

Anaesthetic Management Of Caesarean Section In A Patient With Syringomyelia.

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Abstract

We describe the case of a 32 year old primiparous woman with syringomyelia and Arnold-Chiari type I malformation who presented in early labour at 37 weeks gestation. She was delivered by caesarean section performed under general anaesthesia with a remifentanyl infusion. There was no intra-operative complication or neurological deterioration post-operatively. We discuss the anaesthetic implications of this disease in pregnancy and focus on the advantages of using a remifentanyl infusion as part of a general anaesthetic technique.

INTRODUCTION

Syringomyelia is a rare degenerative neurological condition characterised by the development of a cystic cavity or syrinx within the spinal cord.¹ The exact pathogenesis of syrinx formation and propagation are unknown although many associated factors are recognised. It is frequently associated with an Arnold-Chiari type I (AC I) malformation which describes the condition of elongation of the cerebellar tonsils with their displacement below the foramen magnum.^{2,3} Pregnancy exposes the mother to the risk of neurological deterioration as a result of the physiological changes and medical interventions experienced during labour and delivery.

CASE REPORT

A 32 year old primiparous woman presented at 37 weeks gestation in early labour. She had been diagnosed with syringomyelia and AC I malformation ten years previously. Neurological sequelae had gradually progressed and included left sided paraesthesia of her face, chest and limbs and loss of temperature sensation. Motor function, balance, speech and swallowing were unaffected. Her MRI showed a syrinx between C6 and T9 with a small degree of cerebellar tonsillar herniation which had not required decompressive surgery. During the pregnancy she suffered mild headaches with increasing frequency but no other change in her symptoms.

Delivery by caesarean section (CS) under general anaesthesia (GA) was planned following discussion with a

neuroanaesthetist, neurosurgeon and obstetrician. There was no anticipated difficulty with airway management. The patient was given oral ranitidine 150 mg and 30 mls 0.3 M sodium citrate preoperatively. Peripheral venous and radial arterial cannulae were inserted and routine anaesthetic monitors applied.

At induction the patient's heart rate was 110 min⁻¹ and her arterial pressure was 158/80. She was pre-oxygenated with 100% oxygen (O₂) for three minutes and a remifentanyl infusion started at 0.5 mcg kg⁻¹ min⁻¹. A modified rapid sequence induction was performed with thiopentone 250 mg and rocuronium 40 mg. Following tracheal intubation, anaesthesia was maintained with sevoflurane (end tidal concentration 1-1.5%) in a mixture of O₂/air (FiO₂ 0.5) and a remifentanyl infusion (0.1-0.2 mcg kg⁻¹ min⁻¹). There was minimal cardiovascular response to tracheal intubation or surgical stimulation. Normocapnia was maintained throughout the procedure with a peak airway pressure < 20 cmH₂O. A baby girl was delivered who required supplemental O₂ initially but no further resuscitation. Her Apgar scores were 6 at one and 9 at five minutes. Our patient was then given paracetamol 1 g and morphine 10 mg by intravenous injection. There was one brief episode of hypotension intraoperatively which responded to a bolus of phenylephrine 25 mcg. 2000 mls of Hartmann's solution was given during the procedure. At the end of surgery, the wound was infiltrated with 20 mls 0.5% levobupivacaine. Rectal diclofenac 100 mg was administered and the remifentanyl infusion was discontinued. Neuromuscular blockade was

reversed with neostigmine 2.5 mg /glycopyrrolate 500 mcg and the tracheal tube was removed once full muscle power had returned.

The patient's post operative recovery was uneventful. Analgesia was maintained with a patient controlled analgesia system delivering a standard regime of morphine 1 mg boluses. She demonstrated no change in neurological status and had no further headache. She was discharged home five days after her operation.

DISCUSSION

Syringomyelia is a rare progressive degenerative disease of the nervous system, with a prevalence of 8.4 per 100,000. It is characterised by the presence of a cystic cavity or 'syrinx' within the spinal cord.⁴ The term was coined in 1827 by Ollivier d'Angers from two Greek roots meaning 'to become hollow' and 'marrow'.⁵ Syringomyelia causes a progressive myelopathy and is divided into communicating and non-communicating types. Communicating syringomyelia is associated with congenital AC I malformation.⁶ It is thought that the malpositioned cerebellar tonsils partially occlude the subarachnoid space at the foramen magnum and act as a piston on the spinal subarachnoid space. This creates increased cervical subarachnoid pressure leading to syrinx development and progression. Symptoms worsen when CSF pressure fluctuates, for example during coughing/straining.⁷ Non-communicating syringomyelia occurs when a syrinx develops in a damaged section of spinal cord, for example following trauma or meningitis.⁸

The most common initial complaint is pain at the site of the syrinx radiating to the neck and upper extremities. Sensory loss is classically described as distal reduction in pain and temperature sensation with preservation of proprioception and light touch.⁹ Reduced motor power and muscle wasting are common features caused by destruction of lower motor neurones. Weakness in the paraspinal muscles may lead to thoracic scoliosis and respiratory impairment. Other sequelae depend on the level of the syrinx and include spasticity, hyperhydrosis (above level of injury), loss of sphincter control, sexual dysfunction, Horner's syndrome and autonomic dysfunction. Extension upward into the medulla (syringobulbia) leads to cranial nerve deficits characterised by paralysis of the palate, tongue and vocal cords and loss of facial sensation. Joint deformity or swelling (Charcot joint) may also be present. Signs may be unilateral due to one-sided pathology.¹⁰

Data concerning the preferred mode of delivery in patients

with syringomyelia is scanty but strict avoidance of straining during the second stage of labour is recommended to avoid an increase in intracranial pressure.⁴ To date there has been only one documented successful instrumental vaginal delivery using epidural analgesia in this patient group however the authors admit that the safety and efficacy of this delivery mode is unconfirmed.³ In our patient we chose to perform a CS following interdisciplinary discussion.

The optimal anaesthetic management of such cases has not yet been established. At present there is a limited body of evidence in the form of case reports of CS in patients with syringomyelia demonstrating successful use of both epidural^{2, 3, 11, 12} and general^{1, 4, 13-15} anaesthetic techniques. With epidural anaesthesia respiratory function is less compromised and the potential hazards of securing the airway are avoided. However during epidural insertion there is a risk of dural puncture and altered CSF pressure gradients.¹⁶ Nel and colleagues favoured this technique as they felt it was the best method to avoid aggravating the disturbed cranio-spinal pressure relationship. However, they admitted the possibility of cardiovascular instability and distension of the epidural space causing subarachnoid compression and a potentially damaging pressure wave in the syrinx.² General anaesthesia has been shown to be a safe and effective technique to facilitate CS in this patient group with excellent maternal and neonatal outcomes. The potential hazards of neuraxial instrumentation and consequent neurological deterioration are avoided. However care must be taken to avoid a rise in intracranial pressure caused by coughing or vomiting at induction or emergence.¹⁵

Anaesthetic assessment for CS in these patients must include a full history and examination of the patient's airway, respiratory, cardiovascular and neurological systems. The back should be inspected to assess ease of performing central neuraxial block and identify thoracic scoliosis. Physiological changes in pregnancy may exacerbate respiratory impairment and in the presence of pre-existing respiratory compromise, pulmonary function tests must be carried out. Evidence of autonomic dysfunction should be sought.^{1, 2}

The chosen anaesthetic technique must firstly aim to avoid elevating intracranial pressure or changing cranio-spinal pressure gradients.² Cardiovascular stability should be maintained and hypotension should be promptly treated with fluids and directly acting vasopressors. In autonomic dysfunction the vasodilatory effects of regional anaesthesia

are poorly tolerated and likewise the vasodilatation and cardiac depression of most general anaesthetic agents may be deleterious.¹⁷ Body temperature should be closely monitored especially if autonomic dysfunction is suspected.¹⁰ Patients who demonstrate evidence of syringobulbia with cranial nerve involvement are at a heightened risk of aspiration of gastric contents and consideration should be given to avoiding a GA in these cases.¹⁷ Drugs that cause post-operative respiratory depression should be avoided.

No case report has described the use of a remifentanyl infusion as part of a balanced general anaesthetic technique. Remifentanyl is an ultra short acting opioid receptor agonist with an analgesic potency similar to fentanyl. It has a rapid offset of action thanks to its degradation by plasma and tissue esterases and virtually inactive metabolites with no accumulation. It has been shown to provide more stable maternal heart rate and arterial pressure during elective caesarean section under GA.¹⁸ We found that the remifentanyl infusion effectively attenuated the cardiovascular response to intubation, a response which could potentially stimulate progression of a syrinx.¹⁶ Its rapid offset provides the advantage of minimal post-operative maternal respiratory depression. Remifentanyl crosses the placenta and although it appears to be rapidly metabolised or redistributed by the neonate, it may cause neonatal respiratory depression and should be reserved for cases with clear maternal indications.¹⁹

We used rocuronium to facilitate rapid intubation of the trachea as succinylcholine is associated with hyperkalaemia when given to patients with denervated muscles.²⁰ In some patients with lower motor neurone dysfunction there may be prolonged neuromuscular blockade with non depolarising muscle relaxants.²¹ It is therefore important to monitor the neuromuscular junction throughout the procedure. Our patient demonstrated a normal return to full muscle function following a standard induction dose of rocuronium. Invasive arterial pressure monitoring enables close control of haemodynamic parameters. Normocapnia should be maintained and high airway pressures avoided to prevent any rise in intracranial pressure. Extra care must be taken at extubation in view of possible impairment of protective airway reflexes.¹⁰ The patient should be closely monitored in the immediate 24 hours following surgery because of the risk of sudden apnoea or cardiac arrest in syringomyelia associated with autonomic dysfunction.¹⁶

In patients with syringomyelia, we feel that caesarean section under general anaesthesia avoids potentially hazardous instrumentation of the epidural or intrathecal spaces and minimises the risk of neurological deterioration. We recommend the use of a remifentanyl infusion as part of a balanced general anaesthetic technique in these patients because of its desirable pharmacological profile.

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