Successful Pregnancy in a Patient With Double Outlet Left Ventricle After a Rastelli Operation Using a Prosthetic Valve

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A woman with double outlet left ventricle (DOLV) had undergone a Rastelli operation using a prosthetic Björk-Shiley valve and who was receiving anticoagulant drug delivered a healthy male infant. Oral warfarin was replaced by heparin from the 5th to the 13th week of gestation and for the last 5 weeks of gestation. Successful pregnancy in patients with DOLV after a Rastelli operation using a prosthetic valve is possible with careful maintenance. (Circ J 2004; 68: 501–503)

Key Words: Anticoagulant therapy; Double outlet left ventricle; Pregnancy; Prosthetic valve; Rastelli operation

Double outlet left ventricle (DOLV) is a rare (incidence of fewer than 1 in 200,000) congenital cardiac malformation in which the aorta and the pulmonary trunk both arise entirely or predominantly above the morphological left ventricle. Increasing numbers of children with congenital heart disease (CHD) are surviving to reproductive age, and successful pregnancies in patients who have undergone surgery for CHD are being reported with increasing frequency. However, there have been no reports to date on pregnant patients who have undergone a radical operation using a prosthetic valve for DOLV.

Pregnant women with a prosthetic valve replacement present significant problems in management, primarily because of the high incidence of embolization and therefore they require prophylaxis against thromboembolism.

We report here on a successful planned delivery in a patient with DOLV who had undergone a Rastelli operation using a prosthetic valve.

Case Report

A pregnant 35-year-old woman (gravida 4, para 1, aborta 3) who had been diagnosed with DOLV was referred at 4 weeks' gestation. At age 11, she had undergone a Rastelli operation in which the pulmonary valve was replaced with a 21 mm Björk-Shiley prosthesis. After surgery, anticoagulant therapy with warfarin was initiated, and her heart condition improved from New York Heart Association (NYHA) functional class III to class I. Postoperative cardiac catheterization and angiographic study were performed at 2 and 7 years (Table 2) after operation and the patient’s right ventricular pressure was 56/0 mmHg and 46/0 mmHg, respectively. There was no residual ventricular septal defect shunt.

The patient married at age 24 years and became pregnant at 26, but the fetus died at 8 weeks gestation and the patient subsequently underwent an evacuation without complications. Her second pregnancy occurred at the age of 28 years. At 32 weeks' gestation, the prosthetic click disappeared and at this point, the patient’s warfarin intake was 3 mg daily. The diagnosis of prosthetic obstruction because of probable valve thrombosis was made, and intravenous heparin therapy was initiated. The prosthetic click remained inaudible, but there was no change in the patient’s

### Table 1 Cardiac Catheterization at 2-Years Postoperative

<table>
<thead>
<tr>
<th>Pressure (mmHg)</th>
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<tbody>
<tr>
<td>SVC</td>
<td>s (a)</td>
</tr>
<tr>
<td>(4)</td>
<td>(4)</td>
</tr>
<tr>
<td>IVC</td>
<td>(5)</td>
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<tr>
<td>RA</td>
<td>(4)</td>
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<tr>
<td>RV</td>
<td>56</td>
</tr>
<tr>
<td>LV</td>
<td>110</td>
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<td>Ao</td>
<td>112</td>
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SVC, superior vena cava; IVC, inferior vena cava; RA, right atrium; RV, right ventricular; LV, left ventricular; Ao, aorta.

### Table 2 Cardiac Catheterization at 7-Years Postoperative

<table>
<thead>
<tr>
<th>Pressure (mmHg)</th>
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<tr>
<td>SVC</td>
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<tr>
<td>(8)</td>
<td>(6)</td>
</tr>
<tr>
<td>IVC</td>
<td>(6)</td>
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<tr>
<td>RA</td>
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<tr>
<td>RV</td>
<td>46</td>
</tr>
<tr>
<td>LV</td>
<td>118</td>
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<tr>
<td>Ao</td>
<td>118</td>
</tr>
</tbody>
</table>

SVC, superior vena cava; IVC, inferior vena cava; RA, right atrium; RV, right ventricular; LV, left ventricular; Ao, aorta.
condition and she was kept under observation. At 37 weeks' gestation, she gave birth to a healthy baby by cesarean section under general anesthesia. The baby showed no indications of fetal warfarin syndrome. After the cesarean section, the valve click once again became audible.

Although the patient's next 2 pregnancies ended in spontaneous abortions, she was eager to have one more child. She knew of our earlier case of a successful planned pregnancy in a woman with a prosthetic valve, and wished to follow the same protocol; that is, substitution of heparin for warfarin from 6 to 12 weeks' gestation and at the end of pregnancy. The fetal and maternal risks were thoroughly explained to both the patient and her husband, according to the ACC/AHA guidelines for the management of patients with valvular heart disease and written informed consent was obtained. The protocol for the present patient was based on the accepted guidelines for anticoagulation therapy in pregnancy and on our previous case.

At the 5th week of gestation, the patient was admitted to hospital, warfarin (3.25 mg/day) was discontinued and treatment with subcutaneous heparin was begun. The heparin dose was adjusted to maintain the partial thromboplastin time ratio (patient/control) between 1.8 and 2.3 during this period of gestation. During the 14th week of gestation, warfarin was reintroduced and the dose was adjusted according to the prothrombin time to maintain a British corrected ratio of 2 to 3 times the control value. Subcutaneous heparin therapy was discontinued when the target prothrombin time was achieved. Warfarin therapy (3.25 mg/day) was then continued until the 34th week of gestation. Fetal echocardiography at 20 weeks' gestation showed no evidence of fetal CHD. At the 34th week, the patient was again admitted to the hospital. Warfarin was discontinued, and intravenous heparin was given, adjusted to maintain a partial thromboplastin time ratio (patient/control) between 1.8 and 2.3 for the remainder of gestation.

Elective cesarean section was planned to take place at 38 weeks' gestation, and because she had had a previous cesarean section, intravenous heparin was temporarily reduced from 35,000 IU/day to 7,000 IU/day. The patient successfully delivered a 2,944 g male infant by cesarean section. The infant's Apgar score was 8 and 9 at 1 and 5 min, respectively, and by 10 min, his Apgar score had risen to 10. Cord blood coagulation was normal; the placenta was also normal. In the absence of hemorrhagic complications, intravenous heparin was resumed. Three days after surgery, the patient was again started on warfarin therapy. The infant was normal at birth and has subsequently done well and the patient's postpartum course was uneventful. She was given prophylactic antibiotic therapy perioperatively against infective endocarditis. Mother and child were discharged on the 16th post-delivery day. The patient's heart condition did not change from NYHA functional class I at any point during the pregnancy or delivery.

Discussion

Advances in the medical and surgical management of patients with CHD have increased the number of women with cardiac problems who survive to childbearing age. The impact and outcome of pregnancy in these mothers and the effect of the mother's heart condition on her children have been matters of concern. Today, both improved comprehension of the physiologic and cardiodynamic responses to pregnancy and the use of modern equipment provide greater safety and better results for pregnant CHD women.

In the Rastelli operation, which is suitable for patients diagnosed with large ventricular septal defects and pulmonic stenosis, the left ventricle is used as the systemic ventricle; however, the operation involves the use of ventricular conduits, which may restrict blood flow, and the prosthetic valve in the right ventricle-to-pulmonary artery conduit can become stenotic and/or regurgitant, especially if it is a tissue valve.

Kreutzer et al reported that the Rastelli repair can be performed with low early mortality, but that substantial late morbidity and mortality are associated with conduit obstruction and arrhythmias. There are few previous reports on pregnancy in patients who have undergone the Rastelli operation and there is no published information to date about pregnancy in DOLV patients.

Dore et al, in a report on patients with pulmonary autograft valve replacement, state that no significant progress of aortic regurgitation, pulmonary regurgitation or right-sided obstruction occurred during pregnancy in 14 pregnancies in 8 women. Furthermore, pregnancy appeared to have no effect on the function of the pulmonary autograft valve or on the right-sided homograft. Reoperation for right-sided obstruction was carried out in 2 patients at 4 and 7 years after a second pregnancy. In the present case, there were no clinical problems associated with conduit or valve dysfunction or arrhythmias.

Maternal cardiac function status is known to exert a strong influence on outcome, and the potential for a successful outcome in pregnancy is determined by the level of derangement with which the patient enters pregnancy. The risk of maternal mortality and morbidity is correlated to the type and severity of CHD and to the patient's functional class. Patients at high risk are (1) those showing signs and symptoms of heart failure, (2) those with pulmonary hypertension, (3) those with severe reduction of ventricular contractility and (4) those with outflow stenosis.

Women with CHD may also be at risk for endocarditis, which, although rare, can have dire consequences. Seaworth et al report an overall maternal mortality rate of 29% with a fetal mortality rate of 23%. An uncomplicated vaginal delivery should not pose a significant risk because the incidence of bacteremia is quite low, at approximately 0–5%. Women at risk should receive prophylaxis that continues into the postpartum period and ampicillin and gentamicin are usually recommended for this purpose.

Whittemore et al have reported an incidence of approximately 16% of infants with cardiac anomalies born to mothers with CHD; it is therefore important that expert fetal cardiac scanning be performed. Genetic counseling may serve to relieve the mother’s anxiety and if possible, fetal echocardiography is also recommended.

Patients with a prosthetic valve are able to tolerate the hemodynamic load of pregnancy well. There are few problems in patients who are assigned to NYHA functional class I or II before pregnancy. Cumulative experience suggests that the incidence of thromboembolic events during pregnancy in women on anticoagulants is comparable with that in the nonpregnant population and can be further reduced by careful adjustment of the anticoagulant dosage. Thus our recommended strategy for managing a patient with a mechanical prosthetic valve is as follows: warfarin should be stopped before the 6th week of gestation, and adjusted-dose subcutaneous heparin should be stopped.
started and continued until the end of the first trimester. The activated partial thromboplastin time should be adjusted to 1.8–2.3 times normal. The patient may be started on warfarin during the second trimester. Near term, the patient is admitted to hospital, warfarin is stopped and intravenous heparin is substituted. Heparin is stopped at the onset of labor and restarted after delivery in the absence of maternal hemorrhagic complications. Warfarin is started at 24 h after delivery and overlaps with intravenous heparin for 4 days before heparin is stopped.

The risk of warfarin embolopathy ranges from 6.8% to 7.9% if warfarin is taken between 6 and 12 weeks of gestation. The present patient’s second pregnancy resulted in the delivery of a healthy child in spite of warfarin intake. Nevertheless, it is important to keep in mind that the use of warfarin during pregnancy is associated with a high spontaneous abortion rate ranging between 16.2% and 44%. The patient suffered three abortions that might have been avoided had heparin been substituted for warfarin during the first trimester of gestation. The case reported here proves that it is possible for patients with DOLV that has been surgically repaired with a prosthetic valve (Rastelli operation) and in NYHA functional class I to complete a successful pregnancy. Careful cardiac and obstetric management is necessary for a good maternal and fetal outcome.

References