CASE REPORTS

Living related donor liver transplantation in a patient with severe aortic stenosis

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We report the successful anaesthetic management of a young girl with Alagille’s syndrome and severe aortic stenosis (resting pressure gradient 88 mm Hg) undergoing living related donor liver transplantation (LRDLT). The patient had end-stage liver disease and LRDLT was performed before replacement of the aortic valve. Anaesthesia was conducted uneventfully with the aid of a pulmonary artery catheter. Intra-aortic balloon pumping was used in the perioperative period for protection against myocardial ischaemia. Total clamping of the inferior vena cava was avoided during surgery and volume administration was guided by the pulmonary artery pressure. A stable circulation was maintained in the reperfusion period. The patient was discharged from hospital on day 54 after operation with normal liver function. Two years later her aortic valve was replaced successfully.

Br J Anaesth 1999; 83: 488–90

Keywords: liver, transplantation; complications, Alagille’s syndrome; complications, aortic stenosis

Accepted for publication: March 23, 1999

Although severe aortic stenosis has been reported to increase the risk of non-cardiac operations,1 selected patients with severe aortic stenosis may undergo non-cardiac procedures with a reasonably low risk if cardiovascular status is monitored carefully during anaesthesia.2 The pathophysiological conditions in aortic stenosis dictate that anaesthetic management should be based on avoidance of systemic hypotension, maintenance of sinus rhythm and an adequate intravascular volume, with awareness of the potential for myocardial ischaemia.3

Patients with end-stage liver disease tend to demonstrate low systemic vascular resistance and liver transplantation often causes cardiovascular instability and intractable haemorrhage.4 These factors exacerbate the problems of providing anaesthesia in a patient with aortic stenosis. We report successful anaesthetic management for living related donor liver transplantation (LRDLT) in a patient with Alagille’s syndrome. We report successful anaesthetic management for living related donor liver transplantation (LRDLT) in a patient with Alagille’s syndrome and severe aortic stenosis. Alagille’s syndrome is characterized by a paucity of intrahepatic bile ducts; congenital cardiovascular anomalies consisting mainly of peripheral pulmonary artery stenosis; retarded growth; dysmorphic face; ocular anomaly; mental retardation; skeletal malformation; and xanthomata.5

Case report

The patient was a 9-yr-old female with Alagille’s syndrome, end-stage liver disease and severe aortic stenosis. She was born after a normal delivery but received phototherapy for jaundice in the neonate period. She had a systolic murmur and was diagnosed with aortic stenosis by echocardiography soon after birth. When 2 months old she was diagnosed with Alagille’s syndrome by open liver biopsy, cholangiography and her characteristic dysmorphic face. Jaundice and pancytopenia gradually progressed. Oesophageal varices were noted at 7 yr of age. She was referred to our hospital for LRDLT aged 8 yr. Cardiac function was evaluated by cardiac catheterization. It revealed that the left ventricle–aortic pressure gradient was 62 mm Hg, the left anterior descending artery had 50% stenosis and she had slight peripheral pulmonary artery stenosis. She was judged suitable for LRDLT and was enlisted as a recipient in our hospital. At the age of 9 yr, she was urgently admitted to a local hospital with severe dyspnoea after extraction of a tooth. Heart, liver and renal function deteriorated rapidly. Plasma exchange and haemodialysis were commenced and repeated for 2 weeks. Heart and renal function gradually
improved. Two months later she was transferred to our hospital for LRDLT.

On admission, growth retardation and severe emaciation were observed (height 107.5 cm (–4 SD), body weight 17 kg (–2.3 SD)). She was confined to bed because of dyspnoea but was conscious. A systolic murmur (Levine 3/6) was auscultated at the second intercostal space on the right edge of the sternum. Severe hepatosplenomegaly and ascites were observed on ultrasound examination. Echocardiography revealed severe aortic stenosis (resting pressure gradient 88 mm Hg), slight aortic regurgitation, a small amount of pericardial effusion and myocardial hypertrophy. Left ventricular function was preserved (ejection fraction 0.54). Severe jaundice (total bilirubin 1046 µmol litre⁻¹), coagulopathy (prothrombin time 14.2 s, INR 1.61), anaemia (haematocrit 20.4%) and thrombocytopenia (platelets 47 000 mm⁻³) were noted.

Surgery was performed 2 weeks after admission. Anaesthesia was induced with midazolam 5 mg and fentanyl 200 µg, and maintained with midazolam 0.1 mg kg⁻¹ h⁻¹ and fentanyl 5 µg kg⁻¹ h⁻¹. Neuromuscular block was achieved with vecuronium. Dopamine 2 µg kg⁻¹ min⁻¹ was infused continuously to increase renal blood flow. Monitoring included a pulmonary artery catheter. The pulmonary artery catheter and intra-aortic balloon pump were inserted after induction of anaesthesia via the right internal jugular vein and the right femoral artery, respectively. Intra-aortic balloon pumping (IABP) was started at a 1:2 ratio for protection against myocardial ischaemia before operation. During operation, systolic arterial pressure was 80–140 mm Hg, cardiac index 3.3–6.4 litre min⁻¹ m⁻² (Table 1).

Monitoring included a pulmonary artery catheter. As in all patients with aortic stenosis, the inferior vena cava (IVC) was partially clamped during liver transection and hepatic vein reconstruction. The lateral segment of the liver of her mother was transplanted. At reperfusion of the portal vein, we maintained circulating blood volume (PAWP 13 mm Hg) resulting in only a small reduction in systemic arterial pressure (<10 mm Hg). No additional catecholamines were needed. Total operation time was 10 h 36 min, total anaesthetic duration 14 h 48 min, total blood loss 4800 g, total infused volume 4750 ml (crystalloids 1000 ml, colloids 3750 ml) and total blood transfusion approximately 2500 ml (packed red blood cells 16 u., fresh frozen plasma 8 u. and concentrated platelets 10 u.). Urine output during anaesthesia was 583 ml.

The next day, further surgery was performed to stop intra-abdominal bleeding. The postoperative course was otherwise uneventful. IABP was stopped and removed on the second day after operation. Total bilirubin decreased markedly to 134.3 µmol litre⁻¹ on the second day after operation. The patient was discharged from hospital on day 54 after operation with normal liver function (total bilirubin 8.5 µmol litre⁻¹). At 11 yr of age, aortic stenosis and coronary artery stenosis were corrected successfully with replacement of the aortic valve to pulmonary valve (Ross procedure) and coronary patch angioplasty.

**Discussion**

Severe aortic stenosis is rarely associated with Alagille’s syndrome but the combination of the two was a serious problem for this patient. We chose LRDLT before correction of the aortic stenosis because cardiac function was preserved, in contrast to end-stage liver function.

Haemodynamic disturbances of aortic stenosis are exaggerated by cardiovascular instability during liver transplantation. This cardiovascular instability occurs mainly when clamping the IVC during liver transection and at the time of reperfusion of the transplanted liver.² Both the decrease in venous return caused by clamping the IVC and reduction in systemic vascular resistance as a result of reperfusion of the transplanted liver may cause acute hypotension. In this case, as is usual during LRDLT, the surgeon avoided total clamping of the IVC³ and the change in arterial pressure induced by partial (side) clamping of the IVC was negligible. At the time of portal blood flow reperfusion, we maintained the circulating blood volume, and only slight hypotension was observed. Total blood loss (4800 g) was about four times the circulating blood volume of this patient. However, as the rate of blood loss was slow, we easily maintained circulating blood volume by monitoring cardiac filling pressure.

Perioperative fluid management was aided by a pulmonary artery catheter. As in all patients with aortic stenosis, increased left-sided filling pressure should be maintained appropriately.³ Although there may be some risk of catheter-induced arrhythmia, the pulmonary artery catheter played a very important role in estimating left ventricular end-diastolic pressure (LVEDP) and cardiac output. It should also be noted that even PCWP underestimates LVEDP when ventricular compliance is reduced markedly.¹ In such cases, transesophageal echocardiography demonstrates a closer

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**Table 1** Haemodynamic variables during surgery. HR = Heart rate; SAP = systolic arterial pressure; MAP = mean arterial pressure; DAP = diastolic arterial pressure; CI = cardiac index; SVRI = systemic vascular resistance index; and PCWP = pulmonary capillary wedge pressure

<table>
<thead>
<tr>
<th></th>
<th>HR (beat min⁻¹)</th>
<th>SAP (mm Hg)</th>
<th>MAP (mm Hg)</th>
<th>DAP (mm Hg)</th>
<th>CI (litre min⁻¹ m⁻²)</th>
<th>SVRI (dyn s m⁻² cm⁻⁵)</th>
<th>PCWP (mm Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Baseline</td>
<td>79</td>
<td>93</td>
<td>69</td>
<td>54</td>
<td>5.96</td>
<td>765</td>
<td>12</td>
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<tr>
<td>After IABP</td>
<td>87</td>
<td>116</td>
<td>86</td>
<td>59</td>
<td>6.36</td>
<td>931</td>
<td>16</td>
</tr>
<tr>
<td>Pre-anhepatic</td>
<td>111</td>
<td>103</td>
<td>82</td>
<td>65</td>
<td>4.16</td>
<td>1440</td>
<td>13</td>
</tr>
<tr>
<td>Anhepatic (IVC partial clamp)</td>
<td>108</td>
<td>89</td>
<td>73</td>
<td>56</td>
<td>3.33</td>
<td>1753</td>
<td>6</td>
</tr>
<tr>
<td>Post-anhepatic</td>
<td>96</td>
<td>110</td>
<td>83</td>
<td>59</td>
<td>4.81</td>
<td>1314</td>
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Liver transplantation and severe aortic stenosis
correlation with ventricular volume. However, as the patient had oesophageal varices, ventricular compliance was acceptable and also, because of her small size, we avoided using transoesophageal echocardiography.

At the time of reperfusion, hypotension occurs mainly because of arterial vasodilatation (i.e. reduction in afterload). Acute hypotension may not be tolerated and is potentially hazardous in patients with severe aortic stenosis, as lowered coronary perfusion pressure results in myocardial ischaemia and worsening left ventricular performance. If acute pump failure occurs, efforts to increase cardiac output with inotropic agents or by reduction in afterload with a vasodilator serve mainly to worsen ischaemia. IABP can provide a combination of reduction in afterload with increased diastolic perfusion pressure. We had IABP available during the perioperative period to support cardiac function if necessary, especially around the reperfusion period, although no serious cardiovascular instability occurred at this time.

Plasma exchange and haemodialysis were performed on this patient before admission to our hospital. Preoperative creatinine clearance was 17.5 ml min⁻¹. Infusion of a low dose of dopamine 2 µg kg⁻¹ min⁻¹ was effective in maintaining urinary output, and haemodialysis was not required in the perioperative period.

In summary, we successfully managed anaesthesia for LRDLT in a patient with Alagille’s syndrome and severe aortic stenosis. Careful surgery, which avoided total clamping of the IVC, and volume administration, controlled using a pulmonary artery catheter, produced stable circulation in the reperfusion period.

References