Clinical Report

Anaesthesia for Caesarean section in a patient with Watson's syndrome

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Watson's syndrome is an uncommon genetic disorder whose features include mental retardation and pulmonary valvular stenosis. The purpose of this report is to describe one management strategy used successfully to anaesthetize a woman with this disorder who presented for Caesarean section. Epidural anaesthesia using 0.5% bupivacaine without epinephrine in 2–3 ml boluses to a total dose of 15 ml was administered over 30 min. Invasive haemodynamic monitoring in the form of arterial and central venous catheters were used to guide therapy and help ensure maternal and fetal well-being. Intravenous ketamine in doses of 10–20 mg every five minutes to a total dose of 245 mg ($4.5 \text{ mg} \cdot \text{kg}^{-1}$) was used to overcome the patient's uncooperative nature and facilitate invasive procedures. Postoperative analgesia was provided using 3 mg epidural morphine. The patient was observed in the intensive care unit for the first postoperative day and experienced an uncomplicated intra- and postoperative course. We conclude that this technique represents a safe and effective method for anaesthetizing patients with this complicated problem for Caesarean section.

Le syndrome de Watson est une aberration génétique rare caractérisée principalement, entre autres, par un retard mental et une sténose de la valvule pulmonaire. Cette observation décrit la stratégie mise en oeuvre pour anesthésier une patiente porteuse de ce syndrome soumise à une césarienne. Une anesthésie

épidurale est réalisée avec de la bupivacaine 0,5% administrée en bolus de 2–3 ml sur une période de 30 min pour une dose totale de 15 ml. Le monitoring hémodynamique est constitué d'une tension veineuse centrale et d'une canule artérielle. De la kétamine iv aux doses de 10–20 mg aux cinq min pour un total de 245 mg ($4,5 \text{ mg} \cdot \text{kg}^{-1}$) est administrée pour pallier à l'absence de collaboration et faciliter les mesures effractives. L'analgésie postopératoire est obtenue avec de la morphine épidurale 3 mg. La patiente est gardée sous observation à l'unité des soins intensifs pendant la journée qui suit l'intervention. Aucune complication ne survient pendant et après l'intervention. En conclusion, cette technique représente une méthode d'anesthésie efficace et non dangereuse pour effectuer une césarienne chez une patiente porteuse de ce désordre compliqué.

Key words

ANAESTHESIA: obstetric;

SYNDROMES: Watson's;

ANAESTHETICS, INTRAVENOUS: ketamine;

ANAESTHETIC TECHNIQUES: epidural.

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Watson's syndrome is a rare genetic disorder with an autosomal dominant mode of inheritance. Its most consistent clinical features include café-au-lait spots, short stature, diminished intelligence, and pulmonary valvular stenosis. Women affected by this syndrome are fertile and obstetric management must take into consideration the abnormal characteristics of this condition to ensure maternal and neonatal well being. We describe the anaesthetic management of a woman with Watson's syndrome presenting for Caesarean section.

Case report

A 29-yr-old, 54-kg woman with Watson's syndrome was admitted to hospital complaining of dysuria, abdominal pain, and vomiting of one week's duration. Physical examination revealed a large abdominal mass which on ultrasound examination demonstrated an enlarged uterus and feto-placental unit consistent with a 32 wk gestation.

Past medical history included severe mental retardation and hydrocephalus which had been corrected at birth with a ventriculoperitoneal shunt. A grand-mal seizure disorder had been inactive for a period of eight years. She was

taking no medications and was known to have a penicillin allergy.

Physical examination revealed a blood pressure of 110/70 mmHg and heart rate of 80 bpm. The patient was uncooperative and had a functional level equivalent to that of a five-year-old child. A grade 3/6 systolic murmur was heard at the second left intercostal space, and a grade 2/6 diastolic murmur was detected at the base. Jugular venous distension was normal. Bilateral leg oedema to the knees was noted. Chest and airway examination was unremarkable. Other abnormalities included short stature, numerous café-au-lait spots, and congenital absence of the patellae.

Transoesophageal echocardiography performed under sedation during this admission confirmed the diagnosis of severe pulmonary valvular stenosis. The peak trans-valvular pressure gradient was 130 mmHg and the mean gradient was 70 mmHg. Mild aortic and tricuspid insufficiency were noted, but left ventricular size and function were normal. Her ECG demonstrated a sinus rhythm with right axis deviation and right ventricular hypertrophy. Elevation of hepatic transaminase concentrations was noted with AST = $115 \text{ U} \cdot \text{L}^{-1}$ and ALT = $151 \text{ U} \cdot \text{L}^{-1}$. The urine was free of protein. Platelet count, PTT, and INR were normal.

Fetal assessment revealed a normal fetal heart rate with good beat-to-beat variability. Ultrasound examination was normal and fetal echocardiography failed to demonstrate any fetal cardiac malformation.

At 35 3/7 wk gestation she developed spontaneous rupture of membranes and the onset of contractions. A continuous *iv* infusion of fentanyl was used to provide analgesia for labour. After 36 hr there was failure of labour to progress beyond 8 cm cervical dilatation. Therefore, it was decided to proceed with Caesarean section. Informed consent for this procedure was obtained from the patient's sister.

Large bore *iv* access was established and 500 ml lactated Ringer's solution was administered over 30 min. Vancomycin one gram was also infused as prophylaxis against bacterial endocarditis. Under the influence of ketamine sedation, arterial, central venous, and epidural catheterizations were successfully performed. Ketamine 245 mg *iv* was administered in 10–20 mg increments every five minutes as required. Supplemental oxygen was supplied by face mask and left uterine displacement was maintained. Additional monitoring consisted of continuous electrocardiography and pulse oximetry. An epidural block adequate for surgical anaesthesia was instituted using 2–3 ml incremental doses of bupivacaine 0.5% without epinephrine. A total of 15 ml bupivacaine was required. Central venous pressure ranged between 6 and 9 mmHg. Ninety minutes after the onset of ketamine

sedation a single, live female infant was born. The infant was morphologically normal, required no resuscitation, and received Apgar scores of 6 and 9 at one and five minutes. Three mg epidural morphine were administered after delivery of the fetus and the epidural catheter was removed upon completion of the surgery. The mother was transferred to the ICU for observation where she remained well. Her recovery was uneventful, and she was discharged home six days after surgery.

Discussion

Watson's syndrome was originally described in 1967,¹ and was characterized by the triad of pulmonary valvular stenosis, café-au-lait spots, and dull intelligence. Its clinical phenotype has since been expanded to include short stature, relative macrocephaly, Lisch nodules, and neurofibromas. Recent work suggests that the genetic defect responsible for this rare, autosomal dominant disorder may lie on the long arm of chromosome 17.² This unusual combination of abnormalities poses several difficult challenges for the anaesthetist, particularly when superimposed upon the physiological changes of pregnancy.

The most important clinical feature of the syndrome is pulmonary valve stenosis. This obstruction to right ventricular outflow results in both increased right ventricular work and pressure with compensatory concentric right ventricular hypertrophy. With severe pulmonary stenosis, right ventricular output may be limited thus reducing left ventricular output. This is of special concern in the pregnant patient where the increase in cardiac output associated with labour and delivery may result in acute cardiac decompensation and right-sided cardiac failure. Thus, the goals of haemodynamic management include maintaining an adequate preload, a normal heart rate, and avoiding decreases in afterload and myocardial contractility. Fluid management is of prime importance since excessive preload can precipitate right-sided heart failure, whereas inadequate preload which may occur with aortocaval compression, induction of regional anaesthesia, or severe bleeding at delivery will result in inadequate cardiac output. General anaesthesia using a nitrous oxide-relaxant technique has been recommended to anaesthetize patients with pulmonary stenosis for Caesarean section.³

This patient demonstrated severe pulmonary valvular stenosis and evidence of right ventricular hypertrophy on transoesophageal echocardiography. The presence of bilateral leg oedema to the knees and an elevation of hepatic enzymes was felt to represent evidence of right-sided heart failure rather than preeclampsia. Due to the severity of her pulmonary valvular disease consideration was given to performing balloon valvuloplasty. However, this procedure was felt to put the fetus at considerable risk and was not performed. Additional considerations included

the need for antibiotic prophylaxis against bacterial endocarditis,⁴ and the need for invasive haemodynamic monitoring to ensure optimal peri-partum management. An arterial line was required to facilitate adequate beat-to-beat control of blood pressure and a central venous catheter was inserted via the right internal jugular vein to monitor preload and right heart function. Pulmonary artery catheterization was not attempted due to the severe pulmonary valve stenosis and very low likelihood of successful insertion. The initial central venous pressure was 6 mmHg which likely represents careful antepartum fluid management prior to the onset of labour. Administration of lactated Ringer's solution was carefully controlled to avoid gross changes in the CVP during titration of the epidural block. A total of three litres lactated Ringer's solution was infused during the operation, and the CVP remained between 6 and 9 mmHg throughout.

The use of epidural anaesthesia in this situation is controversial. Potential hazards include precipitous reduction of peripheral vascular resistance with subsequent decreased blood pressure and organ perfusion; possible coagulopathy secondary to hepatic congestion; difficulty with positioning; lack of cooperation; and difficulty in assessing the adequacy of anaesthesia. Nevertheless, we believe that these disadvantages are offset by several important advantages of epidural anaesthesia. In addition to providing excellent surgical anaesthesia the presence of an epidural catheter allows controlled venodilatation and an additional means to treat right heart failure. Given the initial evidence of right heart failure in this patient, this is advantageous not only intraoperatively but also postoperatively when fluid balance may be affected by such factors as autotransfusion of uterine blood. It also offers superior postoperative opioid analgesia and patient comfort and this may help to prevent tachycardia and impairment of cardiac function. Epidural anaesthesia also avoids subjecting the patient to tracheal intubation and the possible aspiration of gastric contents. Reduced neonatal depression is a further possible benefit.⁵ We were successful in inserting a lumbar epidural catheter at the L₄-L₅ level. Bupivacaine, 0.5% without epinephrine, was administered in 2-3 ml boluses to a total of 15 ml over 30 min. Bupivacaine was chosen because of its slow onset of action. It was considered that an epinephrine-containing test dose of epidural local anaesthetic would not be useful due to the confounding effects of intermittent painful uterine contractions and *iv* ketamine boluses on maternal heart rate. Surgical anaesthesia was confirmed using painful stimuli administered during periods of diminishing sedation. Using this method, we avoided hypotension, maintained haemodynamic stability, and preserved satisfactory cardiac function.

The use of ketamine in the parturient has been widely

reported since the original description in 1966.⁶ It has been used to provide both analgesia for vaginal delivery⁷⁻⁹ and as an anaesthetic induction agent for Caesarean section.^{10,11} Intravenous ketamine provides profound analgesia, rapid onset of action, rapid recovery, and general support of the cardiac and respiratory systems. Maternal adverse effects include delirium and hallucinations, amnesia for the birth, increased salivation, and excessive increases in heart rate and blood pressure. Conflicting results have been reported with regard to the oxytocic effects of ketamine.¹²⁻¹⁴ Reduced uterine blood flow secondary to uterine hypertonus is avoided with doses $<1.1 \text{ mg} \cdot \text{kg}^{-1} \text{ iv}$. Several studies have reported on the neonatal effects of ketamine.^{7,9,11,15} Varying degrees of neonatal depression have been reported with doses $>1.5 \text{ mg} \cdot \text{kg}^{-1}$. However, the neonatal effects of doses $<1 \text{ mg} \cdot \text{kg}^{-1}$ have been found to be comparable with those of other anaesthetic agents. The suggested doses for ketamine in parturients are $1 \text{ mg} \cdot \text{kg}^{-1} \text{ iv}$ bolus for induction of anaesthesia for Caesarean section,¹⁶ and 10-15 mg *iv* boluses repeated every two to five minutes to a maximum of 100 mg over 30 min for analgesia during labour and delivery.¹⁷

We administered boluses of ketamine 10-20 mg *iv* every five minutes as required for sedation. The cumulative dose was 245 mg ($4.5 \text{ mg} \cdot \text{kg}^{-1}$), titrated over 90 min. Of this, 175 mg were given before delivery of the fetus. The decision to use ketamine was based on several factors including its ability to elevate systemic and pulmonary vascular tone and maintain heart rate. Its inotropic effects offer a clear advantage in the management of severe pulmonary valvular stenosis. The lack of severe respiratory depression in subanaesthetic doses offers a margin of safety not found with other sedative medication. Profound analgesia was advantageous since assessment of sensory anaesthesia was difficult due to the patient's mental retardation. Neonatal depression was another concern. Assuming the (unknown) father was not affected by Watson's syndrome, autosomal inheritance of the gene allows for a 50% likelihood of the fetus being affected. Such a neonate may require extensive resuscitation which would be hampered by circulating sedative agents. We felt that the likelihood of neonatal depression with ketamine was less than with other sedative agents such as benzodiazepines or opioids given the relatively large doses required in this patient. Although the amount of ketamine used in this case exceeded the recommended guidelines,¹⁷ there was only minimal evidence of neonatal depression at birth. The surgeon reported no subjective impression of increased uterine tone. There was no evidence of hallucinations or vivid dreams after surgery, nor of excessive maternal salivation or airway obstruction.

Given the difficult nature of this problem, adequate preparation is essential to good patient management. Where possible, we believe this should include the presence of a second anaesthetist, a neonatologist experienced in advanced neonatal resuscitation, and the availability of a high-level nursery.

In conclusion, the anaesthetic management of a patient with Watson's syndrome is complicated by potentially severe dysfunction of the central nervous and cardiovascular systems. Anaesthetic management depends upon the severity of organ system involvement. We describe one method using *iv* ketamine sedation to facilitate invasive haemodynamic monitoring and the performance of epidural anaesthesia for Caesarean section which proved safe and effective.

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