Total Anomalous Pulmonary Venous Drainage 
at Cardiac Level

Angiocardiographic Differentiation

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In UNCOMPLICATED total anomalous pulmonary venous drainage, the commonest site of entry into the systemic venous circulation is through the left superior vena cava and the left innominate vein. In a diagnostically less spectacular group, comprising 30 per cent of all cases of the anomaly, the pulmonary veins enter the right atrium either directly or via the coronary sinus.\(^1\) Whereas in the common, supra-cardiac type of connection, diagnosis is usually simple, in those at cardiac level detailed anatomic definition may be extremely difficult.

It has been stated\(^2\) that the contour of the heart in plain x-ray films of the chest is characteristic for total anomalous pulmonary venous drainage into the coronary sinus. In the experience of Bahnson et al.\(^3\) and ourselves, this has not proved to be specific. Cardiac catheterization in both types of anomaly simply localizes a left-to-right shunt of large volume to the atrial level, demonstrates blood oxygen saturations of approximately equal values in all 4 chambers, moderate to marked pulmonary hypertension, and a patent foramen ovale or true atrial septal defect. Indicator-dilution curves of similar contour may be recorded after injections in all of the right heart chambers or tributaries, those following injection in the right ventricle and pulmonary artery having a longer appearance time than those after caval or right atrial injections.\(^4\) A filling defect due to turbulence of contrast material at the site of insertion of the anomalous veins has been noted in venous angiocardiograms of patients in whom pulmonary veins enter the right atrium directly\(^5\) but in a few children with the total anomaly, studied in this manner in the authors' laboratory, the sign was lacking.

None of these methods so far discussed has allowed differentiation of the two types of total anomalous pulmonary venous drainage at cardiac level. The purpose of this paper is to describe an angiocardiographic sign that permits such separation.

Material and Methods

Six cases of this anomaly, 4 with drainage into the coronary sinus and 2 with drainage into the right atrium directly, were studied. The patients were all young, 5 being under 6 months of age. The clinical, electrocardiographic, and radiographic features in all cases were compatible with total anomalous pulmonary venous drainage. Data from right- and left-sided catheterizations in each case prior to angiocardiography confirmed the presence of total anomalous pulmonary venous drainage at cardiac level. At the conclusion of the preliminary study, selective angiocardiography was performed with the catheter in the main pulmonary artery. An average amount of 1.6 ml. of contrast material (50 per cent Hypaque; 70 per cent Diodrast) per Kg. of body weight was injected with use of technics and equipment previously described.\(^6\) The diagnosis was confirmed subsequently at autopsy in 4 patients and at surgical correction in the remaining 2.

Results

In both groups a large pulmonary artery trunk and branches were visualized. Owing to the position of the catheter in 3 cases, only 1 major pulmonary artery branch was heavily opacified. It was in the outline of pulmonary venous return to the right atrium in the anteroposterior view usually between 1 and 2 seconds after the injection that the differences between the two types of connection were observed.

In the coronary sinus variety (fig. 1) pul-

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Selective angiocardiograms of 4 patients with total anomalous pulmonary venous drainage into the coronary sinus. Note in each instance an ovoid opacification within the right atrial segment.

Pulmonary veins from each lung merged into a common vein that arched upward to join its opposite and then descended into the coronary sinus. This latter structure opacified as a vertical, ovoid structure lying over the left side of the vertebral column with its lower margin at the level of the diaphragm. While quite obvious in very young infants, the appearance was more striking in the older patient and looked like an egg lying within the right atrium. The contrast gradually emptied into the right atrium proper, producing a faint opacification of that chamber.

Where all 4 pulmonary veins entered the right atrium directly (fig. 2) pulmonary veins appeared to pass in a horizontal manner at a lower level to form a circular opacity within the right atrium rather more to the right over the spine. The right atrium was more rapidly and completely filled with contrast than in the coronary sinus form.

In neither variety was the lateral projection contributory, nor did the presence of a true atrial defect rather than a foramen ovale appear to influence the degree of opacification of the coronary sinus or right atrium.

Discussion

As in so many cardiac anomalies, the final details of morphology depend in great part on a prior demonstration of the functional pathways of the abnormal circulation. Once it can be ascertained by preliminary cardiac catheterization that pulmonary venous drainage is totally anomalous and that the entry of these veins to the systemic system is at car-
ANOMALOUS PULMONARY VENOUS DRAINAGE

Figure 2
Selective angiocardiograms of 2 patients with total anomalous pulmonary venous drainage into the right atrium. Note in both instances, opacification of the right atrium but absence of the separate ovoid structure visible in the coronary sinus variety.

diac or atrial level, selective angiography will clarify the anatomic type of connection. Of the two types, the coronary sinus variety has the more arresting appearance after angiography. On the basis of present experience it is probable that this angiographic picture of the ovoid structure within the cardiac silhouette in this type is, under the named circumstances, a pathognomonic sign of the malformation. It seems likely from other evidence as yet unconfirmed, that when at least 2 pulmonary veins are connected with the coronary sinus in the mixed variety of total anomalous pulmonary venous drainage, a similar ovoid opacification can be demonstrated at angiocardiography.

When the pulmonary veins enter the right atrium by a final, single channel, as is usually the case, the angiographic appearance is also uniform though less striking. So far we have not demonstrated by this technic the less common examples of the right atrial connection having separate attachment of all 4 pulmonary veins. It may be possible to exclude such an arrangement when the pulmonary veins from either lung are seen to merge into a single vein before entering the right atrium.

With the advent of open-heart methods for their correction, the differentiation between total anomalous pulmonary venous drainage into the coronary sinus or right atrium has rather less practical importance than previously. Technically the coronary sinus variety is easier to repair whereas the type in which all 4 pulmonary veins are connected separately to the right atrium offers formidable surgical problems. Prior knowledge of the type may, therefore, still influence the timing of surgical treatment, especially in infants.

Summary
In 6 patients with total anomalous pulmonary venous drainage at cardiac level, differentiation between pulmonary veins entering the right atrium and those entering the coronary sinus was possible after selective angiocardiography into the pulmonary artery.

The principal feature of the angiocardiogram of the coronary sinus variety is an egg-shaped opacification over the spine within the right atrial contour. In the direct connection of pulmonary veins to the right atrium, the latter chamber may fill promptly after pulmonary venous return, sometimes starting as a circular shadow with the right
atrium but never having the ovoid appearance of the coronary sinus variety.

References

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As a frog's heart has but one ventricle, the blood is thrown by the same ventricle, at the same instant, both into the lungs and all over the body; then since its velocity is, in arteries of equal diameters, five times greater in the lungs than in the muscles, notwithstanding it is impelled by one common impetus; this evidently shows, that it must have freer passage through the lungs. Accordingly the left ventricle of the heart is made much stronger, thereby to impel the blood with a greater force than the right ventricle does.—Stephen Hales, B.D., F.R.S. Haemastatics, Vol. II, London, 1733.
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