Transposition of the Pulmonary Veins

By Warren G. Guntheroth, M.D., Alexandes S. Nadas, M.D., and Robert E. Gross, M.D.

PULMONARY veins entering the right atrium or its tributaries constitute the entity of transposition of the pulmonary veins. The transposition may be complete, when all the veins enter the right heart, or incomplete, when only some of the pulmonary veins drain anomalously. We are reviewing our experiences with total and partial anomalies of pulmonary venous drainage because of the increasing surgical interest in the problem, and because we feel that the clinical profile to be presented is sufficiently characteristic to enable the cardiologist to make the diagnosis with relative ease in most instances.

MATERIALS AND METHODS

Thirty-one patients with uncomplicated transposition of the pulmonary veins admitted to the Children's Medical Center in Boston, were studied clinically, with complete history, physical examinations, x-ray and fluoroscopic examinations, and 12-lead electrocardiograms. The diagnosis was confirmed in each instance by cardiac catheterization, surgery, or autopsy. The technics used in our cardiac catheterization laboratory have been described in previous publications.3

COMPLETE TRANPOSITION OF THE PULMONARY VEINS

Definition

Drainage of the entire systemic and pulmonary return into the right heart is the major abnormality in complete transposition of the pulmonary veins. An atrial septal defect or a patent foramen ovale is necessary for any systemic output, and is therefore an integral part of the lesion. Following Brody's original classification,2 we have not considered a patent ductus arteriosus as a complicating anomaly, but have excluded cases with other major cardiovascular abnormalities from discussion here.

Incidence

Complete transposition of the pulmonary veins is a rare condition. Maude Abbott3 found only 4 cases (including 3 complicated ones) in her series of 1,000 autopsies of congenital heart disease. Brody, in a classical monograph in 1942,5 reviewed 38 cases from the literature; 14 of these were associated with other major cardiac anomalies. Up to the present time there have been 159 uncomplicated cases reported.4-11

Embryology

The developmental anomaly producing this malformation probably occurs in the seventh week of gestation12 and represents either complete failure of absorption of the common pulmonary vein into the dorsal wall of the sinus venosus, or abnormal placement of the ostia of this vein in relation to the septum, which divides the sinus venosus into the right and left atria. In the first instance, the pulmonary veins drain via a persistent, enlarged primitive connection between the pulmonary and systemic circulations. These connections have been demonstrated in normal adults with postmortem injections by Zuckerkandl in 1881, and are reminiscent of the development of the entire pulmonary circulation from part of the presplanchnic plexus.13

Anatomic Considerations

The site at which the pulmonary veins connect to the systemic circulation has been used as the basis of anatomic classification of complete transposition of the pulmonary veins by Darling, Rothney, and Craig.4 This
classification has considerable interest for prognostic and surgical reasons. We have classified 159 uncomplicated cases from the literature according to this method (table 1).

Certain other anatomic considerations are of surgical importance. The anomalous veins, with great consistency, form a relatively large common channel prior to joining the systemic veins except when they join the right atrium directly. In the latter group approximately one third join the right atrium by multiple channels, but more than one half empty into an ante-chamber, described by Kirchmair and Blum, which is, from the surgeon's point of view, conveniently located on the posterosuperior wall of the right atrium, to the left of the superior vena cava and inferior vena cava, and superior to the coronary sinus and the atrioventricular conduction system.

Finally the entire left heart is relatively hypoplastic; and, as Keith pointed out, the circumference of the common pulmonary vein frequently exceeds that of the waist of the left atrial appendage, a formidable disadvantage in attempting surgical correction of this condition.

**Physiology**

Complete transposition of the pulmonary veins is a serious anomaly, profoundly altering the normal circulatory pattern. In this anomaly, the right atrium receives the entire systemic and pulmonary venous return, and supplies the left heart through a patent foramen ovale or an atrial septal defect. As in other types of congenital heart disease with complete mixing, the systemic arterial saturation will depend entirely on the ratio of pulmonary to systemic flow, at a given oxygen consumption.

However, as various investigators have pointed out, mixing at the atrial level may not be complete. The systemic arterial saturation is often slightly less than that of the pulmonary artery; this difference is attributed by the Mayo group to preferential shunting of the inferior vena caval blood through the interatrial communication. This type of preferential shunting has a precedent in fetal circulation.

Occasionally, the pulmonary artery oxygen saturation is lower than that of the systemic artery, perhaps due to preferential flow of blood from the coronary sinus or superior vena cava across the tricuspid valve into the right ventricle.

Although the resting systemic output of these patients is usually normal or only slightly reduced, increasing output 3 or 4-fold (as with moderately severe exercise) would require a corresponding increase in pulmonary blood flow, if the systemic arterial saturation were maintained. With a resting heart rate greater than average and a right ventricular stroke volume 2 to 3 times that of the left, the required pulmonary blood flow would be almost impossible to achieve. Evidence of the resultant limitation of cardiac output is the almost universal history of easy fatigability and the autopsy finding of a hypoplastic left heart.

Although the majority of patients with complete transposition of the pulmonary veins die in infancy or early childhood, a few survive into adulthood with remarkably few symptoms. How this group maintains a compensated cardiovascular system in the face of such profound disorder has never been explained satisfactorily.

**Presentation of Data**

**Clinical Material**

Twenty patients with uncomplicated, complete transposition of the pulmonary veins, are the subject of the present communication. They are seen to have the characteristic clinical and hemodynamic abnormalities and are divided into 4 groups according to the level of drainage of the pulmonary veins, classified into superior, cardiac, infracardiac, and mixed levels.

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<td></td>
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<tr>
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<td>IIIA coronary sinus</td>
<td>14%</td>
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<td></td>
<td>IIIB right atrium</td>
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<td>Infrcardiac</td>
<td>IIIC portal vein or ductus venosus</td>
<td>9%</td>
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<tr>
<td>Mixed levels</td>
<td>IV</td>
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*Dow and Burchell have recently derived formulae to predict the systemic arterial saturation on the basis of these parameters.
complete transposition of the pulmonary veins have been studied at our hospital in the years 1950 to 1956. Because the clinical picture may vary considerably, depending upon the level at which the pulmonary veins drain, we have used Darling, Rothney, and Craig's classification to subdivide our material. The pulmonary veins drained into an ascending left superior vena cava in 9 of our cases, into the coronary sinus in 4, into the right atrium in 3, and below the diaphragm in 4 patients.

The clinical diagnosis was confirmed by operation or autopsy in 15 patients, and by cardiac catheterization only in the remaining 5.

Our patients ranged in age from 10 days to 19 years with an average age of 5 years; 10 of them were less than 6 months old. The group draining below the diaphragm was unique in that none survived more than 7 weeks and 2 survived only 10 days. None of the others died at less than 7 weeks of age.

The incidence of males in the entire group was the same as females; in the infradiaphragmatic group, all 4 patients were males.

The interatrial communication in the 12 autopsied cases was a patent foramen ovale in 10 instances, and an atrial septal defect in 2 (1 ostium primum and 1 ostium secundum). The size of the interatrial opening in our autopsied cases had no correlation with longevity or degree of unsaturation when taken in relation to the circumference of the mitral valve (table 2). Previous reports attributing prognostic importance to the size of this opening failed to correlate it with the over-all heart size.

Similarly, the presence and size of a patent ductus arteriosus had no correlation with longevity in our patients. Eight of the 12 autopsied cases had patent ducti, probe-patent in 5, moderate sized in 1, and large in 2.

Mild pulmonary vascular obstruction was found in 5 autopsied patients, and moderate obstruction in 4. Although the 3 patients in

| Table 2.—Correlation of Age of Death with Size of Interatrial Communication and Patency of Ductus Arteriosus |
|---|---|---|---|---|
| Age | Type | Degree cyanosis | Circumference ASD or PFO (cm.) | Circumference mitral valve (cm.) | Ductus |
| 10 days | III | 3+ | 2.8 | 3.0 | Large |
| 10 days | III | 1+ | 2.5 | 3.0 | Closed |
| 6 weeks | III | 1+ | 4.7 | 3.0 | Probe patent |
| 7 weeks | III | 1+ | 5.5 | 3.5 | Closed |
| 2 months | I | 2+ | 1.5 | 3.0 | Large |
| 2 months | I | 1+ | 1.5 | 3.7 | Probe patent |
| 2 months | I | 2+ | .6 | 3.5 | Closed |
| 3 months | I | 1+ | 1.5 | 3.5 | Probe patent |
| 4 months | II A | 1+ | 3.9 | 4.0 | Closed |
| *1 1/2 yrs.* | II B | 1+ | 5.5 | 5.5 | Probe patent |
| *1 1/2 yrs.* | I | 1+ | 3.1 | 5.0 | Moderate size |
| *4 1/2 yrs.* | I | 2+ | 2.4 | 5.0 | Probe patent |

* Operative deaths. ASD = atrial septal defect. PFO = patent foramen ovale.

the autopsied group surviving the longest had no obstructive changes, there was no correlation, in the other 9 patients, of age at death with severity of medial hypertrophy and intimal hyperplasia of the pulmonary arteries.

**History**

There was no family history of congenital heart disease in any of our 20 patients. In 4 instances, however, there were difficulties in the first trimester of pregnancy; 2 "viral" respiratory infections, 1 threatened abortion, and 1 instance of severe malnutrition.

The symptoms presented by these patients were nonspecific; mild cyanosis, fatigability, dyspnea, and increased frequency and severity of respiratory infections were almost universal complaints. The onset of cyanosis varied from birth to 12 years of age. Squatting and "anoxic" spells were relatively uncommon, occurring in one fifth of the patients.

**Physical Examination**

The findings at physical examination were more specific, in certain instances, than were the histories of these patients. The patients were small and undernourished in general; over two thirds of them fell below the twenty-fifth percentile on our height and weight charts.20

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*The autopsy material of the Children's Medical Center has been presented in detail in an earlier publication.*
The patients without exception were at least mildly cyanotic; more marked cyanosis was found in the group with congestive failure, and in the group with drainage below the diaphragm. Only one half of the patients demonstrated clubbing.

The resting pulse rate was above normal range in 14 patients, including 11 in congestive failure. The blood pressure and pulse pressure did not vary significantly from the normal.

Congestive failure was diagnosed in 16 of 20 cases, manifested in every instance by a large liver, but in only half by distended veins or peripheral edema. Hepatomegaly was particularly marked in the patients with drainage below the diaphragm. The presence of pulmonary congestion was difficult to ascertain by physical examination alone, because of the high incidence of concomitant respiratory infections. In 1 case with drainage into the portal vein, pulmonary edema was diagnosed clinically and radiologically and was confirmed at autopsy.

Left chest prominence was found in 8 of the older children with very large hearts. In every instance, the cardiac impulse was maximal at the lower left sternal border.

On auscultation, the first sound was loud at the apex or lower left sternal border in 6 cases; the second sound was increased in intensity at the pulmonic area in one half the cases, and within normal limits in the remainder. Splitting of the second sound was observed in all but 2 patients. An unusually prominent third sound was present in 13 of 20 patients, and a fourth, or atrial sound was audible in 5 patients.

Systolic murmurs were heard in all except 2 patients, who were 6 and 8 weeks of age respectively. The systolic murmurs were usually soft and heard best at the left sternal border, the majority being maximal at the fourth interspace, but only slightly fewer at the second to third interspaces. This type of soft systolic murmur has been attributed to tricuspid insufficiency by Keith. A palpable systolic thrill and rough, loud murmur were noted in 5 patients; 4 of these were found to have a pressure gradient across the pulmonic valve, and in the fifth, catheterized from the femoral vein, the pulmonary artery was not entered.

Eleven patients had low frequency mid-diastolic murmurs at the apex or lower left sternal border. This type of murmur, frequently found in patients with a large ventricular stroke volume, was observed in every patient over 6 months of age. A presystolic murmur of medium to high frequency at the lower left sternal border, synchronous with atrial systole, was heard in 6 instances (fig. 1). This murmur was usually diamond shaped on the phonocardiogram, and was accompanied by a fourth sound. Examples of this murmur were heard in all types except the group with subdiaphragmatic drainage; within the framework of the other clinical features, we consider its presence to be practically pathognomonic.

Keith and associates have described a pronounced, although transient, venous hum at the upper left sternal border in 4 patients with drainage of the pulmonary veins into a remnant of the left superior vena cava. We were able to find this type of murmur in only 1 case, and then the murmur was maximal at the right sternal border.

Electrocardiograms

The electrocardiograms of patients with complete transposition of the pulmonary veins reflect the pathologic physiology and anatomy, with evidence of right-sided hypertrophy and dilatation. Right axis deviation, right ventricular hypertrophy, and inom-
complete right bundle-branch block were present in every patient (fig. 2). The degree of right ventricular hypertrophy in this group of patients is marked and has been noted by others. Although Keith and co-workers emphasized the frequency of a qR pattern in the right precordial leads, we found this pattern in only 5 cases; and in these there was evidence suggesting that the pattern was actually an rsR' pattern, with an isoelectric r. A more frequent finding in our series was complete reversal of the adult R/S progression in the precordial lead, i.e., qR, rsR', or Rs pattern in the right precordial leads, and qrS in the left precordials, indicating marked right ventricular hypertrophy. Sixteen of our patients demonstrated this phenomenon, and the exceptions were all in the older age group.

Right ventricular intrinsicoid deflection delay correlated closely with age, but was abnormally delayed in 15 cases. The group draining below the diaphragm were an exception, with 3 out of 4 having a normal activation time, which may be correlated with the relatively small size of their hearts by x-ray and postmortem examination.

T-wave abnormalities occurred in 13 cases. An additional 3 cases had T-wave abnormalities, possibly due to digitalis.

Fig. 2. Representative electrocardiogram in complete transposition of the pulmonary veins, illustrating marked right ventricular hypertrophy, incomplete right bundle-branch block, P' pulmonale, and T-wave changes.

A tall, peaked P wave, or P pulmonale, was present in 13 of 20 cases. In the 4 patients with drainage below the diaphragm, there were no instances of P pulmonale.

X-Rays

The radiograms of certain types of complete transposition of the pulmonary veins are practically pathognomonic, as for exami-
ple the figure-eight pattern described by Snel- len and Albers23 (fig. 3). The upper half of the 8 is formed by a remnant of the left super- ior vena cava, as it ascends to join the left innominate, and the right superior vena cava emptying caudally into the right atrium. This venous structure will pulsate, if at all, in an "acv" pattern, whereas the pulmonary artery will expand synchronously with ven- tricular systole. The thymus, in the upper, anterior mediastinal area, does not pulsate intrinsically at all. A wide superior mediastinal pattern also may be produced by a true, persistent left superior vena cava without pulmonary venous anomaly, as it de- scends to drain into the coronary sinus; this group lacks, however, the rounded upper mar- gin characteristic of the figure-eight, and the patients are not ordinarily cyanotic. The full-blown figure-eight pattern is not usually found in infancy. The youngest patient in our series with this contour was 4 years old, although in every case over the age of 3 months, widening of the superior mediastinum was definite.

The group with drainage below the dia- phragm also presents a pathognomonic roent- genologic picture of moderate to marked pul- monary vascular engorgement and congestion with a normal-sized heart (fig. 4). There is
no other type of cyanotic congenital heart disease demonstrating this combination, in the presence of congestive failure. However, the vascular pattern per se is not different from that seen in other heart diseases in which there is passive congestion, such as mitral stenosis.

When drainage occurs at the atrial level, the radiogram is nonspecific in most instances. Two of 4 cases with pulmonary return into the coronary sinus had a widened superior mediastinal shadow due to a true left superior vena cava, which also drained systemic blood into the coronary sinus. One of these patients had a bulge in the posterior aspect of the cardiac shadow, on barium swallow, suggesting by its size and location an enlarged coronary sinus (fig. 5). Another patient with drainage into the coronary sinus had an angular prominence of the upper right atrial border, directly across from the sinus ostium. When the veins drain directly into the right atrium, the picture is characteristic only of the basic disorder (fig. 6). The cardiac silhouette produces a box-like shadow, as Gott et al.6 pointed out, but so do most lesions involving the right side of the heart with increased pulmonary blood flow.

Cardiomegaly was present in our 20 patients in every instance except for the 4 cases with drainage below the diaphragm. In the majority of cases, the enlargement was moderate to marked. In the cases where congestive failure had not obliterated the character-
istic contour, right ventricular configuration was uniformly present. Atrial enlargement could be diagnosed with certainty in 9 patients; there was no demonstrable atrial enlargement in any of the cases draining below the diaphragm. The pulmonary artery was abnormally prominent in over one half, and pulmonary vascular engorgement was marked in all. Passive congestion was present in over half of the children, including all of those with drainage below the diaphragm, in whom it was particularly marked. "Hilar dance" or intrinsic pulsation of the hilar vessels, was found in 8 of the 20 cases, and its absence was correlated in every instance but 1 with an age of less than 6 months.

Angiocardiograms were performed in only a small number of our cases (fig. 7). Since a definitive diagnosis can be made without angiocardiographs, the additional risk of this procedure is not justified, in our opinion, except in those cases being considered for cardiac surgery when additional anatomic detail is desired; and then, it should be done by selective angiocardiography. By venous angiocardiography, however, the exact site of union was never established, when it was not known with certainty prior to the procedure.

Cardiac Catheterization

We have catheterized 10 patients with complete transposition of the pulmonary veins, including 4 patients with drainage into the superior vena cava, 3 draining into the coronary sinus, and 3 into the right atrium, (table 3). None of our 4 cases with drainage below the diaphragm was catheterized (none
survived beyond 7 weeks). The ages of the catheterized subjects ranged from 6 months to 18 years with an average age of 9 years.

Arterial oxygen saturation ranged from 80 to 93 per cent with an average of 87 per cent. (This range excludes 1 patient whose condition deteriorated during the procedure, whose saturation was 47 per cent.) The rise in oxygen saturation at the site of union of the systemic and pulmonary returns, varied from 15 to 29 per cent with an average of 25 per cent. The resting cardiac output ranged from 2.3 to 6.6 L per minute per M² of body surface area, with an average of 3.9 (within normal limits for resting cardiac index). Pulmonary blood flow, however, ranged from 4.3 to 15 L per minute per M², with an average of 8.7. The average ratio of pulmonary to systemic flow, thus, was 2.2 to 1. Although literally all of the systemic flow was via a right-to-left shunt at the atrial level, the effective right-to-left shunt (i.e., the dilution of oxygenated blood with unsaturated venous return) was small relative to the left-to-right shunt. This ratio, on the average, was 1 to 7.*

Right atrial pressures were somewhat increased, on the average, although there was considerable spread, 3 to 15 mm. Hg. In 1 patient the left atrium was entered and was found to have a mean pressure of 1 mm. of Hg less than the right atrium. Pulmonary artery wedge or "P.C." pressure was obtained in 1 patient and was identical with the right atrial pressure. There was no close correlation between the clinical manifestations of failure and the right atrial pressure. For the patients not in congestive failure, there was an interesting inverse relationship between right atrial pressure and left ventricular output; the higher the right atrial pressure, the lower the systemic output (fig. 8). With patients in failure there was somewhat more scatter, but the trend persisted. In the patient with marked deterioration during the procedure, the highest right atrial pressure was obtained; and the relationship to cardiac output was considerably out of line with the other 9 patients. This relationship, although based on a relatively small number of cases and subject to the serious limitation of calculating outputs, has previously been described by Dow and Dexter in patients with atrial septal defects. Dow and Dexter suggested that peripheral tissue requirements determine left ventricular output, which in turn determines the left atrial pressure, and finally, the right atrial pressure, since these 2 chambers have nearly identical pressures in patients with large atrial defects.*

Mild gradients across the pulmonic valve

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*These patients were catheterized from the femoral vein and the pulmonary artery was not entered.
† Actual left atrial pressure.
‡ Probably not representative. This patient's condition deteriorated markedly during the procedure.

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*This relationship does not contradict the findings of Goodale et al.* in similar cases, where there was a direct relationship between right atrial pressure and right ventricular stroke work.

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*Systolic flow, in addition to pulmonary flow, is difficult to estimate with accuracy in these patients, since obtaining a true mixed venous sample is impossible.
ranging from 4 to 17 mm. were observed in 4 patients. All of these had palpable thrills on physical examination.

Although half of our catheterized patients were in frank congestive failure, the calculated pulmonary vascular resistance was not markedly increased in any, with the average ratio of pulmonary to systemic resistance of 1 to 12, the highest being 1 to 7. This is in contradistinction to Burchell's theory that the main factor causing heart failure is the effort of the right heart to maintain a high flow of blood against increasing pulmonary resistance.9 Similarly, all 13 autopsied cases showed evidence of congestive failure, but only 4 showed severe pulmonary vascular obstruction, and 5 showed only slight changes.

Surgery

Surgical correction of complete transposition of the pulmonary veins was attempted in 3 instances between January 1954 and January 1957 at the Children's Medical Center. The first patient was a 4-year-old girl with all pulmonary veins draining into the left superior vena cava. An anastomosis was created between the common pulmonary vein and the posterior wall of the left atrium, in a horizontal fashion, paralleling the atrioventricular ring. The ascending portion of the left superior vena cava was then ligated and divided. The patient developed pulmonary edema, apparently caused by the inadequate size of the left atrium and ventricle, and death followed within 2 hours. Similar experiences have subsequently been reported by others.15, 26 The second patient, an 18-year-old girl with the same anatomic abnormality was operated upon in the same fashion, except that the left cava was not divided, but constricting stitches were placed to reduce the lumen to approximately one fifth of its previous diameter (fig. 9). A 2-stage operation was planned with complete division of the ascending cava to be attempted later. However, the patient has improved so markedly that the second stage has been postponed; she will be re-evaluated shortly.

The third patient, a boy 1½ years old, with drainage directly into the right atrium, was operated upon in an attempt to anastomose the left pulmonary vein to the left atrium. The patient died of blood loss due to complications arising from the small size of the structures.

Difficulties similar to our own in correcting this anomaly surgically, without the use of open heart surgery, have been experienced by others as well. Mustard and Dolan in 1957 reported 1 survival.27 Burroughs and Kirklin26 reported 9 attempts at correction without survival from the literature and one survival of their own.

Open heart surgery by means of the pump oxygenator was first used by Burroughs and Kirklin26 in 1956 in 2 patients with 1 survival. In both these cases a side-to-side anastomosis between the divided common pulmonary vein trunk and the left atrium was carried out with accompanying closure of the patent foramen ovale. Open heart surgery by means of hypothermia has been used by Mustard and Dolan27 in 9 patients with 4 survivors. In 2 of the 4 survivors, the vena cava was not completely occluded distal to the entrance of the pulmonary veins, much as in our second patient. Ehrenhaft et al.28 recently reported the successful correction of one patient with hypothermia in which the ascending cava was
divided and the atrial septal defect was closed.

Our very favorable experience with open heart surgery by means of the pump oxygenator in the correction of various intracardiac anomalies suggests that this method will without question be the procedure of choice for correction of total transposition of the pulmonary veins in the future. If the common pulmonary vein trunk enters the superior vena cava, direct anastomosis to the left atrium will be possible. If the entrance of the veins is into the coronary sinus or the right atrium proper, then by means of shifting the septum, the anomalous structures may be caused to drain into the left atrium.*

**Summary**

The literature on complete transposition of the pulmonary veins is reviewed. Twenty patients studied by us are reviewed in detail. There was no correlation between the relative size of the interatrial communication and the age of death.

Frequent symptoms were easy fatigability, dyspnea, and recurrent respiratory infections. Examination commonly revealed mild cyanosis, congestive failure, and a characteristic triple or quadruple rhythm. Soft systolic and apical middiastolic murmurs were common; a medium-pitched presystolic murmur was present in one third.

Electrocardiograms demonstrated marked right ventricular hypertrophy with complete reversal of the R/S progression. Radiographic patterns usually could be correlated with the level at which the pulmonary veins emptied. Cardiac catheterization data revealed an interesting inverse relationship between right atrial pressure and systemic index.

Patients with drainage into the portal system formed a unique group. All were males, succumbed early, were more intensely cyanotic, and had marked hepatomegaly. X-rays revealed a normal-sized heart with intense pulmonary engorgement.
Reported experiences with surgical correction are reviewed, and 3 of our operated cases are presented. Using closed techniques, we have 1 survivor. Open heart surgery, with a pump oxygenator, is suggested as the optimal approach for these individuals.

**Incomplete Transposition of the Pulmonary Veins**

**Definition**

In incomplete transposition of the pulmonary veins, the major abnormality is the anomalous drainage of part of the pulmonary return into the right atrium or its tributaries. We have excluded from our series all cases with other major cardiovascular lesions, except a patent foramen ovale or a secundum-type atrial septal defect.

**Incidence**

The exact incidence of incomplete transposition of the pulmonary veins is difficult to estimate, since the anomaly may not be suspected clinically and, if the heart is dissected from the lungs in the initial stages of necropsy, anatomic diagnosis may be impossible. There have been well over 100 cases reported in the literature; and, based on careful studies, the incidence is probably 6 to 10 per 1000 routine autopsies. Of the first 37 patients with secundum-type atrial septal defects operated upon by Gross and Watkins, 9 had incomplete transposition of the pulmonary veins.

**Embryology**

Brown, who first described the development of the pulmonary circulation from part of the presplanchnic plexus, also noted the central position of the opening of the common pulmonary vein in the dorsal wall of the sinus venosus, and described the shift of the pulmonary veins to the left with formation of the atrial septum. Davies and MacConaill proposed that a portion of the right wall of the common pulmonary vein actually initiated the formation of the atrial septum, by invaginating the dorsal wall of the sinus venosus. Anomalies of the pulmonary venous return could logically be attributed either to a failure of some of the pulmonary veins to migrate to the left of the developing atrial septum, or to defects of septation, perhaps originating in abnormal invagination from the common pulmonary vein; it is interesting to note in this respect the frequent coexistence of atrial septal defects with anomalies of the right pulmonary veins.

Anomalies of the left pulmonary veins are rarely associated with atrial septal defects, and are presumably due to failure of those veins to connect with the common pulmonary vein, and subsequent persistence of their earlier connections with the branches of the cardinal veins.

**Anatomic Considerations**

Anomalies of the right pulmonary veins are at least twice as frequent as anomalies of the left. The anomalous right pulmonary veins drain most commonly into the superior vena cava or right atrium, and are frequently associated with an atrial septal defect. Anomalies of the left pulmonary veins are more apt to occur as an isolated abnormality, and usually drain into the left superior vena cava, left innominate vein, or coronary sinus. Only 2 cases have been reported of anomalous drainage of part of both lungs.

The associated changes in the cardiovascular system at necropsy vary with the number of anomalous pulmonary veins and with the presence and size of an atrial septal defect. Although, as Brody originally pointed out, "incomplete drainage is not incompatible with long life" the complacency of most subsequent reviewers toward this anomaly is not justified by the case reports in the literature.

Brody further qualified his prognostication with the statement that, of 25 patients in whom more than 75 per cent of the pulmonary flow returned normally to the left side, all but 2 lived to be adults. Brody reviewed 42 patients without other major cardiac abnormalities, excepting atrial septal defects, and the actual age at death varied greatly; however, at least 14 died before the age of 40 (table 4). It should be borne in mind that 40 years is the average life span of patients
with atrial septal defects, coarctation of the aorta, and patent ductus arteriosus. Such prognostications, based on age at death of autopsied patients, have no clear validity, particularly when it is uncertain in a large number of Brody's cases whether there were associated septal defects, and whether the cause of death could reasonably be ascribed to the cardiovascular anomaly under discussion. Certainly, estimates of the volume of the shunt based on the number of anomalously draining veins, or even cross-sectional areas of the veins, are inaccurate, since they fail to account for differential pressure gradients.

In brief, there is no logical basis for the assumption that incomplete transposition of the pulmonary veins is a benign condition, and the significance of the anomaly must be assessed separately in each individual.

**Physiology**

Incomplete transposition of the pulmonary veins usually produces a small to moderate sized left-to-right shunt when not accompanied by an atrial septal defect or other major cardiac abnormality. Right-to-left shunting on the other hand, causing arterial unsaturation, is impossible when the septa are intact, thus, placing this anomaly in a unique position in the group of left-to-right shunts.

Because of the frequent association of anomalies of pulmonary return with atrial septal defects, detailed reports of cardiac catheterization in the isolated anomaly are infrequent, and often proof is lacking that the atrial septum is intact. Jonsson reported 3 proved cases with intact atrial septum, and the ratios of pulmonary to systemic blood flow were 1.9, 1.8, and 1.6. Snellen and Alber's case demonstrated a ratio of 1.4. Friedlich et al. reported a patient who had a pulmonary blood flow of 8.3 L. per minute per M. which is more than twice the normal systemic output, although no calculation of actual systemic flow was presented. Knut's 2 patients had pulmonary flows 1.8 and 2.2 times as large as systemic. An atrial septal defect could not be ruled out in any of the last 4 patients. However, these figures are predominantly in the range of "large" shunts and, in the presence of marked cardiomegaly, congestive failure, or marked limitation, these patients would be candidates for operative repair. Interestingly, when pressures in the right heart were reported, they were normal in 7 of 8 cases, 8.23, 36-38

As was emphasized in the anatomic discussion, the pulmonary blood flow cannot be accurately estimated by the number of anomalously draining veins alone. Since right atrial pressure normally is 4 to 5 mm. Hg less than the left, the pulmonary artery furnishes a pressure head effectively 5 mm. greater for flow through an anomalous vein than through a normally draining vein of comparable cross-sectional area. If the patient has 4 normally draining veins, the left-to-right shunt through one anomalous vein of comparable size would be 29 per cent of the total pulmonary blood flow, and the total pulmonary blood flow would be 1.4 times greater than systemic. Similarly, if 2 and 3 veins drained anomalously with normal pressures throughout both systems, the total pulmonary blood flow would be respectively,

<table>
<thead>
<tr>
<th>Years</th>
<th>Number of deaths</th>
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<tr>
<td>Less than 1</td>
<td>6</td>
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<td>1-20</td>
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<tr>
<td>21-40</td>
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<td>5</td>
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<tr>
<td>51+</td>
<td>9</td>
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<tr>
<td>&quot;Adults&quot;</td>
<td>14</td>
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*With 4 normal and one transposed pulmonary vein, assuming equal cross sectional areas for all 5 veins, and normal mean pressures in pulmonary artery, left atrium, and right atrium (viz., 15, 7, and 2 mm. Hg, respectively), then:

\[
Q_P = Q_s + Q_{apv}
\]

\[
Q_{apv} = Q_s \times 15-2
\]

\[
4 
\]

\[
15-7
\]

\[
Q_P = 1.4 \times Q_s
\]

\[
Q_{apv} = .29 Q_P
\]

Where \( Q_P \) = total pulmonary flow

\( Q_s \) = systemic flow

\( Q_{apv} \) = flow through anomalous pulmonary vein(s).
2.1 and 3.4. With transposition of 3 pulmonary veins, even if the right and left atrial pressures were identical, the total pulmonary flow would be 2.5 times systemic. These calculated figures are of the same order of magnitude as those available from the literature. It would, therefore, seem unwarranted a priori to predict a normal life span for patients with this condition.

The importance of 1 or 2 transposed pulmonary veins may be greatly magnified in patients with pulmonary disease, particularly where resection or collapse therapy is contemplated. Removal or collapse of a normally draining lobe would reduce the useful pulmonary return proportionally, and in situations in which 1 entire lung is drained abnormally, resection of the other lung would create complete transposition of the pulmonary veins, and survival would depend upon a patent foramen ovale or septal defect. Consequently, any candidate for such procedures should be carefully evaluated, if there is even minimal cardiomegaly, engorged pulmonary vasculature, or shadows suspicious of anomalous pulmonary veins.

Also, it should be borne in mind, that a lobe or lung with transposed veins is not necessarily functionless even though inefficient. It may exchange as much carbon dioxide and oxygen as the normally draining lung if the parenchyma is normal, since the concentrations of these gases are the same in both pulmonary arteries and in both sets of pulmonary veins. The anomalously drained lung may contribute 50 per cent or more to the total body gas exchange, which may be critical in some pulmonary diseases. In such patients, resection should be preceded by differential bronchospirometry.*

*Arvidsson reported the results of bronchospirometry in a patient with drainage of all of the right lung into the inferior vena cava, and with apparently normal pulmonary function. The right lung, absorbed 138 ml. oxygen per minute, the left 212 ml., although the vital capacity of the 2 lungs was 600 and 720 ml. respectively. In this case, the left-to-right shunt would be only 40 per cent of the total pulmonary blood flow.

GUNTEROTH, NADAS, AND GROSS

Presentation of Data

Eleven patients with incomplete transposition of the pulmonary veins have been observed by us in the past 6 years. Ten patients were studied by cardiac catheterization, and 9 of these underwent surgical repair. Four patients were autopsied; 1 of these had not been diagnosed by catheterization. Nine patients were found either at surgery or autopsy to have an associated atrial septal defect of the secundum type, and a tenth patient with a calculated pulmonary blood flow 4 times systemic is presumed to have an atrial defect.

Nine patients had anomalous drainage of 1 or more right pulmonary veins into the right atrium. Two of these patients had additional veins from the right upper and middle lobes draining into the superior vena cava. One patient had only 1 vein from the right upper lobe draining anomalously into the superior vena cava. In our series, there was no instance of 2 of the 3 right lobes draining anomalously, but either all the right lobes or only 1 drained anomalously. Four of 10 patients had single vein transposition, and these were all from the right upper lobe.

The 1 patient with transposition of the left pulmonary veins had only a small, patent foramen ovale; the 2 anomalous veins drained into the innominate vein and coronary sinus respectively. This patient died at 2 months of pneumonia, and had moderate dilatation and hypertrophy of the right heart. Catheterization was not performed.

The patients' ages ranged from 2 to 40 years, with an average of 15 years, which is in marked contrast to the ages of patients with complete transposition of the pulmonary veins. The sex distribution reflected the incidence in patients with atrial septal defects: 3 males and 7 females. (The patient with the isolated anomaly was a male.)

History

There was no family history of heart disease from any patient, and only 1 patient's gestational history was abnormal, with maternal rubella in the first trimester.
Dyspnea, fatigue, and frequent respiratory infections were the commonest complaints, occurring in 7 cases. Cyanosis was not noted by any parent. A history of edema was given in 2 instances. Two of the older patients have documented histories of paroxysmal atrial tachycardia, a frequent finding in older patients with atrial septal defects. No patient was completely asymptomatic.

Physical Findings

Our patients with incomplete transposition of the pulmonary veins presented a less striking physical profile than the individuals with the complete anomaly. Six of the patients were well developed and 5 were below the twenty-fifth percentile for height, although 8 of them were thin. None of them was cyanotic or had clubbed nails. Two patients, 8 and 9 years old respectively, were in congestive failure, with hepatomegaly and engorged veins. Precordial bulge was noted in 5 patients; the point of maximal impulse was at the lower left sternal border in all. Two patients had palpable thrill in the pulmonic area; in the first, a systolic thrill was associated with a gradient of 32 mm. Hg across the pulmonic valve; in the second, a 20-year-old patient with pulmonary hypertension, an early diastolic thrill was demonstrated.

On auscultation, the first sound was increased in intensity in 8 patients, and normal in 3. The pulmonic sound was accentuated in 6, normal in 5, and audibly split in all 11. A third sound was detected in 4 individuals and a fourth, or atrial sound was present in only 2. Systolic murmurs at the upper left sternal border were present in all 10 patients with atrial septal defects, but not in the patient with the isolated venous anomaly. The systolic murmur was soft and blowing in character except in 4 instances. These 4 patients had a rough, systolic murmur that transmitted. During cardiac catheterization, a moderate pressure gradient across the pulmonic valve was recorded. Mid-diastolic murmurs at the apex or lower left sternal border, characteristic of large left-to-right shunts were found in 8 patients. Three individuals had early, high-pitched diastolic murmurs at the pulmonic area; these patients were older (20, 22, and 40 years respectively), and 1 had severe pulmonary hypertension. Four patients had presystolic murmurs similar to those present in complete transposition of the pulmonary veins. This murmur, diamond-shaped, of medium to high frequency and synchronous with atrial systole, was accompanied by a fourth sound in 2 patients.

Electrocardiogram

The electrocardiogram of patients with
incomplete transposition of the pulmonary veins reflects the increased volume of the right ventricle although these changes are less marked than in the patients with complete transposition of the pulmonary veins. Right axis deviation was present in 9 patients, normal electric axis in 2. All 11 patients had right ventricular hypertrophy, but only 5 patients, including the 1 with the isolated anomaly, had marked right ventricular hypertrophy, as suggested by complete reversal of the adult R/S progression, or large right ventricular potentials, or both. Incomplete right bundle-branch block was present in every instance; right ventricular intrinsicoid deflection was consistently prolonged, with an average of .06 second. Figure 10 shows a representative electrocardiogram.

The pattern of P pulmonale, so frequent in complete transposition of the pulmonary veins, was present in only 3 patients with the incomplete anomaly. Similarly, T-wave changes were found in only 4 patients, 2 of whom were on digitalis.

X-Rays

The radiographic profile of our patients with incomplete transposition of the pulmonary veins was completely nonspecific, although quite similar to that in patients with atrial septal defects. The specific pattern reported by Steinberg et al.59 observed in patients with 1 or 2 right pulmonary veins draining into the inferior vena cava was not observed in our cases; in fact, there was no radiologic suggestion of the existence of anomalous pulmonary veins in any of our patients.

Cardiomegaly was present in all of our patients: it was slight in 4, moderate in 4, and marked in 3 patients (figs. 11 and 12). In every instance, the ventricular configuration was characteristically right-sided. Atrial enlargement was present in 9 patients, but not in the patient with intact atrial septum. The main pulmonary artery was abnormally prominent in all patients, with slight to moderate enlargement in 8 and marked enlargement in 3. Pulmonary vascular engorgement was present to a moderate degree in 8 of the 10 patients with atrial septal defects; 2 patients had marked engorgement. The patient with an intact septum had mild engorgement. "Hilar dance" was observed in 9 patients; it was not detected in the 2-month-old patient with an intact septum, nor in the youngest patient with an associated atrial septal defect. The aorta was judged to be smaller than average in 5 patients.

Angiocardiograms were not performed in any of our patients with incomplete transposition of the pulmonary veins.

It is interesting that the patient with an intact septum had moderate cardiomegaly,
and marked right ventricular hypertrophy on electrocardiogram with only 2 pulmonary veins draining anomalously. His death at 2 months was secondary to pneumonia, but these changes nevertheless suggest that the isolated transposition of even 2 pulmonary veins may be a moderately serious burden to the heart.

**Cardiac Catheterization**

Ten patients with incomplete transposition of the pulmonary veins were studied by cardiac catheterization because of symptoms and signs suggesting a large atrial septal defect. The 1 patient with an intact septum was not catheterized. Eight of the 10 patients were catheterized in our laboratory; 2 adults were catheterized elsewhere (table 5), although the remainder of their clinical examination and operative procedures were performed by us.

Arterial oxygen saturation in these patients showed no overlapping with those of the total anomaly; 95 per cent was the lowest saturation obtained and the average was 97 per cent.

The rise in oxygen saturation at the right atrial level varied from 15 to 31 per cent, with an average of 20 per cent. This also reflects the left-to-right shunt through the atrial septal defect, of course.

Cardiac output varied from 2.2 to 5.7 L. per minute per M.², with an average of 3.4, within normal limits for systemic index. Pulmonary flow ranged from 6.2 to 19 L. per minute per M.², with an average of 12. There was an average left-to-right shunt of 8.5 L. and no definite right-to-left shunt in any patient. These calculated flows are approximations, at best, because a true mixed venous sample is impossible to obtain.

Right atrial pressures in the incomplete anomaly varied less than in complete transposition of the pulmonary veins, ranging from 2 to 10 mm. Hg, with an average of 5. The interesting inverse relationship between left ventricular output and the right atrial pressure that was present in the complete anomaly, was also present in patients with incomplete transposition of the pulmonary veins (fig. 13). The pressure relationship between the right atrium and left atrium (or wedge pressure) was determined in 7 patients, and similar to the findings in large atrial septal defects, the gradient between left and right atria averaged only 2 mm. Hg.

Four patients had gradients across the pulmonic valve of 10, 18, 28, and 32 mm. Hg. Although none of these patients was autopsied, it seems unlikely that anatomically detectable stenosis was present, since they had flows through the valve of 8, 9, 15, 13, and 19 L. per minute, respectively. It is probable
that, at normal flows, the gradient would be insignificant.

Pulmonary vascular resistance was abnormally increased in only 1 patient, a 20-year-old girl, whose calculated resistance was 328 dynes-second-cm\(^{-5}\), and a ratio of pulmonary to systemic resistance of 1 to 5. This patient died shortly after surgical repair, and was found to have moderately severe medial hypertrophy and intimal hyperplasia of the pulmonary arterioles. The other patients, including a 40-year-old woman, had essentially normal resistances. The average pulmonary vascular resistance in the entire group of catheterized patients is considerably less than normal, with a ratio of 1 to 25, pulmonary to systemic, consistent with the findings of patients with atrial septal defects without pulmonary vein anomalies.\(^{34}\)

Although the possibility of incomplete transposition of pulmonary veins was considered prior to cardiac catheterization, it was not specifically diagnosed in any case. At the time of catheterization, however, an anomalous pulmonary vein was entered, from the superior vena cava in 2 cases, and the right atrium in 4. In the remaining 4 cases, the pulmonary vein anomaly was discovered at the time of operation. Even in those cases in which a pulmonary vein was apparently catheterized from the right atrium, the diagnosis of transposition of a pulmonary vein should be made with reservations, since it is quite possible to slip across into the left atrium through a high, posterior septal defect, into a normally draining vein. Avoiding this error requires careful 3-dimensional defect on position and frequent sampling. Figure 14 illustrates the position of the cardiac catheter in anomalously draining veins, subsequently confirmed at operation.

Although, in some cases, dye-dilution curves may help in the diagnosis of co-existence of incomplete transposition of the pulmonary veins and an atrial septal defect, this technic is as limited as that of passing the catheter into the anomalous pulmonary veins. Due to the proximity of the normally connecting right pulmonary veins to the atrial septal defect, perferential shunting of venous return from the right lung may simulate the curves of anomalous venous connection from the right lung.\(^{40}\) None of our patients had dye-dilution studies.

In view of our present surgical approach to atrial septal defects, the presence of anomalous pulmonary veins is of no major consequence either in selecting candidates for surgery, or in selecting the external approach. In cases of the isolated anomaly, with an intact atrial septum, more exact anatomic information would be crucial, in which case, selective angiocardiology would be of great assistance.

**Surgery**

Seven patients with incomplete transposition of the pulmonary veins have been operated upon at the Children's Medical Center,\(^{31}\) by a method similar to that initially described by Neptune, Bailey, and Goldberg,\(^{41}\) in which pulmonary veins connecting to the right atrium, in association with a posterior defect, are rechanneled by atrioseptopexy. This technic takes advantage of the fact that most, if not all, cases of incomplete transposition of the pulmonary veins to the right atrium are associated with posterior defects, which are
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Fig. 14. Radiogram, illustrating the cardiac catheter in pulmonary veins entered from the right superior vena cava and right atrium.

Ideally suited for closure by invagination of the right atrial wall, thereby enlarging the atrium to include the entry of the anomalously draining veins, but excluding the systemic return from the superior and inferior venae cavae and coronary sinus.

An eighth patient, not included in the clinical part of this discussion, was operated upon recently by means of open heart surgery through a pump oxygenator. This subject had a posterior defect in the atrial septum, 1.5 cm. wide and 2 cm. long. Two right pulmonary veins came into the right atrium. Repair was accomplished in such a way as to bring the anterior edge of the septal defect over against the back wall of the atrium, to the right of the anomalous veins. In this way the defect was closed and all the pulmonary veins were directed into the left atrium.

Various surgical techniques have been proposed to correct the drainage of veins into the right superior vena cava. Since the external approach for atrioseptopexy is through the right chest, actual transplantation of the veins into the left atrium is difficult, if not impossible, without open heart surgery. Resection of a lobe or entire lung should be regarded as a last resort, and should be preceded by careful studies of cardiac and pulmonary functions, including differential bronchospirometry. Two of our patients with atrial septal defects and partial transposition of the pulmonary veins had this anatomic situation. Only the atrial defect was repaired in these children. Ehrenhaft et al. reported 4 patients operated upon with hypothermia, with closure of atrial defects. They transplanted the superior vena cava, and in 2 of these patients, they utilized an arterial graft. Ligation of the anomalous pulmonary veins seems less than completely satisfactory. Ligation of all the veins from 1 lung is frequently fatal. Tying off a single vein, although well tolerated in 1 of our patients, may cause congestion of the involved lobe. Also, the anastomoses connect with the bronchial veins, or with the parietal veins, both of which ultimately drain into the right atrium, since there are no important translobar anastomoses with normally draining pulmonary veins.

Moderate, or even severe pulmonary hypertension in a patient with an intact atrial septum, and incomplete transposition, would not be a contraindication to surgery, since repair would diminish the right ventricular work and increase the left-sided return. This is in contradistinction to patients with atrial septal defect and severe pulmonary vascular obstruction, in whom a right-to-left shunt through the septal defect may allow more adequate systemic output as this disease progresses.

Of the 10 patients with atrial septal defect and transposed pulmonary veins to the present time, 8 had successful closures of the septal defect and correction of all the anomalous
drainage. In the 2 with superior vena cava drainage, only the atrial septal defect was repaired. There were 2 deaths: 1 secondary to infarction of the right atrial wall including the pacemaker, and 1 in whom a plastic button was used in the early stages of developing operative approaches for repair of atrial septal defects. The results obtained in the surviving patients are considered excellent.

**Summary**

The seriousness of incomplete transposition of the pulmonary veins has probably been underestimated in the past.

Data on 11 patients is presented, including 10 with associated atrial septal defects. Common symptoms were dyspnea, fatigability, and frequent respiratory infections. Examination revealed no cyanosis, widely split second sound, soft systolic murmur and mid-diastolic murmur at the apex. One third demonstrated a presystolic murmur of medium pitch. The electrocardiographic and radiographic features were characteristic of atrial septal defects. There were no specific vascular shadows in any of our patients. Catheterization data were also characteristic of atrial septal defects. There was an inverse relationship between the right atrial pressure and systemic index. Anomalous pulmonary veins were entered during the right heart catheterization in 6 of the 10 patients catheterized.

Seven patients have had atrioseptopexy to close the septal defect and to direct the blood from the anomalously draining veins into the newly enlarged left atrium. An eighth patient had repair and shift of the atrial septum by means of open heart surgery through a pump oxygenator. This approach will probably prove optimal for future operations.

**Acknowledgment**

The cardiac catheterizations reported in this paper were performed by Drs. Walter T. Goodale and Abraham M. Rudolph, in the Cardiovascular Laboratory of the Children’s Medical Center, Boston, with the exception of 2 patients with incomplete transposition of the pulmonary veins catheterized by Dr. D. S. Lukas, at the New York Hospital, and 1 patient at the Brooke Army Hospital, respectively. Permission to include these data is gratefully acknowledged. We also wish to thank Dr. M. W. Wittenborg, who reviewed the radiographic findings, and Dr. J. B. Craig, for reviewing the anatomic findings.

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