Anomalous Drainage of Entire Pulmonary Venous System into Left Innominate Vein

Clinical and Surgical Considerations

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Total anomalous drainage of pulmonary venous blood produces a severe circulatory disturbance. In the most common type of this category of lesions, the entire venous return from the lungs enters a common venous channel ascending in the left superior mediastinum where it connects with the innominate vein. Seven cases of this type are described. Successful total correction was possible in 4 of these patients in whom a special technic was used, which incorporated the pump oxygenator for temporary cardiopulmonary bypass. This technic provides optimum conditions for complete correction of this complicated anomaly.

Recent increased interest in open heart surgery has revealed that anomalies of pulmonary venous drainage are much more common than previously realized. Interestingly enough, Winslow first described anomalous connection of the pulmonary veins of 1 lung more than 200 years ago (1739) and in 1798 Wilson recorded a case of total anomalous drainage of pulmonary veins into the systemic venous circulation. At present it is recognized that anomalous drainage of a portion of one or both lungs is relatively common and frequently accompanies atrial septal defect. Hemodynamic effects of the partial anomaly are generally well tolerated and, depending upon the extent of the anomaly, may be compatible with an asymptomatic state.

Total anomalous drainage of pulmonary veins is less frequently encountered and usually is associated with greater morbidity and poor prognosis. For example, in a review of the reported cases Healey discovered an average age at death of 1.8 years in total anomalous pulmonary venous drainage. This fact emphasizes the need for early recognition of the lesion in infants, control of cardiac decompensation, and surgical correction of the lesion. The purpose of this paper is to present certain observations based upon clinical experience with 7 cases of total anomalous pulmonary venous drainage into the left innominate vein and the details of successful surgical management in the last 4 consecutive cases.

Complete anomalous pulmonary venous drainage occurs in several anatomic forms. Common to all forms, however, is the fact that the pulmonary veins from both lungs usually converge to form a chamber or confluence posterior to the left atrium. Most frequently a single anomalous vein emerges from this chamber to join the systemic venous circulation. In a classification based upon the level of emptying of this anomalous vein into the systemic venous circulation Darling and co-workers have recognized 4 types of total anomalous pulmonary venous connections. 1. Supracardiac level: In this type the anomalous connection is usually made with the left innominate vein through a remnant of the left superior vena cava extending superiorly from the common pulmonary venous trunk. This is the anomaly with which this paper is concerned (fig. 1). Infrequently direct connection to the right superior vena cava may occur. 2. Cardiac level: Total drainage occurs in this type with connection into the right atrial cavity directly or into the coronary sinus, which in turn enters the atrium at the usual site but with a greatly enlarged
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ostium. 3. Infra cardiac level: In this type, drainage from the posterior chamber occurs into the systemic venous circulation through an anomalous vein extending below the diaphragm. Pulmonary venous blood returns to the right atrium via the inferior caval system. 4. Combined type: In this type, connection may be made independently at 2 or more levels with total drainage into the right atrium by multiple channels. Occasionally extensive abnormalities in systemic venous drainage are also present.

Of the various types of total anomalous pulmonary venous drainage the most common appears to be supra cardiac (type 1) with connection via a remnant of the left superior vena cava (fig. 1). Keith and associates in a survey of reported cases found 43 per cent to be of this type.

Embryologically the lungs are derived from the foregut with which they share a common blood supply. In early stages the pulmonary veins are derived from the splanchnic plexus and have multiple communications with 2 systems, the cardinal system of veins and the umbilicovitelline system. From the cardinal system the superior vena cava, innominate veins, and coronary sinus are ultimately derived. In the final stages of development the umbilicovitelline system is represented principally by the portal venous system. In this early stage the primordia of the lungs have no direct connection with the heart. Subsequently a direct connection with the heart occurs as a result of the union of these primary lung veins with an outgrowth from the dorsal wall of the sinoatrial region known as the "common pulmonary vein." After the lungs acquire a route of drainage directly into the heart, the connections between the pulmonary portion of the splanchnic plexus and the cardinal and umbilicovitelline veins are lost. Coincident with the interruption of the main anastomoses of the pulmonary vessels with the umbilicovitelline and cardinal venous systems, the common pulmonary vein and its main tributaries become incorporated into the dorsal wall of the left atrium. With completion of this process the principal venous connection of the lungs is directly with the left atrium and no longer with the systemic and abdominal visceral veins.

According to Edwards the underlying cause in most examples of anomalous pulmonary venous connection is either (1) failure of connection of the atrial portion of the heart with the pulmonary portion of the splanchnic plexus or (2) secondary obliteration of normally developed communications between the atrial portion of the heart and the pulmonary portion of the splanchnic plexus. In either event that portion of the pulmonary tissue that fails to make direct connection with the heart has no route for drainage other than the primitive connection between the splanchnic plexus and the cardinal or umbilicovitelline system of veins. In the most common type of total anomalous pulmonary venous return this communication is through the left cardinal system, which ultimately gives rise to the left innominate vein and the coronary sinus. If a particular portion of the left anterior cardinal vein is not obliterated, it persists as a left superior vena cava and connects the left innominate vein to the coronary sinus.

The left superior vena cava is anatomically in the same position as the anomalous vein.
TABLE 1.—Patients with Total Anomalous Pulmonary Venous Drainage into Left Innominate Vein

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age (yrs.)</th>
<th>Sex</th>
<th>Date of operation</th>
<th>Surgical procedure</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1/12</td>
<td>F</td>
<td>9-12-55</td>
<td>Anastomosis of vertical anomalous vein to left atrial appendage. Partial ligation of vertical vein</td>
<td>Died 4 hours later</td>
</tr>
<tr>
<td>2</td>
<td>2/12</td>
<td>M</td>
<td>2-2-56</td>
<td>Anastomosis of vertical anomalous vein to left atrial appendage. Partial ligation of vertical vein</td>
<td>Died 2 hours later</td>
</tr>
<tr>
<td>3</td>
<td>40</td>
<td>M</td>
<td>2-1-57</td>
<td>Exploratory thoracotomy</td>
<td>Died during exploration</td>
</tr>
<tr>
<td>4</td>
<td>6/12</td>
<td>M</td>
<td>7-31-57</td>
<td>Complete correction during cardiopulmonary bypass</td>
<td>Excellent</td>
</tr>
<tr>
<td>5</td>
<td>4/12</td>
<td>M</td>
<td>1-30-58</td>
<td>Complete correction during cardiopulmonary bypass</td>
<td>Excellent</td>
</tr>
<tr>
<td>6</td>
<td>7</td>
<td>M</td>
<td>2-18-58</td>
<td>Complete correction during cardiopulmonary bypass</td>
<td>Excellent</td>
</tr>
<tr>
<td>7</td>
<td>8</td>
<td>M</td>
<td>5-29-58</td>
<td>Complete correction during cardiopulmonary bypass</td>
<td>Excellent</td>
</tr>
</tbody>
</table>

connecting the common pulmonary vein to the left innominate vein in cases of supracardiac connection of total anomalous pulmonary venous drainage. This has given rise to the designation of this anomalous vein as a persistent left superior vena cava. According to Edwards and Helmholtz this is an incorrect term, since this anomalous vein has no connection with the coronary sinus and it may have a different origin than does the true left superior vena cava. They have suggested the term "vertical anomalous pulmonary vein" to differentiate the two. In some instances, however, this anomaly may be associated with a true left superior vena cava connecting with the coronary sinus (table 1, case 3).

As in other forms of total anomalous drainage a communication exists between the atria (fig. 1). In most cases this communication is either a midatrial septal defect or a patent foramen ovale. The size of the atrial communication may be a factor determining survival although this remains an unsettled question. Indeed, a patent ductus arteriosus is frequently present although it is usually of minor physiologic importance. It appears that total anomalous pulmonary venous return without complicating major cardiac defects is twice as common as when multiple cardiovascular anomalies are also present.

The anatomic configuration of the heart in 13 autopsied cases of total anomalous pulmonary venous return was assessed by Keith and associates. The right atrium was 5 to 10 times as large as the left atrial cavity and the right ventricular cavity was 3 to 5 times as large as the left. The circumference of the waist of the left atrial appendage was usually less than that of the anomalous pulmonary vessel, and orifice of the mitral valve was invariably smaller than that of the tricuspid. Keith stated, "the left auricle is underdeveloped and thus cannot be expected to carry the total blood flow adequately."

From a theoretical standpoint he concluded that this anomaly was not anatomically correctible. On the basis of our own experience in which complete correction of this anomaly was accomplished, this conclusion was apparently incorrect. Indeed, from a theoretical consideration this could have been predicted, since the entire cardiac output into the systemic arterial system in these patients depends upon the function of the left ventricle. Thus, the discrepancy in size of the right and left sides of the heart is a reflection only of the difference in volume of
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Fig. 2 Left. Roentgenogram of chest in child with typical figure-eight mediastinum considered diagnostic of total anomalous pulmonary venous return into left innominate vein (case 7). Dotted lines indicate anticipated location of veins.

Fig. 3 Right. Roentgenogram of chest in infant with total anomalous venous drainage into left innominate vein (case 4). Film reveals cardiac enlargement and pulmonary congestion but is not diagnostic of the underlying anomaly.

blood being handled on the 2 sides of the heart, and does not represent a true underdevelopment of the left side.

In our experience 2 clinical patterns have been evident in patients with total anomalous pulmonary venous drainage into the innominate vein (table 1). In infants the principal findings are a result of congestive heart failure, whereas in older children the symptoms and findings consist of cyanosis and exertional dyspnea. The magnitude of the right-to-left shunt at the atrial level could explain the 2 patterns. In infants the right-to-left shunt is usually small and the pulmonary arterial flow greatly increased. Thus, cyanosis was minimal. Cardiomegaly, pulmonary edema, hepatomegaly, and distended peripheral veins were manifestations of the cardiac failure. Repeated respiratory infections were common. Emaciation and retardation of growth and development were usually evident.

In contrast to infants, children with this anomaly had dyspnea and increasing fatigue with slight exertion. Syncopal episodes and squatting occurred occasionally. Growth was usually retarded but not to the striking degree noted in infants. Cyanosis and clubbing of the digits were noted. Cardiac size was moderately increased and right ventricular hypertrophy was present. A systolic murmur at the left sternal border was usually audible.

Roentgenograms of the chest in total anomalous pulmonary venous return into the left innominate vein in children reveal an almost pathognomonic cardiac silhouette. These findings were clearly described by Snellen and Albers,11 who called attention to the figure-of-8 configuration of the mediastinum. The upper half of the 8 is formed by the ascending or vertical anomalous pulmonary vein on the left and the prominence of the distended right superior vena cava (fig. 2). The superior mediastinal vessels pulsate in an "aev" or venous pattern while the pulmonary arterial
pulmonary veins. A hilar dance is usually demonstrable in the pulmonary vessels on fluoroscopy.

Although this roentgenographic appearance is characteristic of the anomaly in children several years of age and older, in infants this pattern is not recognized. In infants cardiac enlargement involving the right ventricle and engorgement of the pulmonary vessels is demonstrated, indicating the presence of a large left-to-right shunt (fig. 3). The superior mediastinal shadow may be widened but the configuration is not at all characteristic of the anomaly. Gott and associates described a box-like appearance of the heart with an almost horizontal take-off of the lower border of the heart below the aortic arch. In this age group angiocardiography is useful in delineating the pulmonary venous collecting system (fig. 4). At cardiac catheterization the catheter may be passed into the anomalous venous connection and both lungs may be entered without the catheter entering the heart.

In one of our patients with total anomalous venous drainage (table 1, case 3), a portion of the blood entered the left innominate vein and the remainder entered the coronary sinus (fig. 5A). Thus, the typical figure-of-8 appearance was not present, since the vertical pulmonary vein did not carry the entire pulmonary venous return. Cardiac enlargement was extreme, and the anomalous superior mediastinal pulmonary vein was evident (fig. 5B). Venous angiocardiography demonstrated the pulmonary venous connection to the left.

![Roentgenograms](http://circ.ahajournals.org/content/490/4/COOLEY.COLLINS)

**Fig. 4.** Roentgenograms made at cardiac catheterization (a and b) and during angiocardiography (c and d) in infant with total anomalous pulmonary venous drainage into innominate vein (case 2). The cardiac catheter in (a) enters the superior vena cava, left innominate vein, and vertical pulmonary vein and in (b) enters the right pulmonary vein. Angiocardiography (c and d) outlines the superior mediastinal course of drainage of pulmonary venous blood.
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superior vena cava (fig. 5C). The unusual roentgenographic findings in this 40-year-old patient may be explained by the type of total anomalous pulmonary venous return that was present. Although pulmonary venous blood entered the left innominate vein through a vessel anatomically similar to that found in the usual type 1 supracardiac level of drainage (Darling), this case may have been an example of type 2 cardiac level. Thus, it is possible that the patient had predominant drainage to the coronary sinus with a typical persistent left superior vena cava. Nevertheless, the patient is included in this report because of the similarity to the other cases.

Electrocardiograms showed right axis deviation, right ventricular hypertrophy, and an impressive degree of right atrial enlargement.

Cardiac catheterization reveals an increased oxygen saturation of right atrial blood, which is equal to or higher than that of the systemic arterial blood. The demonstration of oxygen saturation in the right side of the heart equal to the saturation in a systemic artery is considered almost diagnostic of total anomalous pulmonary venous drainage (fig. 6). Exploration of the superior caval system with the catheter may reveal the connection of the superior caval system of the pulmonary venous trunk with the left innominate vein (fig. 4). Passage of the catheter through the anomalous channel into both lungs is occasionally possible. The pressure within the right side of the heart and the pulmonary artery is elevated, frequently to a striking degree. Difficulty in entering the left atrium from the superior vena cava was characteristic, since the defects were usually of the foramen ovale type with valvelike mechanism favoring entry from the inferior cava.

Fig. 5 A. Diagram of pulmonary venous system in adult cyanotic patient showing drainage from pulmonary veins into coronary sinus and left innominate vein (case 3). RPV, LPV, right and left pulmonary veins; CSO, coronary sinus ostium; IV, innominate vein; RSVC, right superior vena cava; X, possible persistent left superior vena cava.

Fig. 5 B. Roentgenogram of chest in same patient 5A showing extreme cardiac enlargement and pulmonary vascular engorgement. C. Angiocardiogram showing anomalous vein entering left innominate vein filled by reflux of contrast material into anomalous vessel.
Dye-dilution studies reveal a shorter appearance time from the atrium than from the right ventricle or pulmonary artery. Appearance time from the inferior vena cava is shorter than from the superior cava, demonstrating the preferential shunting of inferior caval blood through the foramen ovale.

**Surgical Treatment**

Prior to the development of techniques of cardiopulmonary bypass our experience, like that of others, with attempts at complete correction of total anomalous pulmonary venous drainage into the left innominate vein was unsatisfactory. In those early surgical attempts an effort was made to perform side-to-side anastomosis between the vertical anomalous pulmonary vein and the small left atrial appendage. Not only was this technically difficult in small infants, but it was also evident that even slight anterior displacement of the dilated heart caused cardiac arrest—possibly due to traction on the atrial septum and constriction of the patent foramen ovale. It was thus evident that a successful technique of repair in infants would be possible only if such manipulation was eliminated. Moreover, in most of these cases the size of the venoatrial anastomosis was inadequate, and complete occlusion of the vertical anomalous pulmonary vein was not tolerated. Partial ligation of this structure was usually the final resort. In these cases no attempt was made to close the atrial septal defect. Senning recently reported successful, complete repair of a total anomalous drainage by a right-sided approach for the atriovenous anastomosis in a 21-year-old patient in whom closure of the atrial defect was accomplished by a closed technique. Ehrenhaft has apparently accomplished a satisfactory repair using hypothermia.

Burroughs and Kirklin, using a pump oxygenator in a 6-month-old infant, attempted side-to-side anastomosis between the common venous trunk and the left atrium from the left side of the mediastinum. A right atriotomy was used to close the atrial septal defect. The patient died 8 hours later in pulmonary edema. On July 31, 1957, we employed for the first time a method of complete correction of this anomaly which fulfills certain important criteria for successful repair. These criteria include (1) use of a pump oxygenator to provide cardiopulmonary function during the cardiac manipulations, (2) creation of the largest possible anastomosis between the common venous trunk and left atrium, (3) closure of the septal defect and enlargement of the left atrial cavity by ventral displacement of the septum, and (4) complete closure of the vertical anomalous pulmonary vein emptying into the left innominate vein.

The surgical technique utilized at present in cases of total anomalous pulmonary venous return into the superior vena cava consists first of exposure of the heart and mediastinal

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**Fig. 6.** Diagram showing oxygen saturations and pressures obtained during cardiac catheterization in child (case 7) under general anesthesia and breathing 100 per cent oxygen. Oxygen saturation of blood in right heart identical with saturation in peripheral artery is almost diagnostic of this anomaly.
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FIG. 7 Left. Drawing showing technic of surgical repair of total anomalous venous drainage from the right side using temporary cardiopulmonary bypass. In (a) the right atrium is opened widely. The atrial septum is detached posteriorly and retracted with forceps. A ample anastomosis is created between the anomalous vein and the left atrium. The atrial septum is reattached posteriorly and moved laterally to enlarge the left atrial cavity. In (b) the atriotomy is repaired. This technic avoids unnecessary cardiac displacement during the repair.

FIG. 8 Right. Drawing showing completed repair. The vertical anomalous pulmonary vein is ligated and the left atrial cavity has been enlarged to accommodate the venoatrial anastomosis.

structures through a transverse bilateral thoracotomy transecting the sternum. After incising the pericardium and before any cardiac manipulation, preparations are made for cardiopulmonary bypass. Cannulation is performed of the superior and inferior venae cavae for venous outflow and the femoral artery for return of oxygenated blood by the pump. The vertical anomalous pulmonary vein is temporarily occluded after complete cardiopulmonary bypass is commenced. Actual repair of the anomaly is started by opening widely the right atrium (fig. 7). The atrial communication is identified and the atrial septum is detached from the atrial wall laterally, so that the opening is enlarged. With the left atrium widely opened an ample incision is made parallel to the direction of the common pulmonary vein behind the heart. In order to obtain the largest possible anastomosis the incision is usually continued into the right atrium. Finally a transverse incision is made in the venous trunk and an anastomosis is created between the trunk and the posterior atriotomy (fig. 7a). Upon completion of the anastomosis the atrial septum is transposed ventrally and sutured in such manner that the left atrial cavity is increased in size. The right atriotomy is then closed (fig. 7b). The final step consists of intrapericardial ligation of the vertical anomalous vein, thus restoring the pulmonary circulation to an essentially normal anatomic state (fig. 8).

CLINICAL EXPERIENCE

A total of 7 patients with total anomalous pulmonary venous drainage into the innominate vein have been treated surgically during the past 3 years (table 1). The first 2 were critically ill infants who were operated upon without a pump oxygenator. In both an anastomosis was created, side to side, between
the pulmonary venous trunk and left atrium or appendage. Partial occlusion of the vertical pulmonary vein by partial ligature was used. Operation in both was poorly tolerated and the patients died in pulmonary edema several hours later. Autopsy revealed a relatively small unsatisfactory anastomosis to an underdeveloped left atrium. Pulmonary edema was apparently produced by the obstruction to venous drainage from the lungs. In neither of these patients was correction of the associated atrial septal defect attempted.

The third patient was intensely cyanotic and in intractable cardiac failure. Extreme cardiomegaly, hepatomegaly, and pulmonary edema were present in this patient whose age of 40 years far exceeded the average life expectancy in this anomaly. Preparations had been made for use of the pump oxygenator, but the patient died during the preliminary exploration and before cannulations could be made. This experience convinced us that preliminary exploration and intracardiac palpation through the atrial appendage should not be attempted in these critically ill patients. Accordingly in all subsequent patients immediately upon opening the chest the cannulations of the superior and inferior venae cavae were done for venous outflow to the pump oxygenator. The common femoral artery was intubated for return of oxygenated blood from the pump oxygenator. Final assessment of the anatomic configuration or arrangement of the anomalies was then accomplished with safety, since cardiopulmonary bypass could be instituted if signs of cardiac distress appeared.

The first successful case of complete correction of total anomalous pulmonary venous drainage has been reported in detail elsewhere. This patient 1 year later is developing satisfactorily and shows progressive improvement. Subsequently 3 more patients have been operated upon with complete correction of the anomaly and all 3 are clinically cured.

Summary

Total anomalous drainage of pulmonary veins is a complicated and serious congenital cardiac anomaly. Usually prognosis is poor, and until recently complete surgical correction was not possible. Drainage of the entire pulmonary venous return into the left innominate vein is the most common type of such anomaly.

Clinical features in 7 patients with this lesion are described. A new method of surgical correction during cardiopulmonary bypass is outlined which provides a complete repair of the defect. In all 4 patients operated upon by this technic a successful result was obtained.

Addendum

Since this paper was submitted 3 more patients with total anomalous pulmonary venous drainage into the left innominate vein have been operated upon successfully by the technic described. Ages of the patients were 6 years, 6 years, and 3 months. In one of the 6-year-old patients an associated pure valvular pulmonic stenosis was treated by open valvotomy at the time of cardiopulmonary bypass.

Summario in Interlingua

Drainage anormal del integre systema pulmonary-venose es un complexe e serie anomalia congenite del corde. Usualmente le prognose es pauco promittente, e usque recentemente un complete correction chirurgic non esseva possibile. Drainage a in le sinistre vena innominate es le typo le plus commun de iste anomalia.

Es describite le stato clinic de 7 patientes con iste lesion. Es delineate un novo metodo de correction chirurgic, effectuate con circuito cardiopulmonar e resultante in le complete reparo del defecto. Le technica esseva usate in 4 patientes. Le successo esseva bon in omnes.

References

2. Wilson: Cited by Burroughs.41
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