Isolated Right Ventricular Hypoplasia

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SUMMARY

Hypoplasia of the right ventricle, unassociated with severe pulmonary or tricuspid valvar malformations, is a primary congenital abnormality in which the trabeculated sinus portion of the ventricle fails to develop. An atrial septal defect or a stretched foramen ovale serves as an escape valve. The clinical, hemodynamic, and angiocardiographic features of this rare type of cyanotic heart disease are described in two children and reviewed in 13 previously reported cases. The clinical spectrum varies considerably in accordance with the degree of hypoplasia. Severe forms of hypoplasia, usually presenting in infancy, closely resemble tricuspid or pulmonary atresia. The electrocardiogram in hypoplasia, which shows right atrial enlargement and absence of right ventricular electrical forces, may be indistinguishable from an electrocardiogram in atresia. Lesser degrees of hypoplasia should be differentiated from Ebstein's malformation and anomalies of the systemic venous return. Both our patients, who had only mild underdevelopment of the right ventricle, markedly improved after surgical closure of the atrial septal defect. It is proposed that in mild forms of hypoplasia temporary occlusion of the atrial septal defect by means of a balloon catheter should be used in the preoperative assessment of right ventricular function.

Additional Indexing Words:
Balloon catheter Cyanotic congenital heart disease Ebstein's anomaly
Familial incidence of congenital heart disease Giant a wave Pulmonary atresia
Tricuspid atresia Uhl's syndrome

In 1959 Gasul and coworkers gave a lucid account of a 4-year-old mildly cyanotic girl, thought to have Ebstein's anomaly, in whom at surgery hypoplasia of the right ventricle and an atrial septal defect were found to be the sole lesions.1 The cavity of the right ventricle was noted to be markedly reduced and its walls unusually smooth due to the absence of the trabecular musculature. The tricuspid and pulmonary valves were normal. Twelve similar cases have been described since.2-10 In only half of them were hemodynamic and angiocardiographic studies performed. At cardiac catheterization the main finding is a right-to-left or a bidirectional atrial shunt in the absence of Ebstein's anomaly, tricuspid stenosis, pulmonary stenosis, or pulmonary hypertension. The angiocardiographic findings are characteristic and will be discussed in detail.

From a survey of the literature and the two cases reported here, it appears that this rare type of cyanotic congenital heart disease presents a wide range of severity in accordance with the degree of underdevelopment of the right ventricle. Both children presented in this study belong to a subgroup with mild hypoplasia and relatively adequate right ventricular function. As was demonstrated in this series, such patients may greatly benefit from the surgical closure of their atrial septal defect and abolition of the right-to-left shunt.

Case 1

A 14-year-old boy was referred to the University Clinic of Leuven for evaluation of cyanosis.
Figure 1

Jugular venous pulse recording (middle line) and phonocardiogram at the second left intercostal space (top). A dominant a wave is followed by a deep x descent. In contrast the y trough is shallow due to the limited filling capacity of the right ventricle. The second sound is single.

and a cardiac murmur. He was born after a full-term pregnancy. Birth weight was 3.400 gm. The parents and seven siblings were in good health. According to the family doctor the patient was deeply cyanosed during the first 2 weeks of life. Later on the cyanosis decreased and, for several years, was only visible on exertion. More recently the boy had become cyanotic at rest and complained of dyspnea on effort.

On examination the patient was well developed. Slight cyanosis and digital clubbing were noted. The blood pressure was 120/80. Jugular venous pressure was increased with a waves of approximately 12 cm. This was confirmed by the external jugular venous tracing which showed a dominant a wave and a deep x descent (fig. 1). A hyperdynamic left ventricular impulse was felt well within the midclavicular line. A faint, grade 1/6 early systolic murmur and a loud and single second sound at the pulmonary area were heard. The first sound was normal. Most observers considered this auscultation to be normal and hardly compatible with the diagnosis of cyanotic heart disease.

The electrocardiogram (fig. 2) demonstrated normal sinus rhythm, a QRS duration of 0.08 sec, and a mean QRS axis in the frontal plane of +10 degrees. The P waves were tall (0 and 4 mv) and slightly notched in lead I and peaked in V2 and V3. Deep S waves in V1, high R waves, and depression of the S-T segment in V5 and V6 were interpreted as evidence of left ventricular hypertrophy. The chest X-ray showed questionable cardiomegaly and normal vascular markings. It was felt, at this stage, that the more common types of cyanotic heart disease could be ruled out. Some unusual form of tricuspid atresia or an anomaly of the systemic venous return with drainage into the left atrium seemed the most likely diagnosis.

At cardiac catheterization (table 1) performed from the right saphenous vein, the catheter

Figure 2

Case 1. The electrocardiogram shows a mean QRS axis in the frontal plane of +10 degrees, tall and peaked P waves in leads I, V2, and V3, and left ventricular hypertrophy in the precordial leads.

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Table 1

Hemodynamic Data

<table>
<thead>
<tr>
<th>Case 1</th>
<th>Case 2*</th>
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<tbody>
<tr>
<td>Pressures (mm Hg)</td>
<td>Pressures (mm Hg)</td>
</tr>
<tr>
<td>S</td>
<td>D</td>
</tr>
<tr>
<td>Inferior vena cava</td>
<td></td>
</tr>
<tr>
<td>Superior vena cava</td>
<td></td>
</tr>
<tr>
<td>Right atrium</td>
<td>a = 13–16</td>
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<tr>
<td>Right ventricle</td>
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</tr>
<tr>
<td>Pulmonary artery</td>
<td>20</td>
</tr>
<tr>
<td>Left atrium</td>
<td>a = 12–14</td>
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<tr>
<td>Pulmonary vein</td>
<td>98, 98</td>
</tr>
<tr>
<td>Left ventricle</td>
<td>110</td>
</tr>
<tr>
<td>Systemic artery</td>
<td>88</td>
</tr>
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</table>

*In case 2, O₂ consumption at rest was 228 ml/min, pulmonary blood flow was 5.1 liters/min, systemic blood flow was 5.2 liters/min, left-to-right shunt was 1.7 liters/min, and right-to-left shunt was 1.8 liters/min.

Abbreviations: S = systolic; D = diastolic; M = mean; a = atrial.

passed repeatedly from the right into the left atrium, from which all four pulmonary veins and the left ventricle could be entered. It was more difficult to advance the catheter from the right atrium into the right ventricle and the pulmonary artery. Oxygen saturations demonstrated a right-to-left shunt at atrial level, resulting in a desaturation in the left ventricle (89%) and systemic artery (88%). This was confirmed by dye-dilution curves which showed a significant right-to-left shunt following an injection into the venae cavae and right atrium. The most striking finding on the pressure curves was a high right atrial a wave (14 to 16 mm Hg) which obviously was transmitted to the right ventricle and even up into the pulmonary artery (fig. 3). The early diastolic pressure in the right ventricle was around 8 mm Hg and the end-diastolic pressure, just preceding the superimposition of the atrial a wave, averaged 10 mm Hg. In the left atrium the a waves were of equal amplitude but left atrial mean pressure was slightly lower than in the right atrium.

Angiocardiograms (fig. 4) from the right atrium showed this chamber to be dilated with large atrial systolic and diastolic volume changes. The right ventricle had an unusual appearance. Its cavity was medially situated, small, and consisted only of an inflow portion and a wide infundibulum. Contraction had little influence on the right ventricular volume. Due to the absence of trabeculae the left border of the right ventricle, as visualized in the anteroposterior incidence, was smooth and went straight up to the infundibulum and the main pulmonary artery. The latter was well developed and gave rise to normal branches. Simultaneously with the right ventricle, the left atrium was faintly opacified. It was thought that the patient had hypoplasia of the right ventricle with a right-to-left shunt.

Figure 3

Case 1. Pressure recordings demonstrate elevated atrial a waves (14 to 16 mm Hg) which are transmitted to the right ventricle and pulmonary artery.
through an atrial septal defect or stretched foramen ovale.

In view of the underdevelopment of the right ventricle one was hesitant to propose surgical closure of the atrial septal defect without further evidence that the right atrium and ventricle would be able to cope with the increased volume load. Therefore a second catheterization was carried out with the aim of occluding temporarily the atrial septal defect. A Fogarty balloon catheter was introduced into the saphenous vein and advanced into the left atrium. In this position the balloon was inflated with increasingly larger volumes of contrast medium until it could no longer be withdrawn into the right atrium. By gently pulling the catheter, the balloon was then kept snug against the rim of the septal defect for periods of 15 min. In the meantime right atrial and ventricular pressures were monitored through another catheter.

After a few minutes of occlusion, pressures stabilized at only slightly higher levels: the right ventricular systolic pressure increased from 20 to 23 mm Hg whereas the right atrial a waves, transmitted to the ventricle, went up from 11 to 14 mm Hg. The mean right atrial pressure was not significantly influenced. After deflating the balloon, pressures went back to control levels within a few seconds (fig. 5). The procedure was twice repeated with identical results. Cardiac output or stroke volume was not determined.

The experiment was thought to indicate that the right atrium and ventricle, by an increase in their stroke volume and systolic pressure, were able to handle the extra load represented by the volume of the right-to-left shunt, without compromising the central venous pressure.

At surgery the right ventricle was seen to be small. The anterior aspect of the heart was for the largest part made up of the left ventricle. On opening the dilated right atrium, its wall looked hypertrophied and grossly trabeculated. An atrial septal defect with a diameter of 2 cm, situated in the region of the foramen ovale, was closed by uninterrupted sutures. The tricuspid valve was normal. The postoperative course was uneventful.

Eighteen months after the operation, when the patient was examined at the follow-up clinic, he was completely symptomless and acyanotic. A dominant jugular venous a wave was the only pathological finding. The auscultation and electrocardiogram had not changed.

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**Figure 4**

*Angiocardiogram showing a dilated right atrium and the characteristic appearance of the right ventricle which has a small and medially situated cavity. Its left border is smooth due to the absence of the trabecular portion.*

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**Figure 5**

*Right ventricular pressure (A) at the end of a period of 15 min during which the atrial septal defect was occluded by means of a balloon catheter and (B) following deflation of the balloon. It will be noted that the a waves, transmitted from the right atrium, and the right ventricular systolic pressure, are slightly higher during occlusion. For further comments see text.*
Case 2

This girl was first admitted to the University Clinic of Uppsala at the age of 13 years. Her parents and two siblings were healthy. Growth and development were normal. Cyanosis had been noted since birth. For the last few years the patient complained of exertional dyspnea and palpitations. The physical examination revealed slight cyanosis and moderate clubbing. The cardiac impulse at the lower left sternal border was forceful. On auscultation a soft, grade 1/6 early systolic murmur and a fixed splitting of the second sound were heard. A faint diastolic murmur was audible parasternally at the fourth left interspace. With the exception of cyanosis these findings seemed compatible with an atrial septal defect. The electrocardiogram (fig. 6) showed a mean QRS axis in the frontal plane of −30 degrees and a QRS duration of 0.08 sec. The P wave in lead II was notched but of normal amplitude. There was no indication of either right or left ventricular hypertrophy. A slight intraventricular conduction disturbance was noted over the apex region. On the chest X-ray the cardiac mass was normal (360 ml/m² body surface) and its configuration unremarkable.

Cardiac catheterization (table 1) demonstrated normal pressures. There was a small systolic gradient across the pulmonary valve. Right and left atrial pressures were equal. The right atrial a waves were within normal limits. The main finding was a bidirectional shunt at atrial level. The arterial saturation was 87%. During a working capacity test (6 min at 200 kpm/min) the O₂ consumption increased to 641 ml/min and pulmonary flow to 13.2 liters/min. Right atrial pressures remained unchanged. The arterial saturation dropped to 80%. It was not clear, at this point, why the atrial septal defect, in the absence of pulmonary hypertension, tricuspid or pulmonary stenosis, permitted a right-to-left shunt. In order to evaluate whether surgical closure was advisable or not, we tried to temporarily occlude the atrial septal defect. A balloon catheter, positioned in the left atrium, was filled with 4 ml of contrast medium and pulled snug against the rim of the septal defect. During occlusion the arterial saturation rose to 94% and the left-to-right shunt disappeared. Right atrial pressure did not change.

Biplane angiocardiography with injection into the right atrium showed a dilated right atrium and a small right ventricular cavity. The infundibulum was abnormally wide but the trabecular zone of the right ventricle was lacking. The pulmonary artery was normal. Early opacification of the left atrium and left ventricle produced superimposition and made it more difficult to delineate the border of the right ventricle than in the first case. A second injection, into the inferior vena cava, showed that this vein joined the right atrium more cranially than normal and that the hepatic veins entered the vena cava above the diaphragm.

At surgery a large right atrium and a small right ventricle were seen. The tricuspid valve was normal. A longitudinal secundum type atrial septal defect (2 × 5 cm) was closed by two rows of continuous sutures. The convalescence was uneventful. At the last follow-up examination, 3 years after the operation, the patient was asymptomatic. The auscultatory findings were normal. On the electrocardiogram the intraventricular conduction disturbance had disappeared.

Discussion

Right ventricular hypoplasia is most commonly found in association with severe obstructive valvar malformations. Underdevelopment of the inflow tract and conus of the right ventricle is a striking anatomical feature in tricuspid atresia. In the majority of patients with pulmonary atresia or severe pulmonary stenosis the wall of the right ventricle is thick.
but its cavity diminutive. That the smallness of the right ventricle in these conditions is secondary to poor function and diminished output is shown by the exceptions. Some patients with tricuspid atresia and a large ventricular septal defect have a nearly normal-sized right ventricle. Similarly, pulmonary atresia may be associated with a large ventricular cavity if egress of blood from the right ventricle is possible due to tricuspid insufficiency or large intramyocardial sinusoids.

In contrast to these forms of secondary hypoplasia, underdevelopment of the right ventricle has been described in association with other cardiac malformations without being their direct consequence. At present there is no valid embryological or functional explanation for the right ventricular hypoplasia found in some patients with a large ventricular septal defect or transposition of the great arteries. In these conditions the hypoplasia will only modify the clinical syndrome which is dominated by the major malformation.

A third group of reports deals with isolated right ventricular hypoplasia unassociated with any cardiac anomalies other than a patent foramen ovale or an atrial septal defect. In these instances the underdevelopment of the ventricle may be considered to be a primary and isolated developmental abnormality. Best known, in view of the striking pathological findings, is Uhl's syndrome, characterized by an extremely thin-walled right ventricle (parchment right ventricle) with a normal or dilated cavity. Froment and coworkers recently reported five cases and reviewed the subject extensively. The two cases described in the present paper belong to this group of isolated right ventricular hypoplasia but their anatomical features were quite different from Uhl's syndrome. Indeed, the right ventricle had a normal wall thickness but a small cavity due to the absence of its sinus portion. Only 13 well-documented cases with similar findings were reported in the literature.

A familial tendency has been noted in seven of the 15 patients. Six of them were three pairs of siblings. The daughter of one of the siblings reported by Sackner also had the same malformation. There is an even sex distribution. The time of onset and the severity of symptoms depend on the degree of hypoplasia. Congestive heart failure and cyanosis were encountered in six infants, four of whom succumbed within the first weeks of life, and in two young adults. Less severe cases present with cyanosis, clubbing, and exertional dyspnea. A striking murmur was heard in only three of the 15 patients. In a case of Sackner's it was probably due to associated mild calcific aortic stenosis. In another child it was supposed to be due to a ventricular septal defect, the presence of which, however, was not proved. In the third patient the murmur remained unexplained. In four severely cyanosed infants, no murmur was heard. In the remaining eight patients, including the two children of the present series, a faint ejection murmur was noted. The second sound has been reported to be single or normally split. Fixed splitting of the second sound, which suggested the presence of an atrial septal defect, was heard and registered in our second patient.

Little attention has been paid to the jugular or hepatic venous pulsations. Distention of the neck veins or a pulsating liver have been mentioned in cases with congestive heart failure. If carefully looked for, one would expect to see more patients with dominant or giant a waves, as observed in one of our patients (fig. 1) who was not in congestive failure.

The electrocardiogram may give important clues. Prolongation of the atrioventricular conduction is common and was noted in six of the 15 patients. Right atrial hypertrophy was found in all, with the exception of three patients; two of these had left atrial enlargement and one bialtrial hypertrophy. Eight patients, including our second patient, had left axis deviation, the mean QRS axis in the frontal plane being situated between −10 and −50 degrees. Seven had an axis between +10 and +110 degrees. Direct evidence of left ventricular hypertrophy was
seen in nine patients,\textsuperscript{1, 2, 4, 5, 6, 10} In four additional patients, all neonates,\textsuperscript{3, 7, 8} there was conspicuous absence of right ventricular electrical forces, which was considered to be good evidence of left ventricular preponderance. In our second patient and in one other child\textsuperscript{9} no left or right ventricular hypertrophy was seen. From the diagnostic viewpoint it is important to note that a combination of right atrial enlargement, left axis deviation, and left ventricular hypertrophy, leading to the suspicion of tricuspid stenosis, was encountered in six patients, all of whom had a severe degree of hypoplasia.\textsuperscript{1, 2, 3, 6} The roentgenogram of the heart is of little diagnostic help. The cardiac silhouette is usually normal or slightly enlarged. Diminished vascular markings may be present in the more severe forms with a large right-to-left atrial shunt.

The results of complete cardiac catheterization were reported in eight patients, including the present series.\textsuperscript{1, 2, 6, 9, 10} In five instances an increased right atrial pressure and a dominant or giant a wave were found. An elevation of the early and end-diastolic pressures in the right ventricle reflects its diminished filling capacity. Typical pressure curves were recorded in our first patient (fig. 3). A high a wave (12 to 14 mm Hg) was transmitted through the right ventricle, where it was superimposed on an elevated end-diastolic pressure, up to the pulmonary artery. A similar transmission of a waves to the right ventricle has been described in two previous reports.\textsuperscript{1, 2} Pulmonary or tricuspid valvar gradients have not been registered. The oxygen saturations demonstrated a bidirectional atrial shunt in six individuals\textsuperscript{1, 2, 9, 10} and a right-to-left shunt in two patients,\textsuperscript{6} including one in the present series. It is highly probable that three severely cyanosed neonates who did not undergo cardiac catheterization also had a dominant right-to-left shunt. The arterial oxygen saturation, which was determined in ten patients, ranged between 66% and 90%. It was probably very low in four critically ill infants in whom this value was not obtained. The diagnostic angiocardiographic appearance of the right ventricle has been commented upon,\textsuperscript{1, 2, 9, 10} and is well illustrated in our first patient. From the anteroposterior position the right ventricular cavity is small, medially placed, and obviously smooth walled. The trabecular zone is poorly developed or absent. The infundibulum, on the other hand, is normal or unusually wide.\textsuperscript{8} Right ventricular contraction is poor and systolic-diastolic volume changes are reduced. The right atrium is invariably dilated. Often the characteristic contour of the right ventricle is not distinctly visualized due to a large right-to-left atrial shunt and simultaneous opacification of both ventricles.\textsuperscript{4, 5, 7, 8} Preferably the contrast material should, therefore, be injected into the right ventricle, which was achieved in one patient.\textsuperscript{10} The injection clearly outlined the right ventricle but produced some introduction of contrast material into the myocardium, probably because of the smallness of the cavity.

The anatomical features, as described in surgical\textsuperscript{4, 5, 9} or pathological reports\textsuperscript{1, 2, 3, 6, 7, 8, 10} are relatively uniform. The first detailed account was given by Gasul and coworkers.\textsuperscript{1} Most authors stress the diminutive size of the right ventricle and the dilatation of the right atrium, when the heart is looked at from the exterior. In every case an atrial septal defect or a patent foramen ovale is found. The right ventricular cavity consists of a small inflow portion and a normal or large outflow tract. The trabeculated apical portion is almost completely lacking. In one patient endocardial thickening of the right ventricle was noted.\textsuperscript{7} Histological examination of the right ventricular myocardium did not reveal any abnormality. The tricuspid valve is normally formed. In some cases its orifice is small but appears normal when related to the reduced size of the right ventricular cavity. According to Edwards this simply reflects the basic hypoplasia of the right ventricle.\textsuperscript{7} This is borne out by the observation that a diminutive tricuspid valve was found only in cases with severe right ventricular underdevelopment leading to death in infancy.\textsuperscript{3, 7, 8} The left ventricle is hypertrophied because of the increased volume load.
The basic hemodynamic disturbance may readily be conjectured from the anatomical arrangement and the hemodynamic data. The main problem seems to be the increased resistance to ventricular inflow, due to the small capacity of the right ventricle. This is usually reflected in raised early and end-diastolic pressures. Increased right atrial pressure and a forceful atrial contraction help to overcome this reduced compliance. Moreover, right atrial systole, apart from enhancing ventricular filling, may in some patients take over part of the ventricular function by propelling blood into the pulmonary artery (fig. 3). A similar transmission of a waves has been described in Ebstein's anomaly. At the same time, increased atrial pressure produces a right-to-left shunt through an atrial septal defect or a stretched foramen ovale, which in turn results in arterial desaturation. That the atrial septal defect functions as an escape valve is suggested by the results of its temporary occlusion by means of a balloon catheter. During occlusion a small but significant elevation of right atrial a waves was observed and attributed to an increased resistance to right atrial contraction.

Thus, the two main variables which determine the patient's condition are the degree of right ventricular hypoplasia and the size of the interatrial communication. Obviously the worst combination is severe underdevelopment of the right ventricle with a patent but small foramen ovale. This arrangement was found at autopsy in four infants. If marked hypoplasia is associated with a large atrial septal defect, the patient will be cyanotic but may survive into childhood or even into adulthood.

In milder forms of right ventricular hypoplasia the atrial septal defect will permit a bidirectional shunt, the main direction and magnitude of which will be determined by the relative compliances of both ventricles. These patients will be slightly cyanotic and only mildly symptomatic. Consequently the clinical spectrum varies from severe cyanosis, congestive heart failure, and death in early infancy, on the one hand, to mild cyanosis in patients who otherwise present with signs of an atrial septal defect, on the other.

In the differential diagnosis, tricuspid atresia, pulmonary atresia, Ebstein's malformation of the tricuspid valve, and anomalies of the systemic venous return should be considered. Severe forms of right ventricular hypoplasia, especially, may be very similar to tricuspid or pulmonary atresia with intact ventricular septum. The electrocardiogram, which, in most cases of right ventricular hypoplasia, shows left axis deviation and right atrial enlargement in the absence of right ventricular electrical forces, may be indistinguishable from that in tricuspid atresia. On the other hand, a normal QRS axis or right axis deviation, in the presence of right atrial enlargement and left ventricular preponderance, seen in about one-third of the patients with right ventricular hypoplasia, may be compatible with pulmonary atresia. Cardiac catheterization and angiography will be necessary to differentiate the three malformations. Milder forms, as described in the present report, may mimick Ebstein's malformation. Extra systolic or diastolic sounds on auscultation and the characteristic electrocardiographic features of Ebstein's malformation have, to our knowledge, not been encountered in right ventricular hypoplasia. Slight cyanosis, absence of an obvious heart murmur, and electrocardiographic evidence of left ventricular hypertrophy are also suggestive of partial anomalous systemic venous return to the left atrium. Right atrial enlargement however has not been noted in the latter condition and its presence will be helpful in the differential diagnosis. In our second patient, in whom the P waves were normal, the possibility of an anomalous systemic venous connection was entertained. The observation that temporary occlusion of the atrial septal defect by means of a balloon catheter increased the arterial oxygen saturation to an almost normal level, was considered to be good evidence against anomalous systemic drainage. It is suggested that this procedure should be used as an aid to differentiate both conditions. Obviously the
final diagnosis will rest on the typical angiocardiographic image of the hypoplastic right ventricle.

Several types of surgical treatment have been reported. In severe hypoplasia, in which the hemodynamic situation is similar to that of tricuspid atresia, an anastomosis between the superior vena cava and the right pulmonary artery is the most logical palliative approach and produces good results. In one child this Glenn procedure was attempted but seemed too hazardous in view of the small size of the pulmonary artery. The patient died shortly after a Blalock-Taussig anastomosis had been created. Closure of the atrial septal defect has been reported by Hollmann and was performed in both our patients. All three children became acyanotic and their effort tolerance increased considerably. Admittedly, this operation should be considered only in milder forms of hypoplasia in which the right atrium and ventricle will be able to cope with a larger blood volume once the atrial septal defect has been closed. In the preoperative assessment of the right heart function temporary balloon occlusion of the defect may be helpful, as illustrated in the two patients reported here.

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