TOTAL PULMONARY VENOUS DRAINAGE THROUGH A PERSISTENT LEFT SUPERIOR VENA CAVA

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When Parsons *et al.* (1952) reported two patients with total pulmonary venous drainage into the right heart through a persistent left superior vena cava and the left innominate vein they were able to find accounts of only nine other patients, six of whom were babies diagnosed at autopsy. Another, although only diagnosed in life as having abnormal pulmonary venous drainage into the right superior vena cava by Johnson and McRae (1948), had a teleradiogram similar to those now thought to be characteristic of total pulmonary venous drainage through a persistent left superior vena cava. Two others, reported by Friedlich *et al.* (1950), were the first patients with this congenital anomaly fully diagnosed in life. Adding Taussig's (1947) Case 33, the four patients of Snellen and Albers (1952), and the three patients of Gardner and Oram (1953) the number of published cases is nineteen. From these records it is apparent that before the advent of angiocardiology and cardiac catheterization this congenital anomaly was recognized only at autopsy. Following the introduction of these special methods of investigation it was possible to diagnose the anomaly in life and to recognize it more frequently.

The patients reported by Taussig (1947), Parsons *et al.* (1952), Snellen and Albers (1952), and Gardner and Oram (1953) have similar teleradiograms in which the striking feature is a "cottage-loaf" shaped cardiovascular shadow, formed by an ovoid upper mediastinal mass and an enlarged heart. The two latter groups of observers demonstrated by angiocardiology that the ovoid mediastinal enlargement is produced on the left by a persistent left superior vena cava and on the right by the dilated right superior vena cava, and they supported Taussig's (1947) suggestion that the cardiovascular contour provides a distinctive radiological sign of total pulmonary venous drainage through a left superior vena cava into the left innominate vein.

Brown (1950), who was already aware of the radiological appearance of a persistent left superior vena cava, independently recognized the significance of these distinctive radiological features and diagnosed this anomaly on six occasions at routine clinical and radiological examination of patients in cardiac clinics in Hull and in the Sheffield Region. Three patients (Cases 4, 5, and 6) were already under observation but the others (Cases 1, 2, and 3) were seen for the first time early in 1953, which suggests that this anomaly is not so rare as the published cases indicate. When it is widely known that the radiological features are sufficiently characteristic to allow the anomaly to be recognized at routine examination there will probably be an increase in the frequency of its diagnosis similar to that following the introduction of the special methods of investigation.

CASE REPORTS

Case 1. A man, age 24, was "black at birth" but never more than slightly blue since. He had pneumonia five times between the ages of 4 and 14 and subsequently suffered from a productive cough. He had dyspnoea and palpitation on exertion but was able to walk "miles on the level." He never squatted. He was an epileptic, but his fits were controlled by epanutin. He weighed only seven stone.

Case 2. A girl, aged 7, was the eldest of three children, the second of whom died at birth. She had been under observation from the age of 18 months when her mother had sought advice because of her slow general progress. She did not walk until she was two years old and subsequently failed to grow and gain weight normally, but when seen in hospital she had no symptoms other than nocturnal enuresis. She was undersized, 3 ft. 7 in., and underweight, 33 lb.

Case 3. A woman, aged 22, had pneumonia as a baby and had since suffered from a chronic productive cough with winter exacerbations. Dyspnoea on exertion had been present for 10 years and was increasing in severity although she was still able to walk a mile on the level without becoming breathless. She was not blue at birth but had gradually become cyanosed. For two months she had streaky hæmoptysis and for about the same time her ankles had swollen in the evening. She weighed 6 st. 12 lb. and was 5 ft. 4 in. tall.

Case 4. A boy, aged 8, the second of two children, had pneumonia at 1 and 2 years of age and had since suffered from breathlessness, palpitation, and cyanosis on exertion. He weighed only 3 st. 8 lb.

Case 5. A boy, aged 5, the youngest of three children, had pneumonia at five months, eighteen months, and three years of age. He was more breathless than normal children on exertion but he never squatted. He was small, 3 ft. 1 in., and underweight, 1 st. 13 lb., for his age.

Case 6. A girl, aged 6, a first child, was found to have a heart lesion at the age of six months. She suffered from frequent chest infections. She was occasionally cyanosed but never squatted.

The present communication emphasizes the diagnostic features of this anomaly and reports the patients diagnosed by Dr. James W. Brown, with a summary of the results of angiocardiology and cardiac catheterization in five where investigations were made to confirm the diagnosis.

ANATOMY

The congenital abnormality under discussion is shown diagrammatically in Fig. 1 which was composed from the anatomical descriptions given by the authors quoted previously and from

FIG. 1.—Diagram of the anatomical abnormalities that occur with total pulmonary venous drainage into the right heart through a persistent left superior vena cava and the left innominate vein.

observations at cardiac catheterization and angiocardiology in the present group of patients. The right and left pulmonary veins join a common pulmonary vein behind the heart and this runs into a persistent left superior vena cava which passes upwards in front of the left pulmonary artery into the left innominate vein, or directly into the right superior vena cava. The blood in the pulmonary veins and in the left superior vena cava is almost fully saturated with oxygen and this stream of saturated blood enters the right heart through the left innominate vein. Systemic and pulmonary venous bloods mix in the right atrium and blood from this common pool passes to the systemic and pulmonary circulations. The systemic circulation is supplied exclusively by blood that enters the small left atrium through an atrial septal defect and for this reason the oxygen saturation of blood in a systemic artery and in the pulmonary artery is the same. Only a small fraction of blood from the right atrium passes through the atrial septal defect and because of this the systemic blood flow and blood pressures are sometimes low while the pulmonary blood flow and blood pressures are usually increased.

Gardner and Oram (1953) reviewed the embryology of this congenital anomaly and there are no more recent observations to add to their account.

CLINICAL FEATURES

The clinical features of the present group of patients with total pulmonary venous drainage through a persistent left superior vena cava into the left innominate vein are summarized in Table I. The ages of the patients show that this lesion is compatible with survival to the third decade, which was not recognized before Snellen and Albers (1952) and Gardner and Oram (1953) described their patients, since all previous accounts were of babies and young children. Failure to consider the diagnosis in adult life is probably an important reason why the lesion was previously not more widely recognized. The sexes appear to be equally affected.

Dyspnoea on exertion and recurrent attacks of pulmonary infection are the common symptoms of this congenital anomaly. Four patients in the present series and all four patients reported by

TABLE I

CLINICAL FEATURES OF SIX PATIENTS WITH TOTAL ABNORMAL PULMONARY VENOUS DRAINAGE INTO THE RIGHT HEART THROUGH A PERSISTENT LEFT SUPERIOR VENA CAVA AND THE LEFT INNOMINATE VEIN

Case No.	Age and Sex		Dyspnoea on exertion	Recurrent pulmonary infections	Physical development	Cyanosis	Finger clubbing	Arterial blood pressure (mm./Hg)	Prominent left chest	Murmurs *	Electro-cardiogram
1	24	M.	+	+	Poor	Slight	Slight	110/75	+	S.M. A.D.M. B.D.M.	Incomplete R.B.B.Bl.
2	7	F.	—	—	Poor	Slight	Slight	90/60	—	S.M. B.D.M.	Incomplete R.B.B.Bl.
3	22	F.	+	+	Poor	Slight	Absent	120/60	—	Absent	Incomplete R.B.B.Bl.
4	8	M.	+	+	Poor	Slight	Slight	90/50	—	Absent	R.V.+
5	5	M.	+	+	Poor	On exertion	Absent	115/85	+	S.M. A.D.M.	R.V.+
6	6	F.	—	+	Poor	Slight	Slight		+	S.M.	R.V.+

* S.M. indicates the presence of a basal systolic murmur of maximum intensity in the pulmonary area and widely conducted over the whole chest; A.D.M.=the presence of a rumbling apical diastolic murmur; and B.D.M.=the presence of a short early basal diastolic murmur.

In every case there was clinical evidence of right ventricular hypertrophy, and the radiological examination was diagnostic.

FIG. 2.—Case 1. Electrocardiogram showing ventricular pattern of right ventricular hypertrophy and clockwise rotation of a vertical heart.

FIG. 3.—Case 2. Electrocardiogram showing incomplete right bundle branch block and clockwise rotation of a vertical heart.

Snellen and Albers (1952) had dyspnoea on exertion, while Gardner and Oram (1953) considered this the only important symptom in their series. A history of recurrent pulmonary infection, however, appears to be another feature of the anomaly. Taussig (1947) states that patients with this congenital abnormality commonly have attacks of pulmonary congestion which are diagnosed as pneumonia, and this view is supported by the patients now reported, five of whom gave histories of recurrent pulmonary infections, frequently diagnosed as pneumonia.

The physical findings in these patients are in agreement with the descriptions given by Taussig (1947), Snellen and Albers (1952), and Gardner and Oram (1953), although their poor physical development suggests that the anomaly has much more effect upon general nutrition than is apparent from previous descriptions. Cyanosis and finger clubbing were not prominent in any of our patients. Five were slightly cyanosed at rest and the sixth after exertion, while four had early finger clubbing. Low systemic blood pressures occurred in two. Similar pressures were noted by Gardner and Oram (1953) in two of their patients and by Taussig (1947), who thought they were the result of a diminished systemic circulation. In all six patients the cardiac impulse indicated right ventricular hypertrophy and there was also prominence of the left chest in three. The second heart sound was palpable in one only but in all it was abnormally loud and split on auscultation and suggested the presence of pulmonary hypertension.

Murmurs were not a characteristic feature of these patients. Two had no murmurs, and the dominant auscultatory sign in the four with murmurs was a basal systolic murmur of maximum intensity in the pulmonary area but widely conducted and heard over the whole chest. One of these

FIG. 4.—Case 5. Postero-anterior teleradiogram showing an ovoid upper mediastinal mass and cardiac enlargement which give the cardiovascular shadow a "cottage-loaf" shape. The mediastinal shadow is produced on the left by a persistent left superior vena cava and on the right by the dilated right superior vena cava.

had also a rumbling apical diastolic murmur, a second had a basal diastolic murmur, a third had a rumbling apical diastolic murmur and a basal diastolic murmur, and the fourth had only the systolic murmur. Taussig (1947) considered that the systolic murmur was produced by the flow of blood through the atrial septal defect but it is impossible to be dogmatic about this, or about the cause of the basal diastolic murmurs and apical rumbling diastolic murmurs present in some of these patients and also noted by Gardner and Oram (1953) in two of their patients.

Electrocardiography. The electrocardiograms confirmed the clinical evidence of right ventricular dominance. Three patients had patterns of right ventricular hypertrophy (Fig. 2) and three had patterns of incomplete right bundle branch block (Fig. 3).

Radiology. The radiological examination is the most important step in the diagnosis of this congenital anomaly. Taussig (1947) described the characteristic radiological signs of this anomaly and the importance of these has since been emphasized by Snellen and Albers (1952), Gardner and Oram (1953), and by the diagnosis of the present group of patients on routine radiological examina-

FIG. 5.—Postero-anterior teleradiograms of four patients (Cases 3, 4, 1, and 6) with total abnormal pulmonary venous drainage into the right heart through a persistent left superior vena cava and left innominate vein. They all show an ovoid upper mediastinal mass and cardiac enlargement, which give the cardiovascular shadow a “cottage-loaf” shape, and the pulmonary arteries are abnormally prominent.

tion. At fluoroscopy the distinctive feature is the "cottage-loaf" shape of the cardiovascular shadow, formed by an ovoid upper mediastinal mass and an enlarged heart (Fig. 4, 5, and 6). The pulmonary arteries are abnormally prominent and pulsatile and pulsation is present in the upper mediastinal mass. The ovoid mediastinal enlargement is produced on the left by a persistent left superior vena cava and on the right by the dilated right superior vena cava. The cardiac enlargement is chiefly due to dilatation of the right atrium and hypertrophy of the right ventricle. The apparent hypertrophy of the left ventricle on fluoroscopy is probably due to its backward displacement by the enlarged right ventricle (Fig. 6).

Since the "cottage-loaf" cardiovascular shadow was present on the teleradiograms of all the

FIG. 6.—Case 2. Teleradiograms showing the diagnostic "cottage-loaf" cardiovascular shadow in the postero-anterior position and apparent enlargement of both ventricles in the oblique positions.

patients reported by Taussig (1947), Snellen and Albers (1952), and Gardner and Oram (1953), where there was anatomical proof of total abnormal drainage, and since it did not occur in a patient reported by Conant and Kurland (1947) where only the pulmonary veins from the left lung drained abnormally through a persistent left superior vena cava, this radiological sign appears diagnostic of total pulmonary venous drainage through a persistent left superior vena cava and the left innominate vein into the right heart. There is no anatomical proof that the pulmonary venous drainage in the present patients was totally abnormal but this diagnosis is supported in five by the results of cardiac catheterization and angiocardiography.

ANGIOCARDIOGRAPHY

An angiocardiographic examination was carried out in five of the patients in the present series. The contrast medium was injected from the left arm and simultaneous exposures were made at second intervals for ten seconds in the antero-postero and left lateral positions. The antero-posterior angiograms taken soon after the injection showed contrast medium in a dilated left innominate vein, a dilated right superior vena cava, and a dilated right atrium (Fig. 7 and 8). In most of the patients there was also retrograde filling of the mouth of the left superior vena cava (Fig. 7) and for this reason angiocardiography is more informative when the injection is given from the left arm than from the right. Occasionally left atrial filling was apparent on these early antero-posterior angiograms (Fig. 8). On subsequent angiograms there was simultaneous filling of the right and left hearts and later of the pulmonary arteries and the aorta (Fig. 7, 8, and 9). On the

final angiograms in the series the contrast medium appears in the pulmonary veins, a common pulmonary vein, the left superior vena cave, and again in the right heart (Fig. 7 and 8). From the antero-posterior angiograms it is obvious that the ovoid upper mediastinal enlargement is formed on the right by the dilated right superior vena cava and on the left by the persistent left superior vena cava.

The lateral angiograms were not so informative as the antero-posterior views but they demonstrated very well the simultaneous filling of the right and left hearts and the pulmonary venous drainage into the left superior vena cava without any apparent connection with the left atrium (Fig. 9).

While angiocardiology is not essential for the diagnosis of this congenital anomaly it offers certain confirmatory evidence of the lesion.

FIG. 7.—Case 2. Antero-posterior angiocardigrams. At one second the contrast medium outlines the dilated left innominate vein, the dilated right superior vena cava and right auricle. There is also some retrograde filling of the mouth of a persistent left superior vena cava. At three seconds there is simultaneous filling of the aorta and pulmonary arteries. At four seconds the contrast medium is in the pulmonary veins, a common pulmonary vein, the left superior vena cava and again in the right superior vena cava.

FIG. 8.—Case 5. Antero-posterior angiocardigrams. At one second the contrast medium is present in a dilated right heart and also in the left atrium. At two seconds the aorta and pulmonary arteries have filled. At four second the contrast medium is in the pulmonary veins, a common pulmonary vein, the left superior vena cava, and again in the right superior vena cava.

FIG. 9.—Case 4. Lateral angiocardiograms. At three seconds there is simultaneous filling of the right and left sides of the heart and of the pulmonary and systemic arteries, the aorta apparently filling from the left ventricle. At seven seconds the pulmonary veins are draining into the persistent left superior vena cava, without any apparent connection with the left atrium.

CARDIAC CATHETERIZATION

Cardiac catheterization was carried out in five patients and in all five the abnormal pulmonary venous drainage into a persistent left superior vena cava was demonstrated by passing the catheter from the right median cubital vein through the right and left innominate veins into a left superior vena cava and finally into a pulmonary vein (Fig. 10, 11, and 12). The presence of an atrial septal defect was confirmed in three patients by passing the catheter from the right atrium into the left heart (Fig. 12).

FIG. 10.—Case 2. (A) Antero-posterior and (B) left lateral teleradiograms showing a cardiac catheter lying in a persistent left superior vena cava with its tip in an inferior pulmonary vein.

FIG. 11.—Case 4. (A) Antero-posterior and (B) right lateral teloradiograms showing a cardiac catheter through a persistent left superior vena cava and behind the heart in a common pulmonary vein with its tip in a right pulmonary vein.

FIG. 12.—Case 3. Antero-posterior teloradiograms showing the cardiac catheter lying through the right heart with its tip in the left pulmonary artery (A), in a persistent left superior vena cava (B), and through an atrial septal defect with its tip in the left ventricle (C).

Three of the four patients whose pulmonary arteries were catheterized had pulmonary hypertension, but the fourth had normal pressures in his pulmonary artery and right ventricle; the fifth had a high right ventricular mean blood pressure (Table II). Friedlich *et al.* (1950), Parsons *et al.* (1952), and Gardner and Oram (1953) also recorded pulmonary hypertension in some of their patients and it appears that the range of pulmonary artery blood pressure with this abnormality is similar to that found in other congenital anomalies such as atrial septal defect, ventricular septal defect, and patent ductus arteriosus, where the pulmonary blood flow is increased. It is possible that the severe pulmonary hypertension sometimes found with the present anomaly is associated with organic pulmonary vascular changes as in these other congenital cardiac defects.

Blood samples were taken with the catheter in various sites and the oxygen content of these samples showed that a stream of almost fully saturated blood from the pulmonary veins and left superior vena cava entered the right superior vena cava through the left innominate vein (Table II). Blood samples from the right heart and femoral artery had virtually the same oxygen saturation which was evidence that all the pulmonary venous drainage was into the right side of the heart. If some of the pulmonary veins had entered directly into the left atrium, blood in the left heart and systemic circulation would probably have been more saturated with oxygen than that in the right heart (Table II).

TABLE II

RESULTS OF CARDIAC CATHETERIZATION IN FIVE PATIENTS WITH TOTAL PULMONARY VENOUS DRAINAGE THROUGH A PERSISTENT LEFT SUPERIOR VENA CAVA

Case No.	Blood oxygen saturation per cent									Blood haemoglobin content (g. per 100 ml.)	Oxygen consumption (ml./min.)	Pulmonary blood flow (litres/min.)	Systemic blood flow (litres/min.)	Pulmonary artery blood pressure (mm./Hg)
	Right innominate vein	Inferior vena cava	Right superior vena cava	Left superior vena cava	Pulmonary vein	Right atrium	Right ventricle	Pulmonary artery	Femoral artery					
1	67	75	93	98	99	92-97	91	92	92	16.1	260	17.4	5.1	16/5
2	80	78	—	99	99	97	96	—	92	13.0	105	16.4	4.6	25 *
3	62	67	98	99	—	92	—	93	92	14.3	204	18.5	4.0	54/35
4	42	41	—	—	97	—	—	84	83	15.8	133	4.9	1.5	70
5	74	78			99	93		94		11.9	—	—	—	(Mean) 44/16

* Mean blood pressure in right ventricle.

The small experimental error in the estimation of the oxygen content of a blood sample assumes tremendous proportions in the calculation of the difference in oxygen content of pulmonary arterial and pulmonary venous blood in patients with this lesion, and the estimations of pulmonary and systemic blood flows by the application of the Fick principle are regarded as only rough approximations. Such estimations do, however, indicate a high pulmonary blood flow and a low or low normal systemic blood flow (Tables II).

Cardiac catheterization provides confirmatory evidence of the diagnosis but is not an essential investigation for the recognition of this congenital anomaly.

TREATMENT

The record of surgical treatment of patients with this congenital abnormality is one of disaster. Two patients reported by Friedlich *et al.* (1950) died following operation while one of those reported by Snellen and Albers (1952) and another reported by Gardner and Oram (1953) had unsuccessful thoracotomies. Gardner and Oram (1953) considered that surgical correction by an anastomosis of a thin-walled common venous trunk, which may be transmitting twenty litres of blood a minute, to an atretic left atrium is rarely feasible. The disability of the patients in the present series was not sufficiently serious to justify surgical intervention.

SUMMARY

The diagnosis, on routine clinical and radiological examinations, of six patients with total pulmonary venous drainage through a persistent left superior vena cava into the left innominate vein, is reported.

The most important diagnostic feature of this anomaly is the presence at fluoroscopy of a "cottage-loaf" shaped cardiovascular shadow and pulmonary plethora. This "cottage-loaf" appearance is produced by a pulsating ovoid upper mediastinal shadow, formed by the dilated right superior vena cava on the right and the persistent left superior vena cava on the left, and an enlarged heart shadow, composed largely of the dilated right atrium and the hypertrophied right ventricle.

Other diagnostic features of this congenital abnormality are symptoms of dyspnoea on exertion and recurrent pulmonary infections, and signs of poor physical development, slight cyanosis, early finger clubbing, low systemic blood pressures, right ventricular hypertrophy, prominence of the left chest, and auscultatory evidence of pulmonary hypertension. Cardiac murmurs are not of great diagnostic importance.

The electrocardiograms confirm the clinical evidence of right ventricular dominance.

Angiocardiography and cardiac catheterization, which were carried out in five patients in this series, provide confirmatory evidence of the anomaly but are not necessary for its diagnosis.

Surgery was not advised in any of the patients in this series as their disability was not considered sufficiently severe for treatment offering so little prospect of success.

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