The Narrow Infundibulum in Pulmonary Valvular Stenosis
Its Preoperative Diagnosis by Angiocardiography

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O BSTRUCTION to the outflow of blood from the right ventricle may occur as a result of congenital stenosis of the pulmonary valve, of the infundibulum, or of the supravalvular area. These all result in a fixed constriction of the lumen of the involved region. Recently, considerable interest has been focused upon a more dynamic type of infundibular obstruction that occurs in a number of patients with valvular pulmonic stenosis and an intact ventricular septum. This narrowing becomes most prominent during systole, and is due to hypertrophy of the wall of the right ventricle in the area of its outflow tract. Although some controversy exists as to the importance of this phenomenon, there is evidence that it may occasionally become quite significant if it is not recognized preoperatively and corrected at the time of surgery on the valve.

The purpose of the present paper is to analyze the outflow tract of the right ventricle as studied by routine angiocardiography, and to establish criteria that indicate the presence of an abnormally narrowed infundibulum in patients with pulmonary valvular stenosis and an intact ventricular septum.

Literature

Kirklin et al. in 1953 first reported the occurrence of infundibular muscular hypertrophy and narrowing in cases of valvular pulmonic stenosis, and Brock2–3 discussed this phenomenon in greater detail. He suggested that following valvotomy for congenital valvular stenosis, the hypertrophied infundibulum is "decompressed" causing it to "clamp down," leading to a persistently raised right ventricular systolic pressure. Both at surgery and postmortem examination he found the crista supraventricularis and its muscular bands greatly thickened and noted considerable systolic contraction of the infundibulum at operation.

In reviewing 75 patients with pulmonary valvular stenosis who had undergone valvotomies, Johnson4 found that 58 per cent showed an infundibular gradient postoperatively. The incidence of this phenomenon increased in direct proportion to right ventricular pressure and advancing age.

Some controversy has existed as to the importance of this so-called "acquired" form of infundibular obstruction. Bing et al.,5 from examination of postmortem specimens, thought that this muscle hypertrophy led to no significant obstruction, an opinion also held by Kjellberg et al.6 Engle et al.7 reported three cases showing infundibular obstruction immediately postoperatively, but this regressed in each instance within 18 months. They deduced that surgery on the infundibulum is therefore unnecessary in such cases. Houël et al.8 recommended infundibular resection in cases of extreme hypertrophy, though they found regression complete in most cases following simple valvotomy. Other authors have shown, however, that complete regression may not always occur,4,9,10 some patients maintaining a significantly elevated right ventricular pressure several years following an adequate valvotomy. Johnson11 reports two cases in which severe functional infundibular stenosis has persisted for 2 and 5 years following an otherwise successful operation on the stenotic valve. Furthermore, it has been reported that a hypertrophied infundibulum may be a cause of sudden postoperative death due to complete outflow tract obstruction.12

In these papers the diagnosis of muscular infundibular obstruction has been based on

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cardiac catheterization data and pressure readings during operation. These recordings have shown that though a gradient at the infundibulum may be found preoperatively, more often it does not become apparent until after operation on the pulmonary valve. Johnson, for example, in his series of 75 patients discovered an infundibular gradient of over 20 mm. in only 17 per cent of patients preoperatively compared to 58 per cent following valvotomy, suggesting that “decompression” at the valvular level allowed the latent obstruction in the infundibular area to emerge.

Watson et al. have proposed the use of selective cineangiography as a means of investigating the right ventricle and its outflow tract preoperatively. In two cases of valvular stenosis, they noted a marked systolic narrowing of the infundibulum that was no brief event, but persisted during a greater part of the cardiac cycle. At least momentary dilatation during diastole was regarded as important in differentiating this systolic muscular narrowing from that due to congenital infundibular stenosis. To our knowledge, however, the use of the routine angiogram in the diagnosis of this condition heretofore has not been specifically evaluated.

Materials and Methods

Of 47 cases of congenital pulmonary valvular stenosis with an intact ventricular septum studied at the Massachusetts General Hospital during the last 10 years, adequate angiograms in rapid sequence and cardiac catheterization studies are available in 13 patients. Six of these had small atrial septal defects in addition to the valvular stenosis. Eight patients underwent valvotomies and two died postoperatively. In two additional cases, cineangiographic studies are available. Cases of congenital infundibular stenosis were excluded from the series.

All examinations were performed on an Elema rapid serial film changer with a focal film distance of 40 inches. In most examinations exposures were made at a rate of 6 frames per second.

In order to characterize any changes in infundibular contour in patients with valvular pulmonary stenosis, the angiograms of the above group were compared to those of 30 control patients having no cardiac lesion. In all patients measurements were made of the width of the infundibulum during systole as seen on the lateral view, and these were compared with the diameter of the pulmonary artery. A ratio of these two measurements was then obtained, thereby giving an infundibular measurement independent of heart size.

The pulmonary bulb was chosen as the basis for comparative measurement, because this structure was readily outlined and showed the least variation in its lateral diameter on multiple ex-

Figure 1

Normal right ventricular angiogram during systole. A, left. Anteroposterior projection. B, center. Lateral projection. C, right. Tracing of lateral projection. Note that in both anteroposterior and lateral views systolic width of the infundibulum is similar to that of the pulmonary artery. The crista supraventricularis causes a slight indentation on the posterior wall of the infundibulum in the lateral view, and infundibular width is measured at this point. Site of measurement of pulmonary bulb is also marked.

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posures. Several bulb measurements were averaged and then the ratio of the minimum systolic infundibular diameter to the average pulmonary bulb diameter was obtained. The major error inherent in this method would result from a low frame speed. In this instance one might not visualize the infundibulum during its narrowest point in systole, so that the apparent abnormality might be lessened.

Results

Normals. The normal infundibulum (fig. 1A, B, C) forms a fairly smooth channel in the lateral view (fig. 1B, C) whose width during diastole is approximately the same as that of the pulmonary artery. During systole the crista supraventricularis can be seen causing a slight indentation on the posterior margin, but the channel rarely contracts to less than one half of its maximum diameter. In the anteroposterior view the infundibulum forms a conical shape during diastole, and may contract to a width similar to the pulmonary artery during systole.

In the 30 control patients with no cardiac lesions the average ratio of the minimum systolic infundibular diameter to the pulmonary bulb was 0.63 ± 0.08 (range 0.45 to 0.83).

Pulmonary Stenosis. The maximum and minimum infundibular measurements on the 13 patients with pulmonary valvular stenosis along with the infundibulum-bulb ratios are tabulated in table 1. Also included are right ventricular pressure determinations, and finally whether angiographic or cardiac catheterization findings, or both, indicated the presence of infundibular narrowing.

Since none of the control patients had a minimum systolic infundibulum-bulb ratio of less than 0.45, a ratio of 0.4 has been chosen as critical, and below which abnormal narrowing of the infundibulum is considered present. In the present series, six of the patients with pulmonary stenosis fell into the clearly abnormal range, one was borderline and the remaining six were within normal limits (fig. 2).

The angiocardiograms of patient K.L. are showed in figure 3 and patient L.T. in figure 4. These differ from the normal (fig. 1) in that there is marked narrowing of the infundibulum during the systolic phase. The lateral views (fig. 3C, D) show that this is caused by rounded filling defects, both anteriorly and posteriorly, which represent the hypertrophic crista supraventricularis muscle and its parietal and septal bands. These bands, which enclose the infundibulum as they run obliquely downward and to the left on the septal and anterior walls of the right ventricle, can be seen on the anteroposterior view (fig. 3A, B) as a broad filling defect compressing and narrowing the infundibular channel. During diastole the infundibula in these patients relaxed to normal diameters, differentiating this phenomenon from a fixed congenital stenosis.

It will be noted from table 1 that of the
### Table 1

**Summary of Pertinent Findings in 13 Patients with Valvular Pulmonary Stenosis**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Sex</th>
<th>Age</th>
<th>Lateral infund. meas. (min.-max)</th>
<th>Ratio: minimum infund./ aver. bul</th>
<th>RV pressure</th>
<th>Infund. narrowing present</th>
<th>Assoc. cardiac abnorm.</th>
<th>Operative comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 K.L.</td>
<td>F</td>
<td>9</td>
<td>4½-16</td>
<td>0.17</td>
<td>150</td>
<td>yes yes</td>
<td>none</td>
<td></td>
</tr>
<tr>
<td>2 L.T.</td>
<td>F</td>
<td>14</td>
<td>7-25</td>
<td>0.33</td>
<td>96</td>
<td>yes yes</td>
<td>none</td>
<td></td>
</tr>
<tr>
<td>3 R.L.</td>
<td>F</td>
<td>4</td>
<td>7-18</td>
<td>0.41</td>
<td>80</td>
<td>no ±</td>
<td>none</td>
<td></td>
</tr>
<tr>
<td>4 E.S.</td>
<td>F</td>
<td>17</td>
<td>18-27</td>
<td>0.86</td>
<td>250</td>
<td>no no</td>
<td>none</td>
<td></td>
</tr>
<tr>
<td>5 R.T.</td>
<td>M</td>
<td>27</td>
<td>3-20</td>
<td>0.10</td>
<td>130 (preop.)</td>
<td>yes yes</td>
<td>none</td>
<td>Persistently elevated RV pressure postop. with infundibular gradient. However, no infundibular narrowing noted at valvotomy</td>
</tr>
<tr>
<td>6 M.H.</td>
<td>F</td>
<td>45</td>
<td>5-18</td>
<td>0.14</td>
<td>90 (1952) 175 (1960)</td>
<td>no yes</td>
<td>none</td>
<td>Infundibulum repaired. Found markedly narrowed at valvotomy</td>
</tr>
<tr>
<td>7 C.V.</td>
<td>F</td>
<td>14</td>
<td>13-27</td>
<td>0.46</td>
<td>70</td>
<td>no no</td>
<td>A.S.D.</td>
<td></td>
</tr>
<tr>
<td>8 L.C.</td>
<td>M</td>
<td>40</td>
<td>18-29</td>
<td>0.47</td>
<td>70</td>
<td>no no</td>
<td>A.S.D. (secundum)</td>
<td>Systolic infundibular contraction described at surgery</td>
</tr>
<tr>
<td>9 M.G.</td>
<td>F</td>
<td>30</td>
<td>5-22</td>
<td>0.15</td>
<td>125</td>
<td>no yes</td>
<td>none</td>
<td>Infundibular constriction not noted at valvotomy</td>
</tr>
<tr>
<td>10 P.W.</td>
<td>F</td>
<td>5</td>
<td>12-16</td>
<td>0.80</td>
<td>180</td>
<td>no no</td>
<td>A.S.D.</td>
<td>Outflow tract described as slightly hypoplastic at valvotomy. Died postop. with left ventricular failure</td>
</tr>
<tr>
<td>11 L.R.</td>
<td>F</td>
<td>37</td>
<td>20-33</td>
<td>0.59</td>
<td>180</td>
<td>no no</td>
<td>high A.S.D.</td>
<td>Outflow tract described as hypertrophied at valvotomy. Died postop.</td>
</tr>
<tr>
<td>12 A.P.</td>
<td>F</td>
<td>24</td>
<td>8-22</td>
<td>0.28</td>
<td>130</td>
<td>no yes</td>
<td>A.S.D.</td>
<td></td>
</tr>
<tr>
<td>13 S.S.</td>
<td>F</td>
<td>10</td>
<td>23.30</td>
<td>0.61</td>
<td>170</td>
<td>no no</td>
<td>? small A.S.D.</td>
<td>Infundibulum not described at valvotomy</td>
</tr>
</tbody>
</table>

In this series, there was no apparent difference in the electrocardiograms, preoperative physical findings, right ventricular systolic pressures, and ages of those patients with and without infundibular narrowing by angiogram, a gradient was found at the infundibular level at cardiac catheterization in only three.

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NARROW INFUNDIBULUM

Figure 4

(L.T. #1164007) Demonstrates systolic infundibular narrowing and diaphragm-like pulmonary valvular stenosis. No poststenotic dilatation present. As in patient K.L. (fig. 3) infundibulum dilated out to a normal diameter during diastole.

Discussion

While the existence of associated infundibular muscular hypertrophy in pulmonary valvular stenosis is well established, its importance as a complicating factor following valvotomy is still controversial. This may explain why its preoperative diagnosis has not been generally emphasized. Although this subvalvular obstruction may regress in many cases after simple valvotomy, there is good evidence that, if severe, it can become highly significant. The problems that arise are of two types: first, those occurring in the immediate postoperative period apparently resulting from acute outflow tract obstruction; and second, late effects due to persistent obstruction at the infundibular level when hypertrophy does not regress. Examples of each of these problems were present in our series.

The first is illustrated by the case of a 4-year-old girl with clinical and catheter findings of pulmonary valvular stenosis, and a right ventricular pressure of 285 mm. of mercury. A cineangiogram (fig. 5) confirmed the presence of valvular stenosis. Also present on the study, however, was evidence of marked hypertrophy of the muscular bands of the infundibulum, the importance of which was not recognized before operation. An open pulmonary valvotomy was performed, relieving a tight valvular stenosis. Very shortly after operation acute right heart failure with marked systemic hypotension, tachycardia, and lowered cardiac output occurred; and she died on the day following surgery. On post-mortem examination marked hypertrophy of the infundibular musculature was found.

In reviewing the case it seemed most likely that this sudden deteriorating course immediately after surgery was the result of a "suicidal ventricle" caused by clamping down of the hypertrophic infundibulum with almost complete systolic obstruction of the pulmonary outflow tract. This might have been prevented had it been recognized preoperatively and dealt with appropriately.

Secondly, the fact that muscular infundibular narrowing may not always regress following relief of the valvular stenosis is
illustrated by patient R.T. (table 1), a 27-year-old man first seen in 1948 with clinical and catheter findings of valvular pulmonic stenosis. Preoperative cardiac catheterization also showed a small infundibular gradient. Following valvotomy signs of pulmonary outflow tract obstruction remained, and on physical examination the systolic murmur was found lower in position. Two years postoperatively catheterization showed a persistent infundibular gradient with a right ventricular systolic pressure of 85 mm. Hg, and systolic infundibular narrowing measuring 3 mm. on the lateral view was demonstrated by angiocardiography.

This situation is contrasted by patient M.H. (table 1), in whom definite infundibular narrowing was demonstrated on a preoperative angiocardiogram (fig. 6), though this was not evident from the pressure recording. A hypertrophied infundibulum was confirmed at operation and partial resection with insertion of a Teflon patch was performed. Cardiac catheterization 6 months later revealed only a 30-mm. gradient at the infundibular level.

In this case the angiocardiographic findings alerted the cardiac surgeon to the presence of the infundibular changes associated with the valvular stenosis. Following operation on both valvular and infundibular components, a satisfactory reduction in right ventricular pressure was achieved and the postoperative course proved uneventful.

This last patient underwent cardiac catheterizations on two occasions 8 years apart. It is of interest that between these two studies there was an increase in right ventricular systolic pressure from 90 to 175 mm. Hg. It seems unlikely that the valvular component changed much during this time, and it is therefore most probable that this increase in pressure resulted from progressive infundibular narrowing. Such cases tend to substantiate the current viewpoint that this represents an acquired phenomenon associated with the valvular lesion, and has led to such descriptive terms as "secondary" or "acquired infundibular stenosis." Though it has been reported to occur more frequently in older patients and also to regress postoperatively, the only valid proof for the hypothesis that this is truly acquired would be radiologic demonstration of progressive infundibular constriction.

It is also probable, however, that simple secondary muscular hypertrophy does not wholly explain this phenomenon, and in some instances it may represent a developmental abnormality of the right ventricular outflow tract. This is suggested by its occurrence in very young patients as R.L. and P.W. (table 1), and also by the fact that a considerable group of patients exists, for example E.S., who show no changes despite long-standing right ventricular hypertension.

As for its diagnosis, it might be argued that the infundibulum may be examined directly at surgical exploration rendering previous radiologic studies unnecessary. Exact assessment of the degree of narrowing may be difficult by direct palpation, however, and in our

![Figure 6](M.H. #784464) Pulmonary valvular stenosis with marked infundibular narrowing and poststenotic dilatation. No infundibular gradient was demonstrated at cardiac catheterization.
NARROW INFUNDIBULUM

cases there has been poor correlation between the described infundibular changes at surgery and the angiographic appearance. The distribution of infundibular widths as seen in figure 2 would suggest that the degree of outflow tract hypertrophy is quite variable. Therefore, the selection of a critical ratio is necessarily to some extent arbitrary. One would not, however, expect a patient with an infundibulum-bulb ratio of over 0.4 to have clinically significant outflow tract obstruction, as this degree of narrowing may be seen in normal individuals.

A proportion of cases with significant narrowing will be diagnosed at cardiac catheterization by careful withdrawal tracings through the pulmonary outflow tract. Nevertheless, it seems that radiologic examination offers the best opportunity for preoperative diagnosis of this complicating factor in congenital pulmonary valvular stenosis.

Summary and Conclusions

In six of 13 patients with pulmonary valvular stenosis and an intact ventricular septum studied by angiocardiography at this hospital, significant infundibular narrowing was present. This is due to muscular hypertrophy and systolic contraction of the crista supraventricularis and its parietal and septal bands.

An angiocardiographic infundibular measurement has been established utilizing the ratio of the minimum systolic diameter of the infundibulum on the lateral view to the average pulmonary bulb diameter.

In 30 individuals with no heart disease, this ratio averaged 0.63 ± 0.08 and was never less than 0.45.

In six patients with pulmonary valvular stenosis the infundibulum-bulb ratio ranged from 0.10 to 0.33, indicating abnormal systolic infundibular narrowing. In three of these patients, this narrowing was not evident on pressure tracings obtained at cardiac catheterization.

Cases are described indicating that this phenomenon may be clinically important if not corrected at the time of valvotomy, leading to sudden outflow tract obstruction immediately postoperatively, or to a persistent infundibular gradient several years following valvotomy.

Routine angiocardiography offers an excellent means for the preoperative detection of an abnormally narrowed infundibulum in patients with pulmonary valvular stenosis.

References

The Narrow Infundibulum in Pulmonary Valvular Stenosis: Its Preoperative Diagnosis by Angiocardiography

JOHN B. LITTLE, J. PETER LAVENDER and ROMAN W. DESANCTIS

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