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Brief Report

Unusual interventional management in an adult with tetralogy of Fallot

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Abstract A 53-year-old man with tetralogy of Fallot had been palliated with two classic Blalock-Taussig shunts. Cardiac catheterization in our center revealed acquired atresia of the pulmonary valve. We perforated and dilated the valve as a palliative procedure with a new system using radiofrequency energy.

Keywords: Pulmonary atresia; radiofrequency; tetralogy of Fallot; catheter intervention

Tetralogy of Fallot, the most common cyanotic congenital cardiac malformation encountered in older patients, was the first to be palliated by surgery. Currently, complete surgical correction is performed when patients are very young. Some, nonetheless, still reach an advanced age with only a palliative aorto-pulmonary anastomosis.[1] We describe here the perforation and dilation of the pulmonary valve at an unusual age in a patient with severe tetralogy of Fallot with acquired pulmonary atresia, using the recently available system for perforation that uses radio frequency energy (Baylis Medical Company, Montreal, Canada). This system, having a high impedance cut-off range, permits the atretic valve to be perforated rather than ablated, using low power over a short duration.

Case report

A 53-year-old man recently came to our outpatient clinic complaining of dyspnea at rest and increased cyanosis. Tetralogy of Fallot had been diagnosed during infancy, and had been palliated with two classic Blalock-Taussig shunts, constructed at the age of 5 and 18 years, respectively. A total repair was attempted at the age of 21 years. Dramatic bleeding occurred while opening the chest, secondary to damage to the extensive collateral circulation around the stenotic Blalock-Taussig shunts. When bleeding was finally controlled, the corrective attempt was abandoned, and the chest was closed.

When seen in our clinic, his saturation at rest was 70%, and his tolerance to stress was very reduced. The haemoglobin and hematocrit were measured at 19 g/dl, and 67%, respectively. We proceeded to cardiac catheterization so as to reevaluate the possibilities for treatment. The aortic angiogram showed a left-sided aortic arch, and faint opacification of the pulmonary arteries through minute collateral vessels at the right upper pulmonary lobe. Both Blalock-Taussig shunts were totally occluded, but the intrapericardial pulmonary arteries were of satisfactory dimensions. We decided, therefore, to evaluate the right ventricular outflow tract. After femoral venous puncture, we positioned a 5-French 3.5 right coronary Judkins catheter (Cordis, Issy-les-Moulineaux, France) in the infundibulum. Although the pulmonary valve was atretic (Fig. 1), simultaneous injection of contrast into the infundibulum and into the aorta demonstrated the presence of potential continuity between the infundibulum and the pulmonary trunk, albeit in the absence of anterograde flow. Because of the difficult previous surgical history, and the extreme reluctance of the patient to undergo another operation, we decided to perforate the pulmonary valve as a palliative procedure.

The procedure was carried out under general anesthesia. The femoral vein and artery were punctured

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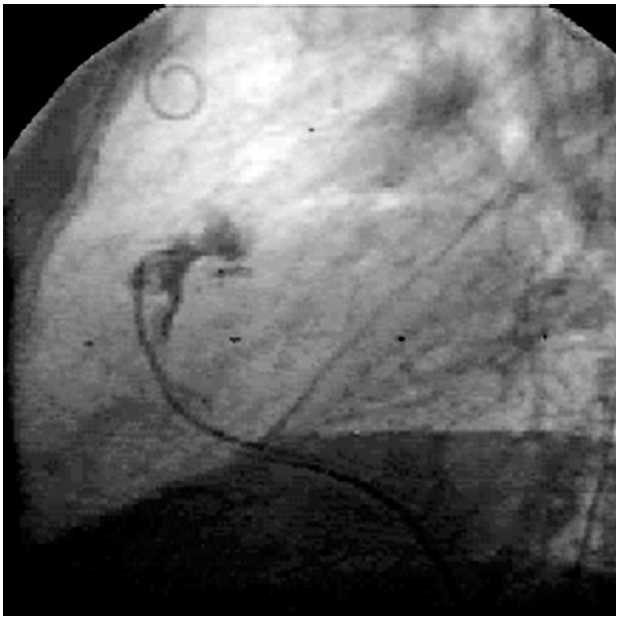

Figure 1.
Lateral angiography showing the acquired pulmonary valvar atresia. Note the 5-French pigtail catheter placed in the aorta.

with 7- and 5-French introducers, respectively. A 5-French pigtail catheter was positioned in the aorta for control angiograms during the procedure. A 5-French right Judkins catheter (Cordis, Issy-les-Moulineaux, France) was positioned in the infundibulum. The radiofrequency system was prepared for perforation after careful positioning of the catheter against the atretic pulmonary valve, assessing the location in 4 chamber and lateral views. An aortogram confirmed appropriate orientation of the catheter relative to the pulmonary trunk. Using power of 5 W over 2 sec, the valve was crossed, and the perforation catheter passed into the left pulmonary artery. After careful withdrawal of the Judkins catheter, the coaxial injectable catheter was loaded over the perforation catheter and easily advanced to the left pulmonary artery, where it was exchanged for a 0.014 inch coronary guide wire. We then dilated the valve using a 4 mm coronary angioplasty balloon (AVE, Medtronic, Boulogne-Billancourt, France). The guide wire was then exchanged again for an extrastiff 0.02 inch wire (Schneider, Boston Scientific Corporation, St Quentin-en-Yvelines, France) using the right Judkins catheter. A further dilation was then done using a 10 × 2 mm Z-Med balloon (Numed Inc, Hopkinton, USA). A final dilation was achieved with a Tyshak II 15 × 2 mm (Numed Inc, Hopkinton, USA) balloon, advanced over the same wire. At the end of the procedure, the ventriculo-pulmonary junction had a diameter of 12 mm (Fig. 2). The saturation of oxygen in the blood increased progressively from 77% to 90%.

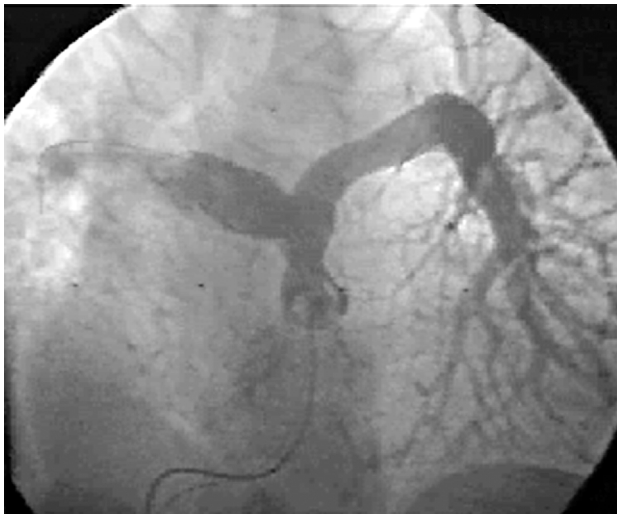

Figure 2.
Four chamber view showing anterograde flow between the infundibulum and the pulmonary arteries subsequent to perforation and dilation of the pulmonary valve.

The mean pulmonary pressure was 12 mmHg after dilation. The patient was discharged on the next day without any medication. Clinical improvement was maintained, with oxygen saturation at rest staying at 90% six months subsequent to the procedure.

Discussion

Although there are sporadic reports of perforation of the atretic pulmonary valve in patients with tetralogy of Fallot and pulmonary atresia,[2–4] as far as we are aware, the procedure has not previously been performed in an adult. In this situation, of necessity there is no anterograde flow into the pulmonary arteries. It is by no means unusual, nonetheless, to find anatomic proximity between the right ventricular infundibulum and the lumen of the pulmonary trunk. In this setting, establishing antegrade flow from the right ventricle to the pulmonary arteries produces the ideal palliative procedure. Growth of the pulmonary arteries is most likely to occur through anterograde flow, and there is no volume overload placed on the left ventricle.[1,5]

We hypothesized that the pulmonary valve was thick and stiff because of the long evolution of the disease. In that circumstance, use of conventional techniques and the regular radiofrequency generator would not open the valve. We decided, therefore, to use the recently available system consisting of a radiofrequency generator, the Nykanen perforation catheter, and a coaxial exchange catheter. The goal of the system is to provide a safer method of perforating the atretic valve using radiofrequency energy without

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mechanical force. The Nykanen perforation catheter is 265 cm in length, with an outer diameter of 0.024 inch, and with a small 0.016-inch diameter active tip. The catheter is not sufficiently stiff to straighten the curve on the guiding catheter, making it easier to position the catheter centrally on the valve. It is designed to be used with the 2.9-French BMC coaxial injectible catheter that has an inner diameter of 0.027 inch and is 145 cm in length. The generator (Baylis Medical Company, Montreal, Canada) has power settings of 1–25 W. When compared to a standard generator, it has a higher impedance cut-off range, perforates using low power over a very short duration, and delivers high voltages so as to perforate rather than ablate the valve. Furthermore, the system is designed to facilitate the exchange of guide wires, thus allowing for relatively easy advancements of balloons of wires in potentially unstable positions. In our patient, we increased pulmonary flow without incrementing the pulmonary pressure. This is consistent with our previous results of dilation in other patients with complex congenital cardiac disease.[6]

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