Pulmonary Atresia With Intact Ventricular Septum
Range of Morphology in a Population-Based Study

Piers E. F. Daubeney, MRCP,*‡ David J. Delany, FRCR,* Robert H. Anderson, MD;‡
George G. S. Sandor, MD,§ Zdenek Slavik, MD,‡† Barry R. Keeton, FRCP,* Steven A. Webber, MRCP,*¶
for the United Kingdom and Ireland Collaborative Study of Pulmonary Atresia with Intact
Ventricular Septum
Southampton and London, United Kingdom; Vancouver, Canada; and Pittsburgh, Pennsylvania

OBJECTIVES
We describe the morphologic variability in pulmonary atresia with intact ventricular septum
(PAIVS) within a population-based study.

BACKGROUND
An uncommon disease, PAIVS shows considerable morphologic heterogeneity. Clinical
reports, based mostly on small samples of patients, may not reflect the true spectrum of
pathology of this condition. We have studied the entire range of morphology in a prospective
population-based study of patients over a five-year period (1991 to 1995).

METHODS
As part of the United Kingdom and Ireland Collaborative Study of PAIVS, all 18 pediatric
cardiac centers were visited by a single investigator. Morphologic features of each case were
determined by direct review of the echocardiograms and angiograms, from surgical and
autopsy reports, and by review of pathology specimens where available.

RESULTS
Among 183 live-born infants, atresia was valvar (membranous) in 74.7% and muscular in
25.3%. Muscular obliteration of the apical trabecular cavity, and in some cases its infundib-
umulum, resulted in “bipartite” right ventricle (RV) in 33.6%, and a “unipartite” chamber in
7.7%. The remaining 58.7% had “tripartite” morphology. Coronary arterial abnormalities
were identified in 45.8%, including arterial stenoses, interruptions and ectasia in 7.6%. Ebohist’s
malformation coexisted in 18 patients. Median tricuspid valvar size and RV inlet
Z-scores were 5.2 and 5.1, respectively.

CONCLUSIONS
This study provides unique data on the diverse pathology of PAIVS in an unselected
population. This will help determine if published reports reflect the true spectrum of
pathology of the condition. (J Am Coll Cardiol 2002;39:1670–9) © 2002 by the American
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Pulmonary atresia with intact ventricular septum (PAIVS) is a rare disease with considerable morphologic diversity (1–3). This has led to many different strategies for treat-
ment. Outcome will depend, in great part, on the underly-
ing cardiac morphology. Thus, reports of unusually good or
disappointing results could reflect a selected population of
patients with favorable or unfavorable pathology, rather
than specific institutional or management factors. Owing to
the rarity of this condition, more than nine-tenths of
published series contain information on
<30 patients. The
range of morphology in a given study, therefore, may not be
representative of the population of infants at large. The aim
of our study was to provide a detailed description of the
range of morphology in a population-based study (the
United Kingdom and Ireland Collaborative Study of Pul-
monary Atresia with Intact Ventricular Septum).

METHODS
The Collaborative Study is a unique, contemporary,

population-based study set up with the cooperation of all 18
pediatric cardiology centers in the United Kingdom and
Ireland. From January 1, 1991, to December 31, 1995, all
infants born with PAIVS and all fetal diagnoses of this
condition were studied (4). Because of the high population
density and small geographic area, it proved feasible for a
single investigator (P. D.) to visit all 18 centers on repeated
occasions. Local databases, admission and operative records
and regional pathology records were directly inspected to
ensure completeness of data collection. Cases were included
if they had PAIVS without associated complex intracardiac
abnormalities (cases with associated Ebohist’s malformation
were included). Cases with tiny coexisting ventricular septal
defects were included where the overall anatomy and phys-

iology reflected that of PAIVS rather than pulmonary
atresia with ventricular septal defect. Cases of critical
pulmonary stenosis were excluded. Cases were also excluded
if they were born outside the United Kingdom and Ireland.

The cases diagnosed prenatally have been reviewed previ-
ously (4). Their range of morphology is not the subject of
this report.

For each patient, the medical records, chest radiographs,
electrocardiograms (ECGs), echocardiograms, hemody-
amic findings, angiograms and the operative and

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autopsy reports were directly reviewed. The echocardiogram at presentation was reviewed by a single investigator (P. D.) (Table 1). Tricuspid valve (TV) diameters and right ventricular (RV) inlet lengths, the latter taken from the midportion of the atrioventricular annulus to the apex of the RV, were measured by postprocessing from the cross-sectional echocardiographic four-chamber view at end-diastole (P. D., Z. S.) as previously described (5). Z-scores representing the number of standard deviations an observation deviates from the normal population mean for a given body surface area, were derived from echocardiographic data collected in 125 normal children (5) (Fig. 1).

Each angiocardiogram was directly reviewed by three investigators (P. D., D. D., G. S.) (Table 1). The RV pressure obtained during initial cardiac catheterization was recorded. The degree of tricuspid regurgitation was estimated from review of the initial angiocardiogram and graded as absent, mild, moderate or severe. To allow statistical comparisons with other variables, patients were categorized into absent or mild, moderate or severe tricuspid regurgitation. Significant RV dilation was recorded when review of the angiocardiograms and echocardiograms demonstrated a hugely dilated and thin-walled ventricle. The angle that the patent arterial duct subtended on the postductal descending aorta was recorded from the echocardiogram as described by Santos et al. (6). The echocardiograms and angiocardiograms were reviewed to ascertain whether the atresia was due to muscular infundibular obliteration (muscular atresia), or complete fusion of the valve leaflets (membranous atresia), with a patent infundibulum existing in the latter setting up to the level of the valvar tissue.

Although appreciating that all three ventricular components are always present in this condition but with variable intracavity muscular overgrowth (7), the number of parts of the RV not obliterated by such muscular overgrowth (so-called tripartite, bipartite and unipartite [8,9]) was recorded. The presence of Ebstein’s malformation, and any additional morphologic abnormalities of the RV, was also recorded. The coronary arterial anatomy was closely studied from RV and aortic root injections, and the presence and position of RV-to-coronary fistulae and coronary arterial stenoses were recorded. When there was slight filling of nondilated coronary arteries from an RV angiogram, RV-coronary communications were termed “minor fistulae.” When there was prominent filling of one or more, usually dilated, coronary arteries associated with retrograde filling of the aorta, the communications were designated as “major fistulae.” Markedly dilated coronary arteries (>3 mm) were described as “ectatic,” and also considered as major fistulae.

Table 1. Sources of Information Available for the Study

<table>
<thead>
<tr>
<th>Source of Information</th>
<th>Total Number of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total patients in study</td>
<td>183</td>
</tr>
<tr>
<td>With one or more angiograms</td>
<td>143</td>
</tr>
<tr>
<td>With angiograms before first procedure</td>
<td>90</td>
</tr>
<tr>
<td>At least one angiogram directly reviewed</td>
<td>137</td>
</tr>
<tr>
<td>With echocardiograms</td>
<td>182</td>
</tr>
<tr>
<td>With echocardiograms before first procedure</td>
<td>177</td>
</tr>
<tr>
<td>At least one echocardiogram directly reviewed</td>
<td>172</td>
</tr>
<tr>
<td>Total patients having a therapeutic procedure</td>
<td>170</td>
</tr>
<tr>
<td>With procedure report available</td>
<td>162</td>
</tr>
<tr>
<td>Total deaths</td>
<td>70</td>
</tr>
<tr>
<td>With postmortems</td>
<td>40</td>
</tr>
<tr>
<td>With postmortem reports available</td>
<td>40</td>
</tr>
<tr>
<td>With specimens examined by author R. A. or regional pathologist for this study</td>
<td>21</td>
</tr>
</tbody>
</table>
“RV dependent coronary circulation” was considered to be present when fistulous communications were associated with either absent aortocoronary connection, coronary arterial interruption or unequivocal stenosis of one or more of the major epicardial coronary arteries. Markedly ectatic coronary arteries were also considered in this category, as RV decompression could result in major coronary steal through the ectatic vessel to the RV (10).

Autopsy reports for all the deceased patients were available for review (Table 1) and, in addition, the cardiac specimen was reexamined for the purposes of this study in 50% of cases by one of the investigators (R. A.), or by a regional pediatric pathologist who recorded findings on a standardized questionnaire. Overall morphology at, or prior to, initial intervention was ascertained by combination of the information from echocardiograms, angiocardograms, operative notes, postmortem records and postmortem specimens (Table 1). Associated cardiac and noncardiac anomalies were also recorded. Throughout the results, the frequencies of specific anatomical findings are expressed as a proportion of the total number of cases for which adequate information was available (Table 1).

**Statistical analysis.** The statistical significance of two morphologic features coexisting in the same heart was examined using Pearson’s chi-square test with Yates’ correction when expected values were small, the Student t test and linear regression depending on whether the data were categoric or continuous. This was expressed as a p value (Table 2). P values <0.05 were considered statistically significant. For subgroups with various significant morphologic characteristics, a 95% confidence interval (CI) was calculated; Z-scores for the TV and RV inlet were expressed as cumulative frequency distributions. All data was stored in coded fashion on a relational database (Filemaker Pro 3.0, Claris, Santa Clara, California), and all analyses were performed using a commercial statistical package (Statview 4.0, Abacus Concepts, Berkeley, California).

**RESULTS**

From 1991 to 1995, there were 183 infants born alive with PAIVS in the United Kingdom and Ireland, giving an overall incidence of 4.5 cases per 100,000 live births. Distribution of births has been described previously (4). There were 86 fetal diagnoses during this period, leading to 53 terminations of pregnancy (61%). Of the 183 live births, 70 have died postnatally.

**Size and morphology of the tricuspid valve.** Median TV Z-score at presentation was -5.2 (interquartile range -8.4 to -2.6) (Fig. 2) as derived using our own normal echocardiographic data (5) (Fig. 1A) or -1.6 (-2.9 to -0.4) using Rowlett’s “normal” data based on postmortem measurement of formalin-fixed samples (11). The relationship between TV Z-score and other morphologic variables is shown in Table 2. Ebstein’s malformation coexisted in 18/183 cases...
based on the normal data of Daubeney et al. (5) (shown in Fig. 1). Valve (TV) diameter measured by echocardiography. The Z-scores are

Size and morphology of the right ventricle. Significant RV dilation was present in 8/183 cases, and this was associated with moderate to severe tricuspid regurgitation in all cases (Fig. 3). Of the eight cases, Ebstein’s malformation was present in three, marked TV dysplasia in two, and a seemingly normal TV, but with a dilated ori

Median RV inlet length Z-score at presentation was –5.1 (interquartile range –7.5 to –2.8) (Fig. 4) as derived using our own normal echocardiographic data (5) (Fig. 1B) or –5.3 (–7.6 to –3.0) using the normal echocardiographic data of Hanseus et al. (12). The relationship between RV inlet Z-score and other morphologic variables is shown in Table 2.

Of the 143 where it was definitely possible to ascertain the RV morphology, muscular obliteration of both apical and infundibular portions of the RV resulted in so-called “unipartite” RV in 7.7% of cases (95% CI, 3.3% to 12.1%) (Fig. 3). Overgrowth of the apical trabecular portion resulted in “bipartite” ventricles in 33.6% of cases (95% CI, 18.6% to 48.6%) (Fig. 3). There were no examples of infundibular obliteration without apical obliteration. The remaining 58.7% (95% CI 50, 1% to 67.4%) had “tripartite” morphology (Fig. 3). The atresia was membranous in 74.7% of cases (95% CI, 68.3% to 81.2%) and muscular in 25.3% (95% CI, 18.2% to 32.4%) (n = 174).

Coronary arterial abnormalities. There was adequate visualization of the coronary arteries in 131 patients by angiography or at autopsy (Fig. 3). The coronary arteries were normal in 71 patients (54.2%) (95% CI, 45.7% to 62.7%), including 16% with prominent but blind-ending fissures in the ventricular wall that filled in ventricular systole without communicating with the coronary arteries.

Minor fistulae were present in 28 patients, and major fistulae in 33 patients, giving a total number of 61 (45.8%) (95% CI, 37.3% to 54.3%) with fistulous communications out of 131 patients. The coronary arterial regions where fistulous connections occurred, and their frequency of occurrence, are documented in the Appendix.

Ten cases, 7.6% (95% CI, 3.1% to 12.2%), were considered to have an “RV-dependent” coronary circulation. Stenosis, interruption or severe ectasia of the right coronary alone was documented in five cases (3.8%), of the left coronary alone in two (1.5%), and of both coronary arteries in another two cases (1.5%). In one case with major fistulous connections to the aorta, there was insufficient information to localize the site of the RV dependence. Fistulous connections were most commonly to the distal right and proximal left anterior descending (LAD) coronary arteries, and rarely to the circumflex artery, when they were extremely distal (Appendix). In one case, there was a connection via the coronary arteries between the RV and pulmonary arteries. Stenoses occurred predominantly in the distal right and distal LAD arteries, usually close to the site of entry of a fistula. Ectasia was found equally in all parts of the right coronary artery and/or LAD, and was often associated with significant luminal irregularity.

One case studied at autopsy had both an atretic orifice of the right coronary artery and absence of the orifice of the left coronary (Fig. 5). The right coronary arterial orifice was represented by only a dimple, and there was no evidence of another orifice. The main stem of the left coronary artery was also absent, and the coronary supply was provided by large fistulae from the RV to the distal right coronary artery, the distal LAD, and the distal left circumflex artery (Fig. 5). Other variants studied by angiography and at autopsy included one case of the right coronary artery originating from the left coronary artery, and another of both coronary arteries originating from a left-sided sinus.

Fistulae tended to occur in the smaller ventricles as judged by TV Z-score (p < 0.0001) and RV inlet Z-score (p = 0.0011). Relations with other variables are shown in Table 2. Of those patients with fistulae, the presence of stenoses, interruptions and ectasia was related to higher RV pressure recorded at catheter prior to initial procedure but no other variables (Table 2). Of 10 cases with RV dependence, 8 cases had membranous atresia and 2 had muscular atresia. In terms of ventricular structure, six cases had so-called tripartite ventricles, three bipartite, and one case was unknown. The largest TV and RV inlet Z-score documented in such a ventricle was −1.7 and +3.5, respectively.

The presence of RV-to-circumflex artery fistulae, as outlined above, was documented before intervention. Subsequent to surgery, there were examples of change in the presence or degree of fistulae. There were three cases of regression of fistulae to a normal coronary arterial arrangement following RV decompression, and two cases of RV-to-circumflex artery fistu-
lae developing stenoses. Neither of these patients had undergone RV decompression.

**Patent arterial duct, aortopulmonary collateral arteries and branch pulmonary arteries.** The angle that the arterial duct subtended with the postductal region of the aorta was obtuse (normal) in 56/112 cases (50%) (95% CI, 40.7% to 59.3%) where it could be ascertained, at right angles in 11 cases (9.8%) (95% CI, 3.95% to 14.6%) and acute in 45 cases (40.2%) (95% CI, 31.1% to 49.8%) (Fig. 6). Acute-angled ducts also tended to arise more proximally on the aortic arch than normal. The pulmonary arteries were always confluent. Aortopulmonary collateral arteries were present in three cases, one with multiple collaterals as the major source of pulmonary blood flow with miniaturized confluent pulmonary arteries and tiny left-sided duct making little contribution. In the other two cases, small collateral arteries were present, but the major source of pulmonary blood flow was via the arterial duct into the pulmonary arteries. Sixteen infants had significantly hypoplastic (<3 mm) pulmonary arteries as assessed by echocardiography and/or angiography (Table 3). The aortic arch was right-sided in three cases with a left-sided duct in one case (unknown in the other two).

**Other abnormalities.** These are documented in Table 3. Fifteen abnormalities of the left ventricle (LV), the subaortic outlet and the mitral valve were found in 11 patients. Although it is acknowledged that many cases of PAIVS have septal bowing into the LV outflow tract, and that this may represent a spectrum, there were four cases where this was extremely pronounced.
DISCUSSION

Review of the surgical literature shows that morphologic information is often scant. This hinders comparison between studies, making it difficult for the reader to determine whether the population described is representative of the population of PAIVS as a whole. Our study describes the range of morphology in an unselected population. This information can now serve as a standard for other studies to ensure that groups are representative of the population of patients with pulmonary atresia with intact septum as a whole. In this manner, it may be possible to decide whether studies with particularly high survival rates represent real advances in management, or whether the population under review is biased in favor of patients with less severe pathology.

Tricuspid valve. Our population-based morphologic data is broadly comparable with the larger nonpopulation-based studies (13,14). Using Rowlatt’s postmortem data to generate Z-scores to aid comparability (11), the median TV Z-score for this study was −1.6. It was −2.2 in the Congenital Heart Surgeons Study (CHSS) (13) and −4.8 in the study reported by Bull et al. (14). We do not, however, advocate use of normal postmortem data to generate Z-scores for echocardiographic measurements of individual patients. Shrinkage occurs to fixed autopsy material, and normal dimensions derived at autopsy will be smaller than those derived from echocardiography. Use of autopsy—rather than echocardiographically derived normal dimensions to generate Z-scores—will, therefore, lead to relatively large (less negative) median Z-scores. Because cross-sectional echocardiography is now the principal tool used for evaluation of infants with PAIVS before initial intervention, we recommend that Z-scores be calculated using normal data based on the same measurement techniques (5).

Coronary arterial abnormalities. In our study, 45.8% of patients had RV-coronary arterial fistulae, virtually identical (p = 0.90) to the proportion of 45% found in the CHSS (13). In the Toronto series, the proportion was 56% (p = 0.13 compared to our proportion) (15), but only 32% (p = 0.05) in Boston (16). Our incidence of RV-dependent coronary circulation, at 7.6%, was similar (p = 0.83) to the 9% found by the CHSS. In the Boston study (16), one or more stenoses occurred in 16/82 (19.5%) patients studied (p = 0.01). The distribution of coronary arterial fistulae was broadly similar to the findings of the Boston group (16). Unlike their study, we found stenoses predominantly in the distal right coronary artery and distal LAD. The Boston group found stenoses most commonly in the mid-LAD (16).

Fistulae tended to occur in patients with the smaller RVs as judged by TV size and RV inlet length, but did occur across the range of ventricular size, and they were present in a quarter of patients with so-called tripartite ventricles with membranous atresia (Fig. 2). Hanley et al. (13) found fistulae correlated with smaller TV Z-score, less TV incompetence, and higher RV systolic pressure. Rychik and colleagues (17) documented that patients with more severe coronary abnormalities had smaller TV Z-scores. We, in addition, showed correlation with smaller RV inlet dimension Z-score, acute-angled duct, muscular atresia, and so-called unipartite and bipartite RV. An RV-dependent coronary circulation (stenoses, interruptions or severe ectasia) correlated only with RV pressure at presentation and not with any marker of ventricular size. A recent study from Boston also failed to demonstrate a statistically significant relationship between RV-dependent coronary circulation and the degree of RV hypoplasia (18). The CHSS did demonstrate a correlation with smaller TV Z-score (13).

Absence of the coronary arterial orifices is a rare but described occurrence in PAIVS. Lenox and Briner (19) first reported a case in a two-month-old who lacked proximal aortocoronary connections with a coronary circulation entirely RV dependent. We found only a single case in our series. This infant died within 24 h of birth despite infusion of prostaglandin.
An epicardial dimple was found in one patient at surgery covering the trabecular portion of RV and close to the LAD. It has been suggested that such dimples may represent subepicardial coronary arteries (20), and may be the external stigmata of coronary artery fistulae indicating the site of such connections (3). The patient in our study was not known to have any fistulae.

**Ductal angle.** The angle that the arterial duct subtends with the postductal descending aorta has been used by some groups as an indicator of whether the pulmonary outflow became atretic earlier or later in gestation (6,21,22). In our study, the ductal angle was acute in 40.2% of cases compared to 45% in the PAIVS patients in the Kutsche et al. study (21) (Fig. 6). This tended to occur in those with smaller ventricles with muscular atresia, and ventriculocoronary arterial connections. It may be that this subgroup represents an earlier occurring lesion compared to those with well-formed ventricles, which may have progressed from pulmonary stenosis to atresia in the last trimester of pregnancy (4,23). The duct provided the main source of pulmonary blood flow in almost all our patients. Two patients had additional small aortopulmonary collateral arteries. Only in one case did multiple aortopulmonary collaterals provide practically all the pulmonary blood flow. This is uncommon, but it has been documented previously (24).

**LV and LV outflow tract.** Abnormalities of the structure of both the LV and mitral valve have been described in PAIVS (1,25,26). In a postmortem study, Akiba and Becker (26) found that in half of the cases studied, the tendinous cords of the mitral valve were dysplastic and shortened. All LVs were hypertrophied and showed an increase in interfove collagen, suggesting chronic ischemia (26). In our series, a lower incidence of mitral valvar abnormalities (1.6%) was
occurred, possibly because most did not come to postmortem examination. Concentric LV hypertrophy was present in only a single patient. Convex bulging of the LV septal surface has been described by Zuberbuhler and Anderson (1), and subsequently by Freedom (25) and Akiba and Becker (26) in patients with small and hypertensive RVs. The subaortic bulge has been reported as promoting severe LV outflow obstruction after the Fontan operation in PAIVS (27), but before this procedure it is highly unusual (17). Subaortic obstruction due to accessory TV tissue prolapsing into the LV outflow has been documented in children with raised RV pressures and septal defects such as complete transposition with ventricular septal defect (VSD), and pulmonary stenosis and VSD (28). To our knowledge subaortic stenosis has not been documented in PAIVS as a consequence of accessory TV tissue walling off a VSD and prolapsing into the LV outflow due to suprasystolic RV pressures.

Noncompactile spongiform myocardium is characterized by a trabecular network with wide Anastomosing sinusoidal blood channels giving a honey-combed, sponge-like appearance on angiography. It can occur in isolation (29,30), or in the setting of PAIVS (31). When it occurs in the presence of PAIVS, it can be present in either ventricle including in the ventricular septum, as in the case we report (Table 3). Although the inclusion of tiny ventricular septal defects

<table>
<thead>
<tr>
<th>Associated Malformation</th>
<th>Number of Cases</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ebstein's malformation</td>
<td>18</td>
<td>9.8</td>
</tr>
<tr>
<td>Hypoplastic pulmonary arteries (&lt;3 mm)</td>
<td>16</td>
<td>8.7</td>
</tr>
<tr>
<td>LV and LV outflow tract abnormality</td>
<td>15</td>
<td>8.2</td>
</tr>
<tr>
<td>Extreme septal hypertrophy with bulging into LV outflow</td>
<td>4</td>
<td>2.2</td>
</tr>
<tr>
<td>Bicuspid aortic valve</td>
<td>1</td>
<td>0.5</td>
</tr>
<tr>
<td>Fibrosed papillary muscle</td>
<td>2</td>
<td>1.1</td>
</tr>
<tr>
<td>Dysplastic mitral valve</td>
<td>1</td>
<td>0.5</td>
</tr>
<tr>
<td>Concentric LV hypertrophy</td>
<td>1</td>
<td>0.5</td>
</tr>
<tr>
<td>Spongiform noncompactile</td>
<td>1</td>
<td>0.5</td>
</tr>
<tr>
<td>Interventricular septum</td>
<td>1</td>
<td>0.5</td>
</tr>
<tr>
<td>Aneurysmal TV tissue prolapsing into LV outflow (walled off VSD)</td>
<td>1 each</td>
<td>0.5 each</td>
</tr>
<tr>
<td>Total patients</td>
<td>183</td>
<td></td>
</tr>
</tbody>
</table>
investigation of each patient. Thus, complete data was not available on all patients despite the investigators’ best efforts. For example, not all patients had a preoperative angiography, nor was an autopsy performed on all patients who died.

The United Kingdom and Ireland Collaborative Study of Pulmonary Atresia with Intact Ventricular Septum is one of the first population-based studies to provide comprehensive data on a large cohort of infants with a rare congenital cardiac malformation. Unlike disease registries, we have not relied on local investigators to forward data to the coordinating center. A single investigator visiting each center on multiple occasions ensured completeness of collection of data and allowed key diagnostic studies, such as echocardiography and angiography, to be directly reviewed and recorded for subsequent review.

Conclusions. We have described the range of morphology in a population-based study of pulmonary atresia with intact ventricular septum. This may serve as a reference source for clinical studies to judge whether their morphology is representative of the population at large.

Acknowledgments

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Reprint requests and correspondence: Dr. Piers Daubeney, Consultant Paediatric and Fetal Cardiologist, Royal Brompton Hospital, Sydney St., London SW3 6NP, United Kingdom. E-mail: p.daubeney@rbh.nthames.nhs.uk.

REFERENCES


Appendix. Distribution of Right Ventricular Coronary Artery Fistulae, Stenoses, Ectasia, Interruption and Their Relative Frequencies

<table>
<thead>
<tr>
<th></th>
<th>Right Coronary</th>
<th>Left Anterior Descending</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Proximal</td>
<td>Mid</td>
</tr>
<tr>
<td>Number of fistulae (%)</td>
<td>16 (15)</td>
<td>7 (6)</td>
</tr>
<tr>
<td>Number of stenoses</td>
<td>1</td>
<td>7</td>
</tr>
<tr>
<td>Presence of ectasia</td>
<td>5</td>
<td>5</td>
</tr>
<tr>
<td>Interruption</td>
<td>1</td>
<td>1</td>
</tr>
</tbody>
</table>

Shows numbers of fistulae, stenoses, etc., not patients. Sixty-one patients had fistulae, of which fistulous connections were accurately delineated in 42. There was one case of an right ventricular-coronary-pulmonary artery fistula (not included in this table). Total number of patients with either stenoses, ectasia or interruption of epicardial coronary arteries was 10. It was possible to delineate position accurately in 9 of 10 cases. Ectasia often included luminal irregularities.