Pulmonary atresia with intact ventricular septum is a category of cyanotic congenital heart disease which constitutes an emergency in critically-ill newborns. These patients are ductus dependent requiring prostaglandin E) infusions until a systemic-to-pulmonary artery shunt is created or they undergo pulmonary valvotomy. either alone or combined with the shunt. Non-surgical (transcatheter) intervention using radiofrequency perforation techniques for the atretic pulmonary valve is now available as an alternative to surgery. We report successful applications of this technique in 3 patients who tolerated the procedure well and had encouraging follow-up results.

We conclude that radiofrequency opening of atretic pulmonary valve will have a role in the treatment of pulmonary valve atresia with intact ventricular septum and will be a valuable alternative to surgical pulmonary valvotomy.

Patients and Methods

Patient 1
A 2-month-old girl was referred from a peripheral hospital as a case of congenital heart disease with cyanosis and tachypnea; she was a product of a full-term spontaneous vaginal delivery after an uneventful pregnancy. Her weight was 4.3 kg, the first heart sound was normal, and single second heart sound with ejection systolic murmur was 3/6 at the left upper sternal border. The liver was 2 cm below the right costal margin. The echocardiogram showed pulmonary atresia, intact septum, patent ductus arteriosus, and a moderate-sized right ventricle. She underwent radiofrequency-assisted pulmonary valve dilation during the same admission.

Patient 2
A 9-month-old girl was first seen in our center at the age of 2.5 months; she presented to her local hospital at the age of 20 days with a heart murmur,
cyanosis on crying, and mild shortness of breath. On examination, the baby was dusky with mild distress, first heart sound was normal, single second heart sound, and a grade 3/6 systolic murmur at the left upper sternal border. The liver was 2 to 3 cm below the right costal margin. The echocardiogram showed pulmonary, atresia with intact ventricular septum, tricuspid valve stenosis, patent ductus arteriosus, severe tricuspid regurgitation, and small ostium secundum atrial septal defect with right-to-left shunt. The patient was admitted at the age of 2.5 months; cardiac catheterization showed pulmonary valve atresia, severe tricuspid regurgitation, and dilated right atrium, to be managed surgically with pulmonary valvotomy. She was re-admitted at the age of 9 months and radiofrequency-assisted pulmonary valve dilation was performed successfully.

**Patient 3**

A 2-week-old boy, the product of a full-term cesarean section delivery, presented at the age of one day with cyanosis and shortness of breath; he was started on prostaglandin E1 infusion. On examination, first heart sound was normal, there was a single second heart sound, a grade 3/6 ejection systolic murmur at the left upper sternal border, and the liver was 3 cm below the right costal margin. The echocardiogram showed pulmonary atresia with intact ventricular septum, patent ductus arteriosus, and moderate tricuspid regurgitation. The patient was intubated electively, and catheterization with radiofrequency-assisted pulmonary valve dilation was performed (Table 1).

### Methods

#### Radiofrequency Equipment

The 0.014-inch radiofrequency guidewire (Osypka, Germany) used in all 3 patients was developed specifically for this procedure. The 120-cm long insulated guidewire terminated in a 3mm long metal tip which was heated during energy delivery. The radiofrequency guidewire was connected to a HAT 200 radiofrequency generator (Osypka) in a unipolar fashion; a large indifferent paddle was placed in contact with the patient's back to complete the circuit after it was soaked in hypersaturated saline solution for better conduction.

**Procedure**

Under general anesthesia and intubation, the femoral artery and vein were cannulated percutaneously in all 3 patients. A SF sheath was placed in the femoral vein and a 4F sheath was passed retrogradely into the femoral artery. The 3 patients were fully heparinized. A SF right coronary catheter was passed through the arterial sheath to the descending aorta and the patent ductus arteriosus into the main pulmonary artery to serve as a landmark. Another SF right coronary catheter was positioned in the right ventricle outflow tract below the atretic pulmonary valve (Figure 1). Simultaneous angiograms were done to orientate the catheter tip. The radiofrequency guidewire was then passed up to the venous catheter into contact with the inferior surface of the pulmonary valve. Radiofrequency energy was then delivered continuously while the radiofrequency guidewire was slowly advanced. However, power was not applied when the machine cuts off because of a very high impedance. After a few seconds power was then re-applied. The energy setting used ranged from 5 to 25 watts. After the valve was penetrated, the radiofrequency wire was replaced with a 0.014inch coronary wire which was snared by a 10-mm Amplatz 2 snare from the arterial side. In all 3

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Age</th>
<th>Weight (kg/sex)</th>
<th>Diagnosis</th>
<th>Balloon size (mm)</th>
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<tbody>
<tr>
<td>1</td>
<td>2mo</td>
<td>4.3/F</td>
<td>PVA, IVS, PDA</td>
<td>2.5, 4, 5, 8, 10</td>
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<tr>
<td>2</td>
<td>9mo</td>
<td>6.7/F</td>
<td>PVA, IVS, severe TR, ASD</td>
<td>5, 10</td>
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<tr>
<td>3</td>
<td>2wk</td>
<td>3.31M</td>
<td>PVA, IVS, PDA</td>
<td>6, 8, 10</td>
</tr>
</tbody>
</table>

PV A = pulmonary valve atresia; IVS = intact ventricular septum; PDA = patent ductus arteriosus; TR = tricuspid regurgitation; ASD = atrial septal defect.
PULMONARY ATRESIA

Figure 1. Lateral projection of right ventricular outflow angiogram showing complete pulmonary valve atresia.

Figure 2. Lateral projection showing the guidewire passing from right ventricle through the hole created in the pulmonary valve to the patent ductus arteriosus and down the descending aorta.

Figure 3. Lateral projection of 4 mm-diameter balloon inflated at the pulmonary valve area.

complete the dilation, starting with a 2.5-mm low profile balloon. This was followed by a 4- or 5-mm diameter balloon and then an 8- or 10-mm diameter balloon (Figure 3).

Results

In all 3 patients the valve was successfully crossed by radiofrequency guidewire (the valve was dilated to 10 mm) (Figure 4), and immediate hemodynamic improvement was evident in all of the patients (Table 2).

Patient 1 was ventilated for 2 weeks due to development of sepsis (Kebcialla species) and left lung collapse. She was treated with antibiotics (ceftiraxone and flucloxacillin) and was started on digoxin and lasix on the second day. Echocardiography revealed some tricuspid regurgitation. She was sent home with oxygen saturation of 78%. Seen 2 months later in the outpatient department, the patient was asymptomatic and gaining weight with no cyanosis. The echocardiogram showed patent ductus arteriosus, pulmonary valve gradient flow of 10 mm Hg, mild pulmonary regurgitation, and mild-to-moderate tricuspid regurgitation. The calculated right ventricular pressure was 40 mm Hg and there was right ventricular hypertrophy.

Patient 2 was extubated immediately after the
Figure 4. Lateral projection of right ventricular cineangiogram showing complete patency of the right ventricular outflow tract following radiofrequency-assisted balloon dilation of the pulmonary valve.

Table 2. Hemodynamic data (mm Hg) before and after radio frequency-assisted pulmonary valve dilation.

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Right ventricle</th>
<th>Pulmonary artery</th>
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<tbody>
<tr>
<td>1</td>
<td>Before</td>
<td>After</td>
</tr>
<tr>
<td></td>
<td>148/0-14</td>
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<td>30/5-15</td>
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<tr>
<td>3</td>
<td>150/0-20</td>
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</table>

procedure with an oxygen saturation of 93% on lasix. Six weeks after the procedure, an echoDoppler examination confirmed right ventricle to main pulmonary artery flow with a gradient of 26 mm Hg with mild tricuspid regurgitation.

Patient 3 was extubated on the eighth day of the procedure with oxygen saturation of 88% to 90% , the echocardiogram showed residual PS gradient of 45 mm Hg and there was mild tricuspid regurgitation. The patient was weaned off prostaglandin EI and discharged home 13 days after the procedure. Seen 6 months later, the patient was growing well, asymptomatic, had an oxygen saturation of 97% by pulse oxymeter, and was not on any medications. The echo cardiogram showed a residual pulmonary valve stenosis gradient of 55 mm Hg and a good-sized right ventricle.

Complications

Patient 1 developed bacteremia. After being treated with antibiotics for 2 weeks, she improved and was then extubated. Patient 2 had pulse less right lower limb immediately after the procedure and was treated by intravenous heparin and streptokinase for 2 days. No significant complications were observed in Patient 3.

Discussion

Pulmonary atresia with intact ventricular septum is a difficult and challenging medical as well as surgical problem. It is very difficult to predict which mode of treatment would be more suitable for each patient.

As a result, research trials were conducted to determine the most effective mode of therapy with the least hospital mortality. At the beginning, surgical palliation (shunt with or without valvotomy) with a mortality rate of approximately 10%4 took the upper hand. But with recent advances in pediatric cardiology during the last two decades, particularly with more invasive interventional procedures being used, perforation of the atretic pulmonary valve was first tried by using direct needle puncture followed by graduated balloon dilation. However, there were non-encouraging outcomes and high mortality.5 In 1974, with the introduction of laser therapy in cardiology as a therapeutic modality, the use of the laser beam to perforate the atretic pulmonary valve intraoperatively was introduced.6 Transcatheter perforation laser-assisted balloon pulmonary valve dilatation in pulmonary valve atresia was used in 5 children (2 had ventricular septal defects and 3 had intact septum); although there was one mortality, 2 there were encouraging results using the Nd- Y AG hot tip laser wire. But the laser beam had the disadvantages of being expensive and hazardous. When radiofrequency energy was introduced in 1991 in the management of cardiac arrhythmia,7 perforation of the atretic pulmonary valve with radiofrequency energy was tried by Rosenthal et al. I They apply it on 4 children as a prospective multicenter study; there were 2 deaths, one from septicemia 6 days later and the other from left ventricular infarction 17 days later. With more understanding of this mode of energy,
radiofrequency-heated guidewire has been found to be an effective modality for treating pulmonary atresia with intact septum.

Conclusion

In this paper, we present our initial experience of this new procedure. Although our patient number was small, our initial results were encouraging. Radiofrequency-heated guidewires offer a promising future for infants who have been diagnosed to have pulmonary atresia with intact septum. The procedure omits the need for surgery at the early stages or decreases the number of surgical steps to be undertaken later.

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References