CASE REPORT

Diagnosis and Management of Postoperative Pulmonary Hypertensive Crisis

JOHN WHeller, M.D., BARBARA L. GEORGE, M.D., DONALD G. MulDER, M.D., AND JAY M. JARMakAni, M.D.

SUMMARY In this paper we discuss two infants and one child who experienced a previously unreported complication after complete correction of a large, unrestrictive ventricular septal defect. Two patients had documented pulmonary hypertensive crises and severe right-heart failure secondary to hypoxia and pulmonary vasoconstriction. These crises were associated with significantly increased right ventricular (RV) peak systolic and end-diastolic pressures and right-to-left shunting via a foramen ovale which, in turn, exaggerated the hypoxia. The crises were treated successfully with tolazoline in the second and third patients. RV pressures returned to normal values and have remained normal up to 12 months postoperatively in the second patient. Although the RV pressures decreased with tolazoline in the third patient, they never reached normal values. Postoperative monitoring of pulmonary artery and RV pressures in infants with large ventricular septal defects is essential when unexplained complications are encountered. Tolazoline proved to be very effective in the treatment of two patients with pulmonary vasoconstriction secondary to hypoxia.

SURGICAL CLOSURE of the isolated, large ventricular septal defect (VSD) is the treatment of choice for the infant with this lesion when either protracted heart failure or increasing pulmonary vascular resistance (PVR) is present. Although the list of possible complications after surgical repair of a VSD is extensive, there have been no reports of postoperative pulmonary hypertensive crises resulting in profound hypoxemia and right-heart failure. It is generally accepted that the PVR of infants with large intracardiac left-to-right shunts remains elevated because of persistent hypertrophy of the medial musculature of the pulmonary arterioles. Increased PVR may also be seen in newborns with otherwise normal hearts. This entity, known as persistence of the fetal circulation (PFC), may result in significant hypoxemia secondary to right-to-left shunting across a patent foramen ovale and/or a patent ductus arteriosus. Tolazoline was used recently to treat infants with PFC, and it produced a significant decrease in the PVR and the right-to-left shunt. This treatment, however, is not always successful and might cause significant decrease in the systemic vascular resistance and an increased right-to-left shunt. The possibility that tolazoline might relieve pulmonary vasoconstriction postoperatively led to its use in two of the three cases presented here. We describe the course and management of pulmonary hypertensive crises after the surgical correction of a VSD.

Case Reports

Patient 1 was seen at 3 weeks of age with the clinical findings of a large VSD and congestive heart failure. Cardiac catheterization (table 1) revealed a nonrestrictive, membranous VSD with a large left-to-right shunt (Qp/Qs 3.28) and normal PVR (Rp/Rs 0.20). Medical management with digoxin, furosemide and spironolactone failed, and the VSD was closed surgically at 3.5 months of age (weight 3.4 kg). The procedure was performed using cardiopulmonary bypass and moderate hypothermia (27°C). The defect was closed with a teflon patch through a right ventriculotomy. There were no intraoperative or immediate postoperative complications except for a persistently low arterial PO2. This significant systemic hypoxemia was present despite excellent ventilation with an FiO2 of 0.90 (table 2). The lungs were clear both clinically and radiographically. A stable hemodynamic state existed for 24 hours. Because of persistent hypoxemia, the infant was maintained on a respirator (Baby Bird) and paralyzed with pancuronium bromide (Pavulon). Various levels of inspiratory and end-expiratory pressures were tried without significant change in arterial blood gases. During the second postoperative day, the infant experienced four episodes of abrupt deterioration characterized by hypoxemia, an acutely enlarged liver and the appearance of a high-pitched holosystolic murmur at the left lower sternal border. During these episodes, oxygen saturation in the left atrium was 10% greater than that in the brachial artery, indicating a
significant right-to-left intracardiac shunt. There were no apparent precipitating causes. The episodes were aborted by the use of hyperventilation sufficient to produce significant respiratory alkalosis. These findings are summarized in Table 2. The episodes increased in frequency and severity during the third postoperative day, became resistant to hyperventilation, and the patient died 90 hours postoperatively. Postmortem examination showed a patent foramen ovale but no residual VSD or other cardiac defects.

Patient 2 presented at 2 months of age with the clinical findings of a large VSD and congestive heart failure. Cardiac catheterization (Table 1) showed a nonrestrictive, membranous VSD with a large left-to-right shunt (Qp/Qs 4.33) and normal PVR (Rp/Rs 0.18). Medical management during the following month failed, and the VSD was closed surgically. The surgical technique and findings were similar to those of patient 1.

There were no intraoperative or immediate postoperative complications except for systemic hypoxemia. On examination, the lungs were clear and the air exchange was excellent. No murmur was noted. The chest film showed clear lung fields and no significant change in the heart size. The infant was paralyzed with pancuronium bromide (Pavulon) and ventilated with a Baby Bird respirator. Despite the hypoxemia, the infant remained stable, with a normal blood pressure, left atrial pressure, urine output and acid-base balance. Twelve hours postoperatively, he developed "hypoxic spells" associated with an abrupt increase in the liver size and the appearance of a grade III/VI high-pitched holosystolic murmur heard best at the left lower sternal border. The clinical findings are summarized in Table 2. These episodes were not preceded by any change in the patient's respiratory or hemodynamic status. After three such episodes, a #5 Berman catheter was introduced via a cutdown in the

### Table 1. Preoperative Cardiac Catheterization Data

<table>
<thead>
<tr>
<th>Patient</th>
<th>Patient 1</th>
<th>Patient 2</th>
<th>Patient 3</th>
<th>Patient 3</th>
<th>Patient 3</th>
</tr>
</thead>
<tbody>
<tr>
<td>O₂ sat (%)</td>
<td>Pressure (s/d)</td>
<td>O₂ sat (%)</td>
<td>Pressure (s/d)</td>
<td>O₂ sat (%)</td>
<td>Pressure (s/d)</td>
</tr>
<tr>
<td>RA</td>
<td>68</td>
<td>10/0</td>
<td>3</td>
<td>67</td>
<td>5/0</td>
</tr>
<tr>
<td>RV</td>
<td>83</td>
<td>90/8</td>
<td>—</td>
<td>83</td>
<td>80/5</td>
</tr>
<tr>
<td>LA</td>
<td>86</td>
<td>85/18</td>
<td>55</td>
<td>87</td>
<td>80/22</td>
</tr>
<tr>
<td>Ao</td>
<td>93</td>
<td>12/3</td>
<td>8</td>
<td>93</td>
<td>9/0</td>
</tr>
<tr>
<td>Qp/Qs</td>
<td>3.28</td>
<td>4.33</td>
<td>—</td>
<td>1.73</td>
<td>2.33</td>
</tr>
<tr>
<td>Rp/Rs</td>
<td>0.20</td>
<td>0.18</td>
<td>0.24</td>
<td>0.23</td>
<td>0.24</td>
</tr>
</tbody>
</table>

Abbreviations: S/D = systolic/diastolic; RA = right atrium; RV = right ventricle; O₂ sat = oxygen saturation; PA = pulmonary artery; LA = left atrium; Ao = aorta; Qp/Qs = pulmonary flow/systemic flow; Rp/Rs = pulmonary vascular resistance/systemic vascular resistance.

### Table 2. Arterial Blood Gases and the Clinical Findings

<table>
<thead>
<tr>
<th>Patient</th>
<th>Patient 1</th>
<th>Patient 2</th>
<th>Patient 3</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fio₂</td>
<td>Pre-op</td>
<td>Steady state</td>
<td>Crisis</td>
</tr>
<tr>
<td>Fio₂</td>
<td>0.50</td>
<td>0.50</td>
<td>0.90</td>
</tr>
<tr>
<td>PₐO₂ (torr)</td>
<td>171</td>
<td>40</td>
<td>27</td>
</tr>
<tr>
<td>O₂ sat (%)</td>
<td>99</td>
<td>76</td>
<td>53</td>
</tr>
<tr>
<td>Pco₂ (torr)</td>
<td>35</td>
<td>40</td>
<td>48</td>
</tr>
<tr>
<td>Base excess</td>
<td>+3.5</td>
<td>+1.0</td>
<td>+2.5</td>
</tr>
<tr>
<td>pH</td>
<td>7.42</td>
<td>7.42</td>
<td>7.38</td>
</tr>
<tr>
<td>Systolic murmur (0 to 6/6)</td>
<td>4</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td>Liver enlargement (cm below costal margin)</td>
<td>3</td>
<td>2</td>
<td>6</td>
</tr>
</tbody>
</table>
left groin and advanced to the right ventricle. The right ventricular (RV) and aortic pressures were recorded simultaneously (fig. 1). During the steady state (panel A), the RV and aortic pressures were 55/5/20 mm Hg (systolic/diastolic/end-diastolic) and 120/70 (systolic/diastolic) mm Hg, respectively. During the hypoxic crises, however, the RV pressures rose to 110/5/35 mm Hg (fig. 1B) and occasionally were as high as 140/5/37 mm Hg (not shown in fig. 1). During two of these episodes the catheter was advanced into the pulmonary artery (PA). Pressure pullback from the PA to the right ventricle showed no significant gradient across the RV outflow tract. Oxygen saturation in the radial artery was 12% less than that of the left atrium, indicating a right-to-left intracardiac shunt. He responded to hyperventilation, but significant systemic hypoxemia (Po2 40–50 torr) persisted despite an Fio2 of 1.00. Tolazoline (1 mg/kg) was injected into the right ventricle and was repeated 5 minutes later. Within 2 minutes after the second injection, the systemic arterial Po2 increased to 174 torr. The lowest postoperative RV pressures were recorded approximately 30 minutes after the tolazoline was given (30/0/6 mm Hg) (fig. 1C). A constant intravenous infusion of tolazoline (1 mg/kg/hour) was continued for 48 hours. The improvement in oxygenation and lowered RV pressures persisted after the tolazoline was discontinued. The patient was extubated 24 hours after starting tolazoline and was discharged on the tenth postoperative day. Subsequent clinical follow-up 12 months postoperatively has shown him to be thriving and off all cardiac medications. Follow-up cardiac catheterization, performed 12 months postoperatively, showed normal PA pressures (28/5 mm Hg) and no residual shunt.

Patient 3, a 3-year-old child with Down’s syndrome, was first seen at 2 months of age for congestive heart failure. The clinical findings were a VSD and a large left-to-right shunt. The family refused cardiac catheterization until the patient was 3 years old. Catheterization at that time (table 1) showed unrestricted VSD with a moderate left-to-right shunt (Qp/Qs 1.73) and increased PVR (Rp/Rs 0.44) Administration of 80% oxygen by mask for 15 minutes resulted in an increased left-to-right shunt (Qp/Qs 2.33) and a decreased PVR (Rp/Rs 0.24). Fifteen minutes after discontinuing the supplemental oxygen, tolazoline (1 mg/kg) was injected into the PA.

**Figure 1.** Right ventricular (RV) pressures (upper panel) and aortic pressures (lower panel) in patient 2 during the steady state (before the pulmonary hypertensive crisis), during the crisis, and after the institution of tolazoline. Peak systolic and end-diastolic RV pressures increased dramatically during the hypoxic crisis and returned to normal values after tolazoline treatment was begun. The peak RV and pulmonary artery pressures were identical.

**Figure 2.** Postoperative right ventricular (RV) pressures (upper panel) and aortic pressures (lower panel) in patient 3 during hypoxia and after tolazoline treatment. RV pressure decreased dramatically after tolazoline treatment was begun. The peak systolic RV and PA pressures were equal.
minutes after the injection, the Qp/Qs (2.42) and Rp/Rs (0.23) ratios were similar to the values obtained during high oxygen inhalation. Because of these data and the clinical findings (long systolic murmur, combined ventricular hypertrophy on ECG and cardiomegaly on chest film), the VSD was surgically closed through a right ventriculotomy, as described in patient 1. After discontinuing the cardiopulmonary bypass, the aortic and RV pressures were 85/55 and 45/5 mm Hg, respectively. After 5 hours of a relatively stable postoperative course, the liver became larger and the arterial Po2 decreased to 29 torr (table 2). The lungs were clear and the air exchange was normal. Oxygen saturation in the left atrial sample was 9% greater than that in the brachial artery sample. The heart size on chest film was unchanged and the lungs were clear. Because we anticipated pulmonary vasoconstriction, a #5 Berman catheter was introduced percutaneously into the femoral vein and maneuvered into the PA. Pressure pullback showed no significant gradient across the RV outflow tract. The RV peak systolic pressure (160 mm Hg) was greater than the aortic peak systolic pressure (fig. 2). Tolazoline (1 mg/kg) was initially injected into the right ventricle and was followed by a constant infusion of tolazoline (1 mg/kg/hour) for 3 days. Twenty minutes after the initial injection, the arterial Po2 rose to 78 torr and the RV peak systolic pressure decreased to 65 mm Hg. The peak systolic RV pressure ranged from 50–65 mm Hg and the patient was extubated 8 hours later. The arterial Po2 and RV systolic pressure remained stable. All catheters were removed on the fourth postoperative day, and the patient was discharged on the tenth postoperative day in mild right-heart failure. Postoperatively, the patient’s exercise tolerance improved significantly but one episode of pulmonary infection was associated with increased right-heart failure. Postoperative cardiac catheterization 3 months later showed no evidence of residual VSD. The RV (70/20 mm Hg) and PA pressures (70/20 mm Hg, mean 35 mm Hg) remained increased. Tolazoline (1 mg/kg into the PA) and oxygen (FiO2 0.70) reduced the PA pressure to 55/15 mm Hg (mean 35 mm Hg).

Discussion

The postoperative pulmonary hypertension in patients 1 and 2 was not expected because these infants had normal PVR preoperatively. The infant with large left-to-right shunt and pulmonary hypertension maintains muscular hypertrophy of the pulmonary arterioles7,8 and may respond to hypoxia with significant pulmonary vasoconstriction. In our patients, the mild hypoxemia caused pulmonary vasoconstriction resulting in severe pulmonary hypertension, tricuspid valvular regurgitation, right-heart failure and intrapulmonary right-to-left shunting. This, in turn, resulted in increased hypoxemia and further aggravation of the pulmonary vasoconstriction. Intrapulmonary right-to-left shunting is known to occur after cardiopulmonary bypass.16-19 This probably explains the hypoxemia noted in our patient during the steady state. Further deterioration in ventilatory status could lead to increased hypoxemia and thereby precipitate a hypertensive crisis.

Initially, hyperventilation alone reversed the crises and improved the clinical status of patients 1 and 2. The crises, however, became more frequent, more severe, and were eventually refractory to hyperventilation. The response to tolazoline in patients 2 and 3 was dramatic (figs. 1 and 2; table 2). Both RV peak systolic pressures and the arterial Po2 returned to normal or near normal values. These findings indicate that the institution of tolazoline resulted in a significant decrease in the PVR with a subsequent elimination of the right-to-left shunt. Tolazoline was infused initially into the right ventricle and subsequently into a peripheral vein without perceptible change in the patient’s response. Patient 2 continued to do well for the 12 months that he has been followed postoperatively, and the PA pressures were normal as determined by follow-up cardiac catheterization. Patient 3 continued to have increased RV and PA pressures, and the effect of tolazoline and oxygen on the PVR was limited. This indicates that the change in most of the pulmonary vascular bed was permanent. Although the postoperative clinical course improved in this patient, the long-term prognosis remains unknown.

Inotropic medication was ineffective in treating these hypertensive hypoxic crises; tolazoline, however, was very effective. The clinical courses of these patients underline the need for PA pressure monitoring in infants with unexplained complications during the first 24–48 hours after the repair of unrestrictive VSDs. Tolazoline proved effective in two cases with postoperative pulmonary hypertensive crises and deserves further careful evaluation.

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References

Two-dimensional Echocardiography and B-mode Ultrasonography for the Diagnosis of Loculated Pericardial Effusion

MARK J. FRIEDMAN, M.D., DAVID J. SAHN, M.D., AND KAI HABER, M.D.

SUMMARY Two cases of loculated pericardial effusion resulting in cardiac tamponade are presented. The loculated nature and extent of the effusion was best defined by two-dimensional echocardiography or B-mode ultrasonography. Cross-sectional images should probably be obtained in all cases of suspected loculated pericardial effusion and in patients in whom the interpretation of the M-mode echocardiogram is equivocal as to the presence or absence of pericardial effusion.

M-MODE ECHOCARDIOGRAPHY is a sensitive and specific technique for the detection of nonloculated pericardial effusion.1, 2 Recent reports of two-dimensional echocardiography suggest that this method is also clinically useful for evaluating pericardial effusion.3 In contrast to the positive experience with these techniques for the detection of nonloculated pericardial effusion, loculated pericardial effusions have frequently resulted in false-negative M-mode echocardiographic studies.4, 6 This report demonstrates the usefulness of two-dimensional echocardiography and B-mode ultrasonography in two patients with loculated pericardial effusion.

Case 1
A 28-year-old Mexican male was admitted to the University of Arizona Health Sciences Center for evaluation of sharp, substernal chest pain, shortness of breath and cardiomegaly. On physical examination, the blood pressure was 92/70 mm Hg with 14 mm Hg pulsus paradoxus. The jugular veins were distended to the angle of the jaw with the patient sitting upright. The first and second heart sounds were normal and a three-component pericardial friction rub was heard over the anterior precordium. A chest radiograph showed marked enlargement of the cardiac silhouette, but was otherwise unremarkable. The diagnosis of pericarditis with cardiac tamponade was made and an M-mode echocardiogram was obtained using a Smith-Kline Ekoline 20A ultrasonoscope with a 2.25-MHz transducer. Standard sweeps from the aortic root to the ventricle were attempted using previously described techniques,7 however, a limited echo window resulted in a suboptimal M-mode echocardiogram. The M-mode echocardiogram (fig. 1) demonstrated a large, echo-free space posterior to the left ventricular wall extending behind the left atrium. A small anterior

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