Case Report

Anaesthetic management for hip arthroplasty in a 46-yr-old patient with uncorrected truncus arteriosus type IV

M. U. Fischer* and H.-J. Priebe

Department of Anaesthesia, University Hospital, Hugstetter Strasse 55, 79106 Freiburg, Germany

*Corresponding author. E-mail: fischerm@ana1.ukl.uni-freiburg.de

In modern adult anaesthetic practice uncorrected complex congenital heart lesions are rare. Persistent truncus arteriosus accounts for only 1.2–3% of all congenital heart malformations. If not corrected, fewer than 20% of these patients survive the first year of life. Here we report the successful management of an adult patient with uncorrected truncus arteriosus who presented for hip arthroplasty.

Br J Anaesth 2006; 97: 329–32

Keywords: anaesthesia, general; complications, truncus arteriosus; heart, congenital defects; monitoring, echocardiography

Accepted for publication: April 20, 2006

In modern adult anaesthetic practice uncorrected complex congenital heart lesions are rare. Persistent truncus arteriosus accounts for only 1.2–3% of all congenital heart malformations.1 If uncorrected, fewer than 20% of these patients survive the first year of life.1–3 We report the successful management of an adult patient with uncorrected truncus arteriosus who presented for hip arthroplasty.

Case report

This 46-yr-old woman with a hip fracture was transferred from a regional hospital to our trauma centre for hip arthroplasty. The patient had a known uncorrected truncus arteriosus type IV, recently confirmed by magnetic resonance angiography. She reported regular hospital admissions because of recurrent pulmonary infections, cerebro-vascular accidents and a cardiac arrest during phlebotomy a few years ago.

Preoperative assessment revealed poor exercise tolerance with New York Heart Association class III. On examination, the patient was cyanotic with marked clubbing and had systolic and diastolic heart murmurs. The heart rate was 90 beats min⁻¹ and arterial pressure 100/60 mm Hg. Arterial blood gas results on room air were: pH 7.42, $P_{a\text{O}_2}$ 5.4 kPa, $P_{a\text{CO}_2}$ 4.5 kPa and peripheral oxygen saturation ($S_{\text{PO}_2}$) 73%.

A 12-lead ECG revealed sinus rhythm, incomplete right bundle branch block and T-wave inversion in leads I, II, aVF and V₁–V₄. Transthoracic echocardiography showed a $20\times25$ mm ventricular septal defect (VSD), a hypertrophic right ventricle with diffuse minor hypokinesia, a normalized left ventricle with good function, and a large truncus arteriosus (45 mm) with mild regurgitation. Neither the pulmonary artery (PA), nor the pulmonary valve could be seen. A large vessel arising from the arch of the truncus was described as ‘presumably the PA’. Blood chemistry revealed a chronic thrombocytopenia of 75 000 mm⁻³, a secondary polycythaemia with a haematocrit of 65%, a serum creatinine concentration of 115 µmol litre⁻¹ (1.3 mg dl⁻¹) and an elevated international normalized ratio of 1.7. Before operation, 1500 iu of coagulation factors II, VII, IX and X (PPSB®, Baxter BioScience, Heidelberg, Germany) were administered to improve coagulation.

The patient was undergoing hip arthroplasty under general anaesthesia. She received midazolam 3.75 mg orally for premedication. No other medication was given. Immediately before induction of anaesthesia, arterial pressure was 140/75 mm Hg, heart rate 90 beats min⁻¹ and $S_{\text{PO}_2}$ 79%. After continuous blood pressure monitoring via a radial arterial line had been established, anaesthesia was induced with a total of flunitrazepam 0.8 mg, fentanyl 0.2 mg, etomidate 10 mg and pancuronium 8 mg. After endotracheal intubation, the right internal jugular vein was cannulated and a transoesophageal echocardiography (TOE) probe inserted for continuous cardiac monitoring. Anaesthesia was maintained with end-tidal isoflurane concentrations of 0.2–0.9% and incremental doses of fentanyl and midazolam.

After insertion of the TOE probe, the four-chamber view showed a markedly hypertrophic right ventricle, a leftward shift of the interventricular septum and a large VSD of 24 mm. With the TOE probe in the mid-oesophageal position and a multiplane angle of 60°, a tricuspid valve with a diameter of 47 mm was seen (Fig. 1). The right ventricular outflow tract and pulmonary valve usually seen in this view were absent. The transgastric long axis view (Fig. 2) showed the VSD and the large truncus overriding the septum. The
colour Doppler revealed minor to moderate incompetence of the truncus valve.

After induction of anaesthesia, transient hypotension to an arterial blood pressure of 85/55 mm Hg was treated with two bolus doses of a cafedrine–theoadrenaline preparation (Akrinor®, AWD.pharma, Dresden, Germany). During the procedure haemodynamic management was guided by TOE and $Sp_o_2$ findings. Decreases in blood pressure responded well to fluid administration. Arterial blood pressure varied between 85–150 mm Hg systolic and 55–85 mm Hg diastolic. Systemic systolic pressures below 110 mm Hg were associated with $Sp_o_2$ values <85%. Central venous pressures and heart rates varied between 14 and 24 mm Hg, and 85 and 115 beats min$^{-1}$, respectively. Obligatory fluid losses and the estimated blood loss of 600 ml were replaced with 2000 ml of a full electrolyte solution (Jonosteril®, Fresenius Kabi, Bad Homburg, Germany), 500 ml of 6% hydroxyethyl starch (Voluven®, Fresenius Kabi, Bad Homburg, Germany), and 400 ml of fresh frozen plasma. Total urine output was 200 ml. A non-cemented prosthesis was inserted to reduce the risk of systemic arterial hypotension and pulmonary hypertension secondary to cement embolism. Surgery lasted 110 min.

After the operation the patient was transferred to our respiratory-surgical intensive therapy unit (ITU) and the trachea extubated 2 h later. A continuous infusion of

---

**Fig 1** The transoesophageal echocardiogram with the probe in mid-oesophageal position and a multiplane angle of 60° shows a large tricuspid valve (truncus valve) with a diameter of 47 mm. PA and pulmonary valve which are usually seen in this view are absent. LA, left atrium; RA, right atrium.

**Fig 2** The transoesophageal echocardiogram with the probe in transgastric position and a multiplane angle of 120° shows a large truncus arteriosus overriding a VSD. LV, left ventricle; RV, right ventricle.
ropivacaine 0.2% via a femoral sheath catheter (placed before induction of anaesthesia) provided postoperative analgesia. After an uneventful recovery, the patient was discharged from the ITU on postoperative day 1 and back to the referring hospital on postoperative day 11.

**Discussion**

Children with congenital heart disease (CHD) routinely receive specialist multi-disciplinary care in specialized centres. In contrast, patients with adult CHD may present to any centre. Although this happens infrequently, every anaesthetist and intensivist must be familiar with the respective management issues. In patients with adult CHD, perioperative mortality during non-cardiac surgery can be as high as 30%. There are few cases of long-term survival in patients with unrepaired truncus arteriosus. To our knowledge, this is the first report of a patient above the age of 45 yr with unrepaired truncus arteriosus undergoing major non-cardiac surgery.

Truncus arteriosus is a rare CHD accounting for approximately 1.2–3% of all forms of CHD. It is characterized by one common vessel (truncus) with a cuspid valve (usually three or four cusps) that originates from both ventricles on top of a large VSD. It supplies blood to the systemic, coronary and pulmonary circulations. Depending on the origin of the pulmonary arteries, four types of truncus arteriosus can be differentiated. The pulmonary arteries may originate as common trunk from the posterior wall of the truncus (type I) or as separate branches dorsally (type II) or laterally (type III) from the truncus. Our patient had a type IV truncus arteriosus in which only major aorto-pulmonary collateral arteries (MAPCAs, Fig. 3) exist (also referred to as pseudo-truncus arteriosus). This type is difficult to differentiate from the tetralogy of Fallot with atresia of the pulmonary valve. It is the least frequent of the four types accounting for only 10–13% of them. The prognosis without surgical intervention is poor: 50% of the affected children die within the first month of life. One-year survival is only 10–25%, and mainly in those with stenosis of the pulmonary arteries.

In patients with this type of adult CHD, the leading clinical signs and symptoms are those of the Eisenmenger complex. Eisenmenger described post-mortem evidence of pulmonary vascular disease in the presence of VSD and right ventricular hypertrophy. More than half a decade later Wood outlined the important findings in Eisenmenger’s complex: pulmonary hypertension at systemic level secondary to high pulmonary vascular resistance (PVR) with reversed right-to-left shunt. The higher the PVR in relation to SVR, the higher the right-to-left shunt. The resulting hypoxaemia is usually not responsive to oxygen therapy.

Secondary to longstanding hypoxaemia these patients frequently have multiple co-morbidities such as hypertension, ischaemic heart disease, lung disease, renal impairment, haematological disorders, electrolyte imbalance and multiple coagulation factor deficiencies. The secondary polycythaemia increases blood viscosity, which renders these patients prone to thrombosis and cerebral vascular accidents. On the other hand, multiple clotting factor deficiencies as well as thrombocytopenia are common.

All patients with adult CHD are best managed in a tertiary referral centre. A multi-disciplinary approach is important. A thorough assessment of the degree of cardiovascular impairment and detailed knowledge of the underlying pathophysiology are mandatory. A cardiologist experienced in managing such complex cases should be consulted. Guidelines for the management of adult CHD have been published.

Patients with Eisenmenger’s syndrome have done well under a variety of anaesthetic techniques. Whereas traditionally general anaesthesia was the preferred technique, the benefits of regional anaesthesia have recently been emphasized. We decided to use general anaesthesia because of patient anxiety, the co-existent coagulopathy, and to allow TOE. The overall anaesthetic management has to be formulated on the basis of the underlying cardiovascular pathophysiology of the defect. As pulmonary and systemic blood flows originate from one common vessel (truncus), blood distribution to either circulation will vary with the respective vascular resistances. As we had to assume that our patient had elevated PVR, we aimed at avoiding any further increase in PVR and to prevent a disproportional decrease in SVR, which would worsen the right-to-left shunt fraction. Factors that increase PVR include hypercarbia, hypothermia, acidosis, pulmonary arterial hypoxia,
increased sympathetic tone and sympathomimetic drugs (e.g. epinephrine and norepinephrine). Positive pressure ventilation itself can increase PVR, with inspiratory time having a greater effect than peak inspiratory pressure.

TOE allows real time assessment of cardiac filling, ventricular and valve function, and regional wall motion. TOE monitoring is, therefore, particularly helpful in the haemodynamic management of this type of CHD as it allows early recognition of the possible aetiology of any haemodynamic instability and decreases in $S_{\text{po}_2}$. In the presence of a small left ventricular end-diastolic area and ‘kissing papillary muscles’, i.v. fluid administration restored blood pressure. When hypotension is associated with a decrease in $S_{\text{po}_2}$, an increase in the right-to-left shunt fraction on the basis of an increase in the PVR/SVR ratio is the probable cause in this type of CHD. Prophylactic phenylephrine and norepinephrine infusions\(^{10}\) have been recommended to prevent decreases in SVR during anaesthesia in patients with Eisenmenger’s syndrome. We decided against such pharmacological prophylaxis as it may increase PVR. On the other hand, when $S_{\text{po}_2}$ and blood pressure decreased simultaneously during induction of anaesthesia, we did not hesitate to administer a cafedrine–theoadrenaline preparation. This intervention restored systemic pressure and $S_{\text{po}_2}$. In case of prolonged hypotension associated with increased right-to-left shunt, we had planned to administer norepinephrine by continuous infusion. However, this did not become necessary. Arginine vasopressin might have been a valuable alternative because it is a systemic vasoconstrictor, which causes pulmonary vasodilation in a variety of experimental models. To our knowledge its use has not been reported in a patient with Eisenmenger’s syndrome undergoing non-cardiac surgery.

The duration of surgery, the amount of fluid replacement and haemodynamic instability directly correlate with perioperative mortality.\(^4\) A non-cemented prosthesis was used to reduce the risk of systemic hypotension and pulmonary hypertension associated with cemented hip arthroplasty.\(^{19,20}\)

The immediate postoperative period requires special attention. Of 12 perioperative deaths in patients with Eisenmenger’s syndrome who underwent major surgery, 4 occurred within 6 h and a fifth within 7 h of anaesthesia.\(^{17}\) Postoperative care should, therefore, preferably be provided in a high-dependency unit.\(^{4,13}\) This facilitates invasive monitoring and early interventions. Early tracheal extubation is recommended\(^{18}\) to reduce the risk of increases in PVR.\(^{19}\) Adequate pain control is essential to avoid pain-induced increases in sympathetic tone.\(^{24}\) We achieved this by continuous femoral nerve block. In general, intensivists involved in the patient’s postoperative care should avoid setting unrealistic therapeutic goals.

In conclusion, patients with adult CHD pose unique management challenges. Non-cardiac surgery is best performed in a centre that has experience in the treatment of such patients. A careful multi-disciplinary approach is required which takes into consideration the complex underlying pathophysiology of the disease entity. This includes a detailed preoperative workup and a careful perioperative and postoperative management plan. The importance of adequate cardio-respiratory monitoring cannot be overemphasized. On the other hand, physicians caring for these patients need to take into account the individual underlying fixed haemodynamic and respiratory pathology to avoid setting unrealistic therapeutic goals.

References
\begin{enumerate}
\item Williams JM, de Leeuw M, Black MD, Freedom RM, Williams WG, McCrindle BW. Factors associated with outcomes of persistent truncus arteriosus. J Am Coll Cardiol 1999; 34: 545–53
\item Litovsky SH, Ostfeld I, Bjornstad PG, Van Praagh R, Geva T. Truncus arteriosus with anomalous pulmonary venous connection. Am J Cardiol 1999; 83: 801–4
\item Ammash NM, Connolly HM, Abel MD, Warnes CA. Noncardiac surgery in Eisenmenger Syndrome. J Am Coll Cardiol 1999; 33: 222–7
\item Bodi V, Insa L, Sanchis J, Ibanez M, Losada A, Chorro FJ. Persistent truncus arteriosus type 4 with survival to the age of 54 years. Int J Cardiol 2002; 82: 75–7
\item Hicken P, Evans D, Heath D. Persistent truncus arteriosus with survival to the age of 38 years. Br Heart J 1966; 28: 284–6
\item Ammash N, Warnes CA. Cerebral vascular events in adult patients with cyanotic congenital heart disease. J Am Coll Cardiol 1996; 28: 768–72
\item Perloff JK, Warnes CA. Challenges posed by adults with repaired congenital heart disease. Circulation 2001; 103: 2637–43
\item Baum VC, Perloff JK. Anesthetic implications of adults with congenital heart disease. Anesth Analg 1993; 76: 1342–58
\end{enumerate}