CASE REPORT

Anaesthetic management of hip arthroplasty in an individual with trisomy 21 and Eisenmenger's syndrome

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SUMMARY

A 49-year-old man with trisomy 21 and Eisenmenger's syndrome presented for hip arthroplasty. Eisenmenger's syndrome is defined by the presence of obstructive pulmonary vascular disease secondary to long-standing left-to-right shunt causing pulmonary hypertension, eventually leading to shunt reversal in to right-to-left direction. Patients with Eisenmenger's syndrome pose a significant perioperative risk because of the physiological alterations induced by anaesthetic agents. The choice of anaesthetic technique in these patients is therefore not straightforward. A successful outcome was achieved with general anaesthesia supplemented with nerve blocks.

BACKGROUND

Trisomy 21 is a chromosomal disorder associated with a higher risk for congenital heart disease (CHD). Septal defects are common, which initially produce a left-to-right cardiac shunt. If uncorrected, the ensuing pulmonary hypertension increases right-sided heart pressures, and ultimately the shunt may reverse to a right-to-left direction. This is known as Eisenmenger's syndrome.¹ Because of its rarity, anaesthetic experience with this condition is limited. Median life expectancy in patients with Eisenmenger's syndrome is reduced by about 20 years.¹ The perioperative death rates in such patients can be as high as 24%.² The American Heart Association (AHA) and the American College of Cardiology (ACC) guidelines on management of patients with congenital heart disease recommend that the anaesthetic approach to patients with Eisenmenger's syndrome should be individualised.³ Case reports are the primary source of information on anaesthetic management of patients with Eisenmenger's syndrome, and several techniques have been described.⁴⁻⁶

CASE PRESENTATION

A 49-year-old man with trisomy 21 was admitted with a hip fracture. He had an uncorrected atrioventricular septal defect, which was monitored regularly by cardiologists. His medical history included hospital admissions for recurrent supraventricular tachycardias and heart failure. He also had scoliosis, spondylosis, hypothyroidism and secondary polycythaemia. He had a left hemiparesis following a suspected embolic stroke. His medications included warfarin, allopurinol, amitriptyline, bendroflumethiazide, codeine, folic acid, levothyroxine, omeprazole, paracetamol and sodium decussate.

He possessed poor exercise tolerance (New York Heart Association class III). His weight was 54 kg, and height of 1.58 m giving a body mass index of 22. On examination the patient had central cyanosis, digital clubbing and a loud pansystolic murmur. His heart rate was 86 bpm and arterial pressure was 130/70 mm Hg.

The perioperative team and the relatives discussed the risks of anaesthesia and surgery and also considered the option of non-surgical management. This would have entailed a long period of immobilisation in traction. Despite the risks, there was a consensus that surgery was the better alternative.

INVESTIGATIONS

A thorough preoperative assessment is essential in patients with Eisenmenger's syndrome, and should include pulse oximetry, ECG, transthoracic echocardiogram, a full blood count and coagulation screen.³ All these tests were performed on this patient including arterial blood gases analysis. The tests revealed pH of 7.29, PaO₂ of 6.9 kPa, PaCO₂ of 4.5 kPa and oxygen saturation of 78%. A 12-lead ECG showed sinus rhythm with left axis deviation, bifascicular block and right ventricular hypertrophy. Transthoracic echocardiography revealed an atrioventricular canal defect with a common atrioventricular valve. The left ventricle was of normal size and systolic function. The right ventricle was thickened and the right atrium was dilated. The magnitude of the shunt was difficult to assess accurately owing to the common atrioventricular valve defect. The shunt was bidirectional with obvious venous and pulmonary flows mixing. The pulmonary artery pressure was estimated by Doppler (84 mm Hg).

His haemoglobin was 15.8 g/dl and haematocrit 46%. His creatine was 84 µmol/l, urea 12.2 mmol/l. The international normalised ratio on admission was 5.0; with vitamin K therapy this had fallen to 1.5.

TREATMENT

The patient's medication list was reviewed as part of the preoperative assessment. Warfarin was stopped 5 days before operation and anticoagulation was maintained with dalteparin 10 000 units once daily. Warfarin was restarted 5 days after the surgery. Codeine was stopped before operation, and restarted postoperatively after the morphine infusion was discontinued. The other medications were not considered to be contraindicated for surgery and anaesthesia, and therefore were continued.

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Various anaesthetic techniques were considered. Ultimately general anaesthesia was chosen supplemented with a combined femoral and lateral cutaneous nerve of thigh block. Standard monitoring (ECG, pulse oximetry, end-tidal CO₂ monitoring, non-invasive blood pressure) was applied. Intravenous access was acquired and invasive arterial pressure monitoring was established via the right radial artery. At induction of anaesthesia the arterial pressure was 120/60 mm Hg, heart rate of 98 bpm and SpO₂ 75%. Induction was achieved using a total of 70 mg of propofol and 25 µg of fentanyl. The propofol was carefully titrated because of its potential to drop cardiac output. A size 4 laryngeal mask airway (LMA) was inserted and the patient breathed oxygen-enriched air with an FiO₂ of 0.5 and up to 2% isoflurane through a circle circuit. End-tidal CO₂ ranged from 5.3 to 6.3 kPa. Additional analgesia was achieved with a femoral nerve block (20 ml of 0.375% bupivacaine) performed using a nerve stimulator. A lateral cutaneous nerve of thigh block (10 ml of 0.25% bupivacaine) was carried out using a landmark technique.

There were intermittent falls in arterial blood pressure the lowest of which was 82/69 mm Hg. These were usually associated with drops in oxygen saturations, the lowest of which was 64%. To stabilise the arterial pressure, a phenylephrine infusion (100 µg/ml) was run at a rate of up to 0.3 mg/kg/h. Intermittent small boluses (0.5 mg) of metaraminol were also given to a total of 3.5 mg. Further boluses of fentanyl were administered to a total of 75 µg. The estimated blood loss was 500 ml. Altogether 2000 ml Hartmann's solution was infused over 120 min of surgery.

At the end of surgery isoflurane was discontinued and the patient rejected the LMA. He was transferred to the intensive care unit breathing spontaneously with 5 litres oxygen via face mask. The phenylephrine infusion was continued for 3 h and he received a morphine infusion at a rate of 1–5 ml/h for 24 h. Analgesia was supplemented with oral paracetamol and codeine.

OUTCOME AND FOLLOW-UP

The patient was discharged uneventfully to the ward on the second postoperative day.

DISCUSSION

Eisenmenger's syndrome is an end stage of a disease process initiated by an anatomic communication between the systemic and pulmonary circulation.⁷ The presence of an intracardiac shunt increases pulmonary blood flow, which if uncorrected leads to pulmonary obstructive vascular disease.⁷ Over time, pulmonary hypertension ensues, which causes reversal of the shunt into a right-to-left direction.⁸ As a result, these patients develop cardiomegaly, chronic hypoxaemia and polycythaemia.⁷ Patients with Eisenmenger's syndrome have a fixed pulmonary vascular bed, and lose the ability to compensate for sudden change in systemic vascular resistance (SVR) and pulmonary vascular resistance (PVR).^{7–8} Factors which cause a rise in PVR or a decrease in SVR, such as hypovolaemia, hypercapnia, hypoxaemia and acidosis are all potentially harmful, and should be avoided.⁹ The cornerstones of anaesthetic management are to promote oxygen delivery by preventing arterial desaturations, maintain balance between the systemic and pulmonary blood flow, optimise haematocrit and monitor for and treat promptly right ventricular decompensation.^{4–9}

The choice of anaesthetic agents and technique should be individualised and tailored to the patient's haemodynamic status and surgical procedure.⁹ Both general⁶ and regional anaesthesia^{4–5} have been used successfully in patients with Eisenmenger's

syndrome. Both techniques have the potential to produce hypotension and vasodilation, which will increase the reversed shunt. Recently, regional anaesthesia has been reported to have a slightly lower mortality compared with general anaesthesia although the authors suggest that most deaths probably occurred as a result of the surgical procedure and the disease, not the anaesthesia.² Spinal and epidural anaesthesia have been used successfully in pregnant patients with Eisenmenger's syndrome.^{2–5} There were communication problems with our patient that led us to favour a general anaesthetic technique.

Most of intravenous induction agents depress the cardiovascular system causing reductions in SVR (propofol and midazolam), sympathetic stimulation (ketamine) and myocardial contractility (thiopentone).^{8–10} However, most can be used safely as long as care is taken with the dose and rate of injection.⁷ We chose a carefully titrated dose of propofol, which produced minimal cardiovascular depression.

Volatile agents also reduce cardiac performance by reducing contractility and causing vasodilatation. They have been shown not to affect left-to-right shunts but whether this is the case for reversed shunt is unknown.¹¹ Inhalational inductions will take longer time because of the intracardiac shunt.¹²

Various vasopressors have been used to counteract falls in SVR during anaesthesia. These agents are likely to be required regardless of the anaesthetic technique used. Norepinephrine,¹³ metaraminol⁴ and phenylephrine⁶ have all been used successfully. Our patient required a continuous background infusion of phenylephrine with intermittent small boluses of metaraminol to maintain stable arterial pressures and oxygen saturations.

Intermittent positive pressure ventilation increases intrathoracic pressures and can compromise venous return, increase PVR and exacerbate right-to-left shunting in patients with cyanotic heart disease.⁸ Laryngoscopy can cause catecholamine release and transient rise in PVR.⁵ We elected not to intubate and ventilate this patient, and instead used a spontaneous breathing technique via a LMA.

Many patients with cyanotic CHD develop hyperviscosity syndrome, characterised by fatigue, dyspnoea, headaches and visual disturbances,¹⁴ and require intermittent venesection. Therapeutic phlebotomy is recommended at haemoglobin levels greater than 20 g/dl and haematocrits greater than 65%.³ Our patient's haematocrit was 46%, as he had undergone venesection 2 weeks before sustaining his hip fracture. Thromboembolic complications can be reduced by preoperative venesection,¹⁴ but excessive reductions in haemoglobin will compromise oxygen delivery in these patients.¹⁵

Conventional non-invasive perioperative monitoring is recommended, which includes pulse oximetry, ECG, and capnography. Pulse oximetry is particularly useful as it can be used to assess the degree of right-to-left shunting because of raised PVR.⁸ End tidal carbon dioxide measurements will underestimate PaCO₂ in the presence of right-to-left shunt because the volume of blood bypassing the lungs creates a dead space effect.¹⁰ The degree of this discrepancy will change as the degree of the shunting changes.⁷

Invasive arterial pressure monitoring allows for rapid detection of changes in SVR. Although not used in our patient, transoesophageal echocardiography can be useful to assess ventricular function, preload and shunting.⁷ There are risks associated with the use of central venous and pulmonary catheters in these patients and the data acquired can be misleading and difficult to interpret.⁴

The early postoperative care of patients with Eisenmenger's syndrome should be provided in an intensive care unit,³ where our patient required an ongoing vasopressor infusion for a few

hours. Adequate analgesia helps to prevent cardiac instability. In our patient's case the regional block inserted at induction helped provide pain relief and was supplemented with an opioid infusion.

Learning points

- ▶ Patients with Eisenmenger's syndrome benefit from good preoperative assessment, a thorough evaluation of risks, vigilant monitoring and professional aftercare in the intensive care unit.
- ▶ The perioperative mortality for these patients is high. We describe a successful outcome following general anaesthesia, but other anaesthetic techniques have been described.
- ▶ During anaesthesia and surgery care should be taken to try to avoid increasing the right-to-left shunt as this will worsen the hypoxia.

Contributors JB and JS have equally contributed to conception and design of the article, acquisition, interpretation and analysis of the data presented. Both the authors equally contributed to writing and revising of the article. JB and JS have also reviewed the final version of the article.

Competing interests None.

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