ANAESTHETIC MANAGEMENT FOR CAESAREAN SECTION IN A PATIENT WITH UNCORRECTED TRUNCUS ARTERIOSUS

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Truncus arteriosus is a rare congenital cardiac malformation in which only one artery arises from the heart. From this common vessel, the systemic, pulmonary and coronary arteries originate [1]. The prognosis for individuals with this lesion is poor. Untreated, the malformation is ultimately fatal, with most patients experiencing progressive congestive heart failure in infancy because of excessive pulmonary blood flow [2]. Occasionally, untreated patients do survive until adulthood, although their prognosis remains poor [3].

There has been only one previous case report of a patient with this lesion becoming pregnant, and although a live infant was delivered vaginally, the mother died early in the post partum period [4]. Information from a large review of patients with Eisenmenger’s syndrome suggested that these patients have a high mortality rate associated with their pregnancy, and that this poor prognosis is worsened if Caesarean section is used to deliver a live infant [5].

We present the successful anaesthetic management of a patient with uncorrected truncus arteriosus who required Caesarean section for the delivery of a live infant.

CASE REPORT

A 25-yr-old woman (weight 77 kg) with a diagnosis of truncus arteriosus was referred to the High Risk Pregnancy clinic. The diagnosis of truncus arteriosus had been made provisionally at age 6 months when the patient developed congestive heart failure. No cyanosis was noted at this time (but increased pulmonary vascular markings were noted on chest x-ray). Cardiac catheterization at age 3 yr, undertaken because of decreasing exercise tolerance, confirmed the diagnosis of truncus arteriosus type III by the Collett and Edwards classification. Surgical intervention was refused because the family members were Jehovah’s Witnesses and would not accept a blood transfusion. The patient was reviewed intermittently until age 17 yr and she remained clinically stable during that time. Repeated chest x-rays continued to show cardiomegaly with increased pulmonary vascularity, and ECG confirmed right ventricular hypertrophy. The patient was then lost to follow-up.

The patient was seen initially by us at 23 weeks gestation, but would not accept termination of pregnancy. Symptoms were mild exertional dyspnoea without evidence of orthopnoea, paroxysmal nocturnal dyspnoea, palpitations or oedema. The patient was admitted to hospital at 29 weeks gestation because of shortness of breath. No other change in her signs or symptoms had occurred.

Physical examination revealed heart rate 80 beat min⁻¹ in sinus rhythm and arterial pressure 114/74 mm Hg. Heart sounds were normal S₁.
CAESAREAN SECTION AND UNCORRECTED TRUNCUS ARTERIOSUS

TABLE I. Haemodynamic and blood-gas data associated with general anaesthesia in a patient with uncorrected truncus arteriosus

<table>
<thead>
<tr>
<th>Time</th>
<th>Arterial pressure (mm Hg)</th>
<th>Heart rate (beat min⁻¹)</th>
<th>CVP (mm Hg)</th>
<th>Oxygen saturation (%)</th>
<th>HCT (%)</th>
<th>pH</th>
<th>Pco₂ (kPa)</th>
<th>Po₂ (kPa)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Before intubation</td>
<td>100/60</td>
<td>100</td>
<td>3</td>
<td>80</td>
<td>49</td>
<td>7.39</td>
<td>5.1</td>
<td>7.8</td>
</tr>
<tr>
<td>After intubation</td>
<td>120/60</td>
<td>100</td>
<td>6</td>
<td>90</td>
<td>47</td>
<td>7.37</td>
<td>5.0</td>
<td>7.8</td>
</tr>
<tr>
<td>After delivery</td>
<td>100/60</td>
<td>70</td>
<td>6</td>
<td>87</td>
<td>45</td>
<td>7.40</td>
<td>4.6</td>
<td>8.3</td>
</tr>
<tr>
<td>After extubation</td>
<td>130/70</td>
<td>100</td>
<td>7</td>
<td>86</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>1 h after op.</td>
<td>140/70</td>
<td>80</td>
<td>3</td>
<td>87</td>
<td>44</td>
<td>7.36</td>
<td>5.0</td>
<td>6.8</td>
</tr>
<tr>
<td>6 h after op.</td>
<td>115/60</td>
<td>70</td>
<td>4</td>
<td>85</td>
<td>44</td>
<td>7.38</td>
<td>4.9</td>
<td>6.8</td>
</tr>
<tr>
<td>18 h after op.</td>
<td>120/70</td>
<td>75</td>
<td>6</td>
<td>83</td>
<td>46</td>
<td>7.42</td>
<td>4.7</td>
<td>6.2</td>
</tr>
</tbody>
</table>

single $S_2$ with an audible ejection click and 2/6 systolic ejection murmur. Central and peripheral cyanosis were present, as was clubbing of the fingernails. The haemoglobin concentration was 17.3 g dl⁻¹ and the haematocrit was 52 %. Arterial blood-gas analysis showed a pH of 7.43, $P_{co_2}$ 4.2 kPa, $P_{o_2}$ 6.3 kPa and oxygen saturation 83 % ($F_{i_o_2}$ 0.21). ECG showed heart rate 80 beat min⁻¹, PR interval 0.16 s, QRS interval 0.10 s with right ventricular hypertrophy. Echocardiogram confirmed truncus arteriosus with evidence of severe pulmonary artery hypertension by Doppler examination. Doppler studies also showed a 20-mm gradient across the truncal valve with mild truncal insufficiency. Ventricular function was normal. Therapy was commenced with digoxin 0.125 mg day⁻¹ and heparin 7000 units s.c. twice daily. For the next 6 weeks, the patient remained in hospital and she received oxygen 2 litre min⁻¹ via nasal cannulae, during which time she became slightly more dyspnoeic. Her haemoglobin oxygen saturation measured intermittently by pulse oximetry was 80–84 %. The patient’s haemoglobin concentration remained between 15.0 and 17.3 g dl⁻¹ and the haematocrit between 44 and 50 %. Repeated ECG showed no worsening of truncal insufficiency or ventricular function. During heparin therapy, a mild thombocytopenia (115–130000) occurred, but the prothrombin time and activated partial thromboplastin time (PTT) remained within normal limits.

The fetus was monitored with a weekly non-stress test (NST), beginning at 30 weeks gestation. At 35 weeks gestation, the NST became non-reactive. An Oxytocin Challenge Test (used to assess the adequacy of fetal oxygenation), revealed decelerations with poor short and long term variability. This suggested suboptimal oxygenation of the fetus [6] and it was decided, therefore, to proceed urgently with Caesarean section before the condition of the fetus deteriorated. Before Caesarean section, the haemoglobin was 16.8 g dl⁻¹, haematocrit was 51 % and oxygen saturation was 80 %. Ampicillin 2 mg and gentamycin 90 mg i.v. were administered to provide antibiotic prophylaxis.

We believed that the usual anaesthetic techniques for Caesarean section would not be appropriate. The patient was sedated with midazolam 7 mg i.v. A right internal jugular catheter was inserted for central venous pressure monitoring and a left radial arterial catheter was also inserted. Thirty millilitre of sodium citrate 0.3 mol litre⁻¹ was administered by mouth. Following 3 min of preoxygenation, induction was performed using ketamine 2 mg kg⁻¹, fentanyl 5 µg kg⁻¹ and suxamethonium 2 µg kg⁻¹ with cricoid pressure. Following intubation, anaesthesia was maintained with 50 % nitrous oxide and 0.2–0.5 % halothane in oxygen to control arterial pressure. Vecuronium was given for neuromuscular blockade. Fifteen minutes after induction of anaesthesia and 2 min after uterine incision, a normal infant was delivered weighing 1.9 kg, with Apgar scores of 4 and 8 at 1 and 5 min, respectively. At the end of the operation an extradural catheter was inserted, and the residual neuromuscular block antagonized with glycopyrrolate 0.6 mg and neostigmine 2.5 mg i.v. Following extubation of the trachea, the patient was given 6 mg of preservative-free morphine in 10 ml of normal saline via the extradural catheter. Blood loss throughout the procedure was estimated at 0.6–0.8 litre and lactated Ringer’s solution 2.2 litre was administered during the immediate preoperative and the intraoperative periods. Intraoperative and postoperative haemodynamic and blood-gas data are shown in table I.
An infusion of heparin was started 12 h after operation at a rate of 1000 unit h\(^{-1}\) and adjusted to maintain the PTT > 1.5 x control. Two units of packed red cells were required during the following 24 h to maintain an adequate haematocrit. The patient received two further extradural doses of morphine, at 18 h and 42 h after operation, and required minimal additional analgesia during this time.

Further recovery was uneventful until 6 days after operation when she developed left arm weakness lasting 3 min. This recurred on the 7th day after operation. Heparin i.v. was continued for a further 4 days, after which time therapy was changed to subcutaneous heparin. The mother was discharged on the 13th day after operation, receiving the following therapy: digoxin 0.125 mg twice daily, calcium gluconate 500 mg three times daily and heparin 10000 units s.c. three times daily. Nine months later, both mother and child were doing well.

DISCUSSION

This report describes the successful anaesthetic management of Caesarean section in a patient with truncus arteriosus. One other pregnancy has been described in a patient with this congenital heart lesion, but she died after giving birth to a live infant [4].

Truncus arteriosus represents a severe form of Eisenmenger's syndrome in that the pulmonary arteries are subject to systemic pressures and high blood flow from birth. Furthermore, as the output from both ventricles enters the common trunk, systemic blood tends to be desaturated and a compensatory polycythaemia develops from birth. From a review of Eisenmenger's syndrome and pregnancy, two important points become apparent: the maternal mortality with Caesarean section (75%) was significantly higher than with vaginal delivery (34%), and patients with a right ventricle exposed to systemic pressure from birth (ventricular septal defect) had a higher maternal mortality than those in whom right ventricular hypertension was likely to be a later phenomenon (atrial septum defect and patent ductus arteriosus) [5].

A survey of the world literature to date reveals 15 reported Caesarean sections in 14 patients with Eisenmenger's syndrome (secondary to various lesions), but none with truncus arteriosus. The overall mortality of these patients was 50%. Of these reported cases, one patient received local anaesthesia only and died [7]. Two received lumbar extradural anaesthesia and survived [8, 9]. Seven received general anaesthesia [10–15] and only three of these survived. The method was not documented in the remaining five, although it is likely that they all received general anaesthesia [16–18].

The anaesthetic requirements and physiological changes in a pregnant patient with truncus arteriosus and Eisenmenger's syndrome undergoing general anaesthesia for Caesarean section differ from those of a normal pregnant patient. Anaesthetic techniques used during Caesarean section are designed to minimize cardiovascular and respiratory depression of the newborn, but marked changes in systemic and pulmonary vascular resistance may occur in the mother.

Oxygenation in a patient with truncus arteriosus depends on an adequate cardiac output and an appropriate balancing of blood flow through the pulmonary and systemic vasculature. Decrease in systemic vascular resistance (SVR) or increase in pulmonary vascular resistance (PVR) tends to decrease pulmonary blood flow, while opposite changes increase pulmonary blood flow. During pregnancy, particularly in the last trimester, the SVR normally decreases, as indicated by an increase in cardiac output despite a mild decrease in arterial pressure [18]. Haematocrit also tends to decrease, thereby further decreasing SVR. It is unlikely that these changes occurred in our patient as the haematocrit did not decrease and the oxygen saturation remained stable at 80–85%, suggesting approximately balanced blood flow through the pulmonary and systemic circulation [19].

With Eisenmenger's syndrome it is not possible to decrease PVR by pharmacological means, therefore the anaesthetic management must be directed toward maintaining SVR and cardiac output while avoiding situations which increase PVR. The use of a Swan-Ganz catheter is not possible in this lesion. Continuous measurement of oxygen saturation by pulse oximetry, however, is particularly useful as it allows continuous assessment of the adequacy of pulmonary and systemic blood flow. Assuming that mixed venous oxygen saturation remains between 50 and 70%, an oxygen saturation of 75–85% represents approximately balanced systemic and pulmonary blood flows [20]. A decrease in oxygen saturation indicates inadequate pulmonary blood flow, whereas persistently increased oxygen saturation.
indicates an excessive pulmonary blood flow. If
the cardiac output from the single ventricle
cannot increase sufficiently, theoretically, sys-
temic hypotension and acidosis may develop.
End-tidal carbon monitoring would also be help-
fuIn this type of patient, in addition to inter-
mittent measurement of arterial PCO₂ by blood
sampling, as the pulmonary vascular resistance is
sensitive to pH changes, particularly in the
presence of hypoxia [21].

Ketamine was used as an induction agent as it
produces minimal changes in PVR and does not
adversely affect the relative blood flows through
the pulmonary and systemic circulation [22, 23].
Fentanyl was used in an attempt to obviate
increases in PVR associated with tracheal intu-
bation and surgical incision [24]. Although a dose
of only 5 μg kg⁻¹ was used, no decrease in systemic
oxygenation occurred, suggesting that marked
changes in PVR did not occur. Nitrous oxide has
been found either to increase [25], or not to affect
PVR [26]; in our patient no adverse effects were
observed as assessed by oxygen saturation. Low
concentrations of halothane were used to ensure
amnesia for the procedure, and this did not
produce untoward haemodynamic changes.

Although extradural anaesthesia has been used
successfully in patients with Eisenmenger’s syn-
drome, both cases reported were elective, and
this allowed gradual onset of analgesia and
accompanying sympathetic block [8, 9]. The main
concern with the use of extradural anaesthesia for
Caesarean section is that the decrease in systemic
vascular resistance that occurs in the presence of
a fixed, increased pulmonary vascular resistance
may lead to a decrease in pulmonary blood flow
with worsening of cyanosis. However, general
anaesthesia allows independent manipulation of
the pulmonary and systemic circulations by
appropriate physiological and pharmacological
means. Pulmonary hypertension and increases in
pulmonary vascular resistance (in particular) are
not amenable to treatment by extradural analgesia,
but may respond to hyperventilation and high
dose opioids.

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