is in the same position as in Fig. 5; left oblique view to show course through the heart.

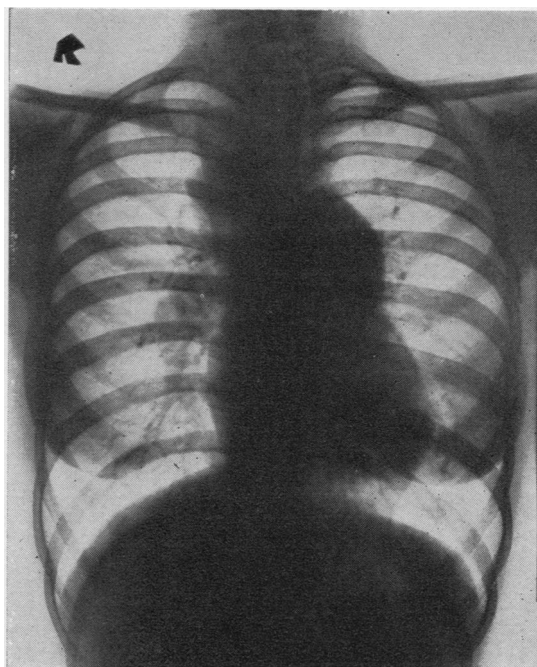


FIG. 5

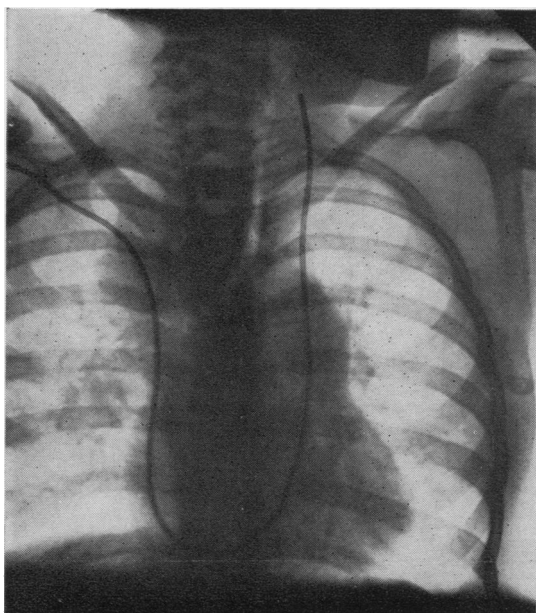


FIG. 6a

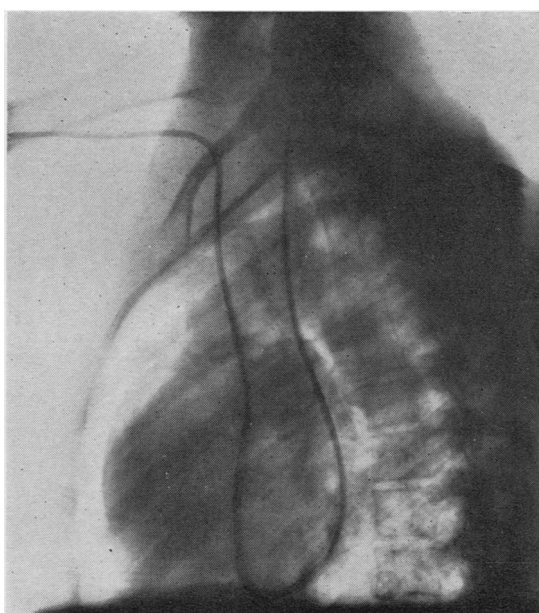


FIG. 6b

hands and feet which at times seemed greater in the hands than in the feet and at others appeared equal in hands and feet. There was no cyanosis of the face except when she was breathless after effort. The cardiac impulse was displaced to the left and a right ventricular heave was easily palpable. There was a loud, rather coarse systolic murmur over the praecordium with maximum intensity at the pulmonic area, and associated with a palpable thrill. When she first came under observation no diastolic murmur was detected, but a few weeks later a diastolic murmur was also present. Blood pressure was 95/75 mm. Hg in the arms, 120/90 mm. Hg in the legs. No collateral arterial circulation was detected.

**Cardiogram.**—There is considerable right ventricular hypertrophy and stress. Splintered QRS in leads 2, 3, VL, and VF, with slight notching of QRS in all chest leads but no conclusive signs of partial bundle branch block.

**Phonocardiogram.**—There is continuous systolic/diastolic murmur at the pulmonic area; a discrete systolic murmur and faint mid-diastolic murmur at the mitral area; split second sound with an interval of 0.04 sec. between the two components.

**Radiology and Fluoroscopy.**—Films showed hypoplastic aortic knuckle; rounded vascular shadow to the right of the sternum in the upper mediastinum, showing very little pulsation, originally thought to be the ascending aorta or thymus but later identified as the dilated superior vena cava (Fig. 7). A straight-edged shadow of similar density running downwards from the left sternoclavicular joint possibly represents a persistent left superior vena cava. Considerable enlargement of both right and left ventricles; no enlargement of left auricle. Engorged pulmonary artery with vigorous pulsation of main pulmonary trunks. Constriction of aorta visible in left anterior oblique position.

**Circulation Time.**—Arm-tongue (Decholin) 10.8 sec.

**Cardiac Catheterization.**—The right arm was used. The catheter passed without difficulty to the right ventricle, and after some difficulty entered the pulmonary artery; it could not be manipulated into the left branch or through a patent duct. In its course through the superior mediastinum it followed the left wall of the superior vena cava (Fig. 9), the right wall of which can be seen as a convex shadow projecting to the right; the width of the superior vena cava at this point is worthy of comment.

Pressures and oxygen content were as follows :

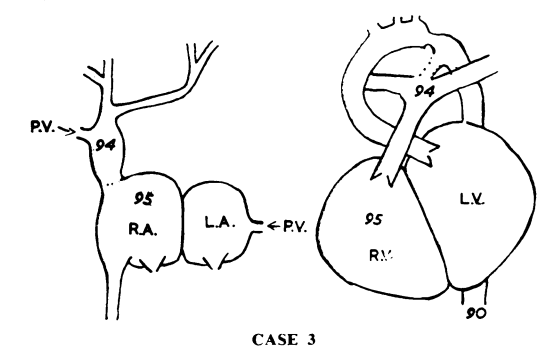
	Oxygen (Vol. per 100 ml.)	Per- centage Saturation	Pressures (mm. Hg)	
			Dynamic	Mean
Peripheral pulmon- ary artery ..	20.23	92.9	32 12	16
Main pulmonary ar- tery ..	20.50	94.2	38 6	18
Right ventricle ..	20.70	95.0	47 5	15
.. atrium ..	20.59	94.7	—	0
Superior vena cava ..	20.55	94.4	—	0
Femoral artery ..	19.53	90.3	—	0
O <sub>2</sub> capacity ..	21.70	100	—	—

(An oxygen sample from a more peripheral vein was unfortunately overlooked, as a pulmonary-systemic venous connexion was not suspected.)

With the catheter in the position shown in Fig. 10 the possibility of passage through the coronary sinus to a pulmonary vein was considered (or through an atrial septal defect to the left atrium and thence to the pulmonary vein), but the pressure curve was similar to that obtained from the main pulmonary artery and on withdrawal during continuous pressure recording the sequence of arterial, ventricular, and auricular pressure curves was obtained. The pulmonary artery pressure curve is unusual, showing four pressure rises with each cycle; two of these correspond to the two peaks of a bifid pressure rise in the right ventricle; the third occurs in early diastole and may be an aortic systolic impulse transmitted back to the pulmonary artery through a patent duct; the fourth corresponds in time to auricular systole. On the other hand these may be a Venturi effect due to "catheter dance."

**Angiocardiogram.**—The left subclavian and innominate veins are normal (Fig. 8). Right-sided superior vena cava considerably widened. Right auricle, right ventricle, and pulmonary artery large. Slow emptying of right auricle and right ventricle and pulmonary artery. No veno-arterial shunt demonstrated.

**Conclusion.**—This girl has : (1) Partial pulmonary venous drainage into a dilated superior vena cava, but a left superior vena cava has not been demonstrated with certainty; (2) coarctation of the aorta below the level of the left subclavian artery; (3) probable patent ductus arteriosus supplying the lower trunk and producing a higher blood pressure in legs than arms, despite the coarctation.

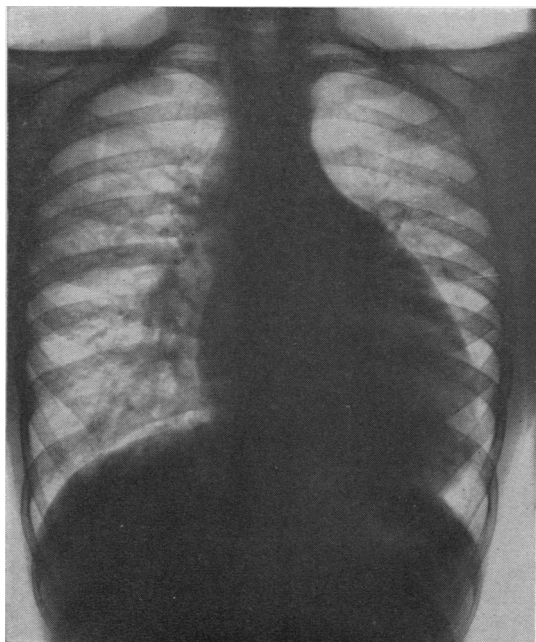


**CASE 4.**—M. T., a schoolgirl aged 10 years, had been cyanosed from birth and activities had always been strictly limited by shortness of breath. She had had measles, chickenpox, and pneumonia, but had never had signs of congestive failure.

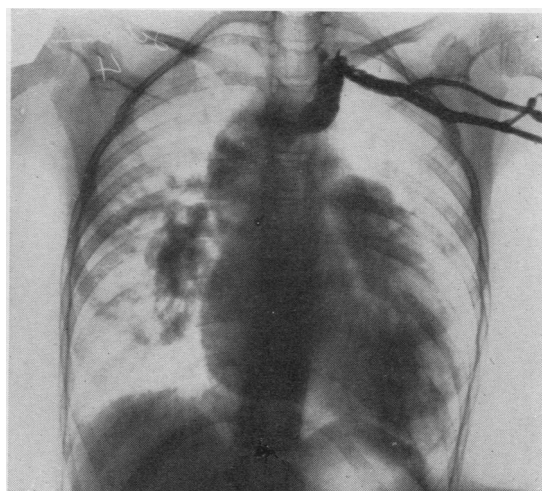
On examination, she was of normal stature and development for her age. There was cyanosis of the face, hands, and limbs at rest, of similar intensity. The toes were clubbed, but not the fingers. Both



*Figs. 7 to 10, Case 3*



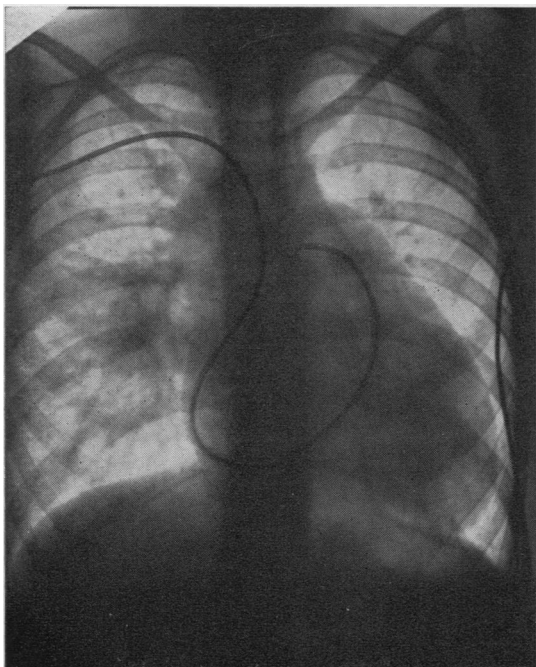
**FIG. 7**



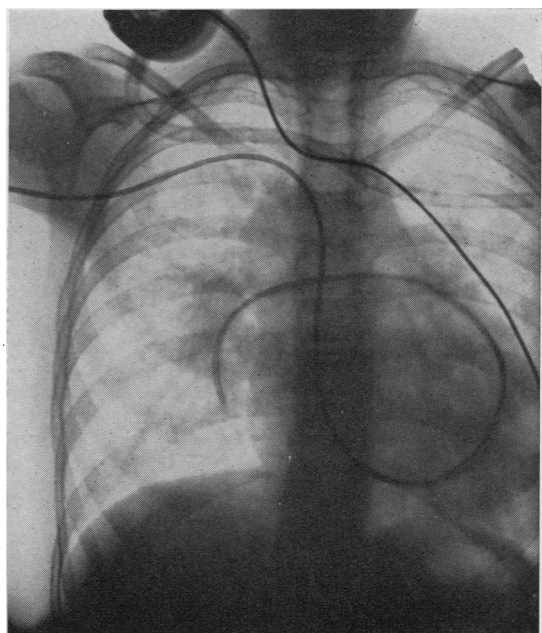
**FIG. 8**

**FIG. 7.**—Postero-anterior radiograph. The heart shadow large and globular; aortic knuckle small. Note prominent convexity of the right border of the superior mediastinum; this was at first thought to be the aorta, but showed only feeble pulsation on fluoroscopy.

**FIG. 8.**—Angiocardiogram at 4 sec., showing a dilated superior vena cava, right auricle, right ventricle, and pulmonary artery with delayed emptying of the right side of heart.



**FIG. 9.**—The catheter passes along the medial wall of the superior vena cava to enter the right atrium, right ventricle, and main pulmonary artery. The convex shadow to the right of the superior mediastinum can now be identified as the dilated superior vena cava; its left edge is outlined by the catheter and the gross dilatation of the vessel is well demonstrated. Oxygen saturations: right superior vena cava, 94.4%; pulmonary artery, 94.2%.



**FIG. 10.**—The catheter has been advanced from the position shown in Fig. 8 to the right lower pulmonary artery. Note the altered position of the catheter within the heart shadow, illustrating how far the right ventricle extends towards the left. A pressure curve in this position was arterial in type, and on withdrawal arterial, ventricular, and auricular curves were obtained in succession, thus excluding the possibility that the catheter had passed through a septal defect to the left atrium and thence to a pulmonary vein.

Figs. 11 and 12, Case 4

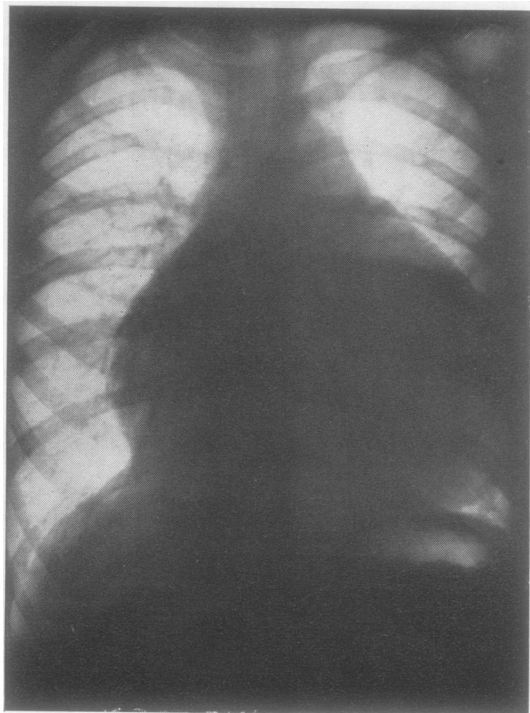


FIG. 11

radial pulses were of poor volume and blood pressure readings were not recordable. Femoral pulses were palpable. The apex beat was palpable in the fifth interspace an inch outside the mid-clavicular line. The heart rate was 110/min., regular with gallop rhythm and a systolic murmur, well heard in the mitral area and at the left sternal border. Liver was palpable two fingerbreadths below the costal margin. The spleen was not enlarged.

**Cardiogram.**—Tall, notched P wave in leads 1 and 2. Well-marked evidence of right ventricular hypertrophy with inverted T waves from V1 to V6.

**Phonocardiogram.**—At the mitral area there is a soft first sound, a mid-systolic murmur, and a loud second sound. At the pulmonic area there is a murmur throughout systole. The second sound is accentuated.

**Radiology and Fluoroscopy.**—There is considerable generalized enlargement of the heart shadow, which is globular in shape. In the left anterior oblique view the right ventricle is particularly enlarged. The aortic knuckle is small and the aorta hypoplastic. In the postero-anterior view the right superior vena cava is clearly shown, while a similar shadow running downwards below the left sterno-clavicular joint is very suggestive of a left superior vena cava (Fig. 11).

FIG. 11.—Postero-anterior radiograph. The heart shadow is globular and grossly enlarged. The right superior vena cava is clearly visible and a corresponding shadow on the left side probably represents a persistent left superior vena cava. The left anterior oblique view in this case showed especial enlargement of the right ventricle and a small hypoplastic aorta.

FIG. 12.—Angiocardiogram at 4 seconds showing filling of the right atrium, right ventricle, and pulmonary artery. The collateral veins in the axilla and root of neck have been entered by the dye, but there is no trace of dye entering a left innominate vein.

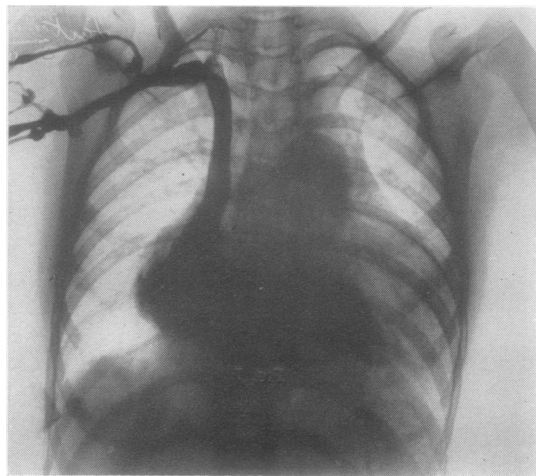


FIG. 12

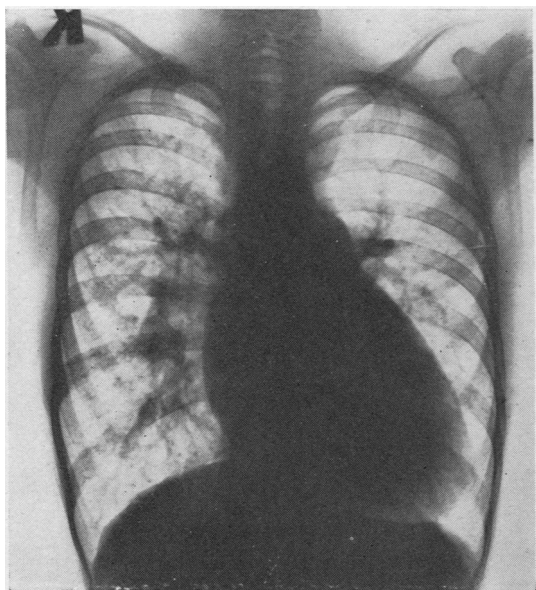
**Angiocardiography.**—The right auricle, right ventricle, and pulmonary artery are enlarged, but the most striking feature is their slow rate of emptying. Another noteworthy feature is the fact that, while tributaries of the subclavian vein are outlined by the dye, there is no evidence of a left innominate vein joining the superior vena cava (Welsh, 1955, personal communication; Fig. 12).

**Cardiac Catheterization.**—This was attempted via the left median basilic vein under general anaesthesia with pentothal, flaxedil, nitrous oxide, and oxygen. The catheter did not pass beyond the subclavian vein and death occurred during the procedure. During attempted resuscitation it was noted that the right arm and face were bright pink while the left side was intensely cyanosed. The left radial pulse was felt while the right pulse was imperceptible. The right jugular vein was engorged, the left empty.

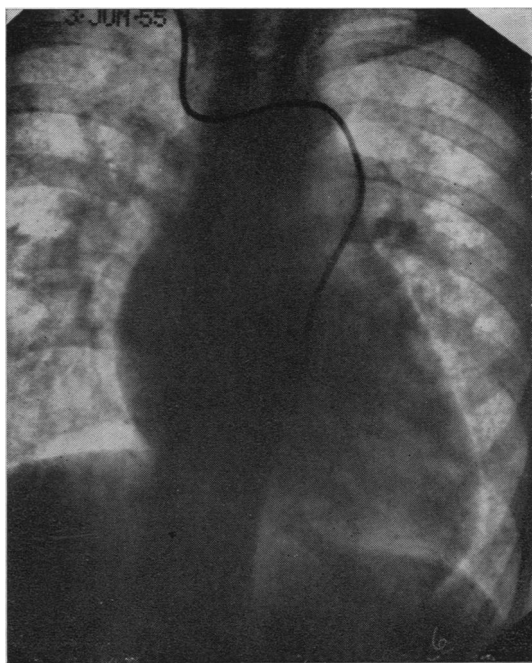
**Necropsy Findings.**—The right subclavian and jugular veins joined a right vena cava which entered the right auricle. The left subclavian and jugular veins formed a left superior vena cava which entered the left auricle. There was no communicating innominate vein and no evidence of catheter vein damage. The inferior vena cava, right auricle, and right ventricle were grossly dilated, but there was no valvular stenosis or significant septal defect. The



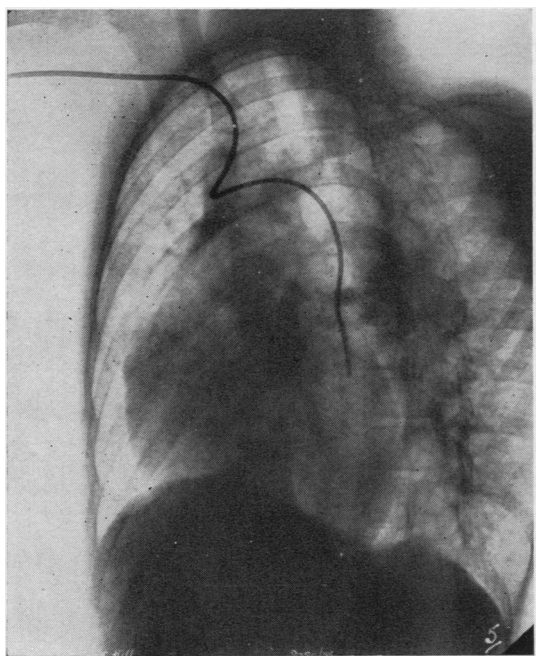
*Figs. 13 to 19, Case 5*



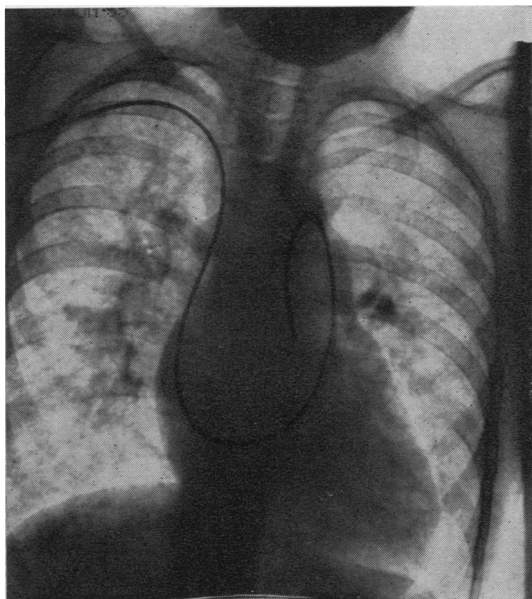
**FIG. 13.**—Postero-anterior radiograph. Right pulmonary artery and hilar shadows enlarged. A shadow running downwards from the left sterno-clavicular joint suggests a left superior vena cava. Fluoroscopy showed well-marked hilar dance, and considerable enlargement of the right ventricle was noted in the left anterior oblique view.



**FIG. 14.**—Catheter has passed from the right superior vena cava through a connecting vein to the left superior vena cava and thence through the coronary sinus to the right atrium.



**FIG. 15.**—Same position of catheter; left oblique view to show its intracardiac course.



**FIG. 16.**—The catheter has been retracted to the subclavian vein and reintroduced. It now continues along the right superior vena cava to the right auricle, right ventricle, and pulmonary artery, from whence it passes through a patent ductus arteriosus to the descending aorta. Note that the intracardiac course leads clearly into the pulmonary artery and not into the ascending aorta.



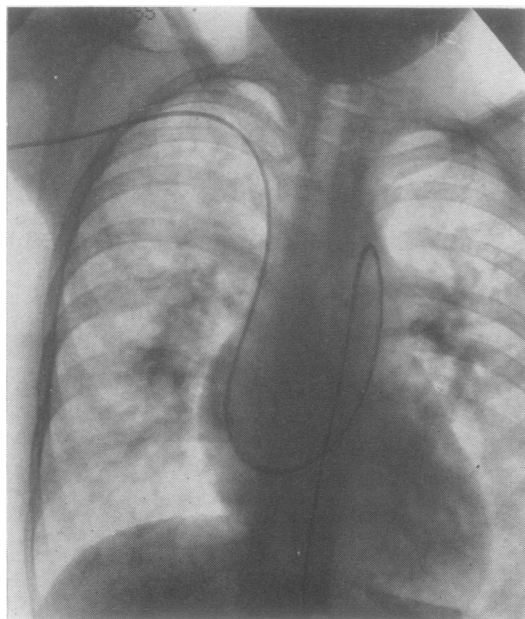


FIG. 17.—The catheter has been further advanced to abdominal aorta.

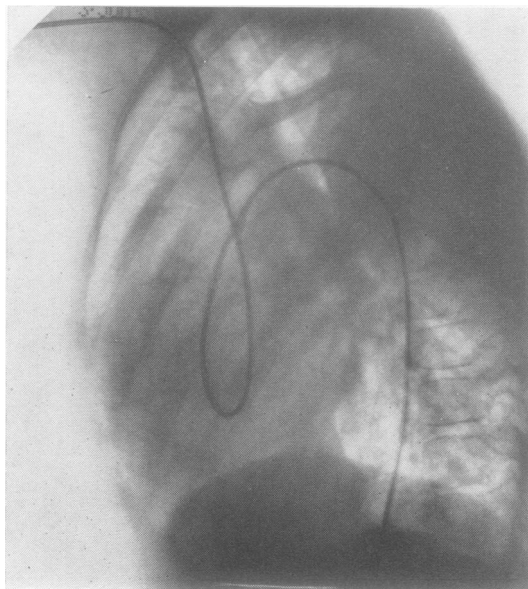
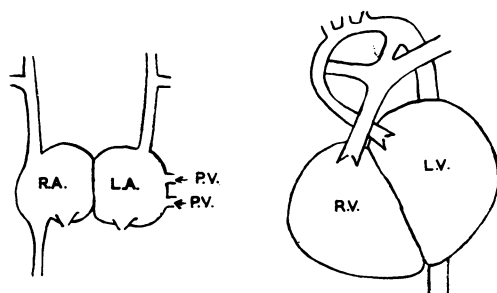


FIG. 18.—Same position as Fig. 16, left oblique view.

pulmonary veins entered the left auricle in the normal manner. The left auricle and left ventricle were small chambers, though thick-walled. The aorta was hypoplastic, hardly larger than a normal femoral artery, but it widened considerably after the entry of the patent ductus arteriosus.

*Comment.*—This girl had a left superior vena cava draining into the left auricle, a hypoplastic aortic arch, and a patent ductus arteriosus. Catheterization produced spasm and closure of the left superior vena cava with consequent diminished filling of and output from the left heart, which was thus unable to supply the brain under sufficient pressure through the hypoplastic aorta. The case is described in greater detail and discussed in a separate article (Peel, Kelly, Blum, and Semple, 1956).



CASE 4

CASE 5.—R. H., a schoolboy aged 15 years, was known to have a cardiac lesion at birth. He was slow in developing, being bottle-fed till the age of 4 years and unable to walk until nearly 3 years. He vomited after feeds until about the age of 10 years. He had never squatted. Mental development was normal and he was not slow in talking. He had a good performance at school and he was an excellent pianist.

He had always been breathless on exertion, small for his age, and slightly cyanosed. The parents and two older siblings were healthy. On examination he was a small intelligent lad, weighing 5 stones. Radiological examination of bones showed some delay in development, about two years younger than his age. There was slight cyanosis of the face, lips, and fingers, but no clubbing. In contrast, his feet and toes were normal in colour. All peripheral pulses were palpable. The blood pressure in both arms was 150/96 mm. Hg, in both legs 116/90 mm. Hg. He was pigeon-chested with a diffuse apex beat, 96 per minute and regular. There was a right ventricular impulse and a palpable pulmonic second sound. A loud blowing systolic murmur was present at the apex and soft systolic murmur over the base and into the neck. Other systems were normal. The urine was clear. The E.S.R. was 2 mm. in the first hour. Haemoglobin was 116% ; R.B.C. 5,750,000 per c.mm.

*Cardiogram.*—P waves were slightly notched in lead 1, prominent in leads 2 and 3. PR interval 0.16 sec. There is right axial deviation and chest leads

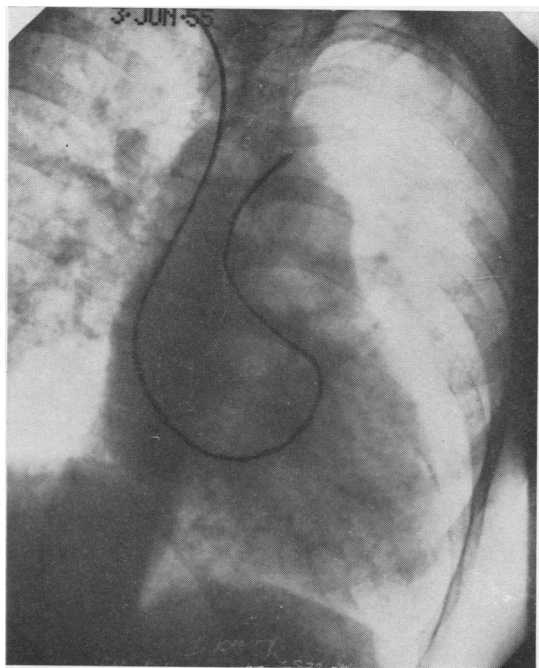


FIG. 19.—The catheter has been retracted to the right ventricle and then advanced into the ascending aorta. Oxygen saturations: coronary sinus, 27.2%; right atrium, 26.8%; right ventricle (low), 58.2%; pulmonary artery, 87.7%; ductus arteriosus 91.9%; abdominal aorta, 92.8%.

show considerable right ventricular hypertrophy and stress.

**Radiology and Fluoroscopy.**—The heart is considerably enlarged in all diameters (cardio-thoracic ratio 14.5:21.5, oblique diameter 17.8 cm.). Both ventricles are enlarged, as also are the pulmonary artery and branches. Hilar dance is well demonstrated. A shadow suggesting a persistent left superior vena cava is visible below the left sterno-clavicular joint (Fig. 13).

At this stage a diagnosis was made of partial transposition associated with a patent ductus and persistent left superior vena cava; the disparity in cyanosis of hands and feet was considered most significant.

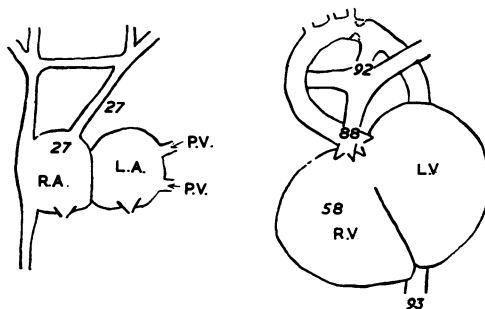
**Cardiac Catheterization.**—(1) The catheter passed from the right arm into the superior vena cava, thence across by the innominate vein to a left superior vena cava and into the right auricle via the coronary sinus (Figs. 14 and 15). (2) The catheter was retracted and passed directly via the right superior vena cava into the right auricle, right ventricle, and pulmonary artery. It then entered a patent ductus and proceeded downwards into the abdominal aorta (Figs. 16, 17, and 18). (3) Once more the catheter was retracted into the right ventricle and passed via a high ventricular septal defect into the ascending aorta. Unfortunately, owing to catheter blockage no pressure readings or samples were possible from this site (Fig. 19).

M

Pressures and oxygen content were as follows:

	Oxygen (Vol. per 100 ml.)	Per- centage Saturation	Pressures (mm. Hg)	
			Dynamic	Mean
Left superior vena cava ..	5.48	26.9	—	—
Coronary sinus ..	5.56	27.2	—	2
Right atrium ..	5.48	26.9	—	—1
„ ventricle ..	11.87	58.2	100 0	45
Pulmonary artery ..	17.88	87.7	75 55	60
Ductus arteriosus ..	18.72	91.9	80 50	65
Abdominal aorta ..	18.92	92.8	85.65	73
Venous oxygenated	20.40	100		

**Conclusion.**—Catheterization confirmed the clinical impression of partial transposition of great vessels, with a patent ductus arteriosus, and a persistent left superior vena cava. This was found to drain via the coronary sinus into the right auricle. Blood pressure readings lower in the legs than in the arms might also indicate the presence of a degree of coarctation of the aorta.



CASE 5

**CASE 6.**—J. McC., a labourer aged 18 years, was rejected at a medical examination for the Royal Navy in August, 1954, on account of a "heart murmur." He was referred to hospital on this account, but admitted to no symptoms. He played football and did heavy work without breathlessness. Apart from the usual childhood illnesses, he had always been in good health. Five sisters and four brothers were healthy; one sister aged 13 years had been blue since birth; one brother died of gastro-enteritis aged 3 months.

On examination he was a healthy-looking lad with a fresh complexion. His hands were highly coloured with moist palms. He had no cyanosis. The apex beat was in the normal position. Heart sounds were 84/min., regular. Sounds were normal at the mitral area. A soft systolic murmur at the base was abolished on full inspiration and thought at the time to be innocent in character. Blood pressure was 156/80 mm. Hg. Other systems were healthy. The blood picture was normal: haemoglobin 97%; R.B.C. 5,060,000 per c.mm.

**Cardiogram.**—This showed partial right bundle branch block.



Figs. 20 and 21, Case 6

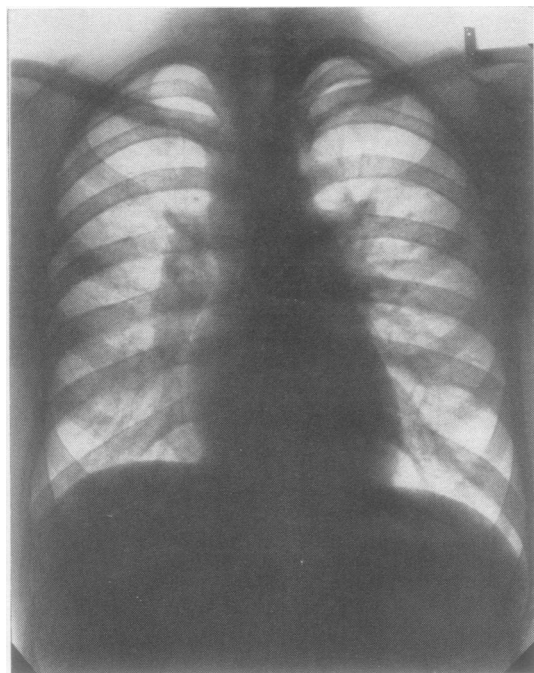


FIG. 20.—Postero-anterior radiograph. Heart shadow within normal limits; hilar shadows enlarged; superior vena cava prominent.

**Phonocardiogram.**—This showed slight splitting of both first and second sounds: systolic murmur, no diastolic murmur.

**Radiology and Fluoroscopy.**—The heart on measurement was within normal limits (Fig. 20), apart from broadening of the right upper mediastinum. Moderate enlargement of right ventricle on fluoroscopy and considerable branching and digitation of pulmonary arteries, suggesting the presence of an atrial septal defect. The barium-filled oesophagus shows marked indentation at the pulmonary conus level.

As the cardiogram and fluoroscopy were suggestive of a congenital cardiac lesion, it was decided to proceed with catheterization.

**Cardiac Catheterization.**—Three attempts at catheterization via a left median basilic vein were unsuccessful, the catheters on each occasion sticking near the junction of the left subclavian with the jugular.

On the other hand, the catheter passed easily via a right median basilic vein into the superior vena cava and right atrium. Here its end became bent into a "J-shaped" loop which could not be undone, and in this condition it entered the right ventricle and pulmonary artery. During manipulation of the patient to obtain right and left oblique views, the catheter slipped back into the right atrium and thence entered

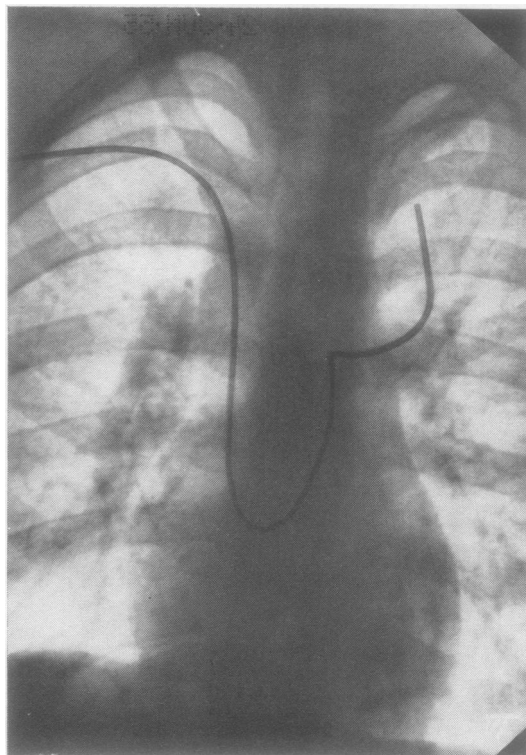


FIG. 21.—The catheter passes from the right superior vena cava into the right atrium and thence to a persistent left superior vena cava from which it enters a pulmonary vein draining the left upper lobe. The oxygen saturations confirm a connexion between the right and left superior venae cavae. Oxygen saturations: right superior vena cava, 93.6%; right atrium, 92.0%; pulmonary vein, 92.9%; pulmonary artery, 95.8%.

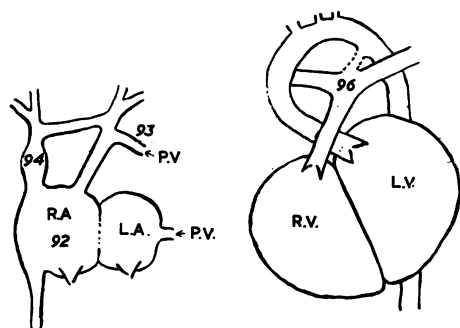
a left superior vena cava, from where it passed into a pulmonary vein tributary from the left upper lobe (Fig. 21). Blood oxygen figures from these sites (see diagram) confirmed that a pulmonary vein entered the left superior vena cava which also communicated with the right superior vena cava.

	Oxygen (Vol. per 100 ml.)	Per- centage Saturation	Pressures (mm. Hg)	
			Dynamic	Mean
Right superior vena cava	16.85	93.6	—	—
Left superior vena cava	—	—	—	—
Left upper pulmon- vein	16.73	92.9	6.2	4
Right atrium	16.57	92.0	3.0	1½
ventricle	—	—	30-1	7
Pulmonary artery	17.24	95.8	30.10	18

**Conclusion.**—This lad, who had no symptoms and whose heart was passed clinically as probably normal, had a left superior vena cava into which there was pulmonary venous drainage from the left lung. It was not possible to exclude the presence of additional



defects; indeed a very high oxygen content of blood from the pulmonary artery suggested the possibility of an associated patent ductus, while the presence of right bundle branch block was in keeping with atrial septal defect.



CASE 6

#### DISCUSSION AND SUMMARY

The literature concerning anomalous systemic and pulmonary venous drainage is reviewed and six illustrative cases are added. These illustrate:

(1) Right pulmonary veins entering the right atrium in a man of 25 years with an aorto-pulmonary communication and pulmonary stenosis but little disability.

(2) Pulmonary veins joining a persistent left superior vena cava, draining to the right atrium via the coronary sinus and also connected to the right superior vena cava, in a young woman of 19 years with aortic coarctation and patent ductus arteriosus. Despite the anatomical findings she is not at present seriously disabled, but the outlook is considered poor.

(3) Pulmonary veins entering a dilated right superior vena cava. This girl of 14 years also has coarctation of the aorta and a patent duct; she is undersized and is capable of less exertion than Cases 1 or 2.

(4) Persistent left superior vena cava entering the left atrium and producing cyanosis in a girl of 10 years with a hypoplastic aorta and a patent duct. Attempted catheterization through the left arm induced spasm of the jugular vein and caused death from reduced left ventricular output and cerebral anoxia.

(5) Persistent left superior vena cava draining into the right atrium through the coronary sinus and connected also to the right superior vena cava. Pulmonary venous draining normal, to the left atrium. This boy of 15 years has partial transposition of the great vessels with a patent ductus arteriosus. He is undersized, breathless on exertion, and slightly cyanosed but with normally coloured feet and toes.

(6) Left pulmonary veins entering a left superior vena cava which drains into the right atrium. An atrial septal defect is suspected in view of partial right bundle branch block in the cardiogram, but has not been proved with certainty. This lad of 18 years is symptom-free, and his heart would have been passed as normal clinically and radiologically but for the presence of hilar dance and partial bundle branch block.

A persistent left superior vena cava can usually be suspected from careful examination of the postero-anterior radiograph. It usually enters the right atrium, either directly or via the coronary sinus. In these circumstances, and provided it does not receive pulmonary venous drainage, it is of academic rather than practical importance. Frequently, however, it receives pulmonary venous drainage, in which case an arterio-venous shunt is present; this in turn may lead to right-sided cardiac hypertrophy with pulmonary plethora and ultimate right heart failure. Rarely the left superior vena cava enters the left atrium, producing a veno-arterial shunt and cyanosis. Additional malformations are usually present.

Catheterization through the left arm may be difficult in patients with a left superior vena cava; obstruction is often met at the junction of the subclavian and jugular veins where they cross the first rib. The attempt can be highly dangerous in cases where the left superior vena cava enters the left atrium, especially if there is coarctation or hypoplasia of the aorta, but we feel that in such a case catheterization through the right arm would carry no more than the usual risk.

Anomalous pulmonary venous drainage can sometimes be suspected by noting unusual dilatation of the superior vena cava in the radiograph, by the presence of a left superior vena cava, or by shadows curving downwards along the left or right heart border. When it is suspected, blood samples should be obtained at catheterization from the subclavian vein as well as from the superior vena cava, which has a significantly higher oxygen content. The presence of a pulmonary-systemic communication renders the diagnosis of a concomitant intracardiac shunt more difficult owing to the presence of oxygenated blood in the superior vena cava and right atrium; unless the catheter enters a septal defect or patent duct, the concomitant lesion may remain suspected rather than proven. Finally, anomalous pulmonary venous drainage into the right atrium via a left superior vena cava in the absence of a communication with the right superior vena cava could mimic

an atrial septal defect in so far as blood oxygen figures are concerned.

It has been shown that pulmonary venous drainage to the right heart, systemic venous drainage to the left heart, and transposition may all in turn mask the cardinal physical sign of infantile type coarctation with patent ductus, that is, cyanosis confined to the lower trunk and limbs.

From the surgical standpoint, three procedures suggest themselves. Where one or more pulmonary veins drain into the right atrium producing signs of right heart strain or failure, one might consider lobectomy or pneumonectomy to relieve the right heart of the additional load of oxygenated blood. In some such cases also it may be possible to transplant the offending pulmonary vein into the left atrium or into a normally draining pulmonary vein. Lastly, in the rare event of a left superior vena cava draining into the left atrium, ligation might be considered, but only after careful search for additional abnormalities.

We are much indebted to Dr. Ian Anderson, Mr. A. P. Kenny, and Mr. A. Jamieson for the blood oxygen analyses, to Dr. J. G. Stevenson for angiocardigrams, and to Mr. H. C. Gray for the photographic reproductions of the radiographs. We also gratefully acknowledge the assistance of the nursing staff and the radiographers during catheterizations.

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