Combined Double Chambered Right Ventricle and Discrete Subaortic Stenosis

ANTHONY BAUMSTARK, M.D., KENNETH E. FELLOWS, M.D., AND AMNON ROSENTHAL, M.D.

SUMMARY Nine cases of combined double-chambered right ventricle (DCRV) and discrete subaortic stenosis (DSAS) are presented. A review of 1077 consecutively catheterized patients indicates an association of these two uncommon anomalies nine times greater than expected. One or both obstructive lesions may be hemodynamically significant and require surgery. Whenever DCRV or DSAS is suspected or identified, the cardiac catheterization should include studies of both the right and left ventricles.

DISCRETE SUBAORTIC STENOSIS (DSAS) may occur as an isolated anomaly or in association with other left heart abnormalities such as aortic insufficiency, valvar aortic stenosis, coarctation of the aorta, patent ductus arteriosus, and mitral insufficiency. Double chambered right ventricle (DCRV), although occasionally accompanied by left-sided lesions, is also most often associated with ipsilateral cardiac defects such as pulmonary valvar stenosis and peripheral pulmonary stenosis.

We have recently studied nine patients in whom both DCRV and DSAS were present and diagnosed by cardiac catheterization. Although these two entities have each been the subject of numerous reports, attention has not been drawn to their combined occurrence. The purpose of this paper is to present the clinical, hemodynamic and angiocardiographic characteristics of these patients and to provide evidence of a statistically significant association between the two anomalies.

Method

Nine patients with combined DCRV and DSAS were hospitalized at the Children's Hospital Medical Center in Boston between 1972 and 1976. Right and left heart catheterizations and angiocardiograms were performed in all patients, surgery in seven, and postmortem examination in one (table 1).

To evaluate the statistical association between DCRV and DSAS, the cardiac catheterization reports of all patients studied at Children's Hospital Medical Center between January 1, 1975 and December 31, 1976 were reviewed. Care was taken to identify all patients with DCRV, right ventricular anomalous muscle bundles, and subaortic stenosis regardless of measured gradient. Patients with subaortic stenosis of the septal hypertrophic type were excluded from the statistical analysis summarized in table 2. Three of the nine patients with combined DCRV and DSAS have been previously reported.

Results

Clinical and Hemodynamic Data

The combination of DSAS and DCRV was not suspected before cardiac catheterization in any of the patients. The diagnosis was established in each case by pressure recordings and biplane ventriculography. In two patients (T.A. and S.C. --- table 1), the DSAS was overlooked at the first study and discovered at a subsequent cardiac catheterization.

The severity of the obstructing lesions was variable. In all but one patient (L.C., table 1), one or both of the outflow tract obstructions were hemodynamically significant at first catheterization. Preoperative right ventricular peak systolic gradients ranged between 14 and 150 mm Hg (mean 48). Left ventricular gradients varied between 15 and 100 mm Hg (mean 57). Pulmonary to systemic flow ratios (Qp/Qs) ranged from 0.8 to 3.3 in the eight patients who had ventricular septal defects (VSD).

Angiographic Findings

The anatomy of the obstructing lesions in both the right and left ventricular outflow tracts was the same as that found in isolated DCRV and DSAS. On right ventriculography, five of the nine patients had high (subinfundibular) and four had low (mid sinus) anomalous muscle bundles (fig. 1). Left ventriculography revealed three distinct forms of subaortic stenosis. A subaortic membrane (fig. 2) was present in four patients, a thick fibrous ring (fig. 3) in three patients, and a long fibromuscular segment in two patients. No association between the different types of right and left ventricular obstructions was observed.

A VSD was present in eight of the nine patients. Angiocardiographically, the VSD was proximal to the right ventricular obstruction (fig. 4), conforming to the VSD in isolated DCRV. In seven of the eight cases, the VSD on the left ventricular side was between the stenosis and the aortic valve (figs. 2, 3). In the remaining patient, the defect was in the middle of a long segment stenosis.

Surgical and Autopsy Observations

Of the seven patients who were operated upon, four had biventricular, two right ventricular, and one left ventricular repair. Of the four patients with bilateral repair, in only one (J.Z., table 1) was this accomplished at the first procedure. In the remaining three patients, the DSAS was not resected at the first operation because it was either missed at catheterization (S.C. and T.A.) or not identified surgically through an aortic incision (A.D.).

Postoperative cardiac catheterizations, obtained in four patients, showed absence of the right ventricular gradients (A.D., T.C., T.A., S.C.). One left heart catheterization has been performed following resection of the DSAS (T.A.) and no residual subaortic obstruction was present.

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TABLE 1. **Clinical, Hemodynamic, and Angiographic Data in Patients with Combined DSAS and DCRV**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age yrs-mo</th>
<th>Angiographic type of stenosis</th>
<th>Outflow tract gradient (mm Hg)</th>
<th>VSD</th>
<th>Qp/Qs</th>
<th>Associated anomalies</th>
<th>Surgical results</th>
</tr>
</thead>
<tbody>
<tr>
<td>SL</td>
<td>3-7</td>
<td>high membranous</td>
<td>35</td>
<td>+</td>
<td>1.9</td>
<td>MS, PS</td>
<td>No surgery</td>
</tr>
<tr>
<td>LC</td>
<td>2</td>
<td>high membranous</td>
<td>20</td>
<td>+</td>
<td>2</td>
<td>SI</td>
<td>No surgery</td>
</tr>
<tr>
<td>AD</td>
<td>1</td>
<td>low long segment</td>
<td>25</td>
<td>+</td>
<td>3.3</td>
<td>AR</td>
<td>DCRV and VSD repaired; DSAS not seen</td>
</tr>
<tr>
<td>TC</td>
<td>4-4</td>
<td>low membranous</td>
<td>56</td>
<td>+</td>
<td>1.6</td>
<td>None</td>
<td>DCRV and VSD repaired</td>
</tr>
<tr>
<td>SJ</td>
<td>7-5</td>
<td></td>
<td>0</td>
<td></td>
<td>0</td>
<td></td>
<td></td>
</tr>
<tr>
<td>JZ</td>
<td>2-9</td>
<td>high fibrous ring</td>
<td>150</td>
<td></td>
<td>0</td>
<td>PS</td>
<td>DCRV and DSAS repaired</td>
</tr>
<tr>
<td>DS</td>
<td>5-6</td>
<td>low fibrous ring</td>
<td>14</td>
<td>+</td>
<td>2</td>
<td>AR</td>
<td>Intra-operative death</td>
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<tr>
<td>TA</td>
<td>6-2</td>
<td>high long segment</td>
<td>40</td>
<td></td>
<td></td>
<td>PDA</td>
<td>DCRV, VSD and PDA repaired</td>
</tr>
<tr>
<td>7-2</td>
<td></td>
<td></td>
<td>0</td>
<td></td>
<td>0</td>
<td></td>
<td>DSAS resected</td>
</tr>
<tr>
<td>MG</td>
<td>14</td>
<td>low fibrous ring</td>
<td>18</td>
<td></td>
<td>1.7</td>
<td>AR</td>
<td>VSD and DCRV repaired</td>
</tr>
<tr>
<td>SC</td>
<td>12-7</td>
<td>high membranous</td>
<td>80</td>
<td>+</td>
<td>0.8</td>
<td>AR, ASD</td>
<td>DCRV and VSD repaired</td>
</tr>
<tr>
<td>28</td>
<td></td>
<td></td>
<td>0</td>
<td></td>
<td>0</td>
<td></td>
<td>ASD repaired</td>
</tr>
</tbody>
</table>

*Abbreviations: MS = mitral stenosis; PS = pulmonary stenosis; SI = situs inversus; AR = aortic regurgitation (trivial); PDA = patent ductus arteriosus; ASD = atrial septal defect.*

TABLE 2. **Incidence of Combined DCRV and DSAS**

<table>
<thead>
<tr>
<th>No. patients</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>DCRV</td>
<td>28</td>
</tr>
<tr>
<td>DSAS</td>
<td>24</td>
</tr>
<tr>
<td>Combined DCRV-DSAS</td>
<td>5</td>
</tr>
</tbody>
</table>

*Expected incidence = 0.005 x 0.022 = 0.000067; the actual incidence of 0.5% is nine times greater than the expected (P = <0.0001).*

An autopsy was performed on one patient who died intra-operatively. Examination of the left ventricle revealed a fibrous ring approximately 0.5 cm below the aortic valve. Between the aortic valve and the fibrous ring were multiple defects perforating the ventricular septum at the conal and membranous levels. On the right ventricular side, the defects communicated with a high pressure chamber created by a prominent moderator band arising high from the septal band.

**Statistical Data**

Of a total of 1077 patients consecutively catheterized at CHMC between January 1, 1975 and December 31, 1976, 24 (2.2%) were diagnosed as DSAS and 30 (2.6%) were classified as DCRV. Included in each group were five cases in which the two lesions were combined. The incidence of the combined malformations (0.5%) shown in table 2 is nine times the rate expected on a chance basis (P = 0.0001). 21

Of all patients with DSAS, 25% had right-sided lesions (five DCRV and one pulmonary valvar stenosis). Among all patients with DCRV, 28% had left-sided lesions (five DSAS; one case each of aortic regurgitation, patent ductus arteriosus, mitral valve prolapse and mitral regurgitation).

![Figure 1. Biplane right ventriculogram (patient S.C.) illustrating right ventricular anomalous muscle bundles at the subinfundibular level (small arrows) and in the mid sinus (large arrows). RV = right ventricle, PA = pulmonary artery.](image-url)
Lesions most commonly associated with DCRV are VSD (80%) and pulmonary valvar stenosis (33%).

Apart from the patients presented in this study, we are aware of only three other cases in which DCRV and DSAS were found in combination.

The relatively few previously reported patients may be explained partially by incomplete cardiac catheterization. As demonstrated in two of our patients (T.A. and S.C.), the diagnosis of DSAS was missed initially because left heart studies were not performed.

Even when right and left heart studies are performed, the association of DCRV and DSAS may be overlooked. Both
DCRV and DSAS may progress with time, and what may be considered a trivial stenosis at one catheterization may be increased in severity on follow-up study. Another source of error is the ease with which subvalvular obstructions may be mistaken for valvar disease. Despite careful pressure recording in the outflow tract, some cases will be misinterpreted when the low pressure chambers are very small. This is a particular problem in DSAS in which the obstruction may be just millimeters below the aortic valve. Ventricular angiography has been emphasized as a crucial adjunctive procedure in these difficult cases. Still, short segment subvalvular stenoses may not be visible in standard angiographic projections and their demonstration may require angled-oblique views.

Eight of the nine cases of combined DCRV and DSAS were accompanied by a ventricular septal defect. Its position, connecting the high pressure RV chamber to the low pressure LV chamber, is illustrated in figure 5. The direction of intracardiac shunting is independent of the severity of the subaortic stenosis since the VSD is distal to it and faces the systemic pressure of the subaortic chamber. The Qp/Qs ratio is determined by the size of the VSD and the degree of right ventricular outflow obstruction. If the stenosis caused by the right ventricular anomalous muscle bundles is severe enough, right to left shunting will occur.

**Etiologic Considerations**

In 1909, Sir Arthur Keith devoted his Hunterian lecture to the description of two congenital cardiac malformations: subdivision of the right ventricle and subaortic stenosis. He concluded that both conditions are caused by incomplete atrophy of the primitive bulbus cordis. Although this theory of common origin is attractive, it is based on two embryologic concepts that are no longer well supported: 1) that the aorta and pulmonary artery are normally aligned over their respective ventricles through a process of selective atrophy and fusion of the bulbus cordis with the primitive ventricle, and 2) that the bulbus cordis gives rise to only the infundibular portion of the right ventricle. More recent investigations in cardiac embryogenesis have contradicted both of these assumptions. According to Van Mierop and Odgers, the channel connecting the left ventricle to the aorta is formed by persistence of the primary interventricular foramen and not by atrophy of tissue. The bulbus cordis is now generally thought to give rise to the right ventricular sinus.

A contemporary explanation of the origin of DSAS implicates anomalous endocardial cushion tissue as the source of the obstruction. Although the etiology of DCRV is a matter of some controversy, it appears that there is now no obvious common denominator in the development of DCRV and DSAS. The statistical association found in our review is unexplained by current embryological knowledge.

**References**

18. Keith A: Fate of the bulbus cordis in the human heart. Lancet 2: 1267, 1924

**Figure 5. Schematic diagram illustrating the relationship of VSD to the intraventricular chambers in combined DCRV and DSAS. The direction of interventricular shunting through the VSD (arrow) is determined by the severity of obstruction produced by the anomalous muscle bundle within the right ventricle (RV). Solid line represents DSAS in the left ventricle (LV). Shunting is left to right until pressure within the right ventricular chamber below the anomalous bundle exceeds systemic pressure.**
Left Ventricular Anomalies
Associated with Ebstein's Malformation of the Tricuspid Valve

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SUMMARY Seventeen patients with Ebstein's malformation of the tricuspid valve have been evaluated. The majority of patients (11/17) presented in the newborn period with cyanosis and cardiomegaly. The remainder (6/17) were referred because of cardiomegaly on chest X-ray and/or a heart murmur heard on routine examination. Three patients have died; one in the newborn period and two suddenly at the age of 11 and 15 years. Thirteen patients have undergone right and left heart cardiac catheterization within the last seven years. Twelve of these 13 patients (92%) had angiocardio graphic left ventricular contraction abnormalities. Five patients also had mitral valve prolapse associated with left ventricular dysfunction. Since many patients with Ebstein's anomaly may have significant left ventricular abnormalities, careful and systematic evaluation of the left ventricle is warranted.

EBSTEIN'S MALFORMATION of the tricuspid valve (EBS) is most commonly associated with a patent foramen ovale or fossa ovalis type of atrial septal defect. Other associated lesions such as pulmonic stenosis, pulmonary atresia, ventricular septal defect, and rarely transposition of the great arteries or tetralogy of Fallot have also been described. In contrast, abnormalities of the left side of the heart have not been well documented. Over the last few years, some unusual left ventricular (LV) abnormalities have been noted on angiocardiology in patients with EBS seen at Children's Hospital of Pittsburgh. This report will describe the findings in these patients.

Materials and Methods

Over the 71/2 year period from January 1970 through June 1977, 17 patients with EBS were seen at Children's Hospital of Pittsburgh: 9 males and 8 females. Thirteen patients had cardiac catheterization during this study period while the remaining four patients had cardiac catheterization prior to this study. All patients who underwent catheterization during this period of time had evaluation of both left and right heart hemodynamics and cineangiocardiography.

The criteria for inclusion of patients in this study were the availability of adequate cardiac catheterization and cineangiocardiographic data and also current clinical information. In each case the diagnosis of EBS was established by cineangiocardiography.

Three patients died during this study period. The ages of the surviving patients ranged from 10 months to 22 years (mean age 10.8 years). In all patients the clinical records as well as previous catheterization data and cineangiocardiograms were reviewed. Autopsy specimens were available in two of the three patients who died.

Results

Clinical Data

A majority of the patients (11/17) presented in the newborn period with cyanosis and cardiomegaly. Three patients between 1 and 12 months of age and three over one year of age were referred because of cardiomegaly on chest roentgenogram and/or a heart murmur heard on routine examination.

Of the 14 patients currently being followed, nine have no symptoms and five have mild exercise intolerance. Nine patients are acyanotic, three have mild to moderate cyanosis and two have severe cyanosis (table 1). Three patients who are currently 10, 11, and 19 months of age presented as newborns. They each had clearing of their cyanosis during the newborn period and are currently doing well without symptoms or cyanosis. Two of four patients between 6 and 10 months of age have signs of cyanosis followed by clearing at 8 and 10 months of age. Three patients who had cyanosis as newborns have since cleared. The patient who presented at 1 month of age with cyanosis has cleared by age 4 months.

Three patients died during this study period. The ages of the surviving patients ranged from 10 months to 22 years (mean age 10.8 years). In all patients the clinical records as well as previous catheterization data and cineangiocardiograms were reviewed. Autopsy specimens were available in two of the three patients who died.
Combined double chambered right ventricle and discrete subaortic stenosis.
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