Right Ventricular Growth in a Case of Pulmonic Stenosis with Intact Ventricular Septum and Hypoplastic Right Ventricle

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SUMMARY Adequate growth of the hypoplastic right ventricle in a patient with severe pulmonary stenosis with an intact ventricular septum was documented after pulmonary valvotomy in infancy. It is postulated that the growth of the ventricular chamber is largely the result of pulmonary regurgitation resulting from successful pulmonary valvotomy. Based on this and the observations of others on the growth of the hypoplastic right ventricle in pulmonary atresia (with intact septum) cases, an organized approach to eventual total surgical correction is recommended.

SIGNIFICANT PULMONARY STENOSIS with an intact ventricular septum is usually associated with a hypertrophied right ventricle. Only rarely is this lesion associated with a hypoplastic right ventricle. The long-term prognosis in these cases, and in cases with pulmonary atresia with an intact ventricular septum and hypoplastic right ventricle, depends upon initial successful palliation and ultimate growth of the right ventricular cavity to a reasonable size, so as to handle an adequate cardiac output. The purpose of this communication is to illustrate the growth of the hypoplastic right ventricle in a case of severe pulmonary stenosis with an intact ventricular septum, to discuss its therapeutic implications in the management of hypoplastic right ventricle (with pulmonary stenosis or atresia), and to comment on an interesting physiologic finding in this case, namely interatrial right-to-left shunting in the presence of a normal right ventricular pressure.

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FIGURE 1  Posteroanterior view of the chest X-ray (A) obtained at one year of age (at the time of first catheterization) shows normal cardiac size with decreased pulmonary vascular markings. The chest X-ray taken at third catheterization (B) shows minimally enlarged heart with prominence of the proximal pulmonary arteries (the main pulmonary artery cannot be clearly identified).

FIGURE 2  Standard electrocardiograms (ECGs) of the patient obtained at ages 1 (1), 4½ (2), 10½ (3) and 13½ (4) years. All but the leads marked '½' were obtained at full standardization.

The first ECG reveals a mean frontal plane vector of +40° with counterclockwise loop, right atrial enlargement and left ventricular hypertrophy. Right ventricular hypertrophy may be suggested because of upright T waves in the right chest leads. The ECG at 4½ years (2) is essentially unchanged from the first except for possible additional left atrial enlargement. The third ECG continues to show right atrial enlargement, possible left atrial enlargement and left ventricular hypertrophy. The ECG at the last clinic visit (4) remains essentially unchanged except for more pronounced ST-T wave changes in the left chest leads.

moderate cyanosis, a large "a" wave in the jugular venous pulse, a single second sound of normal intensity, and a grade II/VI systolic ejection murmur at the upper sternal border. Chest X-ray showed moderate cardiomegaly and prominent proximal pulmonary arteries. The electrocardiogram (fig. 2-2) showed right atrial enlargement and left ventricular hypertrophy. Cardiac catheterization (table 1) showed an increase in right ventricular chamber size (fig. 3C), normal right ventricular pressure, no tricuspid stenosis, and persistence of the right-to-left shunt at the atrial level.

He was followed clinically to ten years of age when he had an increasing hematocrit. Physical examination showed severe cyanosis and clubbing. There was a prominent left ventricular impulse. S1 was normal. S2 was loudest at the upper left sternal border, of normal intensity, and single. There was a grade II/VI systolic ejection murmur at the upper left sternal border. There was no diastolic murmur. Chest X-ray (fig. 1B) showed mild cardiomegaly and increased proximal pulmonary arteries. The electrocardiogram was unchanged (fig. 2-3). Cardiac catheterization (table 1) showed normal right ventricular and pulmonary artery systolic pressures,
with no significant gradient across the pulmonic valve. The pressure tracings in the right ventricle and pulmonary artery were similar, suggesting that they were acting as a single chamber. There was persistence of the right-to-left shunt at the atrial level. A right ventricular cineangiogram revealed an increase in right ventricular chamber size (fig. 3D) in comparison to the study at one year. (Growth of the right ventricular chamber since four and one-half years could not be evaluated because of different magnifications in the cineangiograms.) There was no evidence of tricuspid stenosis. Free pulmonary regurgitation was noted in a separate pulmonary arterial cineangiogram. There was good transmission of the "a" wave into the right ventricular tracing.

The patient underwent surgical closure of the atrial septal defect. In the immediate postoperative period he developed signs of mild congestive heart failure, which improved with medical management. Subsequent follow-up for three and one-half years after the surgery has shown improved exercise tolerance and good physical development without cyanosis. A striking abnormality of his cardiac examination remains that of a jugular venous "a" wave. The electrocardiogram (fig. 2-4) was essentially unchanged.

<table>
<thead>
<tr>
<th>Table 1. Cardiac Catheterization Data</th>
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<tr>
<td>Age (yr)</td>
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<tr>
<td>Site</td>
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<tr>
<td>Superior vena cava</td>
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<td>Right atrium</td>
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<td>Right ventricle</td>
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<td>Left ventricle</td>
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Discussion

Hypoplasia of the right ventricle in association with pulmonic stenosis and an intact ventricular septum is a well documented clinical and pathological entity.\textsuperscript{1-4} The case presented belongs to this group. There is no existing classification for pulmonic stenosis with intact ventricular septum, but when a classification similar to that used for pulmonary atresia with intact ventricular septum\textsuperscript{7} is applied, the present case could be grouped with Type I pulmonary atresia.

At the initial cardiac catheterization and angiography the size of the right ventricular cavity was clearly small and hypoplastic (figs. 3A and B). Subsequent to pulmonary valvotomy, the right ventricular cavity increased in size (figs. 3C and D), so that eventually it was decided that this ventricle could handle the entire cardiac output into the pulmonary circuit. Hence, the atrial defect was surgically closed to prevent right-to-left shunting at the atrial level. After a brief initial period of cardiac decompensation, the right ventricle maintained an effective pulmonary output. It might be worthwhile to point out that the size of the tricuspid orifice increased in proportion to the size of the right ventricular cavity (fig. 3D).

The factors causing the enlargement or dilatation of the right ventricular cavity are not entirely clear. The clinical experience suggests that pressure overload (e.g., pulmonic and aortic stenosis) increases the total ventricular muscle mass without increasing the size of the cavity of the ventricle. Indeed, the cavity size may even be reduced. In contrast to this, volume overload (e.g., mitral and aortic insufficiency) is definitely associated with increase not only of the ventricular muscle mass but also of the size of the ventricular cavity. This might lead one to suspect that the free pulmonary regurgitation, which was created by isolated pulmonary valvotomy in this case, was responsible for increasing the size of the cavity of the right ventricle. A similar increase in the size of the cavity of the right ventricle has been reported to be produced by pulmonary valvotomy in patients with pulmonary atresia and an intact ventricular septum.\textsuperscript{6,7} Bowman et al.\textsuperscript{7} suggested that forward decompression of the right ventricle is responsible for the enlargement of the chamber. Several patients in an older age range have been reported, in whom the right ventricular size remained small, despite years of forward flow across the stenotic pulmonic valve.\textsuperscript{1,3,4} Therefore, it is unlikely that simple forward decompression is responsible for the increase in the size of the right ventricular cavity.

Hypoplasia of the right ventricle occurs with pulmonary atresia or pulmonary stenosis or it may be an isolated anomaly.\textsuperscript{9,12} It is most common with pulmonary atresia. Although the following discussion on the therapeutic approach to the hypoplastic right ventricle may pertain to all three groups, it mainly applies to pulmonary atresia with intact ventricular septum. The prognosis in cases of pulmonary atresia with intact ventricular septum is poor with or without conventional surgery.\textsuperscript{9,10,11} Therefore, a comprehensive program of medical and/or surgical treatment to eventually result in complete surgical correction of the lesion should be planned for these patients. The objectives of such a program are: 1) to relieve hypoxia and acidosis by a timely and appropriate procedure to increase pulmonary blood flow at the time of initial presentation (usually in the newborn period); 2) to facilitate adequate egress of blood from the right atrium; and 3) to stimulate growth of the right ventricle so that it can eventually handle the cardiac output into the pulmonary circuit or to perform an operation to bypass the right ventricle. To attain this goal several therapeutic alternatives appear to exist.

A. Pulmonary Valvotomy: Pulmonary valvotomy alone at the time of initial presentation in the neonatal period has been advocated by several groups of workers. However, pulmonary valvotomy alone often resulted in death.\textsuperscript{7,16} The reason for this might be that the right ventricular cavity is too small and noncompliant to support the pulmonary circuit. Therefore, only patients with normal or large right ventricular cavities with a short segment of atresia\textsuperscript{7} should undergo pulmonary valvotomy. It should be noted that isolated pulmonary valvotomy may not produce good results even in patients with what appears to be an adequate-sized right ventricular cavity. For this reason, despite a good result in the patient presented in this report, the authors do not recommend valvotomy alone as the surgical procedure of choice.

B. Staged Surgical Correction Utilizing the Right Ventricle: As mentioned above, the prognosis of patients with pulmonary atresia (with intact ventricular septum) is poor with or without palliative surgery.\textsuperscript{3,13-15,17} The conventional palliative surgery, consisting of a systemic-to-pulmonary artery anastomosis, was only successful for moderately long-term palliation. A combination of the shunt (along with ligation of the patent ductus arteriosus) and adequate decompression of the right atrium (by balloon atrioseptostomy or surgical atrioseptostomy) appeared to markedly improve the survival rate.\textsuperscript{18} Even with this regimen the ultimate outcome was poor because total correction was not feasible due to the small size of the right ventricular cavity. Recent reports\textsuperscript{6,8} suggested that the right ventricular cavity size could be stimulated to increase by additional pulmonary valvotomy either at the time of initial systemic-pulmonary arterial anastomosis\textsuperscript{7} or at a subsequent time.\textsuperscript{8} (Our case adds further support to this hypothesis.) These patients subsequently had corrective surgery.\textsuperscript{7,8} Based on these studies, it is recommended that all cases of pulmonary atresia with an intact ventricular septum and hypoplastic right ventricle have 1) initial systemic-to-pulmonary artery shunt and atrioseptostomy (balloon or surgical),\textsuperscript{7,8} 2) pulmonary valvotomy in the first few months of life\textsuperscript{6} (or at the time of shunt and atrioseptostomy if possible),\textsuperscript{7} and 3) resection and reconstruction of the right ventricular infundibulum and pulmonary valve along with the closure of the atrial septal defect at the age of five to ten years.

1) The need for combining atrioseptostomy with a surgical shunt for treating this lesion is amply documented by the study of Shams et al.\textsuperscript{19} They showed from their experience with a large group of patients that isolated valvotomy or a shunt procedure uniformly resulted in death; the only survivors were from the group that had both the shunt and atrioseptostomy. Several types of systemic-to-pulmonary shunt operations are currently available. These are Blalock-Taussig operation (subclavian artery-to-
pulmonary artery), Potts anastomosis (descending aorta-to-left pulmonary artery), Waterston shunt (ascending aorta-to-right pulmonary artery), and Glenn procedure (superior vena cava-to-right pulmonary artery). As mentioned previously, one of these operations has to be performed very early in life, most usually in the neonatal period. The Glenn operation may not be successful in increasing pulmonary blood flow in the neonatal period and even up to the age of six months. Most groups of workers had either prohibitive mortality or superior vena caval obstruction. These are presumably related to the elevated pulmonary vascular resistance in early life and/or the small size of the right pulmonary artery. Additionally, it is a destructive operation and may not be advisable when the aim is ultimate total surgical correction. Furthermore, late complications of the Glenn operation are rather significant. Potts or Waterston anastomoses have a tendency to produce elevated pulmonary arterial pressure (and consequently a higher chance of developing increased pulmonary vascular resistance and even pulmonary vascular obstructive disease) or congestive heart failure apart from other reported complications. Therefore, the Blalock-Taussig operation, where the size of the shunt is limited by the size of the subclavian artery, is probably the palliative shunt of choice. It should be mentioned, however, that this operation is technically difficult to perform in the neonatal period. But, recent experience (unpublished reports) suggests that this operation is increasingly used in neonates and that it is successful.

In regards to the atrioseptostomy, Rashkind’s balloon atrioseptostomy at the time of diagnostic catheterization is preferable. This will avoid the surgical septostomy and thus shorten the duration of the operation and possibly decrease the mortality and morbidity of the shunt procedure. However, critically ill infants may have to be taken to surgery urgently to relieve hypoxia and the balloon atrioseptostomy may not be possible; in such cases surgical septostomy is indicated.

2) It appears that the data from the literature support the contention that right ventricular growth can be encouraged by pulmonary valvotomy. Our case further supports this hypothesis. Valvotomy can be done at the time of the shunt and atrioseptostomy, as recommended by Bowman and associates, if the infant’s status permits and if there is good exposure of the area concerned. Otherwise, it could be performed a few months after the surgical shunt and atrioseptostomy.

3) The repair of the pulmonary valve and right ventricular infundibulum along with closure of the interatrial communication is the next logical step in the process of obtaining normal cardiovascular status. The age at which this procedure should be done varies depending on the degree of polycythemia and exercise limitation. In most cases this could be around 5-10 years. Balloon occlusion of the atrial defect prior to surgical correction may be used to test if the right ventricle could handle the pulmonary output, although such testing may not always be technically successful.

Thus, a comprehensive approach to this lesion may be required for eventual correction. It is likely that even with this regimen, the severely hypoplastic ventricle will still not grow.

C. Staged Surgical Correction by Right Ventricular Bypass: For the cases not suitable for either primary pulmonary valvotomy or staged surgical correction utilizing the right ventricle, right ventricular bypass operations like the Fontan procedure may be performed. Fontan and Baudet devised this operation to physiologically correct tricuspid atresia. It consists of creation of superior vena cava-to-right pulmonary artery shunt (Glenn), anastomosis of the right atrium to the main pulmonary artery by means of an aortic valve homograft, insertion of a pulmonary valve homograft into the inferior vena caval orifice, and closure of the atrial septal defect. Either a deliberate Glenn type of anastomosis was performed at the time of surgical repair of tricuspid atresia or the patients had a prior Glenn anastomosis performed for palliation. The results of the Fontan operation without a Glenn shunt at the time of or prior to the Fontan operation are encouraging and therefore, it appears that a Glenn anastomosis may not be a necessary prerequisite to the procedure. If the right ventricle fails to grow after the regimen outlined in B, the Fontan operation, which can be successfully performed at or after eight years of age, can be attempted.

In the usual case of severe pulmonic stenosis, there is increased right ventricular systolic pressure proportional to the degree of obstruction. Consequently the right ventricle hypertrophies. The thickened right ventricle is less compliant and offers increased resistance to diastolic filling. This is usually associated with elevated right ventricular end-diastolic pressure. The factor other than the right ventricular compliance that determines the degree of right-to-left shunting is the size of the interatrial communication itself. Since our case, at ten years of age, had normal systolic pressures in the right ventricle, the presence of right-to-left shunting via the patent foramen ovale is interesting. The exact reason for this is not clear, but it might be speculated that the degree of decrease of right ventricular compliance is such that it does produce right-to-left shunting (by increased resistance to diastolic filling) but the compliance is not sufficiently low to cause elevation in the right ventricular end-diastolic pressure. Though the pressures in both atria are normal by themselves, their interrelationships are abnormal in that the right atrial pressures are higher than the left atrial pressures. We feel that this pressure difference can be explained at least in part by decreased right ventricular compliance.

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