Congenital absence of pulmonary valve leaflets

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SUMMARY Congenital absence of pulmonary valve leaflets is an uncommon condition usually associated with ventricular septal defect and an obstructive pulmonary valve ring. Twenty-one patients with these malformations are described. Twenty had an associated ventricular septal defect with ventriculoarterial concordance, and one also had transposition of the great arteries, ventricular septal defect, and obstructive pulmonary valve ring. The clinical features, cardiac catheterisation findings, and angiocardiographic results are presented. Twelve patients underwent cardiac surgery. Three patients died, one in the early, and the other two in the late postoperative period. The results, according to the surgical technique employed and postoperative cardiac catheterisation findings, showed that patients in whom the bioprostheses were implanted in the pulmonary position had a better late follow-up.

Congenital absence of the pulmonary valve leaflets is a rare cardiac malformation that can be seen as an isolated lesion or, more commonly, in association with ventricular septal defect. Other malformations such as atrial septal defect, double outlet right ventricle, persistent ductus arteriosus, endocardial cushion defect, transposition of the great arteries, and other complex cardiac malformations have also been described in patients with congenital absence of the pulmonary valve leaflets. There are at present 266 reported cases of these malformations.

The purpose of this paper is to present the clinical findings in 21 patients with congenital absence of the pulmonary valve leaflets and the surgical results in 12 of these children who underwent elective cardiac repair.

Subjects and methods

From April 1963 to December 1980, 21 patients with congenital absence of the pulmonary valve leaflets (12 male and nine female) aged from 3 months to 41 years were studied. All patients were referred to our institution because of the presence of cardiovascular symptoms or the detection of a cardiac murmur at a variable age, but, for unknown reasons, many of them came months or years later. They were followed up in the outpatient clinic until December 1982 in order to obtain at least 24 months of additional clinical data, particularly in those who underwent surgery to record the fate of the bioprosthetic valves. The diagnosis was suspected clinically and in all, cardiac catheterisation was performed. The clinical data, physical findings, chest x-ray films, electrocardiograms, and angiocardiographic data were analysed. Six patients were studied by M-mode and cross-sectional echocardiography, and bronchographic studies were made in five cases with lobar emphysema. Cardiac catheterisation included measurements of QP/QS ratio and of systolic and diastolic pressures in the right ventricle and main pulmonary artery. Systemic oxygen saturation was determined in all patients via a femoral artery tap simultaneously with cardiac catheterisation. Angiocardiograms in the right ventricle were performed in 20 patients. The size and orientation of the infundibulum of the right ventricle, the characteristics of the pulmonary ring, and the anatomy of the pulmonary arteries were analysed in the angiocardiogram. The ventricular septal defect was diagnosed by demonstrating a left to right shunt at the ventricular level either by left ventriculogram or laevophase after right ventricular injection. We also looked for the downward displacement of the left
<table>
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<tr>
<th>Case No.</th>
<th>Age (s)</th>
<th>Sex</th>
<th>Physical examination</th>
<th>X-ray CTR</th>
<th>ECG</th>
<th>Hemodynamic features</th>
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<tr>
<td>1 3/12 mth</td>
<td>F</td>
<td>LRTI and cyanosis since birth, murmur detected at 2 months of age, to-and-fro murmurs in 3rd LIs, SS2 cardiac failure</td>
<td>0-52</td>
<td>-155° RVH</td>
<td>92/1-5</td>
<td>33/13</td>
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<td>2 8/12 mth</td>
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<td>LRTI since age of 3 months, murmur detected at 8 months, to-and-fro murmurs in 3rd LIs, SS2</td>
<td>0-66</td>
<td>+165° RVH</td>
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<td>LRTI since age of 40 days, admitted to hospital with cardiac failure, to-and-fro murmur in 2nd LIs, SS2</td>
<td>0-50</td>
<td>-120° RVH</td>
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<td>LRTI since age of 6 months, murmur detected at 16 months; to-and-fro murmurs in 3rd LIs, SS2</td>
<td>0-48</td>
<td>+120° RVH</td>
<td>110/10</td>
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<td>5 3 y</td>
<td>M</td>
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<td>+130° RVH</td>
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<td>27/10</td>
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<td>+140° RVH</td>
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<td>M</td>
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<td>+150° RVH</td>
<td>140/7</td>
<td>38/12</td>
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<td>Asymptomatic, precordial bulging since age of 1 year, to-and-fro murmurs in 2nd LIs, SS2</td>
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<td>+150° RVH</td>
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<td>60/18</td>
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<td>+100° RVH</td>
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<td>26/11</td>
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<td>+160° RVH</td>
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<td>19/9</td>
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<td>+130° RVH</td>
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<td>16 11 y</td>
<td>F</td>
<td>LRTI in first year of life, cyanosis at age of 3 years; cyanosis, to-and-fro murmur in 2nd LIs, SS2</td>
<td>0-44</td>
<td>+150° RVH</td>
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<td>+130° RVH</td>
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<tr>
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<td>+125° RVH</td>
<td>76/0</td>
<td>44/0</td>
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<td>0-58</td>
<td>-150° RVH</td>
<td>76/4</td>
<td>65/11</td>
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<td>20 17 y</td>
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<td>Effort dyspnoea and shortness of breath during feeding since age of 3 months, LRTI in first year of life; cyanosis, to-and-fro murmur in 3rd LIs, SS2</td>
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<td>+170° RVH</td>
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<td>46/14</td>
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<td>21 41 y</td>
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<td>Asymptomatic till age of 40 years, dyspnoea and oedema in lower limbs since then; to-and-fro murmur in 2nd LIs, SS2</td>
<td>0-56</td>
<td>-170° RVH</td>
<td>104/7</td>
<td>104/6</td>
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CTR, cardiothoracic ratio; ECG, electrocardiogram; RV, right ventricle; PA, pulmonary artery; LV, left ventricle; QP/QS, pulmonary/systemic flow ratio; LRTI, lower respiratory tract infection; LIs, left intercostal space; SS2, single second heart sound; RVH, right ventricular hypertrophy; RBBB, right bundle-branch block. Pressures in mmHg.
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atrium produced by dilatation of the right pulmonary artery.

Twelve patients between 5 and 17 years of age underwent elective surgical repair; in all but one, postoperative catheterisation was performed between 10 days and one year after treatment.

Results
All cases had a ventricular septal defect, one of them with absence of the left pulmonary artery, and another with transposition of the great arteries. Table 1 shows the clinical data of all patients. Twelve had a history of recurrent lower respiratory tract infections; eight presented late with cyanosis, and only one had cardiac failure. Physical examination disclosed a pansystolic thrill and a harsh systolic ejection murmur maximal at the second and third intercostal spaces at the left sternal border. The second heart sound was single. All patients but one (case 14) had a decrescendo early diastolic murmur, which was best heard over the same area. The murmurs were not continuous but had a to-and-fro quality. Phonocardiography was carried out in 12 patients and no pulmonary closure sound was recorded in any of them.

The electrocardiographic features were analysed in 21 patients. The AQRS ranged from +100 to −170. All cases showed right ventricular hypertrophy and in 11 of them an associated incomplete right bundlebranch block was evidenced.
Cardiothoracic ratio ranged from 0.43 to 0.66. Cardiomegaly was produced by dilatation of both right cardiac chambers. The middle segment of the left heart border was always prominent except for the patient with transposition of the great arteries. In eight cases, its convexity was pronounced. Dilatation of the right pulmonary artery was clearly seen on the frontal view in all cases, but the left pulmonary artery was usually easier to identify in the left anterior oblique projection. In nine patients both the right pulmonary artery and left pulmonary artery shadows reached as far as the middle of the right and left lungs. The pulmonary vasculature varied according to the degree of stenosis present in the pulmonary annulus. Sixteen patients presented an important degree of right ventricular outflow tract obstruction and in all of them the chest x-ray film showed the sudden change in calibre between the right pulmonary artery and the peripheral vessels (Fig. 1).

In the five patients with lobar emphysema, bronchial obstruction was shown by bronchographic studies (Fig. 2). In case 16, with absence of the left

![Fig. 1](image1.png) Posteroanterior chest x-ray of case 18 showing distinct bulging of main, right, and left pulmonary arteries. Observe the sudden change in calibre between the main branches and peripheral vessels. Important cardiomegaly is seen. A foreign body is located in the upper right bronchus.

![Fig. 2](image2.png) (A) Chest x-ray of case 8. The transverse diameter of the heart is increased. The right pulmonary artery is somewhat enlarged and the right peripheral pulmonary vasculature is within normal limits. In the upper lobe of the left lung obstructive emphysema, produced by the dilated pulmonary artery, can be visualised. (B) Tracheobronchogram of the same case. The left bronchus is compressed and displaced downwards by the dilated pulmonary artery.
pulmonary artery, the bronchogram showed a normal left bronchial tree and the right bronchus was found to be compressed by the dilated right pulmonary artery (Fig. 3).

M-mode and bidimensional echocardiograms were performed in six children using the parasternal and suprasternal approaches. Fluttering of the tricuspid valve, linear echoes originating either in the pulmonary valve or in the valvular ring, and a dilated right pulmonary artery were shown in each of these studies. The obstructive valvular ring and a to-and-fro movement of the contrast bubbles were seen to occur between the right ventricle and pulmonary artery using the two-dimensional technique (Fig. 4).

All patients were studied by cardiac catheterisation and 20 by means of angiocardiography (Table 1). No significant gradients were detected between the mean right atrial pressure and the end-diastolic pressure of the right ventricle. In five patients there was equalisation of right ventricular and pulmonary arterial pressures at the end of diastole. A diastolic gradient between the pulmonary artery and the right ventricle was found in 13 patients, and in two cases the right ventricular end-diastolic pressure was higher than the pulmonary arterial diastolic values. Withdrawal pressure recordings from the pulmonary artery to the right ventricle did not show gradients at the infundibular level. Twelve patients presented dominant left to right shunts, and eight had dominant right to left intracardiac shunts.

Right ventricular angiograms showed several interesting features concerning the level and anatomical characteristics of the pulmonary annulus, the spatial orientation of the infundibulum, and the dilatation of the pulmonary arteries. In all patients the outflow tract stenosis was situated at the level of the pulmonary valve ring and was formed by a ridge of tissue displaced upwards and backwards. There was no stenosis of the infundibulum; in fact, it was dilated in all patients. Pronounced thickening of the infundibular septum that did not cause obstruction was visualised in 16 studies (Fig. 5). The infundibulum was abnormally oriented in 15 patients. In seven, the entire infundibulum was directed from left to right; in four, it was vertical, and in four it was oriented slightly towards the left (Fig. 6). In the remaining cases its direction was normal. In all cases the pulmonary artery was dilated, and in 12 of them it was aneurysmal (Fig. 7).

Pulmonary arteriograms were performed in six cases and in all of them pulmonary regurgitation was shown. In four, the massively dilated right pulmonary artery displaced the roof of the left atrium downwards (Fig. 8). Clearance of the contrast material was slow in the five patients with equalisation of pulmonary and right ventricular diastolic pressure. The aortic arch was normal in all but one of the patients (case 14).

Surgical correction was performed in 12, with extracorporeal circulation and moderate hypothermia. Cardiac catheterisation was done in 11 after operation (Table 2).

The ventricular septal defect was closed via a right ventriculotomy in all cases except one. Infundibular resection was performed in 10 cases because the infundibular septum was found to be thickened during surgery. Four patients underwent resection of the right ventricular infundibulum and enlargement of the pulmonary annulus. Two of these died. One, case 7, died two months after operation; in the postoperative period she developed severe cardiac failure and respiratory distress, which were produced by massive pulmonary regurgitation; reoperation was carried out in order to insert a valvular prosthesis but the patient died 14 days after. Case 8 died six months after operation because of congestive heart failure and respirat-

Fig. 3 (A) Posteroanterior selective right ventriculogram (RV) of case 16. Note the huge right pulmonary artery (RPA) and absence of the left branch. (B) Tracheobronchogram of the same patient showing a normal left bronchus and compression of the right bronchus by the right pulmonary artery. 1, right ventricular infundibulum.
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Infection, and necropsy showed regional emphysema produced by bronchial compression resulting from dilatation of the pulmonary artery. The remaining two (cases 9 and 16) are symptom free at present, even though they had right ventricular systolic pressures of 47 and 67 mmHg, respectively, after surgery. In cases 3, 5, 18, and 20 in which the infundibulum was resected without enlargement of the pulmonary annulus, we observed only an insignificant reduction in right ventricular systolic pressure. In three (cases 3, 5, and 18) the pulmonary artery systolic pressure was normal and in the other one it was slightly increased. Despite the surgical results, at the present time all of them have only mildly decreased exercise tolerance.

Fig. 4 M-mode and cross-sectional echocardiograms of a patient with congenital absence of the pulmonary valve leaflets. (A) Parasternal M-mode scan from the pulmonary artery (PA) to the aorta (Ao). The pulmonary root has an increased anteroposterior diameter. Echoes originating in the rudimentary pulmonary valve (arrow) are seen near the posterior wall of the pulmonary artery. The aorta is located behind and to the right of the pulmonary artery and is normal in size. (B) M-mode suprasternal echogram. The aorta (Ao), right pulmonary artery (rpa), and left atrium (LA) are recorded cephalocaudally. The diameter of the pulmonary branch is larger than that of the aorta. (C) Short-axis cross-sectional echocardiogram at the level of the great arteries. Anteroposteriorly the right ventricular infundibulum (i), aorta (Ao), and both atria (ra and la) were recorded. The obstructive pulmonary annulus can easily be seen (arrow). (D) Short axis contrast cross-sectional echogram at the level of the great arteries. The pulmonary annulus obstructs the passage of contrast material from the infundibulum into the pulmonary artery.
Fig. 5  Lateral view of selective right ventriculogram (RV) in a case of congenital absence of the pulmonary valve leaflets with a ventricular septal defect. Same case as Fig. 2. Observe the hypertrophied crista supraventricularis, the dilated infundibulum, and main pulmonary artery (PA). The arrow points to the valvular ring, which is displaced upwards and backwards.

Fig. 6  Selective right ventriculograms (RV) in two patients with congenital absence of the pulmonary valve leaflet. (A) The right ventricular outflow tract (I) is directed from left to right with a massively dilated right pulmonary artery (RPA). (B) The infundibulum (I) is oriented from right to left and the main and left pulmonary artery (LPA) are dilated. Both studies show the association between infundibular orientation and pulmonary arterial dilatation.
Fig. 7  Pulmonary angiogram in the same patient as in Fig. 1. Notice the aneurysmal dilatation of the pulmonary branches. LPA, left pulmonary artery; RPA, right pulmonary artery.

Fig. 8  Pulmonary (PA) angiogram in a patient with congenital absence of the pulmonary valve leaflets, transposition of the great arteries, and ventricular septal defect (case 14). Compression of the roof of the left atrium (LA) by a dilated pulmonary artery is observed during laevophase.
Repair of the malformation using pulmonary valve prostheses was performed in three patients. Ionescu-Shiley bioprostheses were used in cases 12 and 15; case 14 underwent correction by the Rastelli technique. In the former two patients, important reduction of the right ventricular systolic pressure was found postoperatively. The case with transposition had residual high right ventricular and pulmonary systolic pressures and is symptom free.

Case 6 was diagnosed as having congenital absence of the pulmonary valve leaflets and atrial septal defect. At operation, during which the patient died, the atrial defect was closed and the pulmonary annulus enlarged. At necropsy, a ventricular septal defect was found.

When comparing the pre- and postoperative values of the systolic pressure of the right ventricle we obtained significant differences (p=0.001). In cases 7, 8, and 16 we observed an increase in the pulmonary systolic pressure after operation. These three cases underwent annular enlargement during surgery and developed important pulmonary insufficiency.

Discussion

In congenital absence of the pulmonary valve leaflets the critical period for survival is during infancy. Most of the symptoms are caused by respiratory distress or infection secondary to airway obstruction produced by the dilated pulmonary artery compressing the bronchus. Bronchial compression is a problem in early infancy and may improve spontaneously during the first years of life. This improvement is accounted for by several factors that are related primarily to maturational changes in the tracheobronchial tree and lungs. The compression involves the anterior aspects of the lower trachea and major bronchi, particularly the right bronchus. Depending on the severity of the bronchial obstruction, emphysema or atelectasis may develop.

Although most authors consider congenital absence of the pulmonary valve leaflets with ventricular septal defect a form of tetralogy of Fallot, none of our cases fulfils the anatomical or angiographic characteristics of tetralogy, because none of the patients presented deviation of the infundibular septum and its consequences.

The combination of a pansystolic murmur at the left sternal border, a single second heart sound of normal intensity, and a low pitched crescendo-decrescendo diastolic murmur over the pulmonary area starting after the second heart sound are pathognomonic of this malformation. Absence of the pulmonary leaflets necessarily results in pulmonary regurgitation, which is manifested by the diastolic murmur. The presence of cyanosis depends on the size of the ventricular septal defect and the degree of pulmonary obstruction.

The electrocardiogram shows non-specific changes in congenital absence of the pulmonary valve leaflets.
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The presence of incomplete right bundle-branch block associated with right ventricular hypertrophy can be of value in differentiating congenital absence of the pulmonary valve leaflets from tetralogy of Fallot.

In most patients the cardiac shadow is moderately enlarged, occasionally it is very enlarged, and in a few cases it is normal. We assume that cardiomegaly is related to the severity of pulmonary regurgitation. The middle segment of the left heart border is almost always prominent and in many cases its convexity is marked. Dilatation of the right pulmonary artery is clearly seen on the frontal view, but the left pulmonary artery is usually easier to identify in the left anterior oblique projection. The cause of the aneurysmal dilatation of the pulmonary artery is uncertain; it may result from increased right ventricular stroke volume caused by pulmonary regurgitation and a left to right shunt through the ventricular septal defect. The dilatation of the pulmonary artery and proximal branches contrasts with the peripheral pulmonary vasculature, that is either normal or slightly increased.12 38-43

Pulmonary vascularity depends upon the degree of stenosis in the pulmonary outflow tract, the presence of a ventricular septal defect, and the anatomical characteristics of the pulmonary artery. Differences in the vascularity of the two lungs were observed in patients with absence of one pulmonary artery. The cause of the emphysema is the obstruction of the respective bronchus resulting from compression by the dilated pulmonary artery, as shown by the fact that the homolateral bronchus was not obstructed in patients with absence of one of the pulmonary artery branches.

Although the diagnosis of congenital absence of the pulmonary valve leaflet can easily be made on the basis of auscultatory and radiological findings, M-mode and cross-sectional echocardiographic studies offer interesting data.44-46 It is easy to identify the dilated pulmonary artery, the diameter of which is greater than that of the aorta. Other important features include the findings of the obstructive pulmonary annulus, the echoes originating from the rudimentary leaflets, and the to-and-fro movement of the contrast material in the right ventricle and pulmonary artery. These findings, together with the non-specific signs of right ventricular diastolic overload, are of value in the accurate diagnosis of this malformation.

Cardiac catheterisation is important to identify the site of the right ventricular outflow tract obstruction which is usually situated at the level of the valve ring.41 In several patients, however,7 16 the obstruction was shown to exist either at the infundibulum or at both the infundibular and annular levels. Withdrawal pressure recordings from the pulmonary artery to the right ventricle do not always distinguish between these two entities, and, because of this, angiography is mandatory in order to assess the anatomy of the right ventricular outflow tract as well as that of associated lesions. Data obtained from cardiac catheterisation are also important in planning the surgical correction since, as we found in our patients, the long term results depend to a large extent on the type of operation performed. The pulmonary artery angiographic features of this syndrome are impressive. Gross dilatation of the pulmonary artery and its branches with an abrupt transition in calibre between the main trunks and their immediate branches are typical of this malformation.16 33 39 Equally striking is the compression and downward displacement of the left atrium by the dilated right pulmonary artery. We found a relation between the degree of dilatation of the branches of the pulmonary artery and the orientation of the infundibulum.33 When the infundibulum was oriented towards the right, the right pulmonary artery was aneurysmal. Similarly, vertical and leftward orientation was responsible for bilateral and/or left pulmonary artery dilatation.

The surgical results in congenital absence of the pulmonary valve leaflets depend on the age at operation, the presence and degree of residual regurgitation, and pulmonary hypertension.19 27 30 47 Both medical treatment and intracardiac repair in infants with this syndrome are associated with a high mortality rate.19 30 48 Some authors recommend vigorous respiratory treatment in severely affected infants and children after operation because surgical repair does not always alleviate the respiratory symptoms.30 Probably this may be because of anatomical changes in the bronchial generations and alveolar multiplication as was evidenced by Rabinovitch and her colleagues.49

In older patients the surgical treatment of choice has been closure of the ventricular septal defect and relief of the outflow tract obstruction. Patients in whom the operation included enlargement of the pulmonary annulus, but without implantation of a bioprosthesis, showed massive regurgitation with an increased systolic pressure of the pulmonary artery. Follow-up results in these cases are similar to those observed in patients who were not operated on, in whom pulmonary insufficiency produces progressive dilatation of the pulmonary artery with compression of the bronchi, regional emphysema, respiratory distress, and death.27 30 50 Our patients in whom the ventricular septal defect was closed and the infundibulum was resected showed a better evolution despite the fact that a high mortality rate has been reported in other patients undergoing this type of repair.7 13-16 27 40 42 51 The diastolic gradient between
the pulmonary artery and right ventricle probably protects the pulmonary arterial tree and the right ventricle against the consequences of massive regurgitation. Nevertheless, in these patients a residual systolic gradient remains because of the obstructive pulmonary annulus.

Valve insertion at the pulmonary annulus during the initial operation has been a controversial subject because of early calcification and dysfunction in young patients. Despite the limited durability of bioprostheses in children, repair of the malformation using these valves seems to offer the best results when implanted at the time of the initial repair because they prevent pulmonary regurgitation and preserve the contractile characteristics of the right ventricle as well as the pumping action of the pulmonary arteries.

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