Echocardiographic Evaluation of Partial Anomalous Pulmonary Venous Drainage

MAY LING WONG, MBBS, MMED(PAEDS),* BRIAN W. MCCRINDLE, MD, FACC, CLEONICE MOTA, MD, JEFFREY F. SMALLHORN, MD

Toronto, Ontario, Canada

Objectives. This study was undertaken to determine the accuracy of routine echocardiography in the detection of partial anomalous pulmonary venous drainage.

Background. Although there are occasional case reports of the echocardiographic appearance of partial anomalous pulmonary venous drainage, no large series have addressed the accuracy of this technique in a large cohort of patients.

Methods. Between January 1983 and December 1993, 50 patients with partial anomalous pulmonary venous drainage (with or without an associated atrial septal defect as the only other significant intracardiac defect) were identified from the data base at the Hospital For Sick Children, Toronto. Routine echocardiographic reports were reviewed, and the results were compared with surgical or catheterization findings. Risk factors related to diagnostic errors were sought using a Fisher exact test, chi square analysis, t test and Kruskal-Wallis analysis of variance.

Results. Confirmation of the diagnosis was available in 45 patients whose data were subsequently used for risk factor analysis. The median age at echocardiography was 4.1 years (range 1 month to 18 years). Right-sided drainage was present in 43 patients (86%), with left-sided drainage in 7 (14%). Thirteen patients had an intact atrial septum, 7 a patent foramen ovale and 30 a secundum atrial septal defect. Right ventricular dilation was observed in 46 patients. Two had normal dimensions (two not assessed). The diagnosis was missed by echocardiography in 15 (33%) of the 45 patients with a confirmed diagnosis. Year of study and use of color flow mapping were the only significant variables related to detection rate (7% missed diagnosis with vs. 62% without the use of color flow, p < 0.0005). The median year of missed diagnosis was 1985 versus 1990 (p < 0.002). Transesophageal echocardiography accurately defined the site of drainage in all three patients in whom it was utilized.

Conclusions. Two-dimensional echocardiography in conjunction with color flow mapping is a valuable tool for the diagnosis of partial anomalous pulmonary venous drainage.

(J Am Coll Cardiol 1995;26:503-7)
Measurements and methodology. The following information was abstracted from routine echocardiographic reports and clinical records of the remaining 50 patients: date and age at initial echocardiogram; the echocardiographic imaging modalities that were used; characterization of partial anomalous pulmonary venous drainage, including side and site of drainage; the presence of associated pulmonary venous obstruction; an associated atrial septal defect and right ventricular dimension. Confirmation for all cases was sought from cardiac catheterization, surgical or autopsy records. In addition, the original video recordings were available for 35 (70%) of echocardiographic studies and were reviewed in blinded manner and independently by one of the investigators (J.F.S.).

Analysis. Descriptive statistics were used to characterize the study patients and measurements. For patients whose diagnosis had been confirmed by cardiac catheterization, surgical or autopsy records, factors related to diagnostic errors on echocardiography were sought. Partial anomalous pulmonary venous drainage that was missed by echocardiography was defined as follows: 1) missed on initial report, and video recording not available for independent review; 2) missed on both initial report and on second review; or 3) detected on initial report and missed on second review. Risk factors for missed partial anomalous pulmonary venous drainage were sought by means of the Fisher exact test, chi-square analysis, t test and Kruskal-Wallis analysis of variance; p < 0.05 was considered significant.

Results

Characteristics of study patients. There were 50 patients with partial anomalous pulmonary venous drainage. The mean age at echocardiography was $5.3 \pm 5.1$ years (median 4.1, range 1 month to 18 years). Confirmation by cardiac catheterization, surgical or autopsy records was obtained in 45 patients.

For the total cohort the connection of the partial anomalous pulmonary venous drainage was right-sided in 43 patients (86%) and left-sided in 7 (14%). Drainage was to the right superior vena cava in 9 patients (18%) (Fig. 1), superior vena cava right atrial junction in 4 (8%) (Fig. 2), right atrium in 18 (36%) (Fig. 2), infracardiae in 12 (24%) (Fig. 3) and left vertical vein in 7 (14%).

Thirteen patients (26%) had isolated partial anomalous pulmonary venous drainage; 30 (60%) had a secundum atrial septal defect; and 7 (14%) had a patent foramen ovale. The presence of an obstructed flow pattern was noted in five patients (11%), where the diagnosis of anomalous veins was made by echocardiography.

Right ventricular dilation for age was noted in 46 patients (96%), with 2 (4%) having normal right ventricular dimensions for age. In two patients right ventricular size was not assessed. In one of these patients, despite total unilateral drainage diagnosed at 3 months of age, the pulmonary vein was significantly stenotic, hence a small left to right shunt with a normal-sized right ventricle. The second child with normal right ventricular dimensions had Down syndrome and isolat
anomalous pulmonary venous drainage diagnosed at 7 months of age, with no cardiac murmur; the abnormality had been detected during a study to exclude congenital heart disease.

Risks factors for missed diagnosis. Of the 45 patients with confirmation by cardiac catheterization, surgical or autopsy records, the diagnosis of partial anomalous pulmonary venous drainage was missed by echocardiography in 15 (33%). The diagnosis had been missed on both the initial report and the independent review in 10 patients and, noted on the initial report but missed on independent review in 5.

Risk factors associated with a missed diagnosis on echocardiography were sought (Table 1). Characteristics of the partial anomalous pulmonary venous drainage, such as the side and site of drainage, presence of venous obstruction, associated defects and right ventricular dilation, were not significantly related to a missed diagnosis, nor was the age at echocardiography.

Both year of echocardiography and use of color flow mapping were significantly related to detection rates. Patients whose diagnosis was missed were less likely to have studies with color flow mapping (7% vs. 62%; p < 0.0005) and were earlier in the experience (median year at echocardiography 1985 vs. 1990; p < 0.002) (Fig. 4). However, these variables (year of study plus the use of color) were interrelated, with more diagnoses missed in earlier years when color flow mapping was not available (first available at our institution in 1987, all machines converted in 1989), with fewer diagnoses missed in later years with the increased use of routine color flow mapping (Fig. 4). There were no significant changes in the characteristics of partial anomalous pulmonary venous drainage over the study period. The use of transesophageal echocardiography resulted in the accurate detection and characterization of partial anomalous pulmonary venous drainage in three patients (Fig. 1), where transthoracic echocardiography was inconclusive but demonstrated a volume overloaded right ventricle.
Table 1. Risk Factors for Missed Diagnosis on Echocardiography in 45 Patients

<table>
<thead>
<tr>
<th>Missed Diagnosis</th>
<th>No. of Pts</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Side of drainage</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right</td>
<td>12/37</td>
<td>32</td>
</tr>
<tr>
<td>Left</td>
<td>3/8</td>
<td>38</td>
</tr>
<tr>
<td><strong>Site of drainage</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>RSVC</td>
<td>1/7</td>
<td>14</td>
</tr>
<tr>
<td>RSVC/RA junction</td>
<td>0/4</td>
<td>0</td>
</tr>
<tr>
<td>RA</td>
<td>7/17</td>
<td>41</td>
</tr>
<tr>
<td>Infracardiac</td>
<td>4/11</td>
<td>36</td>
</tr>
<tr>
<td>LSVC</td>
<td>3/6</td>
<td>50</td>
</tr>
<tr>
<td><strong>Presence of associated defects</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Isolated PAPVD</td>
<td>1/7</td>
<td>13</td>
</tr>
<tr>
<td>Secundum ASD</td>
<td>8/19</td>
<td>42</td>
</tr>
<tr>
<td>Scimitar vein</td>
<td>4/11</td>
<td>36</td>
</tr>
<tr>
<td>Patent foramen ova ile</td>
<td>2/7</td>
<td>29</td>
</tr>
</tbody>
</table>

ASD = atrial septal defect; LSVC = left superior vena cava; PAPVD = partial anomalous pulmonary venous drainage; Pts = patients; RA = right atrium; RSVC = right superior vena cava.

Accuracy of echocardiographic localization of partial anomalous pulmonary venous drainage. The precise site of drainage of the partial anomalous pulmonary veins on echocardiography was compared with that identified by cardiac catheterization or surgical records in 34 patients. The echocardiogram was accurate in 28 patients (82%); in those with missed diagnosis, drainage was to the right atrium or superior vena cava. In all but one patient, the disagreement revolved around whether or not drainage was to the superior vena cava or superior vena cava/right atrial junction. The use of color flow mapping did not affect the accuracy of precise echocardiographic localization (84% accuracy with and 80% without the use of color flow mapping, p = NS).

Discussion

To our knowledge our study represents one of the largest series of patients seen to have relatively isolated anomalous pulmonary venous drainage. The patients in our series may not be completely representative of all possible patients with this diagnosis because it can be hypothesized that many asymptomatic patients never come to clinical attention.

In our study we found right-sided partial anomalous pulmonary venous drainage to be more common, with only seven patients (14%) having left sided drainage. This is comparable to other reports, where anomalous drainage from the left side is infrequently encountered (3,4).

**Previous echocardiographic data.** Previous reports of the role of echocardiography are limited (5), with little data on its accuracy in patients with this condition. Both surgical or catheter closure of an atrial septal defect (6,7) may be influenced by the presence of associated partial anomalous pulmonary venous drainage; hence, accurate assessment by echocardiography is essential.

Apart from those situations where the diagnosis of partial anomalous pulmonary venous drainage (8) was likely, such as in the presence of a sinus venosus atrial septal defect, a dilated coronary sinus or a left vertical vein, our experience has thus far indicated that the diagnosis was invariably missed during the process of routine echocardiography, particularly before the introduction of color flow Doppler.

The present study demonstrates that routine imaging and color flow Doppler of the potential drainage sites should permit the echocardiographer to make an accurate diagnosis in the majority of cases. The diagnosis in our cohort was most unlikely if right ventricular dimensions were within normal limits.

**Technique of echocardiographic assessment.** One of the first signs in patients with right-sided partial anomalous pulmonary venous drainage can be seen from the subcostal view, where there is a blunt appearance at the junction of the atrial septum and the right-sided free wall of the left atrium (Fig. 5). In our series, this sign was only reliable in patients with total right-sided partial anomalous pulmonary venous drainage. However, careful inspection of this site, by a sweep of the transducer in a superoinferior plane should permit future identification of those patients with partial unilateral drainage; as in the normal heart, veins can be seen entering the right side in both positions.

Color flow interrogation of this area may be helpful; however, unless analyzed carefully, it may give a false impression of pulmonary venous flow originating normally from this area.

![Figure 4](image-url)
Figure 5. Subcostal view of total unilateral anomalous right-sided pulmonary venous connection demonstrating blunt appearance of the right side of the left atrium. Abbreviations as in Figure 1.

site. This appearance is the result of flow from the left-sided pulmonary veins, hugging the superior aspect of the left atrium and then changing direction at the blunted junction of the atrial septum and free wall.

Those anomalous veins entering the proximity of the superior vena cava right atrial junction, as well as the body of the right atrium, can be identified as the transducer beam is angled from the subcostal position in a superoinferior direction, to image the atrial septum. From this portion the color flow patterns from the individual veins can be seen. Those veins closer to the inferior vena cava right atrial junction are best imaged at the same time as the inferior vena cava is being interrogated, by continuing the sweep to include the lower portion of the right atrium. By contrast, those veins that drain directly into the superior vena cava are in general best imaged from the suprasternal view, with the transducer beam rotated clockwise from the frontal plane, with subsequent angulation to image the total length of the superior vena cava right atrial junction. From this position, the color flow pattern of the anomalous pulmonary veins is toward the transducer, in the opposite direction of the superior vena cava flow, apart from the normal brief period of reversed flow during atrial systole.

In the present series, age had only a marginal effect on the diagnostic accuracy; this would probably be unlikely in a large adolescent or adult population, where inferior imaging is usual. In this older age group, transesophageal echocardiography (9-11) should permit the echocardiographer to secure a correct diagnosis in the majority of cases. Thus far there have been no large series using this technique; however, our limited experience, plus previous published reports (9,10) demonstrate the feasibility of this technique. Our main experience has been in patients with drainage to the superior vena cava or its junction with the right atrium. Indeed, because this technique provides superior imaging of the actual pulmonary veins, it is relatively straightforward to follow them to their abnormal site of drainage. Armed with a knowledge of the potential sites of drainage, along with a sound understanding of the normal pulmonary venous pattern, it should be relatively straightforward to achieve similar results in other patients using transesophageal echocardiography.

Conclusions. The diagnosis of partial anomalous pulmonary venous drainage requires a high degree of suspicion, a thorough knowledge of both the normal and abnormal patterns, along with a systematic approach to the diagnosis. If the diagnosis cannot be excluded by transthoracic echocardiography alone, particularly in the presence of a volume-loaded right ventricle, then an assessment by transesophageal echocardiography should be undertaken. This approach, along with the use of color flow Doppler, should make possible an accurate diagnosis in the majority of patients.

References

4. Brody H. Drainage of the pulmonary veins to the right side of the heart. Arch Pathol 1942;33:221-40.