Anomalous Pulmonary Venous Return

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Many detailed studies concerning isolated aspects of anomalous pulmonary venous return are available. Surveys encompassing the entire spectrum of such anomalies are, however, generally lacking. For purposes of this presentation, 113 selected instances of anomalous pulmonary venous return encountered at Brooke General Hospital, Walter Reed General Hospital, and the Registry of Cardiovascular Pathology of the Armed Forces Institute of Pathology have been reviewed and correlated with related material from the literature. To obtain completeness, tempered with brevity, the better known anomalies, while mentioned, are discussed only briefly, whereas the less well described transitional forms are discussed in detail. Highly schematic diagrams are used to portray some 27 variations in this theme. It is hoped the wide anatomic spectrum shown will promote understanding and interest.

Figure 1A illustrates the most frequently described variety of anomalous pulmonary venous return in which all or a part of the veins from the right lung drain directly into the right atrium. In two of 26 such instances the septum interestingly was intact. It should be emphasized that a normal connection does not always mean normal drainage. We, like others, have seen examples in which indicator-dilution curves indicated normal drainage and at surgery the vein connected anomalously.1, 2

Figure 1B, a situation encountered twice, we think important as it represents a paradox not easily explainable embryologically. Entry of the right pulmonary veins into the left atrium and entry of the left pulmonary veins into the right atrium seemingly defy ex-
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Figure 1

Schematic representation of types of anomalous venous return.

associated with high, sinus venosus-type atrial septal defects.\textsuperscript{4, 5} Such high defects were observed in 17 instances, typical secundum defects occurred twice, and in one instance the atrial septum was intact.

All pulmonary veins entering the right superior vena cava to date have done so at or below the azygos entry. This finding is interesting in view of the frequency of anomalous pulmonary venous return on the left finding its way to the left jugular-subclavian juncture. Higher, more peripheral connections suspected on catheterization, but not proved at surgery, are explained by the perfused unexercised right arm giving unusually high oxygen-saturation values. Entry at or below the azygos level enhances surgical repair.

Complete anomalous pulmonary venous return to the right superior vena cava, found seven times, is depicted in figure 1G. In this small group, three anatomic features were apparent. First, the lowermost end of the cava was consistently dilated. Secondly, the anomalous orifice was usually quite large, placed directly posterior, and single. Thirdly, the accompanying septal defect was highly placed. These factors again favor surgical repair.

Anomalous pulmonary venous return through the azygos vein may exist in part or
in whole. Figure 1H depicts partial return of all the right pulmonary veins to the right superior vena cava through the azygos. One might also suspect an absence of the suprarenal portion of the inferior vena cava to accompany this defect.6 This was not true in this case.

Total anomalous pulmonary venous return to the superior vena cava through the azygos and persistent left innominate veins was seen but once and is depicted in figure II. The term persistent left innominate vein is preferable to persistent left superior vena cava when this remnant of the left anterior cardinal vein fails to maintain a connection with the coronary sinus or left atrium.

With these four descriptions completed, the three remaining variations of anomalous pulmonary venous return to the right superior vena cava will be inserted at points considered more advantageous in regard to continuity.

Low cardiac and immediately infracardiac connections are the next two groups to be considered. In these categories, five interesting and often extremely complex connections were encountered. These were (1) partial to the inferior vena cava, (2) partial and (3) complete to the coronary sinus, and (4) partial and (5) complete to the portal system.

First discussed are partial connections from the right lung to the inferior vena cava. The seven instances encountered have proved interesting in that they are somewhat in variance with published data. In five of them, there was an associated atrial septal defect, in two there were none. Additionally, both supradiaphragmatic and infradiaphragmatic inferior vena cava connections were seen. The anatomic configuration of these channels has been studied in detail, as correction usually entails transplantation to a higher level. In two of the four cases suitable for study, approximately one third of the circumference of the anomalous channel was visible superficially as it lay grooving the lung parenchyma. From the embedded portion of the circumference, small pulmonary venous branches serially joined the main trunk in a manner analogous to the coniferous branching of the spruce. In the remaining two cases, a large dichotomous branching took place immediately prior to this channel joining the inferior vena cava. This form of branching, as opposed to the smaller serially branching type, introduces the danger of a lower lobe venous infarction should either of these large limbs be divided to permit mobilization and higher reimplantation of the anomalously entering pulmonary vein.

Figure 2A demonstrates the partial supradiaphragmatic drainage of the right lung into the inferior vena cava encountered six times. Drainage of blood from the left lung into the inferior vena cava is apparently rare. D'Cruz has reported a recent case.7 Interestingly, in our small group, only two cases demonstrated the features of the vena cava bronchovascular syndrome described by others.8–11 Likewise, the value of “comma-like,” “sickle-shaped,” “smitar-shaped” paracardiac shadows in the lower right lung field as a pathognomonic indication of anomalous pulmonary venous return into the inferior vena cava should probably be de-emphasized. Figure 3 shows such a paracardiac shadow, which though it was a long redundant pulmonary vein emptied normally into the left atrium.12 Pulmonary veins of this size with elongation, redundancy, and a normal left atrial point of entry probably best fit the category of “varicosities” of the pulmonary vein.13

Figure 2B demonstrates the singly encountered partial drainage of the right lung into the inferior vena cava below the diaphragm. As the diaphragm was not opened at surgery, the anomalous pulmonary venous channel's course after piercing the central leaf of the diaphragm is obscure with but catheterization evidence of its high entry into the inferior vena cava. Cooke has described a somewhat similar case.14 Total infradiaphragmatic pulmonary venous connections to the inferior vena cava have been reported and twice successfully repaired.15, 16 One should distinguish this form prognostically from infradiaphragmatic drainage into the portal

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system, as the physiologic disturbance produced by the latter is infinitely greater.

Semantically representing low cardiac, rather than infracardiac drainage, anomalous return into the coronary sinus varies from one vein to that of all the veins returning by this route. Of the former, a single case, illustrated in figure 2C, was encountered. Bilateral lower lobe drainage into the coronary sinus has occurred. It should be noted that infrequently the coronary sinus fails to drain into the right atrium. In atresia of the orifice of the coronary sinus, the coronary sinus drains retrogradely into the left atrium through a persistent Bochdalek’s foramen. Figures 4F and G later illustrate forms of this anomaly. Figure 2D represents a case of total coronary sinus drainage. The anatomic proximity of the enlarged orifice of the coronary sinus to the tricuspid valve presumably encourages streamlining of the coronary sinus-pulmonary venous return. This may result in the catheterization finding of higher oxygen saturations in the right ventricle and pulmonary artery than in the left atrium, left ventricle and the aorta, a point of diagnostic importance.

Repair of such an anomaly is shown in figure 5. The narrow intervening bridge of tissue lying between the atrial septal defect
and the enlarged coronary sinus orifice is excised and the two then confluent openings are roofed over diverting the pulmonary venous and the coronary sinus return (the volume of the latter's desaturated blood is considered to be inconsequential) into the left atrium. Production of surgical heart block is to be avoided. \(^{21}\)

A variant of coronary sinus drainage is depicted in figure 2E. It has been inferred that mixed levels of anomalous pulmonary venous drainage do not occur or are quite rare. Gott et al. reporting 30 cases of total anomalous pulmonary venous drainage stated, "Of the cases cited in the literature as having drainage via the left superior vena cava, none appears to drain in this fashion—communicating proximally with the coronary sinus and distally with the innominate vein." \(^{22}\) Similarly, Darling's group in a review of 17 personal cases and a survey of the available literature (80 cases) could find but a single case of mixed levels of drainage. \(^{23}\) Interestingly, nine instances of such double levels of drainage as illustrated were encountered in this series.

In a single additional case, the left upper lobe vein drained normally into the left atrium as shown in figure 2F.

A transitional form leading towards the classic "figure-of-eight" anomaly is illustrated in figure 2G. In this, and the following anomaly, the clinical and radiologic pattern can apparently be modified considerably, depending upon whether or not the persistent left caval remnant passes as it usually does anterior to the pulmonary artery or posteriorly as is occasionally seen. In passing posterior to the pulmonary artery, the anomalous channel may become obstructed with impeded venous outflow from the lungs simulating an infradiaphragmatic type connection. \(^{24}\)

Figure 2H represents the classic form of total anomalous pulmonary venous return. Diagnostic-wise, wide variations have occurred in the time of appearance of the classic chest roentgenogram. The chest roentgenogram of a 3-month-old infant was most suggestive, whereas in a 13-year-old the configuration was not particularly impressive. The monotonously equal, markedly elevated oxygen-saturations throughout all chambers, pulmonary and peripheral arteries is highly suggestive of total anomalous pulmonary vein drainage, especially of the supracardiac type. The values found in one such case are shown.

Two associated anomalies have greatly influenced the clinical courses followed by these patients. We have repaired such an anomaly in an older child in which there was only the slightest clinical disability. This clinical well-being apparently resulted from the coexistence of a mild pulmonary stenosis which beneficently prevented pulmonary flooding and encouraged right-to-left shunting through a large atrial septal defect. At the other extreme, marked early deterioration has been seen in quite young infants when the atrial communication was small, allowing little blood to reach the left side of the heart. In a 9½ pound infant recently operated upon the diameter of this communication

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**Figure 3**

Arrows identify a vertically coursing, right paracardiac pulmonary vein. After a circuitous route, this vein entered the left atrium normally.
(patent foramen ovale) was a mere 5 mm.

In figure 2I an anomaly encountered three times is shown. Should disease or removal of the right lung occur, survival would be possible only in the presence of an atrial septal defect.25 The variant encountered once, seen in figure 4A, does not, of course, carry these dire implications.

In figure 4B a transitional form combining supracardiac and infradiaphragmatic portal drainage is illustrated. The heart and lungs showed no abnormality other than the anomalous pulmonary venous drainage and the atrial septal defect. This technically correctable association of anomalies, with the lack of a severe intracardiac malformation, invites staged surgical repair.

Total infradiaphragmatic pulmonary venous drainage, as illustrated in figure 4C, was encountered twice. The pulmonary veins formed a common trunk which passed through the esophageal hiatus to enter the portal vein by way of an enlarged gastric (coronary) vein. The clinical and radiologic picture is that of obstructed pulmonary venous outflow.26, 27 The better prognosis of an infradiaphragmatic drainage emptying into the inferior vena cava or the hepatic veins has already been mentioned.

Figure 4D, an example of mixed levels of
Total anomalous pulmonary venous drainage into the coronary sinus. In (1) the intraatrial anatomy is shown. In (2) the thin septum of tissue dividing the coronary sinus and the atrial septal defect is being divided. In (3) the resultant confluent complex has been roofed over diverting the pulmonary venous and coronary sinus return into the left atrium.

Figure 5

The anomaly depicted in figure 4E is interesting embryologically. The heavily dotted circle diagrammatically represents a thick muscular diverticulum found growing outward from the back wall of the left atrium. Had this diverticulum succeeded in connecting with the common pulmonary venous trunk, two possibilities can be envisioned. If the connection were sufficiently large, the infradiaphragmatic channel would likely disappear. If the connection were a small or constricted one, a cor triatriatum might have resulted upon involution of the portal connection.

The patient represented by figure 5F had atresia of the coronary sinus orifice, a primum atrial septal defect, and a left superior vena cava (receiving one anomalous pulmonary vein) entering the left atrium directly.

In figure 4G a tiny 2-mm. slit marked the hypoplastic coronary sinus orifice. Atresia of the mid-portion of the sinus is postulated with retrograde flow into the left atrium through the orifice also discharging blood from the persistent left superior vena cava. The correctness of this assumption, however, could not be fully ascertained by drainage, differs only in the presence of a left upper lobe vein draining normally. Figure 6 is nearly identical except that there are two inferior venae cavae present.

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The next three figures (4F, G, and H) represent left superior venae cavae emptying directly into the left atrium. In many hearts, immediately above the mitral mural leaflet, a large thebesian vein orifice is seen, which Bochdalek, Langer, and Tandler have noted as an almost constant feature in the left atrium. In atresia of its ostium, the coronary sinus generally drains retrogradely to enter the left atrium at this point (fig. 7). A left superior vena cava directly entering the left atrium also does so by this venous orifice.

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For clarity, the heart excepting a posterior shell of the essentially common atrium has been removed. Three cavae—a right superior, and a right and left inferior enter the atra. Transparency of the liver and atra permits visualization of the posterior-lying common pulmonary venous trunk's course and connection with the coronary vein along the lesser gastric curvature.

Cor triatriatum is usually an abnormality of the size, not the site, of the left atrial pulmonary venous connection. In figure 4H a cor triatriatum is combined with the presence of a "levo-atrial cardinal vein" as described by Edwards.28 Because of the small common pulmonary vein-left atrial communication, a venous connection between the mid and the anterior cardinal systems persisted. As the major pulmonary venous flow was directed into the right superior vena cava, the case hemodynamically represents one of anomalous pulmonary venous return and not one of cor triatriatum, thus accounting for its inclusion in this presentation.

Summary

One hundred and thirteen selected anomalous pulmonary venous returns have been well documented anatomically. Twenty-seven variations were seen. With the use of simplified diagrams, these have been correlated with related cases in the literature. Emphasis has been placed on transitional forms with one anomaly shading off into the next, rather than upon a series of neat, compartmentalized, classic types of anomalous pulmonary venous return. It is hoped that the broad anatomic spectrum shown will promote additional understanding and interest.

References

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