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Brief Report

Transcatheter reconstruction of the right heart

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Abstract We report the interventional procedures performed on a 12-year-old child with obstruction of the right ventricular outflow tract, pulmonary valvar insufficiency, pulmonary arterial stenosis, and an atrial septal defect. A staged repair of all anomalies was performed successfully using transcatheter techniques.

Keywords: Catheter intervention; valve; stent; atrial septal defect

Failure of the right ventricle is a frequent end-point for patients with congenital cardiac disease.[1] It usually occurs as a result of the combination of obstructive lesions, often with dysfunction of the pulmonary and tricuspid valves.[2] Such patients can become severely symptomatic when there is also right-to-left shunting possible at atrial level. Many of these problems, at least in isolation, are now amenable to treatment by interventional cardiology. Obstructive lesions were the first to become accessible, by balloon dilation and stenting.[3] More recently, the closure of atrial septal defects has become a well-established technique,[4] and now the treatment of pulmonary valvar dysfunction by replacement of the valve has become possible by transcatheter means.[5,6] We describe in this report a severely symptomatic patient, in whom we used interventional techniques to treat all these lesions.

Case report

A 12-year-old girl with tetralogy of Fallot came to our attention in her early childhood. Her clinical history included total surgical repair at the age of 1 month. At 6 months, she required further surgical enlargement of the bifurcation of the pulmonary trunk, and a conduit was placed from the right ventricle to the pulmonary arteries. At the age of 5 years, this conduit was replaced with a second valved conduit of 16 mm diameter. Six years later, this conduit had also become obstructive, and important insufficiency of the valve was detected, with major dilation of the right ventricle. At the age of 12 years, the child had severely reduced exercise tolerance due to severe cyanosis, no more than 65%, at minor physical exercise of 90 W. Echographic findings showed a severely dilated right ventricle with tricuspid valvar insufficiency. Right ventricular pressures were estimated to be at systemic levels. These hemodynamic findings were confirmed at catheterization. Angiographies of the right ventricular outflow tract were performed using a Multitrack catheter (Numed). They confirmed severe pulmonary regurgitation, the conduit being seen to be obstructed at its implantation on the infundibulum. There was severe stenosis at the origin of both pulmonary arteries, along with hypoplasia of the right pulmonary artery. A gradient of 40 mmHg was measured between the trunk behind the conduit and the distal pulmonary arteries. Distal pulmonary arterial pressures, however, were normal. It was decided to replace the pulmonary valve as a first stage, to be carried out under general anesthesia.

Materials

A valve harvested from a bovine jugular vein was mounted and prepared in a platinum stent as previously reported.[5,6] A delivery system (Numed) was used that mounted a "balloon in balloon" catheter. The catheter itself is mounted in a sheath of 11 French dimensions proximally, and 18 French at the last five

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distal centimeters. This part shelters the valved stent when crimped on the balloon catheter. A 3-cm long dilator is fixed on the extremity of the catheter. This assures the progressive dilation of the vessel to the largest diameter of the delivery system when inserted through the skin. The sheath can be pushed and pulled to cover or uncover the valved stent.

Valvar implantation

A 0.035-inch guide wire (Meditech) was positioned in the distal right pulmonary artery. The dilator was then connected to the guide wire and inserted through the skin. The delivery system was advanced across the right ventricular outflow tract. Despite multiple attempts, we were unable to cross the obstructed conduit over its entire length, but we decided to deliver the device. The sheath was pulled back to uncover the device, and the balloons were subsequently inflated, deploying the valved stent. After the deflation of the balloons, the delivery system was retrieved, leaving the valved stent in a slightly low position, with 0.5 cm of the stent in the infundibulum. Evaluation at this stage showed a perfectly functioning implanted valve, a negligible para-prosthetic leak, and a systolic right ventricular pressure at two-thirds of systemic values (Fig. 1). The patient was discharged the day after the implantation. During 5 months of follow-up, the implanted valve was functioning perfectly, without any leak, leading to an improvement in clinical status and right ventricular function, and a normalization of right ventricular size. Chest X-rays, however, showed a fracture of the ventricular part of the stent. The right ventricular pressure evaluated echocardiographically was measured at 80 mmHg for 100 mmHg systolic aortic pressure, with a pressure gradient to the two pulmonary arterial branches. In spite of a marked subjective improvement, repeat exercise testing continued to demonstrate desaturation, with right-to-left shunting through an atrial septal defect responsible for the desaturation. The patient was re-catheterized under general anesthesia to treat the pulmonary arterial stenosis, and to close the atrial septal defect.

Pulmonary arterial stenting

The technique used for implantation has been described previously in detail.[3] In brief, stents were mounted on 12 mm diameter balloon (Z-Med 12*4, Numed) and placed to straddle the lesions with a 9 French long Mullins sheath (Arrow) over a 0.035 inch Amplatz extrastiff exchange interventional wire (Cook). To conform to the configuration of the pulmonary vessel both proximally and distally, a 30 mm long stent (P308E, Cordis), and an 18 mm long stent (P188, Cordis), were used respectively for the right and left pulmonary arteries. Selective angiography was obtained in axial projections to maximize the profile of the stenosis and pulmonary arterial topography. The angiographs showed a long stenosis of the proximal right pulmonary artery extending into the superior lobar branch, and a stenosis at the implantation of the left pulmonary artery on the conduit.

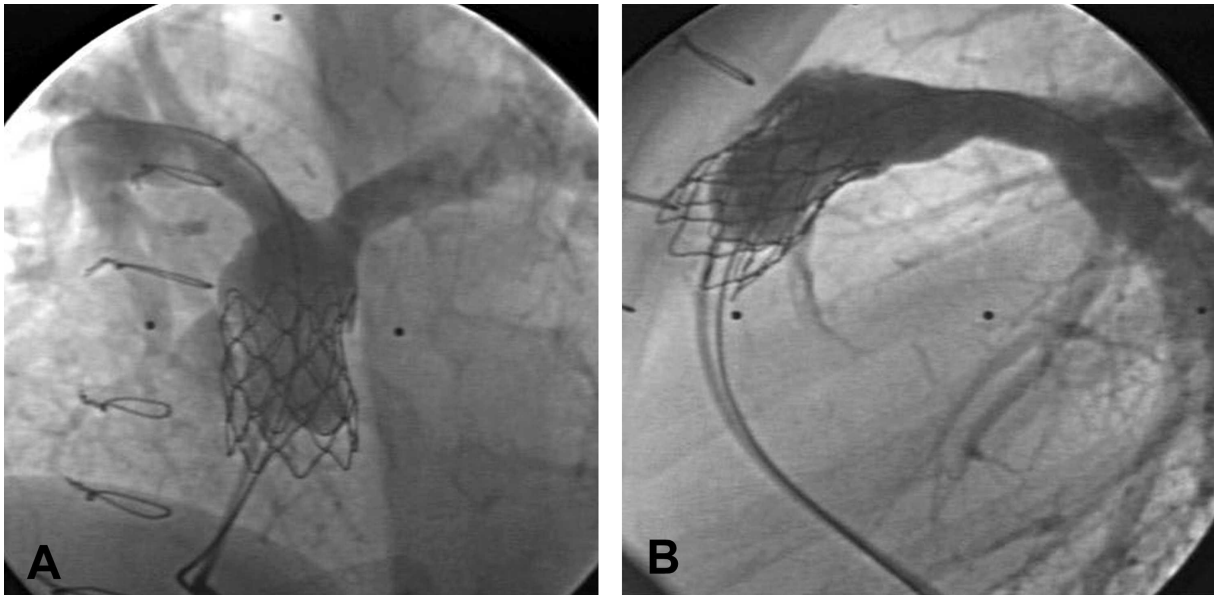

Figure 1.
The four-chamber view of the angiogram (A) shows stenosis of the right pulmonary artery. The lateral view of the same angiogram (B) reveals the competence of the implanted pulmonary valve.

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Distal to the stenosis, the pulmonary artery measured 11.5 mm. The diameter at the site of stenosis was 6 mm for the right pulmonary artery, and 8 mm for the left pulmonary artery, increasing to 12 mm after implantation of the stent. Peak systolic pressure gradients fell from 40 to 24 mmHg, with a mean decrease from 20 to 16 mmHg. Right ventricular systolic pressure fell from 90 to 50 mmHg. The ratios of right ventricular to systemic pressure decreased from 0.80 to 0.48 mmHg.

Closure of the atrial septal defect

An experienced echocardiographer assisted with the closure of the septal defect. The defect was firstly sized with a 30 mm NMT medical balloon (Numed). The stretched diameter was measured at 14 mm. A 14 mm Amplatzer septal occluder (AGA Medical Corporation) was implanted as reported previously.[4] After placement of the device, fixation and stability of the device were proven using the "Minnesota Wiggle" technique. The transesophageal echocardiogram confirmed the position of the device, with no residual shunting, and the occluder was thereafter released. Intravenous heparin (100 IU/kg) was administrated twice during the procedure. The day after the procedure, and during follow-up at 3 weeks, transthoracic echocardiography showed the absence of any atrial shunt. Systolic right ventricular pressure was estimated at 50 mmHg. The four-chamber view after the procedure is shown in Figure 2.

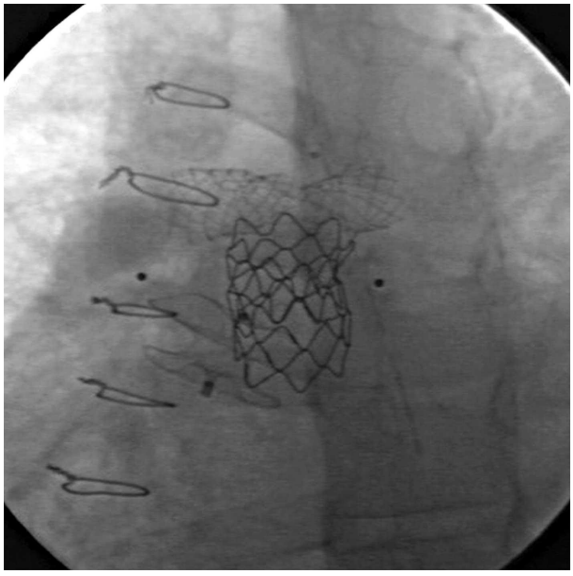

Figure 2.
The four-chamber view shows the stents inserted for the treatment of stenosis at the pulmonary bifurcation, the valved stent, and the Amplatzer device closing the atrial septal defect.

Discussion

The failing right heart has become the major issue in the long-term follow-up of patients who underwent surgery for congenital cardiac disease in early childhood.[1] Pulmonary regurgitation is a common cause of right ventricular deterioration, in particular when associated with obstructive lesions.[2] The standard surgical treatment with valvar replacement usually starts a vicious cycle whereby implanted valves will degenerate and require repeated surgical interventions. The option of replacing a pulmonary valve without surgery should influence the timing of intervention, as early valvar replacement will probably avoid or limit right ventricular damage.[7,8]

The patient we have described is a good example of such an approach. She had a history of multiple operations for tetralogy of Fallot with severe hypoplastic pulmonary arterial branches. At the time of referral, her conduit was failing, with severe pulmonary regurgitation and obstruction. We aimed to "repair" the right heart to enable the patient fully to recover her right ventricular function. The issue was how to define the structural anomalies with which we should begin. As a first step, we decided to treat the failing conduit, because preliminary stenting could have ruptured the balloons during inflation for deployment of a valved stent. After a few months, the right ventricle improved, but its compliance was still altered due to the high right ventricular pressures generated by the stenotic pulmonary arterial tree. This subsequently led to a right-to-left shunting through the atrial septal defect. Before closing the atrial septal defect, to improve the working conditions for the right ventricle, it was decided to stent both pulmonary arterial branches. Haemodynamic evaluation after stenting showed that the right ventricular pressures dropped to half of the systemic pressures. Under these conditions, we decided to make a test occlusion of the atrial septal defect. There was no increase in central venous pressure after 10 min of occlusion. The saturations were at 100%, so we decided to occlude this defect with a 14 mm Amplatzer device.

The configuration of the bovine valve allows further catheterization through the valve without any impairment of the implanted valve. The fracture of one weld of the stent had no clinical impact, and did not disturb the function of the newly implanted valve. This occurrence was probably related to the low implantation of the stent, and its contact with the moving infundibulum.

In conclusion, the patient we report had typical features of right ventricular failure within the context of congenital cardiac disease. Pulmonary valvar replacement, stenting of the arterial pulmonary branches, and closure of her atrial septal defect were

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achieved by transcatheter techniques, enabling us to reconstruct her right heart to an almost normal configuration.

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