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Anomalous pulmonary venous return into inferior vena cava and associated bronchovascular anomalies (the scimitar syndrome)

Report of three cases and review of the literature

J. MATHEY, J. J. GALEY, Y. LOGEAIS, E. SANTORO, A. VANETTI, A. MAUREL, AND R. WUERFLEIN

From the Clinique de Chirurgie Thoracique, Hôpital Laennec, 42 rue de Sèvres, Paris 7

Among the different anatomical forms of anomalous pulmonary venous return, that of the inferior vena cava is of particular interest for the following reasons: the special radiological pattern which is referred to as the scimitar sign and the associated anomalies which often occur in the lungs. We have successfully operated on three[1] patients who are reported in this paper, and we have included a review of the literature.

CASE REPORTS

CASE 1 Liliane D., aged 9, presented because of repeated episodes of 'bronchitis' since infancy. At the age of 2 years a diagnosis of atrial septal defect (A.S.D.) had been made. On examination she was noted to have retarded development and some bulging of the left hemithorax. On auscultation she had a systolic murmur of moderate degree in the second and third intercostal spaces on the left, radiating towards both axillae and the back and of maximum intensity in the right axilla. On radiography (Fig. 1) the right border of the heart was predominantly enlarged, showing increased vascularization and also an opacity along the right cardiac border. The electrocardiogram showed right axis deviation and hypertrophy of the right ventricle.

Catheterization confirmed the existence of an A.S.D. of ostium secundum type. There was a left-to-right shunt of 3 volumes, and moderate elevation of the pulmonary artery pressure. The right ventricular pressure was 48 mm. Hg. Stenosis of the right pulmonary artery with a gradient of 20 mm. Hg across the obstruction was also found. Right anomalous pulmonary venous drainage was found, but the exact site was not known.

Operation was performed on 18 September 1963, using a mid-sternal incision and extracorporeal circulation with a moderate degree of hypothermia. Findings at operation included a huge right atrial chamber, and a small aorta and superior vena cava. On opening the right pleural space it became evident that the venous return from the entire right lung was channelled through a common duct entering into the inferior vena cava above the diaphragm. Following institution of bypass the right atrium was opened and the A.S.D. was found to be high and small. The anomalous pulmonary trunk entered low into the inferior vena cava (I.V.C.), and it was judged impossible to include its opening in a corrective patch covering the defect. Therefore the anomalous trunk was implanted high in the right atrium near the A.S.D., which was enlarged, and the entire defect was patched with a piece of Teflon.

The post-operative course was marked by some difficulty with re-expansion of the right lung. On discharge the child was in excellent condition and radiography showed a small discrete opacity at the right base.

CASE 2 L. Annie was aged 7 years. A short period of cyanosis had been noted at birth. At the age of 2 she was noted to have a slight degree of dyspnoea on exertion. She continued to develop normally. On examination a systolic murmur was heard over the entire precordium with maximum intensity in the left second intercostal space, with radiation to both axillae. A soft diastolic murmur was heard in the left fourth intercostal space. On radiography there was generalized enlargement of the cardiac shadow with a density on the left due to a persistent left superior vena cava (S.V.C.). At the right cardiac border was the typical opacity of the scimitar sign (Fig. 2). The E.C.G. showed right axis deviation with right atrial hypertrophy and an incomplete right bundle-branch block.

Catheterization and angiocardiography confirmed a

[1]Recently, a fourth patient has been successfully operated on.

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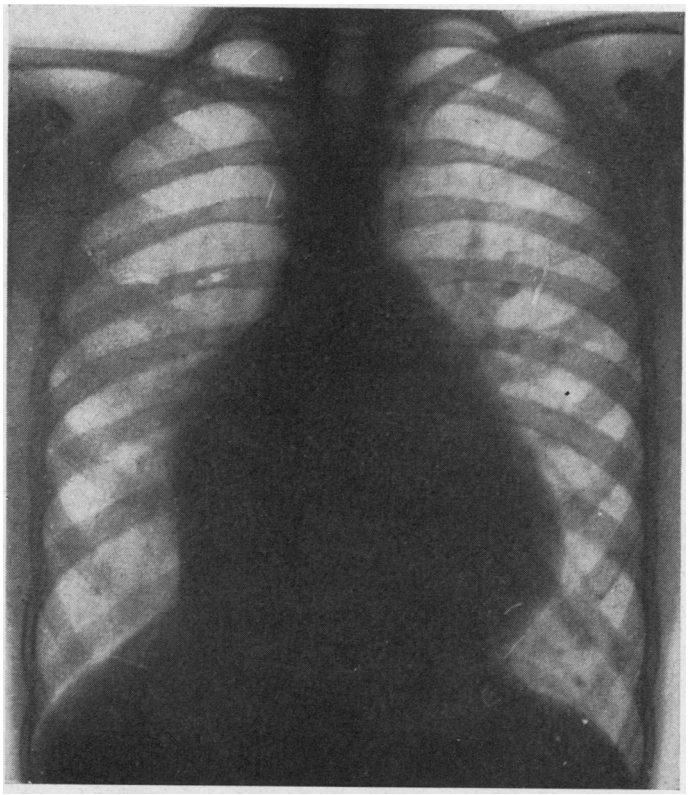

FIG. 1. *The opacity of the anomalous vein is hidden by the right cardiac border due to dextrocardia.*

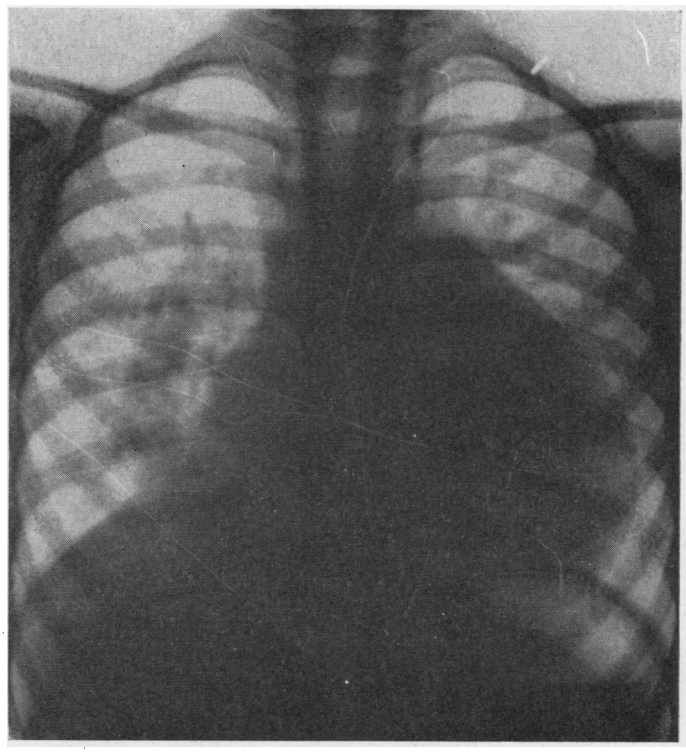

FIG. 2. *Typical scimitar image.*

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persistent left superior vena cava with a large communication at the atrial level. Also the entire right lung appeared to drain into a common channel entering the I.V.C. below the diaphragm (Fig. 3). There was a left-to-right shunt of 3·8 volumes at the atrial level. The right ventricular and pulmonary artery pressures measured 40 mm. Hg.

On 1 October 1964 a right anterolateral thoracotomy was performed in the fourth intercostal space; this was enlarged to include transsection of the sternum. The findings included a large anomalous trunk (Fig. 4) draining the whole of the right lung and entering the I.V.C. below the diaphragm, an early division of the pulmonary artery within the pericardium, and a persistent left S.V.C. draining into the right S.V.C. through the innominate vein.

FIG. 3. *Case* 2. *Pre-operative angiogram showing anomalous vein.*

Bypass was started and the left S.V.C. was clamped. A large high A.S.D. was found. The anomalous venous trunk was too short to be transplanted directly into the left atrium, which was small. The anomalous venous trunk was therefore implanted into the left atrium by means of a 20-mm. Dacron prosthesis which was brought through the right atrial chamber and sutured directly into the A.S.D. (Fig. 5). The edges of the atriotomy were sutured around the prosthesis. The last anastomosis (extracardiac) between the vein and the peripheral end of the prosthesis was performed after bypass had been stopped, with the right pulmonary artery clamped. After release of the clamps the pulmonary venous drainage appeared to be adequate and the course of the prosthesis did not interfere with circulation in the right atrium. This was confirmed post-operatively by angiography (Fig. 6).

CASE 3 V. Roland, aged 15 years, had developed normally since infancy, the only finding being repeated pulmonary infections and a mild degree of dyspnoea on effort. On auscultation a systolic murmur of moderate degree was found with splitting of the second heart sound over the second intercostal space. Radiography (Fig. 7) showed moderate cardiac enlargement with displacement to the right and a double

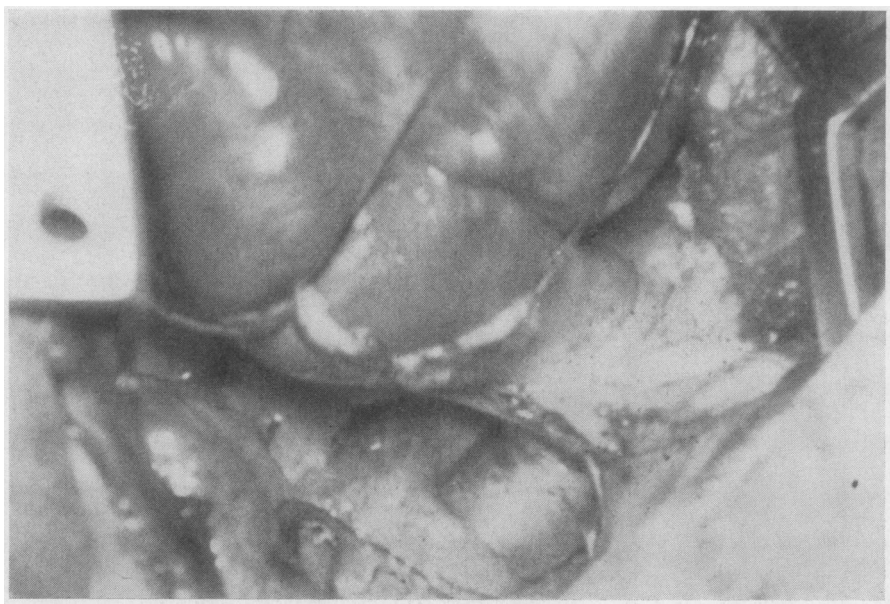

FIG. 4. *Case* 2. *Findings at operation.*

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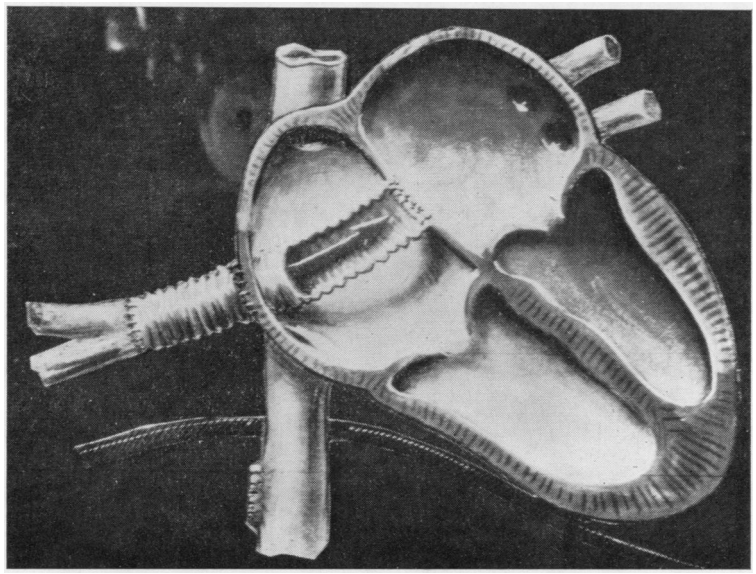

FIG. 5. *Case 2. A model of the operation performed.*

contour shadow along the right cardiac border. The scimitar sign was not seen on this film. Hypervascularization was present. The E.C.G. showed right axis deviation and hypertrophy of the right ventricle.

On catheterization a communication at the atrial level was found, with a left-to-right shunt of 3 volumes. The pulmonary artery pressure was 38 mm. Hg. Angiography revealed anomalous right pulmonary venous drainage entering the I.V.C. at the level of the diaphragm. At bronchoscopy the main right bronchus was of exceptional length, dividing into two parallel channels. Bronchography (Figs 8 and 9) confirmed the

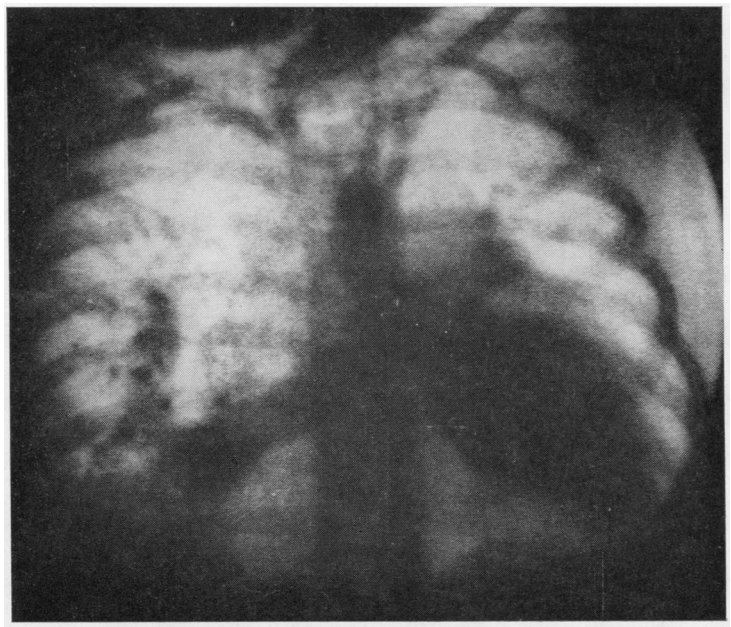

FIG. 6. *Case 2. Post-operative angiogram.*

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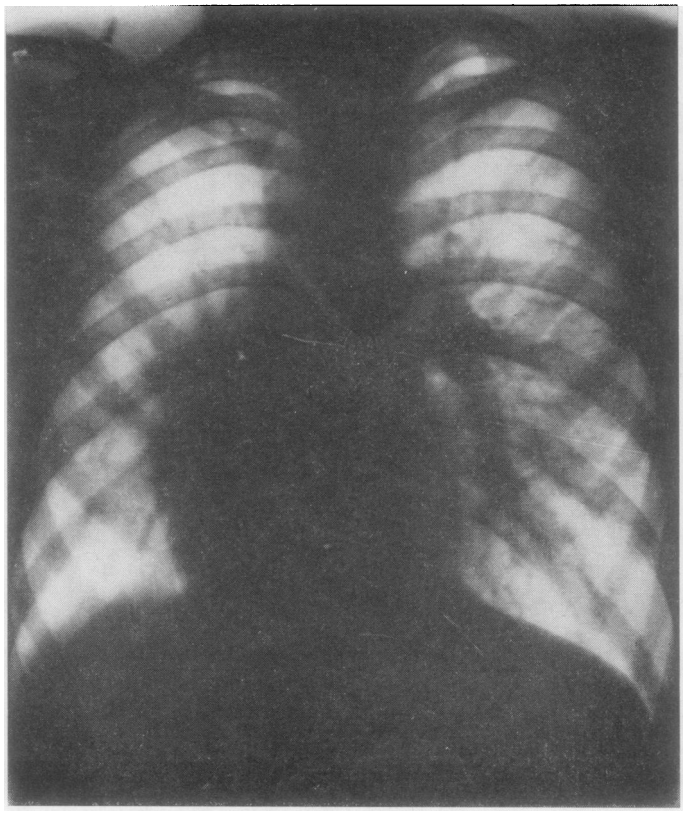
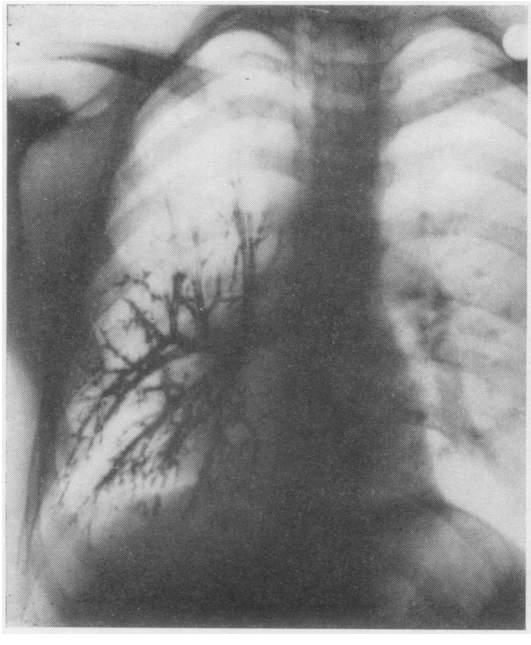


FIG. 7. *Case* 3. *Displacement of the heart to the right makes it impossible to observe the scimitar image in the postero-anterior view. The left anomalous pulmonary artery is seen.*



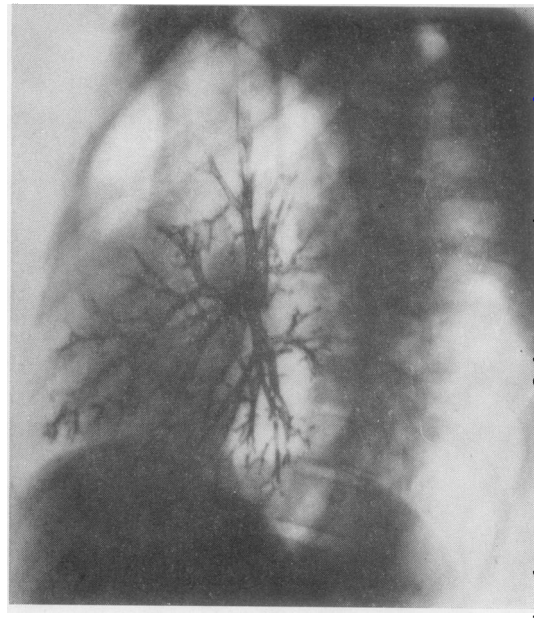


FIG. 8

FIG. 9

FIGS 8 *and* 9. *Case* 3. *Anomalous segmentation of the bronchial tree as seen on bronchography.*

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segmental anomaly. Pulmonary function tests were performed and found to be within normal limits.

On 28 March 1963 a right anterolateral thoracotomy in the fifth intercostal space was performed. The right lung consisted of two lobes, a large posteroinferior and a small anteromedial lobe. Total anomalous pulmonary venous drainage was found, with a common trunk entering the I.V.C. below the diaphragm. After extracorporeal circulation had been started a high A.S.D. was found. The anomalous pulmonary trunk was divided and was long enough to be sutured directly into the left atrium behind the septum. The A.S.D. was closed primarily. Following bypass the small previously described anteromedial lobe of the lung was removed to facilitate proper drainage of the trunk into the left atrium.

REVIEW OF THE LITERATURE

The first descriptions were by Chassinat (1836) and Cooper (1836) from observations made at necropsy. Radiological recognition was described by Grishman, Poppel, Simpson, and Sussman (1949), Runström and Sigroth (1950), and Welti and Nedey (1950).

A few years later other signs of this defect were described along with associated anomalies, principally by Findlay and Maier (1951) and Snellen and Albers (1952), with appreciation of the anatomical syndrome by Halasz, Halloran, and Liebow (1956) and Steinberg (1957). In 1960, Neill, Ferencz, Sabiston, and Sheldon described the specific radiological characteristics of the anomaly and gave the name 'scimitar syndrome' which is now in common use.

TABLE I

Author/Year	No. of Cases	Author/Year	No. of Cases
Alexiu and Stan (1965)	1	Longin and Peppmeier (1958)	1
Arvidsson (1954)	1	McCormack, Marquis, Julian, and Griffiths (1960)	2
Bourrassa (1963)	2	McKusick and Cooley (1955)	1
Bruwer (1956)	5	Michaud, Saubier, Maret, Viard, and Termet (1963)	3
Cabrol, Merlier, and Morel (1957)	1	Neill *et al.* (1960)	2
Chassinat (1836)	1	Papagni and Rovelli (1962)	1
Cooke, Evans, Kistin, and Blades (1951)	1	Park (1912)	1
Cooper (1836)	1	Runström and Sigroth (1950)	1
Cornet, Laboux, and Dupon (1963)	1	Sanger, Taylor, and Robicsek (1963)	5
Dalith and Neufeld (1960)	5	Sepulveda, Lukas, and Steinberg (1955)	1
Dotter, Hardisty and Steinberg (1949)	1	Snellen and Albers (1952)	1
Drake and Lynch (1950)	2	Steinberg (1957)	3
Dubost, Blondeau and Piwnica (1963)	1	Steinberg (1959)	3
Fiandra, Barcia, Cortes, and Stanham (1962)	1	Torner-Soler, Balaguer-Vintro, and Carrasco-Azemar (1958)	1
Findlay and Maier (1951)	1	Törnvall, Jackson, Alvayay, Vargas, Koch, and Zárate (1961)	1
Frye, Marshall, Kincaid, and Burchell[1] (1962)	3 (5)	Van Praagh, Van Praagh, Vlad, and Keith (1964)	1
Gilman, Skowron, Musser, and Bailey (1957)	1	Welti and Nedey (1950)	1
Grishman *et al.* (1949)	3	Wood, Conrad, and Morrow (1957)	1
Halasz *et al.* (1956)	3		
Hollis (1964)	1		
Jue, Amplatz, Adams, and Anderson (1966)	4		
Kirklin, Ellis, and Wood (1956)	1		
Kittle and Crockett (1962)	1		

[1] Among the five cases published by Frye *et al.* one had previously been reported by Bruwer and another by Kirklin.

INCIDENCE Total anomalous right pulmonary venous drainage to the I.V.C. is rare. A review of the literature has disclosed 74 case reports, not including our three (Table I).

This particular defect is one of the least frequent among the many varieties of anomalous pulmonary venous drainage. Between January 1958 and May 1966 these three cases were found among 55 others of anomalous venous drainage (Tables II and III).

TABLE II

ORIGIN OF ANOMALOUS VEIN IN 55 CASES OF ANOMALOUS RETURN OBSERVED FROM 1958 TO 1966

Origin	No.
Right lung	51
Left lung	2
Both lungs	2
Total	55

TABLE III

TERMINATION OF ANOMALOUS VEIN IN 55 CASES OF ANOMALOUS RETURN OBSERVED FROM 1958 TO 1966

	No.
Superior vena cava	17
Right atrium	23
Left innominate vein	3
Inferior vena cava	3
Multiple channels (superior vena cava and right atrium)	9
Total	55

DESCRIPTION OF THE SYNDROME A number of defects were consistently found in this anomaly and together comprise the syndrome. In addition to the persistent anomalous venous return to the I.V.C., there were frequent anomalies of the right lung (bronchial and arterial) and dextroposition of the heart. Other congenital anomalies of the heart and diaphragm have been associated with this defect but are rare and are not included in the classical description.

SPECIFIC FINDINGS OF THE SYNDROME

Anomalous venous drainage of the right lung into the inferior vena cava The detailed anatomy of the deformity was described in 57 of the reported cases. In the majority the anomalous trunk repre-

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TABLE IV

VOLUME OF LUNG DRAINING INTO ANOMALOUS TRUNK

Origin of Anomalous Trunk		No. of Cases
Total right lung		36
Total lung except 1 or 2 segments		6
Two lobes		
Superior and middle	1	3
Middle and inferior	2	
Superior lobe		2
Inferior lobe		
When 3 lobes were present	1	10
When 2 lobes were present	2	
Inferior part of lung		
Only one lobe	2	
Not described	5	
Total		57

sented the return from the entire right lung (Table IV). The anomalous trunk has been single in the majority of cases; multiple trunks were found in three. The trunk formed in the region of the hilum and passed inferiorly and to the left, generally along the greater fissure when this was present. The trunk generally left the lung at the cardiophrenic angle.

The point of entrance into the I.V.C. was variable. The common trunk always entered the I.V.C. above the renal veins and most frequently below the diaphragm and above the attachment of the hepatic veins. Sometimes the trunk remained intrathoracic and ended above the diaphragm. In these cases it entered the I.V.C. at the level of the right atrium. In a few cases the trunk actually terminated within the diaphragm.

By definition, the anomalous trunk must enter the I.V.C. In a number of cases the I.V.C. did not represent the only termination of the anomalous trunk. In 21 out of 57, the anomalous trunk ended in two or more branches with variable entrances. However, in the majority the vessel to the I.V.C. carried most of the blood from the lung (Table V).

Anomalies of the right lung Table VI analyses the findings reported in 21 cases where the lung was precisely described. Generally, the volume of

TABLE V

SITE OF TERMINATION OF ANOMALOUS TRUNK IN 57 CASES

Total Drainage into	No. of Cases
Inferior vena cava	36
Inferior vena cava and left atrium	12
Inferior vena cava and right atrium	1
Inferior vena cava and superior vena cava	2
Inferior vena cava and azygos vein	1
Inferior vena cava and (not precisely described)	5
Total	57

TABLE VI

STUDY OF FISSURE MORPHOLOGY OF RIGHT LUNG IN 21 CASES

		No. of Cases
3 lobes present		7
2 lobes present:		
Large superior lobe and small inferior lobe	3	7
Large inferior lobe and small superior lobe	3	
2 lobes of equal volume	1	
Single lobe (complete absence of fissure)		7
Total		21

the right lung appeared to be reduced. Microscopic examination of the lung generally did not reveal any abnormal findings. Bronchoscopy and bronchography revealed a frequent incidence (77%) of anomalies of the lung fissures. These are described in Table VII. The most frequent anomaly found was a mirror image of the left

TABLE VII

SEGMENTATION OF RIGHT BRONCHIAL TREE IN 22 CASES

Bronchial Segmentation		No. of Cases
Normal		5
Absence of superior lobe bronchi	2	3
Absence of middle lobe bronchi	1	
Mirror image of left bronchial tree		11
Atypical and not precise		3
Total		22

lung. Sometimes the anomalies were haphazard and not described. In other cases, atresia of lobar or segmental branches was found. Some cases showed alterations in the calibre of the bronchi, often showing localized or diffuse bronchiectasis or hypoplasia (Table VIII).

TABLE VIII

ANOMALIES OF BRONCHIAL CALIBRE IN 22 CASES

Bronchial Calibre		No. of Cases
Normal		17
Ectasia		
Diffuse	1	3
Localized	2	
Stenosis		
Superior lobe bronchi	1	2
Superior and inferior lobe bronchi	1	
Total		22

Anomalies of vascularization of the right lung Vascular anomalies associated with this defect were frequent and mainly involved the pulmonary artery or an aberrant origin of systemic vessels

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On angiography of the pulmonary artery in 27 cases, 60% had anomalies of the calibre of the vessel, usually hypoplasia (Table IX). In those cases where the bronchial tree was a mirror image of the left lung, the pulmonary artery pattern likewise had a distribution similar to that of the left side.

TABLE IX

STUDY OF RIGHT PULMONARY ARTERY IN 27 CASES OBSERVED ON ANGIOGRAPHY

Calibre of Right Pulmonary Artery	No. of Cases
Normal	12
Hypoplasia	11
Atresia	2
Ectasia	2
Total	27

Angiography frequently showed pulmonary vessels arising from systemic origins. In 29 cases (25 thoracotomies and 4 necropsies) there was a 48% incidence of abnormal vascular communication from the aorta or its branches. These represented one or more vascular channels accompanying the anomalous venous trunk, generally supplying the inferior lobe (Table X).

TABLE X

SYSTEMATIC VASCULARIZATION OF RIGHT LUNG: OBSERVATIONS FROM 14 CASES

Vessels	No. of Cases
Number	
One artery	4
More than one artery	8
Not described	2
Origin	
Below diaphragm	11
Above diaphragm	2
Above and below diaphragm	1
Amount of lung supplied	
Right inferior lobe	9
Entire right lung	4
Not described	1

Dextroposition of the heart Displacement of the heart to the right hemithorax was generally not prominent. This does not represent a true dextrocardia, but the term is commonly used.

RARE ASSOCIATED ANOMALIES

Cardiovascular anomalies Although an A.S.D. is generally regarded as a rare anomaly in this defect it was present in all three of our cases. Other cardiovascular anomalies have been observed, as recorded in Table XI.

TABLE XI

INCIDENCE OF ASSOCIATED CARDIOVASCULAR ANOMALIES OBSERVED IN 75 CASES

Associated Cardiovascular Anomaly	No. of Cases
A.S.D.	8
V.S.D.	3
Tetralogy of Fallot	1
Patent ductus arteriosus	3
Coarctation of aorta	1
Persistent left superior vena cava	1
Pulmonary stenosis	1
Total	18

Diaphragmatic anomalies Significant anomalies of the diaphragm were found in 6 out of 75 cases. The usual anomaly was eventration of the right cupola. In one case there was a persistence of the foramen of Bochdalek. In another there was duplication of the right cupola.

SYMPTOMATOLOGY The defect was often well tolerated, and of 53 observations the patient was asymptomatic in 22 (40%). In 31 (60%) the patient had some trouble with the cardiorespiratory system. The difficulties were similar to those observed in patients with large left-to-right shunts and some degree of pulmonary hypertension. In addition, a few patients presented because of complications resulting from the bronchial malformations. Table XII shows the different symptoms presented by 31 patients.

TABLE XII

INCIDENCE OF SYMPTOMS OBSERVED IN 31 PATIENTS

Symptom	No. of Cases
Bronchopulmonary infection (repeated)	16
Cough	4
Haemoptysis	3
Dyspnoea on exertion	16
Palpitations	5
Intermittent cyanosis	5
Pain in right hemithorax	3

Clinical examination was not specific for this syndrome. A systolic murmur of moderate intensity was usually heard but was not constant. This was observed in 23 cases. Table XIII shows the age on discovery of the malformation in 53 cases.

Diagnostic features The E.C.G. generally showed hypertrophy of the right chambers. The radiographic findings were of primary importance and the scimitar sign was characteristic of the disease. There was always a long opacity along the right

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TABLE XIII

AGE ON DISCOVERY OF DEFECT: ANALYSIS OF 53 CASES

Age (Years)	No. of Cases
0–10	16
11–20	6
21–30	17
31–40	3
41–50	6
51–60	5

pericardiac border extending towards the cardiophrenic angle. This opacity appeared to be vascular and when seen it demanded further investigation. On some occasions this image was not clearly seen because of the dextroposition of the heart, and the anomalous trunk could best be seen on the right anterior oblique film or on lateral views. The course of the anomalous trunk could be outlined precisely by tomography. The exact termination of the vein, however, had to be confirmed by angiography, which often showed additional anomalies of the pulmonary artery. At catheterization the anomalous trunk could be entered by way of the saphenous vein.

Bronchoscopy, bronchography, and pulmonary function tests are necessary for complete investigation. Radiography of the right hemidiaphragm after injection of air into the peritoneal cavity and aortography may be considered.

TREATMENT Although this defect is usually well tolerated for a long time, progression towards the development of pulmonary hypertension requires early intervention.

Of the 75 cases found in the literature 25 have undergone surgery. Table XIV describes the surgical approach in 25 cases. Chiefly, two types of intervention have been performed, one being removal of the affected lung and the other correction of the anomalous vein.

In choosing the correct operation for a given patient, the degree of associated bronchopulmonary anomaly is most important. In the absence of significant bronchopulmonary malformations, the treatment of choice is reimplantation of the anomalous venous trunk into the left atrium. Anterolateral thoracotomy is the incision of choice, primarily because of the possible need for associated pulmonary surgery. Extracorporeal circulation is necessary except in those cases where an A.S.D. can be positively ruled out.

TABLE XIV

SURGICAL TREATMENT IN 25 CASES

Type of Surgery	No. of Cases
Exploratory thoracotomy	3
Decortication of right lung	1
Ligature of anomalous trunk	1
Right pneumonectomy	6
Right inferior lobectomy	3
Right superior lobectomy associated with reimplantation of anomalous vein	1
Reimplantation of anomalous vein	10
Total	25

Dissection of the anomalous pulmonary trunk is sometimes difficult due to the large systemic bronchial circulation in conjunction with the vein. Different techniques may be used to correct the defect. If the left atrium is large enough the anomalous vein may be implanted directly, or with a prosthesis if the vein is too short.

Another technique consists of implanting the vein into the right atrium, adjacent to the A.S.D., with correction by a patch or direct suturing. If necessary, the A.S.D. may be enlarged or even created.

In one of our cases (case 2) another type of repair was used. A Dacron prosthesis was passed through the right atrium and sutured along the edges of the A.S.D. without impairment of the circulation. Post-operative angiograms confirmed the success of this technique.

The results of these operations are good. Only one case of thrombosis has been reported. In the absence of significant bronchopulmonary malformations, a total correction should always be considered.

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ADDENDUM

Other cases have been recently reported by: Enjalbert, Gédéon, Eschapasse, Mathe, and Puel (1966); Massumi, Alwan, Hernandez, Just, and Tawakkol (1967); Kiely, Filler, Stone, and Doyle (1967) and Brocard, Gallouedec, and Vannier (1967).

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