PERSISTENT LEFT SUPERIOR VENA CAVA DRAINING THE PULMONARY VEINS

BY

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Persistence of the left superior vena cava is not uncommon in children with congenital heart disease and is occasionally found in adults at thoracotomy. The anomalous vena cava usually carries venous blood from the upper part of the body to the right atrium via the coronary sinus and has little functional significance. In the four patients we describe, however, the left superior vena cava drained arterial blood from some or all of the pulmonary veins into the left innominate vein, and thence into the right atrium. In three of them it was the sole channel by which oxygenated blood could reach the systemic circulation.

We record these cases because we believe that the anomaly is more common than has been thought, and gives a clinical picture sufficiently distinctive to be recognized in life, and because it is one for which surgical relief may prove feasible. We have found only two similar cases of which an account of the physical signs has been given and in neither was the diagnosis made in life (Taussig, 1947).

Before describing the clinical picture it is necessary to consider briefly the development of the superior vena cava.

EMBRYOLOGY

Marshall (1850), a century ago, appears to have been the first to give an embryological explanation of the persistence of a left superior vena cava.

A modern account of the anomaly (Hamilton *et al.*, 1945; Patten, 1946) is as follows. The venous system arises from the same network as the arterial system and the capillaries. Very soon three pairs of venous channels are recognizable, namely (i) the two vitelline veins, which pass from the yolk sac along the roof of the primative gut to meet (ii) the umbilical veins at the sinus venosus and (iii) the cardinal veins which also enter the sinus venosus. Each cardinal vein consists of two parts, the anterior cardinal or precardinal vein and the posterior cardinal or postcardinal vein. The precardinal and postcardinal veins unite on each side of the primative heart tube to form a short vessel, the common cardinal duct or duct of Cuvier which enters the horn of the sinus venosus (Fig. 1).

The lung bud arises as an outgrowth from the primitive foregut. Both these structures are covered by a common venous network, the splanchnic plexus. The contiguous primitive pulmonary venous plexus and primitive splanchnic venous plexus drain into the paired precardinal and postcardinal veins and into the vitello-umbilical veins (Fig. 3A). The paired precardinal veins of the embryo supply the venous drainage of the part of the body above the level of the sinus venosus. The postcardinal veins, by a complicated series of changes, give rise to the inferior vena cava and its tributaries. We are only concerned here with the precardinal veins.

Initially the precardinal veins are short, but they elongate as the neck develops and as the heart recedes into the thorax. During this time an obliquely directed transverse vein arises, which connects the two precardinal veins just caudal to the entrance of the veins of the forelimb. This transverse anastomosis forms the left innominate vein. The left precardinal vein, from the point where the left innominate vein leaves it to the left duct of Cuvier, normally becomes obliterated except for its most cephalad part which

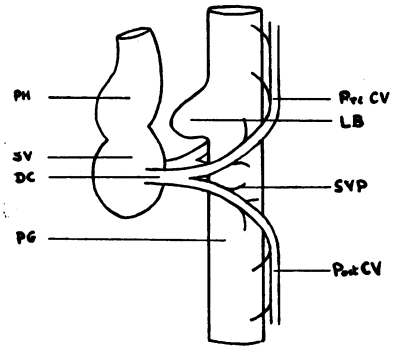


FIG. 1.—Diagram to show how the paired pre- and postcardinal veins receive blood from the venous plexus covering the primitive gut and lung bud, and deliver it by means of the duct of Cuvier into the sinus venosus. PH, primitive heart; SV, sinus venosus; DC, duct of Cuvier or common cardinal duct; Pre CV, precardinal vein; Post CV, postcardinal vein; PG, primitive gut; LB, lung bud; SVP, splanchnic venous plexus communications.

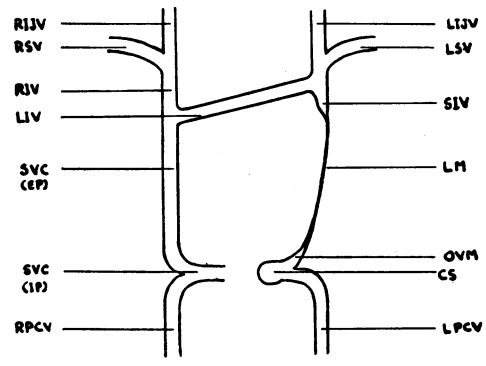


FIG. 2.—Diagram illustrating normal fate of the precardinal veins in man (see text for description). RIJV and LIJV, right and left internal jugular veins; RIV and LIV, right and left innominate veins; SVC(EP) and SVC(IP), superior vena cava, extra- and intrapericardial portions: RPCV and LPCV, right and left postcardinal veins; SIV, superior intercostal vein; LM, ligament of Marshall; OVM, oblique vein of Marshall; CS, coronary sinus.

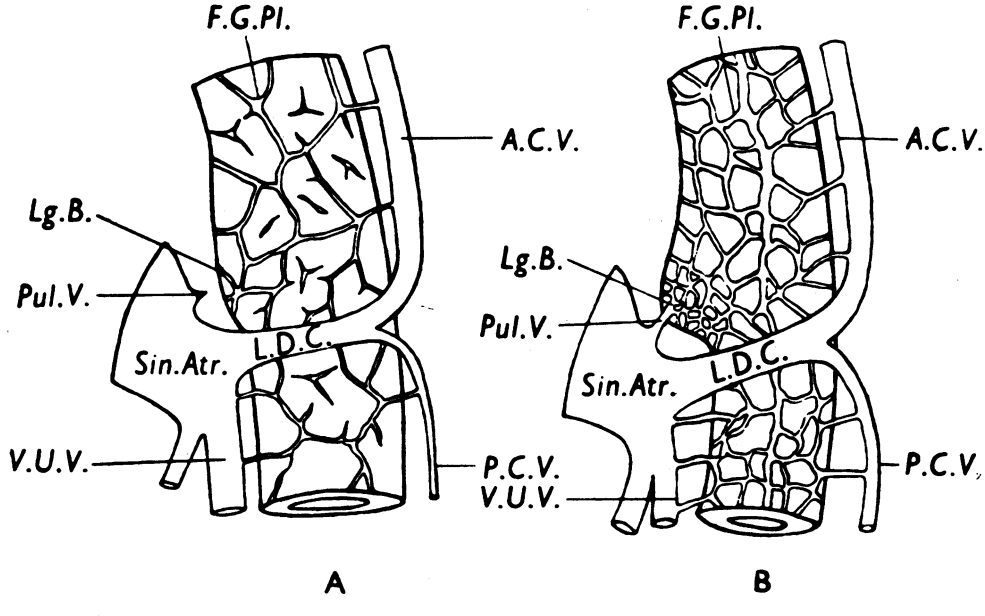


FIG. 3.—Diagram from H. Butler illustrating the formation and routes of drainage of the foregut venous plexus in the rat. (A) 3 *mm. rat embryo.* The foregut plexus (F.G.Pl.) drains into the cardinal and vitello-umbilical veins (V.U.V.). The cardiac part of the pulmonary vein has not yet "tapped" the foregut plexus. (B) 4·5–6 *mm. rat embryo.* The foregut plexus drains into cardinal, vitello-umbilical and pulmonary veins.

gives rise to the left superior intercostal vein (Fig. 2). The obliterated portion forms a fibrous band represented in the adult by the ligament of Marshall. The left duct of Cuvier and the left horn of the sinus venosus persist as the oblique vein of Marshall and the coronary sinus. That part of the right precardinal vein between the left innominate vein and the right duct of Cuvier forms the extrapericardial part of the superior vena cava, while the right duct of Cuvier forms its intrapericardial portion. The right horn of the sinus venosus is absorbed into the wall of the right atrium.

From the foregoing it will be seen that if the left precardinal vein is not obliterated it will persist as a left superior vena cava, receiving blood from the left innominate, internal jugular, and subclavian veins, and emptying into the right atrium at the coronary sinus. This is the commonest anomaly. The transverse anastomosis between the paired superior venæ cavæ may be absent and when this is so the venous drainage from each side of the upper part of the body is independent and the coronary sinus is large. At times the right superior vena cava is absent and the right innominate vein drains into the left superior vena cava.

Closely connected with persistence of the left superior vena cava are malformations of the venous return from the lungs. The stem of the pulmonary vein arises as an outgrowth from the dorsal wall of the sinus venosus (Butler, 1952) and, growing dorsally, forms the pulmonary part of the splanchnic venous plexus (Fig. 3B). Thereafter the pulmonary venous plexus drains directly into the heart and its communications with the cardinal and vitello-umbilical veins become obliterated. Failure of the cardiac part of the pulmonary vein to develop or to join the pulmonary venous plexus may result in persistent drainage of part or all of this plexus into the cardinal veins and thence either directly or indirectly into the right atrium.

Classification of Abnormalities

There are three ways by which the pulmonary veins can drain into the right atrium: they may open directly into its posterior wall; they may open into the coronary sinus; or they may drain into a persistent left superior vena cava and thence into the innominate vein, the right superior vena cava, and the right atrium. In any of these circumstances patency of the atrial septum is essential for life because it is the only way by which blood can reach the left heart.

McManus (1941) classified the various types of persistent left superior vena cava as follows.

I. Persistent left superior vena cava (*S.V.C.*) with right *S.V.C.* (bilateral *S.V.C.*).
   (A) Persistent left *S.V.C.* connected to the coronary sinus, and either with a cross anastomosis (left innominate vein) or without this.
   (B) Persistent left *S.V.C.* draining the pulmonary veins.

II. Persistent left *S.V.C.* without right *S.V.C.*

Our four cases are examples of Group I(B), namely persistent left superior vena cava draining the pulmonary veins.

Case Records

Case 1. M.M., a woman, aged 24, was admitted in January, 1949, complaining of dyspnœa on exertion. Breathlessness after feeds had first been noticed at the age of three months. She had attended a special school and had always been somewhat short of breath on walking uphill, though she could walk on the level without discomfort and had even played badminton. In July, 1948, she had become pregnant and at 22 weeks abdominal hysterotomy and sterilization had been performed.

On examination she was of small build (weight 93 lb.; height 5 ft., 1 in.). There was slight cyanosis and minimal finger clubbing. The pulse was 78 and regular; the blood pressure was 95/65 in the arms.

The left side of the chest was prominent and associated with considerable scoliosis. The apex beat was in the sixth intercostal space, 12·5 cm. from the mid-line and there was a forcible impulse over the outflow tract of the right ventricle and the pulmonary artery. The first sound at the apex was loud and widely split. A systolic murmur was audible over the whole præcordium, maximal in the second space near the left sternal border and louder in the upright position. An early diastolic murmur was heard in the third left space.

On fluoroscopy the right ventricle was considerably enlarged (Fig. 4A). The aortic knob was small. The pulmonary artery was prominent and its branches were large and showed expansile pulsation. There was a rounded pulsating shadow enveloping the great vessels in the superior mediastinum. A barium swallow was normal.

The electrocardiogram showed sinus rhythm, slight latent A-V block (P–R interval 0·22 sec.), and

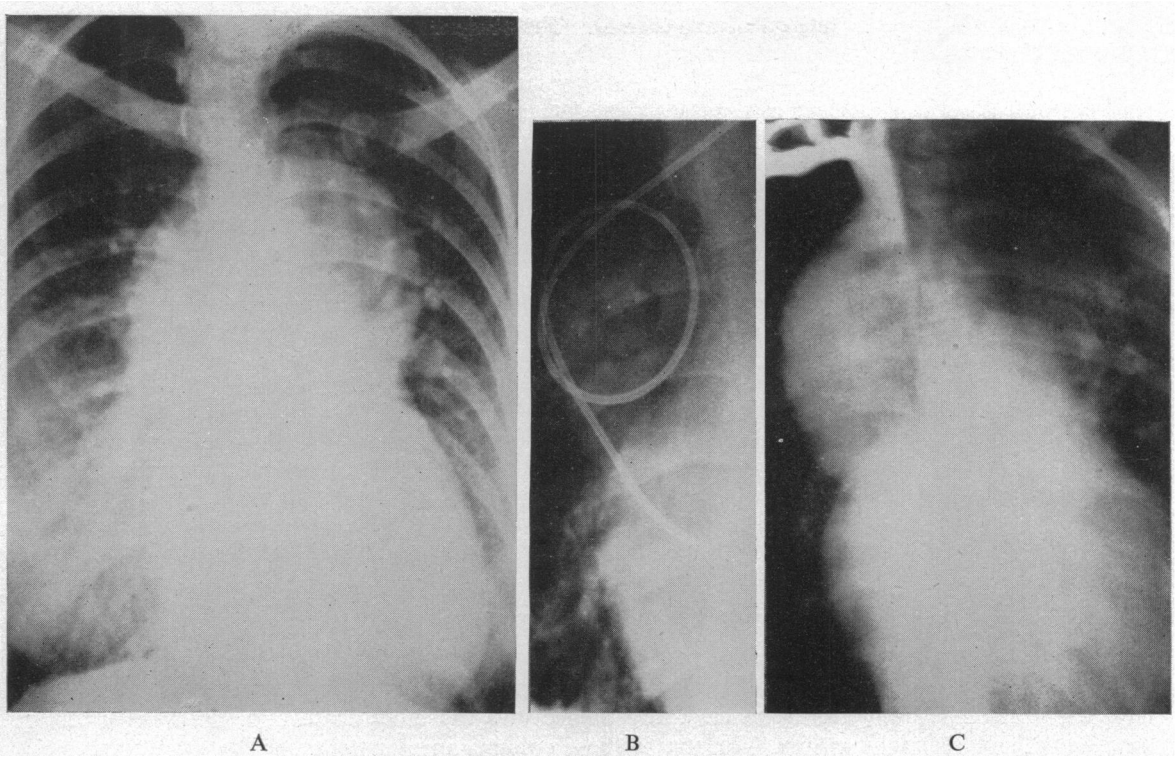


FIG. 4.—Case 1. (A) Teleradiogram showing the rounded shadow of the venæ cavæ in the superior mediastinum enveloping the aortic knob and pulmonary trunk. (B) A–P film showing cardiac catheter coiled in the right superior vena cava. (C) Angiocardiogram, taken 2 sec. after the beginning of injection, showing the dilated right superior vena cava and simultaneous filling of the small aorta and large pulmonary trunk.

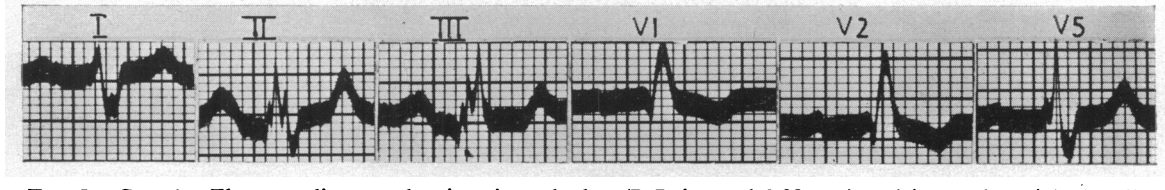


FIG. 5.—Case 1. Electrocardiogram showing sinus rhythm (P–R interval 0·22 sec.) and incomplete right bundle branch block.

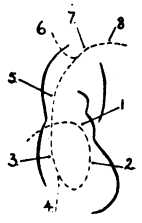
incomplete right bundle branch block (Fig. 5). Red blood corpuscles 5·5 million per cu. mm.; hæmoglobin 112 per cent.

The results of cardiac catheterization are summarized in Table I. The catheter coiled up in the greatly dilated right superior vena cava (Fig. 4B). The left atrium was reached without difficulty but the pulmonary veins could not be entered. The mean pressures in the right ventricle and pulmonary artery were a little high. The oxygen saturation of blood in the left innominate vein was 91 per cent and in the right internal jugular vein and the left subclavian vein it was 68 and 84 per cent respectively. The arterial blood oxygen saturation was 85 per cent.

The angiocardiogram taken 2 seconds after the injection of diodone showed dilatation of the right *S.V.C.* and simultaneous filling of the hypoplastic aorta and the large pulmonary arteries. The 5-second film showed diodone in the left *S.V.C.* and refilling of the right *S.V.C.*

In this patient it is thought that all the pulmonary veins drain into the left superior vena cava and that the systemic circulation is entirely dependent on the shunt through a patent foramen ovale or an atrial septal defect.

TABLE I. CASE 1, M.M., ÆT. 24



Sample of blood from:	O_2 content /100 c.c. bld.	O_2 capacity /100 c.c. bld.	Percentage sat'n.	Pressure mm. Hg. above skin of back
1. M.P.A.	16·1	17·2	93	24
2. R.V.			93	24
3. R.A.	—	—	79–87	13
L.A.	—	—	84	13
4. I.V.C.	12·0	—	70	—
5. S.V.C.	—	—	93	—
6. R.Int.Jug.V. ..	11·73	—	68	—
7. L.Innom.V.	—	—	91	—
8. L.Subclav.V. ..	14·4	—	84	—
Brachial artery ..	14·66	—	85	—

Pulmonary A-V oxygen difference (assuming P.V. 95% sat'd.)	=	3·3 ml./litre blood
Peripheral A-V oxygen difference	=	35·0 ml./litre blood
Pulmonary blood flow	=	48·5 litres/min.
Peripheral blood flow	=	4·6 litres/min.

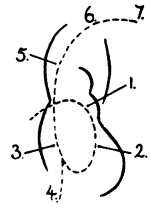
Case 2. R.T., a man, aged 24, was first seen in July, 1949. He had been short of breath all his life. He attended a special school till 14 and was unable to take part in games because of dyspnœa. He could walk only a few yards on the level and had been unable to work for the past eighteen months because of dyspnœa.

On examination he was a fit looking man with no cyanosis or finger clubbing. At rest there was no dyspnœa. The pulse was 84 and regular; the blood pressure in the arms was 120/80. There was recession of the intercostal spaces over the region of the apex beat and in the posterior axilla. The apex beat was palpable in the eighth left intercostal space in the mid-axilla some 17·5 cm. from the mid-line. The cardiac impulse was forcible and there was systolic expansion over the outflow tract of the right ventricle and the pulmonary artery. A moderately loud systolic murmur could be heard all over the præcordium and at the back and was maximal in the third left interspace near the sternum. The second sound in the pulmonary area was loud and widely split. A low pitched mitral diastolic murmur was audible at the apex.

On fluoroscopy the heart was displaced to the left and the right ventricle was greatly enlarged. The branches of the pulmonary artery were large and expansile pulsation was easily seen (Fig. 6). The vascular pedicle was wide forming an elliptical shadow within which the pulmonary artery and aortic knob could be seen. A barium swallow was normal. The cardiogram showed complete right bundle branch block (Fig. 7). Red blood corpuscles 5·7 million per cu. mm.; hæmoglobin 110 per cent.

The results of cardiac catheterization are summarized in Table II. The catheter coiled up in the right *S.V.C.* as in Case 1; attempts to enter the left *S.V.C.* and the left atrium were unsuccessful.

TABLE II. CASE 2, R.T., ÆT. 24



Sample of blood from:	O_2 content /100 c.c. bld.	O_2 capacity /100 c.c. bld.	Percentage sat'n.	Pressure mm. Hg. above skin of back
1. M.P.A.	19·49	21·1	92	25
2. R.V.	—	—	95	26
3. R.A.	—	—	93–95	10
4. I.V.C.	19·01	—	90	—
5. S.V.C.	—	—	95	—
6. L.Innom.V.	—	—	95	—
7. L.Subclav.V. ..	17·92	—	85	—
Femoral artery ..	20·22	—	96	—

Pulmonary A-V oxygen difference (assuming P.V. 96% sat'd.)	=	7·3 ml./litre blood
Peripheral A-V oxygen difference	=	38·0 ml./litre blood
Pulmonary blood flow	=	34·0 litres/min.
Peripheral blood flow	=	6·6 litres/min.

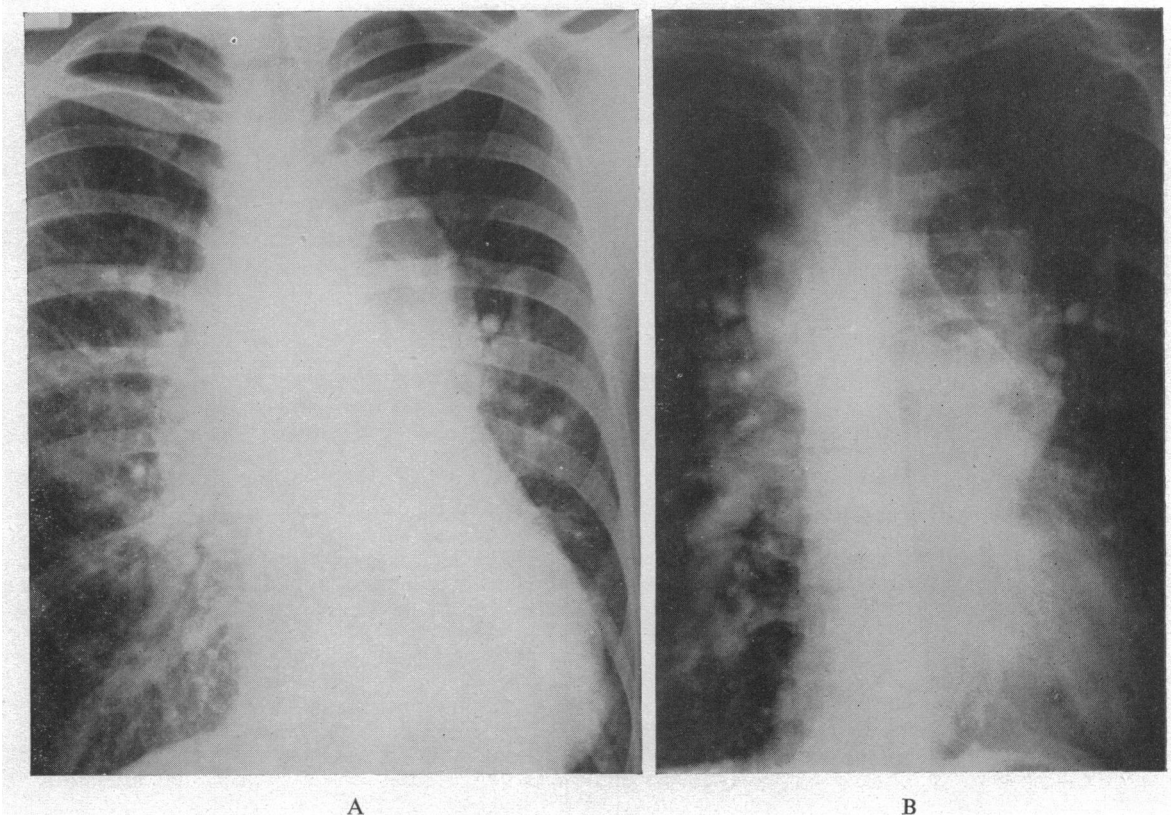


FIG. 6.—Case 2. (A) Teleradiogram showing the heart displaced to the left and large branches of the pulmonary arteries in the lung fields. The superior venæ cavæ form an elliptical shadow in the superior mediastinum enveloping the aortic knob and pulmonary trunk. (B) Angiocardiogram. Film taken 5 sec. after injection of diodone into right ventricle, showing the large vessel formed by the union of the right pulmonary veins passing to the left and then upwards into the left superior vena cava. The left pulmonary veins appear to enter the lower part of the left superior vena cava. The left innominate vein and the right *S.V.C.* also contain diodone.

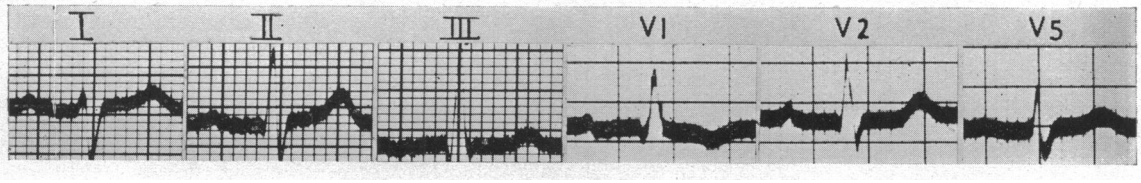


FIG. 7.—Case 2. Electrocardiogram showing complete right bundle branch block.

Mean pressures in the right ventricle and pulmonary artery were a little high. The oxygen saturation of blood in the left innominate vein was 95 per cent and in the left subclavian vein it was 85 per cent. The arterial blood oxygen saturation was 96 per cent.

The angiocardiogram taken 5 seconds after the injection of diodone by catheter into the right ventricle showed the right pulmonary veins uniting to form a single large vessel that ran transversely across the mid-line and turned upwards to be continued as the left *S.V.C.* The left pulmonary veins appeared to enter the latter at its lowermost part. Diodone could also be seen in the left innominate vein and the right *S.V.C.*

In this patient the fully oxygenated arterial blood and the failure to enter the left atrium at right heart catheterization were thought, in spite of the angiocardiographic evidence, to indicate

that some but not all of the pulmonary veins drained into the left *S.V.C.* and that the atrial septum was intact.

In view of the young man's considerable disability thoracotomy was advised in consultation with Sir Clement Price Thomas, with the object of anastomosing the left *S.V.C.* to the left atrial appendage. At operation it was found that a single large vein passed from the right lung transversely behind the heart towards the left hilum where it united with a similar single vein from the left lung to form the commencement of the left *S.V.C.* which thus drained all the pulmonary veins.

Unfortunately the left atrial appendage was abnormally small and for this reason its anastomosis to the *S.V.C.* proved technically impossible.

Case 3. C.B., an infant, aged 18/12, was physically and mentally backward; she was unable to sit up and could only just hold her head up. Nothing was known of her previous history except that she was the youngest of nine illegitimate children.

She was 26 in. long and weighed 14 lb. There was no finger clubbing but screaming induced slight cyanosis. The pulse was 138, regular, and of small volume; the blood pressure was 90/? in the arms. There was bulging of the præcordium and vigorous pulsation was visible and palpable all over the left side of the chest, particularly in the third and fourth spaces. The apex beat was in the fifth left space in the anterior axillary line. The second sound in the pulmonary area was loud and split. There was a loud harsh systolic murmur along the left sternal border, maximal in the third space, and a rumbling mitral diastolic murmur at the apex.

The teleradiogram showed that the heart was greatly enlarged and the main pulmonary artery and its branches were enormous (Fig. 8). There was an almost circular shadow in the superior mediastinum

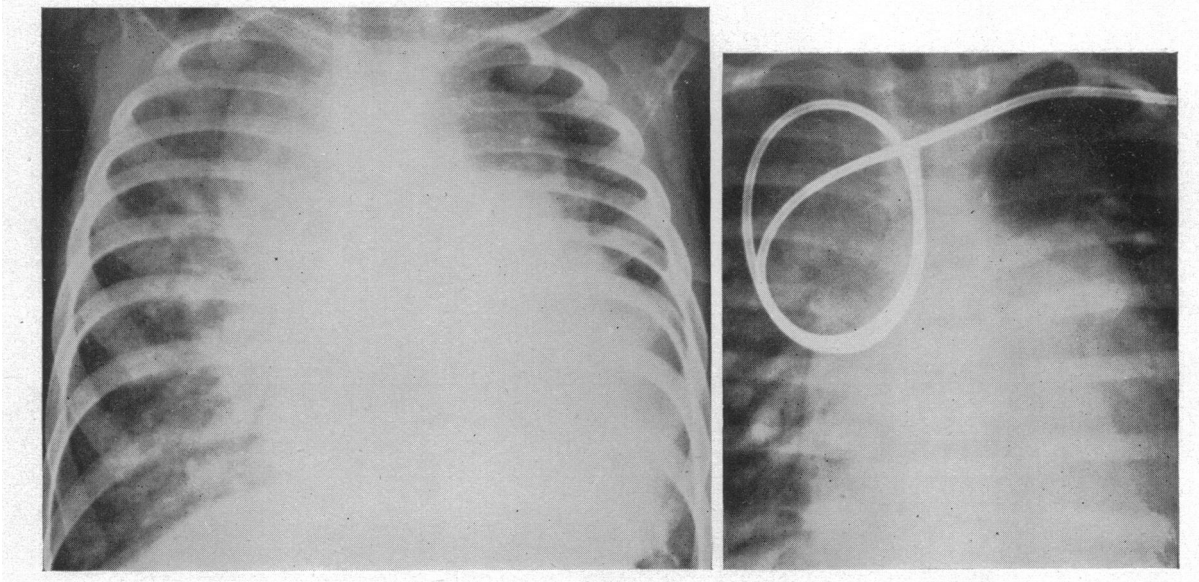


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FIG. 8.—Case 3. (A) Teleradiogram showing the large heart and pulmonary trunk and enormous branches of the pulmonary artery in the lung fields. The shadows of the superior venæ cavæ envelop the pulmonary trunk and aortic knob. (B) A–P film showing cardiac catheter coiled in the right *S.V.C.*

within which, on the left side, the main pulmonary artery and what appeared to be the aortic knob could be seen. The cardiogram (Fig. 9) showed incomplete right bundle branch block with right ventricular enlargement. Hæmoglobin 77 per cent.

The results of cardiac catheterization are summarized in Table III. The left atrium was not entered. The mean pressures in the right ventricle and pulmonary arteries were extremely high. The oxygen saturation of blood in the left innominate vein was 94 per cent and in the left subclavian vein it was 66 per cent. The arterial blood oxygen saturation was 89 per cent.

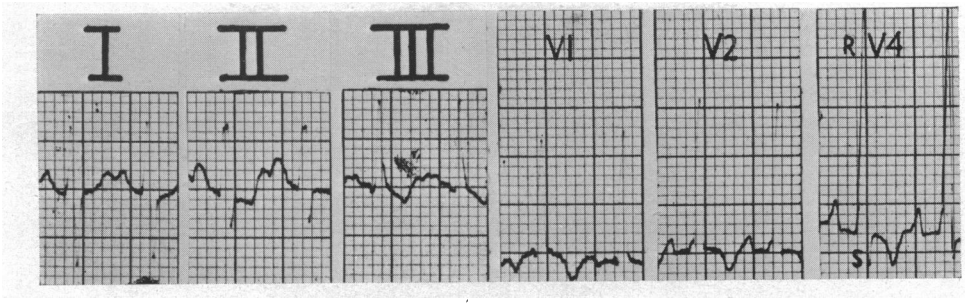
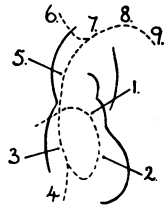


FIG. 9.—Case 3. Electrocardiogram showing incomplete right bundle branch block.

The angiocardiogram taken one second after injection of diodone by catheter into the right atrium showed an enormous right ventricle and simultaneous filling of the hypoplastic aorta and the large pulmonary artery (Fig. 10A). The 2-second film (Fig. 10B) showed diodone in both superior venæ cavæ and in the left innominate vein.

The child was anæsthetized for the angiocardiogram and immediately thereafter pulse and respiration were satisfactory. A few minutes later, however, she stopped breathing and no heart sounds could be heard. Restorative measures were unavailing and the child died.

TABLE III. CASE 3, C.B., ÆT. 18 MONTHS.



Sample of blood from:	O_2 content /100 c.c. bld.	O_2 capacity /100 c.c. bld.	Percentage sat'n.	Pressure mm. Hg. above skin of back
1. M.P.A.	14·02	15·9	88	70
2. R.V.	—	—	86	42
3. R.A.	—	—	87	10
4. I.V.C.	10·53	—	66	—
5. S.V.C.	—	—	92	—
6. R.Int.Jug.V. ..	—	—	55	—
7. L.Innom.V. ..	—	—	94	—
8. L.Subclav.V. ..	9·87 (Mean of 6, 8, 9)	—	66	—
9. L.Axillary V. ..	—	—	65	—
Femoral artery ..	14·17	—	89	—

Pulmonary A-V oxygen difference (assuming P.V. 95% sat'd.) = 10·8 ml./litre blood
Peripheral A-V oxygen difference = 39·7 ml./litre blood

Post-Mortem Examination (Fig. 11 and 12). The right atrium was large. Its cavity measured 45 mm. from the right wall to the tip of the atrial appendage, and 28 mm. from the orifice of the *S.V.C.* to the tricuspid valve ring. The average thickness of the wall was 2 mm. The foramen ovale, 16 × 15 mm., was closed except for a narrow crescentic opening, 8 mm. in length at its anterior end.

The right ventricle formed the apex and nearly the whole of the anterior surface of the heart and was 55 mm. from base to apex. Its cavity measured 38 mm. from tricuspid ring to apex and 55 mm. from apex to the pulmonary valve ring. The wall of this ventricle was 6–8 mm. thick and the papillary muscles were hypertrophied, the larger measuring 6 × 4 mm. in cross-section. The tricuspid and pulmonary valves were normal.

The pulmonary trunk measured 22 mm. in diameter and its right and left branches were 15 mm. and 13 mm. wide, respectively. There was a dimple at the point of insertion of the obliterated ductus arteriosus.

The left atrium was small, 15 mm. from its summit to the mitral valve ring and 21 mm. from the interatrial septum to the root of the diminutive atrial appendage. Apart from the mitral ring, the only opening into this atrium was the crescentic aperture at the anterior end of the foramen ovale.

The left ventricle appeared as a small oval appendage applied to the left border of the right ventricle and was 38 mm. from base to apex. Its cavity measured 33 mm. from the mitral valve ring to the apex and its wall was only 4 mm. thick. The aorta was 9 mm. in diameter.

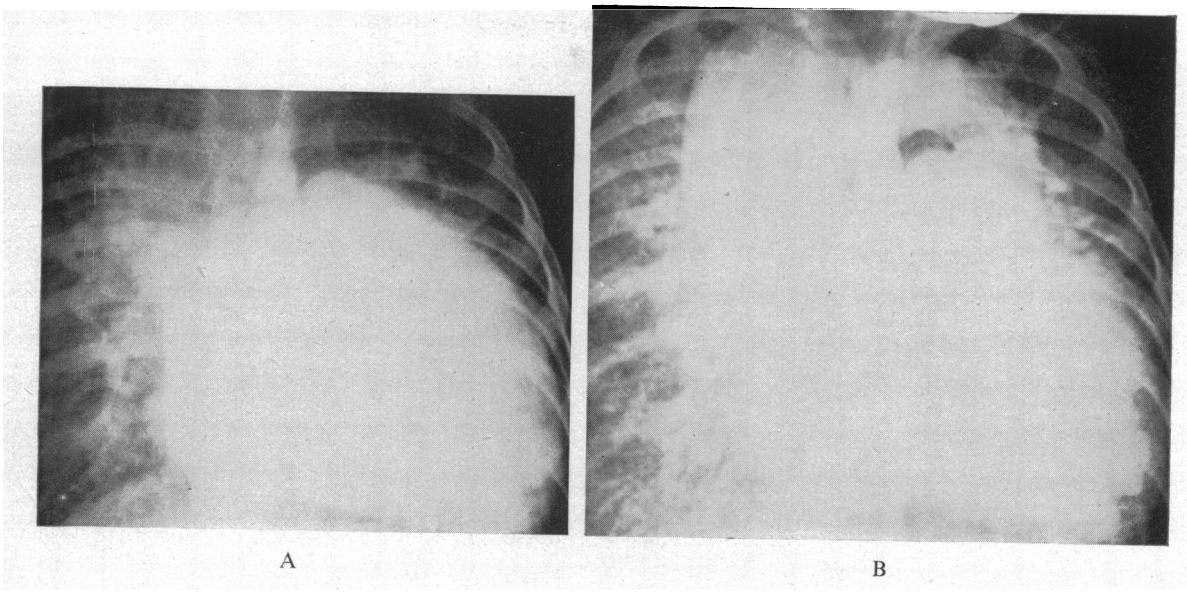


FIG. 10.—Case 3. Angiocardiograms. (A) Film taken 1 sec. after injection of diodone into right atrium, showing enormous right ventricle and pulmonary trunk and the hypoplastic aorta. (B) Film taken 2 sec. after injection of diodone into right atrium, showing the heart and great vessels still filled with diodone and diodone in both superior venæ cavæ and the left innominate vein.

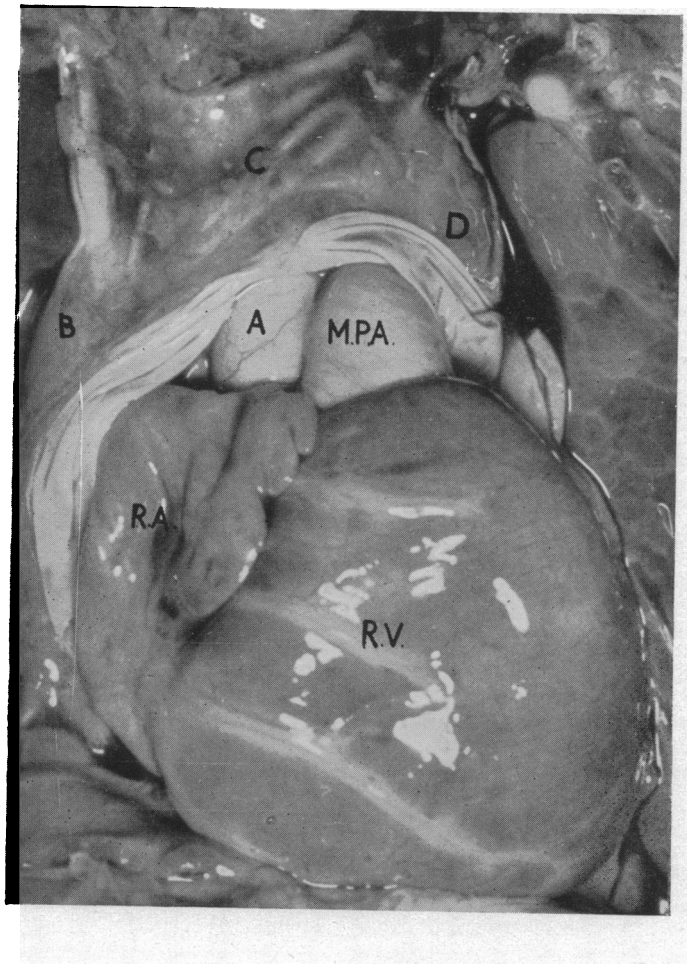


FIG. 11.—Case 3. Photograph of the heart and great vessels in situ, after removal of the anterior chest wall, showing the right ventricle (R.V.) occupying the whole anterior surface of the heart and the aorta (A) and pulmonary trunk (M.P.A.) enveloped above and on either side by the left innominate vein (C) and the right and left superior venæ cavæ (B, D respectively).

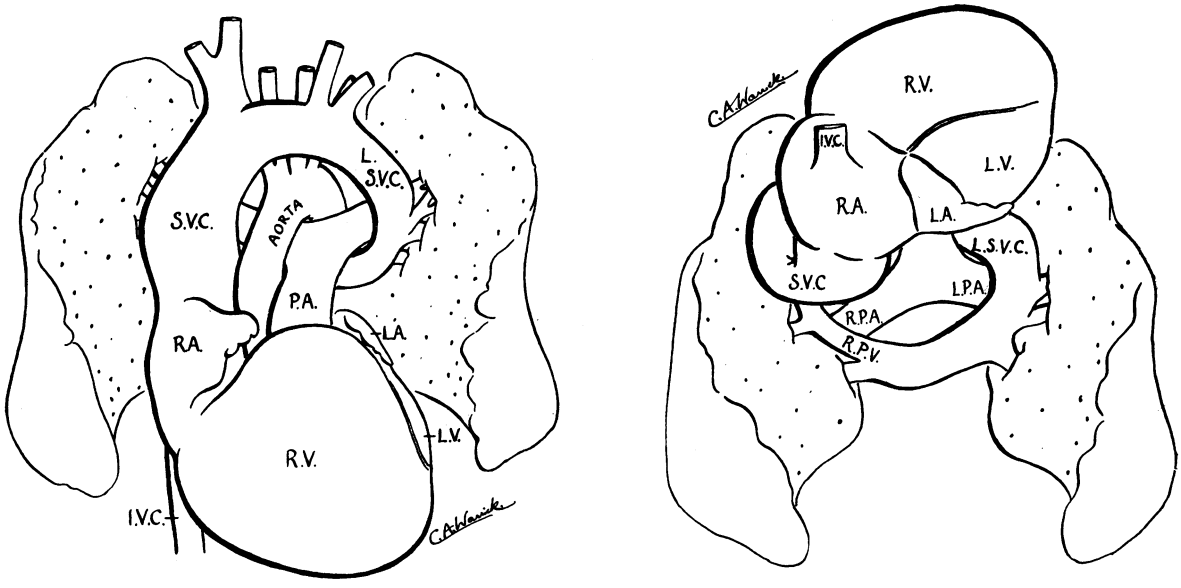


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FIG. 12.—Case 3. Drawings of the heart and great vessels. (A) Viewed from the front. R.A., right atrium; I.V.C., inferior vena cava; R.V., right ventricle; P.A., pulmonary artery; L.A., left auricle; L.V., left ventricle; L.S.V.C., left superior vena cava; S.V.C. superior vena cava. (B) Viewed from the front with the heart lifted forwards, showing the pulmonary veins uniting to form the left superior vena cava. R.P.V., right pulmonary vein; R.P.A. and L.P.A., right and left pulmonary arteries.

The upper and lower right pulmonary veins (Fig. 12B) united to form a single large vessel, 12 mm. in diameter, which crossed transversely behind the heart towards the hilum of the left lung where it was joined by a large vein from the left lower lobe. Thereafter this vessel turned upwards and, having received the left upper pulmonary veins, became continuous with the left *S.V.C.* The cava was 18 mm. in diameter and at its upper extremity (Fig. 12A) it received the left subclavian and internal jugular veins and thereafter turned medially to become the left innominate vein which subsequently united with the right innominate to become the right *S.V.C.* The diameter of the latter in its widest part was 35 mm.

Thus, in this infant, all the pulmonary veins drained into the right atrium via the left *S.V.C.* and existence was maintained by what must have been a diminutive blood flow through the small crescentic opening in the foramen ovale.

Case 4. D.C., a boy, aged 13, was referred to hospital because on two occasions he had become cyanosed and severely dyspnœic after moderate exertion. He had always been small for his age but he led a normal life and took part in all the usual school activities.

In 1948 he was 50 in. high and he weighed 49 lb. There was no cyanosis or finger clubbing. The pulse was 80 and regular; the blood pressure was 110/70. The apex beat was in the fifth left space in the mid-clavicular line. There was a right ventricular type of cardiac impulse and palpable expansile pulsation over the outflow tract of the right ventricle and pulmonary artery. A third heart sound and a variable low-pitched mitral diastolic murmur were audible at the apex. The second sound in the pulmonary area was accentuated and widely split. A harsh, not very loud, systolic murmur was maximal in the second left space and audible over the whole præcordium and between the scapulæ.

On fluoroscopy (Fig. 13) the right ventricle was enlarged and there was increased pulsation in the pulmonary artery and its branches. There was a dilated right *S.V.C.* but no left *S.V.C.* was seen. Barium swallow was normal. The cardiogram showed incomplete right bundle branch block (Fig. 13). Hæmoglobin, 80 per cent.

The results of cardiac catheterization are summarized in Table IV. The left atrium was not entered. The mean pressures in the right ventricle and pulmonary artery were a little high. The

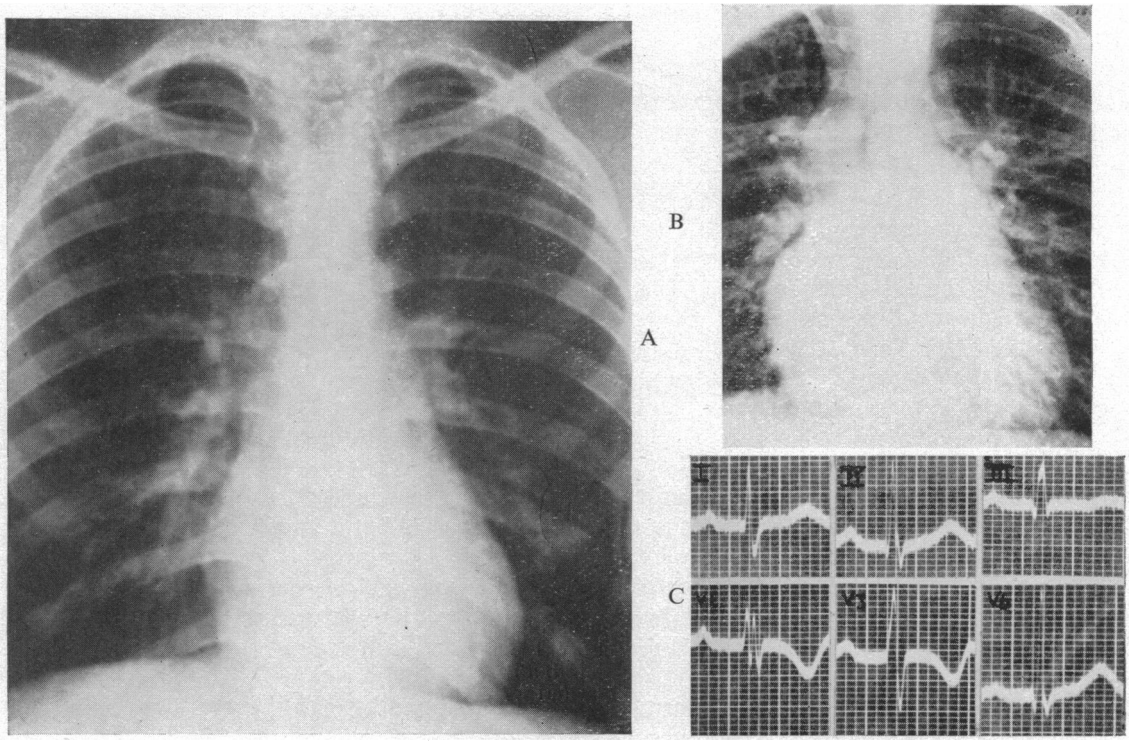
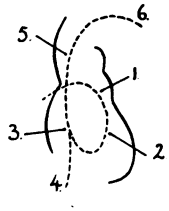


FIG. 13.—Case 4. (A) Teleradiogram showing the dilated right superior vena cava. (B) Angiocardiogram, taken 2 sec. after the beginning of injection, showing diodone in pulmonary artery and aorta. The left upper pulmonary vein passes upwards into the left superior vena cava and the left innominate vein contains diodone. The inferior pulmonary veins appear to drain into the left atrium. (C) Electrocardiogram showing incomplete right bundle branch block.

TABLE IV. CASE 4, D.C., ÆT. 13



Sample of blood from:	O_2 content /100 c.c. bld.	O_2 capacity /100 c.c. bld.	Percentage sat'n.	Pressure mm. Hg. above skin of back
1. M.P.A.	14·08	16·45	86	24
2. R.V.	—	—	84	21
3. R.A.	—	—	81–87	10
4. I.V.C.	12·63	—	77	—
5. S.V.C.	—	—	97	—
6. L.Subclav.V. ..	13·54	—	82	—
Femoral artery ..	15·9	—	97	—

Pulmonary A-V oxygen difference	=	18·2 ml./litre blood
Peripheral A-V oxygen difference	=	28·1 ml./litre blood
Pulmonary blood flow	=	9·9 litres/min.
Peripheral blood flow	=	6·4 litres/min.

oxygen saturation of blood in the right *S.V.C*, was 97 per cent, and in the left subclavian vein, 82 per cent. The arterial blood oxygen saturation was 97 per cent.

The angiocardiogram taken two seconds after the injection of diodone showed the left upper pulmonary vein passing upwards into a left *S.V.C.*, a large left innominate vein, and refilling of the large right *S.V.C.* Diodone was absent from the right atrium but present in the left, and the

inferior pulmonary veins from both sides appeared to drain into the latter chamber. The exact anatomy of the right upper pulmonary veins could not be determined with certainty but they were thought to drain into the right *S.V.C.*

In this child only the upper pulmonary veins have anomalous drainage and it is thought that the atrial septum is intact.

Clinical Picture

When the pulmonary veins drain into the left *S.V.C.* the clinical picture is characteristic and easily recognized in life. From an analysis of the physical signs in our four cases and of the two described by Taussig, the following clinical picture emerges.

The frequency of this particular anomaly it is difficult to assess. Earlier papers on this subject (McCotter, 1916; Papez, 1938) record some 200 examples of persistent left *S.V.C.* but do not distinguish the different types. It cannot be extremely rare, however, since we have encountered four examples in the past three years.

Symptoms and Signs. Surprisingly enough, this anomaly does not necessarily affect the general development although the left heart receives so little blood that it is commonly atrophic. Case 1 was a small asthenic woman, Case 2 was a large well-grown man, and Case 4, coincidentally, was a pituitary dwarf. Case 3 was mentally and physically defective, but it is impossible to say this was due to the cardiovascular abnormality.

The only important symptom is breathlessness on exertion, and this was noticed during the first year of life in our first two patients where all the pulmonary veins drained into the left *S.V.C.* In the fourth, where only the upper pulmonary veins had this anomalous drainage, dyspnœa was absent except for two attacks after severe exertion.

Cyanosis and finger clubbing may or may not be present and are never severe; both were slight in our first patient and absent in the other three.

The pulse volume tends to be small and the blood pressure somewhat low. Prominence of the left chest was obvious in our first three patients. All four had a well developed right ventricular type of impulse and in all there was expansile pulsation and a diastolic shock palpable over the pulmonary artery. The second pulmonary sound was accentuated and widely split in all four patients. Murmurs are not diagnostic. The only constant one is systolic and heard best to the left of the sternum; a basal diastolic and an apical mitral diastolic murmur may also occur.

The electrocardiogram showed complete or incomplete right bundle branch block in all four patients.

Radiology. The radiological appearance is characteristic. When all the pulmonary veins drain into the left *S.V.C.* there is a large ovoid shadow in the superior mediastinum, within which the aortic knob and the shadow of the pulmonary artery are clearly seen. This shadow is formed by the enormous right and left *S.V.C.* and the left innominate vein. Together with the heart shadow it gives an almost dumb-bell shaped silhouette.

When, as in Case 4, only some of the pulmonary veins have this anomalous drainage the appearances are far less striking but there is still considerable widening of the superior mediastinal shadow due to the dilatation of the right *S.V.C.*

The pulmonary arteries are large and show expansile pulsation. As would be expected the right ventricle is always enlarged.

Cardiac catheterization confirms the diagnosis. Blood samples from the left innominate vein and right *S.V.C.* are highly oxygenated and venous blood can be obtained only from the more peripheral veins.

Angiocardiography may demonstrate the abnormal course of the circulation and the right to left shunt through the atrial septal defect.

Prognosis. Brody (1942) states that the prognosis depends largely on the amount of blood diverted to the right atrium. In his series, of all the patients in whom the entire pulmonary drainage was into the right side of the heart, only three lived beyond early childhood. Taussig (1947)

considers the prognosis poor and states that death usually occurs between four and six months of age, although if the foramen ovale remains patent life may be prolonged for several years.

Our first two patients, both aged 24, are in reasonably good health though their activities are considerably restricted by dyspnœa, and in the first early congestive failure has been precipitated on one occasion by pregnancy. In our third patient existence, at the best, seemed precarious and it was thought that the mechanical interference only hastened the inevitably early fatal outcome. The fourth patient, aged thirteen years, leads a normal life.

The usual mode of death is said to be congestive failure and it is claimed, though the evidence is unconvincing, that this is brought about by the gradual closure of the defect in the atrial septum.

Differential Diagnosis. Atrial septal defect and large ventricular septal defects are the conditions most difficult to distinguish clinically from the variety of persistent left *S.V.C.* described here. Fluoroscopic examination, once the significance of the superior mediastinal shadow has been appreciated, makes the diagnosis obvious. Where doubt exists the finding of highly oxygenated blood in the left innominate vein will establish the diagnosis.

The fluoroscopic appearances may be confused with persistent azygos vein, aneurysmal dilatations of pulmonary artery or aorta, or in infants, an enlarged thymus gland. When there is a persistent azygos vein the shadow of the left *S.V.C.* is absent and catheter studies show that the abnormal vein contains venous blood. Aneurysmal dilatation of the pulmonary artery or aorta is easily excluded if the main pulmonary artery and the small aortic knob can be recognized within the shadow of the left *S.V.C.* An enlarged thymus may cause difficulty but it is usually possible to distinguish the lower poles of this organ, the so-called " sail-shadow," lateral to the cardiac silhouette.

Treatment. The only radical measure is an anastomosis of the left *S.V.C.* or the innominate vein to the left atrium or its appendage. This has never been done so far as we are aware. In our Case 2 the left atrium was found to be hypoplastic and could not be brought into apposition with either the left *S.V.C.* or the innominate vein. Brantigan (1947) has suggested that, provided it is certain that some of the pulmonary veins drain into the left atrium, the load on the right heart may be diminished by lobectomy and ligation of the anomalous veins.

Discussion

Failure of the left precardinal vein to become obliterated to form the ligament of Marshall results in persistence of the left *S.V.C.* This anomalous vessel may drain some or all of the pulmonary veins which therefore yield their blood to the right atrium instead of the left. If all the pulmonary veins empty into the right heart in this fashion a defect in the atrial septum is essential for life as it is the only pathway by which blood can reach the left side of the heart.

The most striking signs of this anomaly indicate the large left to right shunt, and in this respect resemble those of atrial septal defect and large ventricular septal defects, only they are exaggerated. The right to left shunt, which must be present when all the pulmonary veins drain into the right atrium, may be impossible to detect clinically since the presence or absence of cyanosis depends on the oxygen saturation of blood in the right atrium from which the left heart is entirely supplied, and in our patients this ranged from 79 to 95 per cent.

This highly oxygenated right atrial blood suggests that the pulmonary flow is many times greater than the systemic. That the pulmonary flow is large is evident from the considerable expansile pulsation, palpable over the right ventricle and pulmonary artery and visible fluoroscopically in the branches of the pulmonary artery in the lung fields. The load on the right heart must be extremely heavy because it receives almost the entire systemic and pulmonary venous blood. This results in very great hypertrophy of the right ventricle which is recognized clinically by the forceful right ventricular thrust, the loud and widely split pulmonary second sound, partial or complete right bundle branch block, and the radiological appearances. The small pulse volume and low blood pressure is evidence that the systemic flow is small, and the atrophic left atrium

and ventricle and hypoplastic aorta found at autopsy likewise suggest that the left heart receives very little blood.

The catheter findings provide some idea of the order of the discrepancy between pulmonary and peripheral flows. The figures can only be regarded as very rough approximations, however, because in no case was it possible to sample blood from the pulmonary veins, and it was quite impossible to obtain an accurate figure for the oxygen content of systemic venous blood.

The severe pulmonary hypertension present in our third case may be compared with the findings in atrial septal defect, where in the majority of patients the mean pressure in the pulmonary artery is within normal limits but in a small minority there is pulmonary hypertension sufficient, on occasion, to result in tricuspid incompetence and partial reversal of the inter-atrial shunt. The poor physical state of this infant suggests that the prognosis may be adversely affected by the presence of much pulmonary hypertension.

The diagnostic value of right heart catheterization in this condition is unquestionable, since in no other anomaly is arterial blood found in the left innominate vein. By contrast angiocardiography is far less useful because the immense pulmonary flow quickly dilutes the contrast media, even if it be introduced directly into the right ventricle, and in consequence the abnormal anatomy of the venous side of the pulmonary circulation is poorly defined.

The obvious radical therapeutic measure is anastomosis of the left *S.V.C.* to the left atrium or its appendage, but our experience, gleaned from one exploratory thoracotomy and one autopsy together with the evidence from anatomical descriptions (Papez, 1938), suggests that these structures may not uncommonly be atresic so that surgical correction of the anomaly may rarely be feasible.

Summary

The development of the superior vena cava and pulmonary veins is outlined and the types of persistent left superior vena cava are classified.

Four cases of a persistent left superior vena cava draining pulmonary veins are reported. In one the diagnosis was confirmed at operation, and in a second at autopsy.

The clinical picture is described. The essential features are evidence of an enormous left to right shunt, considerable right ventricular enlargement, little or no cyanosis, and fluoroscopically the wide mediastinal shadow enveloping the aortic knob and pulmonary arc. The diagnosis is confirmed by cardiac catheterization when arterial blood is found in the left innominate vein.

The differential diagnosis is discussed. The condition is not rare, does not necessarily lead to death in infancy or childhood, and can be diagnosed during life. Surgical correction of the anomaly was attempted in one patient but was impracticable. The scope of surgical measures is briefly considered.

It is a pleasure to record our thanks to Dr. Clifford Hoyle for his help and advice in the preparation of this paper and for allowing us to study one of his patients (Case 1). We are also indebted to Dr. B. Gans, who kindly referred Case 3 to us, and to Dr. Ann Warwick and the Photographic Department of the Royal Free Hospital for the illustrations.

Since this paper was completed we have seen a report on this subject by Snellen and Albers (*Circulation*, Dec., 1952). Their findings and conclusions agree closely with our own.

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