Anaesthesia for scoliosis correction surgery complicated by severe recalcitrant bradycardia upon prone positioning in an adolescent with Prader-Willi syndrome

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Keywords: anaesthesia, cardiovascular, scoliosis, bradycardia, pacing

Abstract

A 13-year-old adolescent presented for correction of an 85-degree idiopathic thoracic scoliosis. She was known to have Prader-Willi syndrome. Previous general anaesthesia for non-spinal surgery had been uneventful. On two occasions following uneventful induction and total intravenous anaesthesia (TIVA) maintenance, she developed severe recalcitrant bradycardia with hypotension that was resistant to anticholinergics, inotropes and vasopressors upon prone positioning. Immediate resolution occurred upon a return to the supine position. On each occasion, she emerged from anaesthesia with no untoward sequelae. Cardiac investigations, including echocardiography, electrocardiography (ECG), troponin and creatine kinase-MB fraction levels were all within the normal range, and ventricular function was good. It was necessary to urgently proceed with the surgery as the scoliosis was progressive, with risks of cardiovascular and respiratory compromise. Additionally, she was scheduled to recommence growth hormone therapy postoperatively to treat her growth retardation. Ultimately, she received propofol for induction of anaesthesia and TIVA with propofol and remifentanil infusions for the maintenance of anaesthesia. Post-induction, a transvenous pacing wire was placed under ECG guidance. A transoesphageal probe was inserted and cardiac function monitored throughout the procedure. Upon prone positioning, she again developed a bradycardia which responded to pacing and surgery was carried out uneventfully. Clinical examination and extensive investigations had failed to demonstrate any specific underlying cause for her repeated positional arrhythmia. Therefore, we deduced that the bradycardia was due to a hypervagal response provoked by prone positioning and because of the severity of the scoliosis. We are unaware of reports of such a complication in the literature.

℗ Peer reviewed. (Submitted: 2012-10-02. Accepted: 2013-03-04.) © SASA South Afr J Anaesth Analg 2013;19(3):171-173

Case study

We present the case of a 35-kg, 13-year-old adolescent who presented for correction of an 85-degree thoracic scoliosis. She was known to have Prader-Willi syndrome and had been treated over a two-year period five years previously with growth hormone. This therapy was discontinued because of deterioration of her scoliosis, but was scheduled to recommence following surgical correction of her spinal curvature. She had previously been diagnosed with a ventricular septal defect which was asymptomatic. Previous general anaesthesia for non-spinal surgery had been uneventful. A comprehensive preoperative workup was carried out, including a multigated acquisition scan of her heart which demonstrated good cardiac function of left ventricular ejection fraction of 62% (normal range 65% ± 9%) and right ventricular ejection fraction of 58% (normal range 54% ± 3%). Transthoracic echocardiography demonstrated a small ventricular septal defect. Pulmonary function tests demonstrated normal forced expiratory volume in one second (1.1 l, 83% predicted), and mildly reduced forced vital capacity (1.2 l, 78% predicted).

She was transferred to theatre for operative correction of her severe scoliosis. Anaesthesia was induced with intravenous (IV) propofol 2 mg/kg, followed by IV atracurium (0.5 mg/kg). Endotracheal intubation was carried out uneventfully with a size 6 cuffed endotracheal tube. Maintenance of anaesthesia was provided by means of total intravenous anaesthesia (TIVA) in the form of propofol (Marsh pharmokinetic model) and remifentanil infusions, titrated to effect. Two additional large-bore peripheral cannulae, a right internal jugular triple-lumen central line, and a radial arterial line were inserted. Initially, she was stable. However, shortly after turning prone onto the Risser frame, she developed recalcitrant bradycardia with associated hypotension. This was unresponsive to boluses of ephedrine (3 mg and 6 mg IV) and adrenaline 100 µg IV.

She was returned to the supine position and immediately her heart rate recovered. A decision was made to abandon the surgical procedure. She was transferred to the paediatric intensive care unit (PICU). She was subsequently extubated with no untoward sequelae and an electrocardiography (ECG) that was carried out in PICU demonstrated normal anatomy, function and no evidence of dyskinesis. Troponin and creatine kinase-MB fraction levels were normal. There was no evidence of an underlying rhythm abnormality. She was discharged home and surgery was rescheduled. Some months later, a subsequent attempt at surgery resulted in the same complications and outcome.

Concern existed that if the scoliosis was not corrected, ongoing progression would result in fatal respiratory and cardiovascular compromise. In addition to this, recommencement of growth hormone therapy which was necessary to allow her to achieve maximal height was being delayed until the correction of her scoliosis. Seven months later, she was readmitted for surgery. On this (third) occasion she once again received an IV induction of anaesthesia with propofol 3 mg/kg and a neuromuscular blockade with atracurium 0.5 mg/kg. She was intubated uneventfully. TIVA was used for the maintenance of anaesthesia with propofol and remifentanil anaesthesia in the form of propofol and remifentanil infusions. Following induction, two large-bore peripheral venous cannulae and a radial arterial line were inserted, as well as a transvenous pacing wire, with the aid of ultrasound guidance and ECG monitoring via the right internal jugular vein. A transoesphageal echocardiographic probe was inserted to allow continuous monitoring of cardiac function and detection of regional wall motion abnormalities intraoperatively. Upon turning prone onto the Risser frame, the patient once again developed profound bradycardia. This was successfully treated with pacing (using the VVI mode) at a rate of 90 beats per minute. The patient's blood pressure was restored. Surgery proceeded uneventfully and she was successfully extubated at the end of the procedure.

The patient was subsequently recommenced on growth hormone therapy. She was seen postoperatively in the cardiology outpatient department and follow-up echocardiography demonstrated left ventricular systolic function in the normal range, with a measured shortening fraction of 40%, and no evidence of cardiomyopathy.

Discussion

This case highlights a highly unusual and difficult predicament. It was necessary to urgently proceed with surgery in this young girl, given the severity of the scoliosis, with risks of progression and cardiorespiratory compromise. In addition, the recommencement of the growth hormone therapy to allow achievement of maximal height was dependent on scoliosis correction. Extensive investigations failed to elucidate a cause for the occurrence of bradycardia upon prone positioning. It is postulated that because of the severe nature of the scoliosis, prone positioning resulted in vagus nerve impingement.

Scoliosis is defined as "any lateral curve, tilt, or angular deviation of one or more vertebral segments of the spine from the normal vertical positioning".[1] Spinal surgery to correct scoliosis deformities presents a number of challenges to the anaesthetist because of the extensive nature of the surgery, with risks of large blood loss and fluid shifts, often in the context of multiple patient co-morbidities, in addition to the physiological consequences of scoliosis, with effects on both the cardiovascular and respiratory systems.

As curvature of the spine progresses, there is a resultant restrictive lung defect. With severe curvature (greater than 100 degrees), severe respiratory compromise can occur with ventilation or perfusion mismatching, respiratory failure and pulmonary hypertension.[2] The indications for surgical correction of idiopathic scoliosis are progression in a patient receiving nonoperative treatment with curvature of greater than 40 degrees, curvature of 40-45 degrees in a non-skeletally mature patient, or greater than 50 degrees in a patient who is skeletally mature.[3]

The Prader-Willi syndrome is caused by a defect in the expression of genes on chromosome 15. Features include neonatal hypotonia, growth retardation, characteristic facies, hypogonadism, skeletal deformities and developmental delay. In later years, characteristically, there may be hyperphagia with resultant obesity. Obstructive sleep apnoea may occur in association with both obesity and hypotonia. Scoliosis is a common finding and patients frequently have a restrictive pulmonary defect. Anaesthetic implications include complications of obstructive sleep apnoea, in addition to problems that relate to disordered thermoregulation.[4]

There have been reports in the literature of cardiovascular abnormalities, such as hypertension and arrhythmias (premature ventricular contractions), in patients with this syndrome.[5] However, we could find no reports of recalcitrant bradycardia occurring in association with the Prader-Willi Syndrome upon prone positioning.

Transcutaneous pacing has been used with success in the emergency management of bradycardia in adults, but is a less common modality of treatment in children. Beland et al[6] demonstrated how noninvasive transcutaneous pacing may be safely and successfully used in children. In our case, bradycardia and hypotension rapidly corrected upon return to the supine position. Therefore, transcutaneous pacing was not necessary. As a means of facilitating the surgery, it was apparent in this complex case that the use of anticholinergic, chronotropic and inotropic drugs would not ensure maintenance of heart rate and blood pressure. The use of transcutaneous pacing was not considered to be a viable option because of difficulties with electrode positioning in prone positioning, particularly in an emergency situation and owing to the risk of skin burns, should the need have arisen for the electrical output to be increased to achieve capture.[7] For this reason, a transvenous pacing wire was inserted post-induction under ECG guidance, prior to attempts at prone positioning.

The choice of TIVA for the maintenance of anaesthesia was not unusual in such a case. There is no definite association between the Prader-Willi syndrome and malignant hyperpyrexia. However, in our institution, all children with musculoskeletal abnormalities receive trigger-free anaesthesia as a precautionary measure. A propofol infusion has the advantages of being antiemetic and rapidly cleared with minimal postoperative residual effects. When combined with an opioid infusion, a lower dose of propofol is required,[8] with less haemodynamic compromise and faster drug clearance. However, there are reports in the literature of propofol being vagotonic, in particular when administered in association with other vagotonic drugs.[9] Remifentanil is associated with the development of bradycardia. However, on each occasion, the development of bradycardia in our patient occurred in the same pattern: immediately following prone positioning on the Risser frame, prior to commencement of the remifentanil infusion (the propofol infusion having being commenced post-induction without untoward effects on heart rate or blood pressure). Therefore, we were confident that the bradycardia in our patient was not a drug-related effect.

The prone position is essential for scoliosis correction surgery. However, prone positioning carries risks of significant complications which were reviewed recently by Edgcombe et al,[10] and include airway device dislodgement, nerve injury, pressure injuries, eye injuries, air embolism, arterial and venous occlusion and injury to the cervical spine. There are reports in the literature of rare, but serious, cardiovascular complications that occur upon turning prone. Bafus et al[11] reported a case of severe hypotension upon prone positioning in a child with pectus excavatum undergoing spinal fusion. This necessitated correction of the pectus excavatum prior to proceeding with scoliosis surgery. Although there are numerous reports of bradycardia occurring during prone positioning in association with presumed air embolism,[12-14] the development of profound bradycardia being experienced on positioning prone is rare. Van Aken et al reported the development of fatal neurogenic pulmonary oedema and bradycardia following prone positioning in a child with neurofibromatosis, who had an undiagnosed posterior fossa neurofibroma that caused compression of the midbrain when the child was turned prone.[15] However, we could find no reports in the literature of severe bradycardia occurring in a patient with scoliosis upon turning prone.

Prone positioning of the child must be carried out meticulously. The vertebral column should be slightly flexed and adequately supported by means of a padded frame. Patients with scoliosis are particularly difficult to position optimally because of anatomical distortion. A study by Von Ungern-Sternberg et al[16] compared the effects of "flat" and "augmented" prone positioning in children. The "augmented" prone position involved the use of supports for the pelvis and upper thorax. Functional residual capacity and ventilation distribution were improved significantly when the "augmented" prone position was used. This suggests that adequate positioning has a significant impact on pulmonary mechanics. The Risser frame is used in our institution to attempt to achieve optimal patient position. However, this does not allow for extremes of augmented positioning, should it be required.

In conclusion, this was an unusual case of a girl with Prader-Willi syndrome with associated scoliosis and growth retardation presenting for scoliosis correction, against a background of previous recalcitrant bradycardia on prone positioning. This case highlights a rare complication of excessive vagal tone upon prone positioning which necessitated perioperative transvenous pacing to allow operative intervention. To our knowledge, this is as yet unreported in the literature.

Conflict of interest

The authors declare that they have no financial or personal relationships which may have inappropriately influenced them in writing this paper.

Declarations

This research was carried out without financial assistance.

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