Extradural anaesthesia for Caesarean section in a patient with syringomyelia and Chiari type I anomaly

M. R. Nel, V. Robson and P. N. Robinson

Summary
We describe elective Caesarean section performed under extradural anaesthesia in a parturient with symptomatic syringomyelia and co-existing Chiari type I anomaly. Syringomyelia is reviewed and the anaesthetic implications of the condition discussed. Anaesthesia should be directed primarily at avoidance of increased intracranial pressure, which can cause sudden deterioration in these patients. (Br. J. Anaesth. 1998; 80: 512–515)

Keywords: complications, syringomyelia; complications, Chiari anomaly; anaesthesia, obstetric; anaesthetic techniques, extradural

Syringomyelia is an unusual neurological condition characterized by cystic degeneration within the spinal cord. The Chiari type I anomaly (also termed Arnold–Chiari type I anomaly) is one of a group of congenital abnormalities associated with descent of the hindbrain structures through the foramen magnum. We report the successful use of extradural anaesthesia for elective Caesarean section in a woman with both of these conditions.

Case report
A 31-yr-old woman, para 1, known to have syringomyelia and a related Chiari type I anomaly, was admitted for elective Caesarean section at 38 weeks’ gestation. Her neurological condition had been diagnosed after an MRI scan (fig. 1), performed 3 months before she became pregnant. Her previous obstetric history included spontaneous vaginal delivery of a healthy infant at term, using only i.m. pethidine as analgesia.

The MRI scan was performed to investigate a history of continuous pain in the right side of the neck, radiating to the right shoulder, which had begun approximately 2 yr earlier. The patient also had recurrent occipital headaches, aggravated by coughing or sneezing. Careful neurological examination after the scan had revealed reduced sensation to pain and temperature from C2 to approximately T8 on the left side, correlating with the appearance on the scan of the extent of the syrinx. She also had depressed reflexes in the left arm and wasting of the left thenar eminence.

After the MRI scan, the patient was assessed by a neurosurgeon who recommended urgent foramen magnum decompression to relieve her neck pain and headaches, and prevent her neurological deficit from worsening. However, she declined surgical intervention and subsequently became pregnant. During the second and third trimesters her headaches intensified, but she continued to refuse surgery. After advice from the patient’s neurologist and neurosurgeon, it was decided that an elective Caesarean section would be the method of delivery least likely to aggravate the syrinx.

At the preoperative visit the patient’s body weight was 58 kg and height 155 cm. Neck mobility and mouth opening were assessed as normal. Her lumbar spine had a mild scoliosis, concave to the right, but the spinous processes were easily palpable. Premedication comprised ranitidine 150 mg orally the night before the operation. The patient received epidural anaesthesia at the L2–L3 interspace, using 0.25% bupivacaine 15 ml, with a loading dose of diazepam 5 mg and fentanyl 50 μg. The patient remained haemo- and normotensive throughout the procedure.

Figure 1 T2-weighted sagittal MRI of the upper spinal cord and brainstem. The syrinx (small arrows) appears discontinuous because of the thin slice. The Chiari type I anomaly is also indicated (large arrow).
Extradural anaesthesia in syringomyelia with Chiari type I anomaly

before surgery, repeated on the morning of surgery together with metoclopramide 10 mg.

On arrival in the operating theatre a 16-gauge peripheral i.v. cannula was inserted, and the patient was given Hartmann’s solution 1000 ml as a preload. Monitoring consisted of pulse oximetry, ECG and non-invasive automated arterial pressure. The sitting position was chosen because of patient preference. Using a 16-gauge Tuohy needle, the extradural space was located by loss of resistance to air and catheterized at the L2–3 interspace. The patient was then placed supine with left lateral tilt. Extradural anaesthesia was established by slowly injecting 3–4 ml bolus doses at 5-min intervals, using a total of 20 ml of 0.5% bupivacaine, to which had been added fentanyl 50 μg. Supplementary oxygen was given by face mask and 35 min after the first bolus the height of the block was assessed as T4 bilaterally. Surgery was started and a healthy baby weighing 2680 g was delivered 12 min later; this was followed by slow i.v. injection of oxytocin 10 μ. Caesarean section proceeded uneventfully, and Hartmann’s solution 1000 ml was given i.v.; blood loss was estimated at 350 ml. The patient remained haemodynamically stable and no vasopressors were required.

Postoperative analgesia consisted of morphine 7.5–10 mg i.m. every 3 h as required, rectal diclofenac up to 150 mg per 24 h and oral co-dydramol. Her neurological condition remained unchanged. She recovered well from the Caesarean section and was discharged on day 4 after operation.

Seven weeks after delivery the patient was reviewed: she continued to have neck pain and headaches but there had been no neurological deterioration. She remains reluctant to undergo neurosurgery, but is currently awaiting elective admission for sterilization.

Discussion

Syringomyelia is characterized by the presence of an expanding, longitudinal cystic cavity (syrinx) within the spinal cord. Ollivier d’Angers (1827) coined the term syringomyelia1 from two Greek roots meaning “channel” and “marrow”. It is an uncommon condition, with a prevalence of 8.4 per 100 000 identified by MRI, particularly in evaluating scoliosis.11

The foramen magnum abnormalities cause intermittent obstruction to CSF outflow from the fourth ventricle, with the development of craniospinal pressure dissociation, a relatively higher CSF pressure in the head and lower pressure in the spine. This pressure dissociation may not be apparent if only baseline intraventricular and lumbar CSF pressures are studied, but is usually measurable after an episode of increased thoracoabdominal pressure, for example after a cough or Valsalva manoeuvre. The resultant increase in CSF pressure is more pronounced and more sustained in the intraventricular CSF.5 Development of a craniospinal pressure gradient is believed to be associated with intermittent propagation of pressure waves into the central canal, which eventually causes dissection into adjacent neural tissue.4 The resultant syrinx typically arises in the lower cervical or upper thoracic region; expansion is characteristically very gradual, initially into the anterior and lateral cord. Extension upwards into the medulla often occurs (syringobulbia), whereas involvement of the lumbar cord is uncommon.

In the non-communicating form the syrinx may develop anywhere in the cord. Factors involved in initiating cavitation may include haematoma formation, ischaemia, venous obstruction, shearing stress with mechanical disruption and secretion of proteaceous fluid.145 Subsequent progression of the syrinx may depend on a CSF pressure gradient developing across the lesion, as in the communicating form.5

The clinical picture is consistent with atrophy of anatomically susceptible spinal cord and brainstem tissue. A classic case of communicating syringomyelia presents with asymmetrical loss of pain and temperature sensation in the upper limbs (lateral spinothalamic tracts), lower motor neurone signs ascending from the hands (anterior horn cells) and upper motor neurone signs in the lower limbs (corticospinal tracts). The presence of posterior column signs usually signifies advanced disease.4 Involvement of the autonomic nervous system is not uncommon. Trophic changes may be striking, particularly the development of gross osteoarthropathy (Charcot’s joints). Syringobulbia is most commonly denoted by trigeminal or vagus nerve dysfunction; other cranial nerves which may be affected include the third, sixth, seventh, ninth and 12th.46 Symptoms may arise independently from a coexisting Chiari type I anomaly; these are often related to Valsalva-type manoeuvres, and include paroxysmal headaches, neck pain, syncope, vertigo, weakness and sensory disturbance.578

Currently, the most common operation performed for communicating syringomyelia involves decompression at the foramen magnum.910 Best results are obtained when surgery is performed relatively early in the course of the disease. Without surgical intervention, the natural history follows one of three patterns: chronic progressive disease, chronic stable disease or, occasionally, rapid onset of severe disability within a few years of diagnosis.6

Mean age at onset of symptoms is 25–40 yr.246 However, cases of syringomyelia are now detected frequently in childhood as a result of increasing use of MRI.
fore, it is expected that a number of female patients with known syringomyelia will require obstetric anaesthesia, although there appears to be only one previous case report in the literature.12

Before an anaesthetic plan can be formulated, a thorough neurological appraisal is necessary, including MRI scan. Several associated abnormalities may have specific implications for anaesthesia—kyphoscoliosis, pes cavus, cervical ribs, fusion of cervical vertebrae (Klippel–Feil syndrome), hydrocephalus and spina bifida,4,6 in addition to the presence of a Chiari type I anomaly or other foramen magnum disorder.7

Neurological assessment must incorporate a check for evidence of autonomic neuropathy; cardiac autonomic neuropathy is more likely when there is coexisting syringobulbia.13 The diagnosis is best made by performing a series of simple bedside tests,12–14 although the presence of a prolonged corrected QT interval (QT) has proved a useful single indicator in diabetic patients.14 Prior identification of autonomic involvement is important as these patients commonly exhibit tachyarrhythmias and a widely fluctuating arterial pressure in response to the stress of anaesthesia and surgery. Moreover, sudden cardiac or respiratory arrest has been attributed to autonomic neuropathy in syringomyelia patients after posterior fossa surgery,13 and in diabetic patients after general or regional anaesthesia.14,15 Further considerations include possible delayed gastric emptying, urinary bladder dysfunction and impaired thermoregulation.

If syringomyelia involves the respiratory muscles or there is significant associated kyphoscoliosis, pulmonary function tests should be performed. These tests can also indicate the presence of unsuspected vocal cord abductor paresis by a characteristic flattening of the inspiratory curve on a flow–volume loop.8

Where trophic changes in the skin are widespread, peripheral venous access may be limited. Acrocyanosis of the hands may be a feature,5 with the potential for confusion over central oxygenation status. There may be disorganized joints, demineralization of bone and fixed flexion deformities,5 requiring careful movement and positioning of these patients to avoid pathological fractures and soft tissue ischaemia. Areas of sensory loss increase the potential for inadvertent injury to occur, even in the conscious patient.

In our opinion one of the most important goals of anaesthesia is to avoid aggravating the already disturbed craniocervical pressure relationship. Even in patients without symptoms of increased intracranial pressure, and without clinical or radiological hydrocephalus, it can be assumed that intracranial CSF pressure is relatively high as this is fundamental to the pathophysiology of communicating syringomyelia. Sudden clinical deterioration in syringomyelia after coughing or straining is well recognized.4,9 It follows that an appropriate anaesthetic technique has to avoid any further increase in intracranial pressure.4,9

General anaesthesia requires careful prior airway assessment and planning. Intubation may be complicated further by the need to avoid succinylcholine if significant muscle denervation has occurred, because of the risk of a hyperkalaemic response.16 In addition, increased sensitivity to non-depolarizing neuromuscular blocking agents has been demonstrated in syringomyelia.17

An earlier case report ruled out regional anaesthesia for medico-legal reasons.12 However, the presence of active neurological disease is no longer considered an absolute contraindication to regional anaesthesia.18 We believe an extradural anaesthetic offers several advantages over general anaesthesia in these patients: the potential hazards of securing the airway are avoided, respiratory function is less compromised and the existing craniocervical CSF pressure relationship is better preserved. Extradural block should be established very slowly: first, to avoid the precipitate decrease in arterial pressure that may occur if there is autonomic neuropathy and second, to avoid sudden distension of the extradural space. Such distension may induce significant subarachnoid compression, thereby both increasing intracranial pressure19 and creating a potentially damaging pressure wave within the syrinx.7

We suggest that spinal anaesthesia is best avoided in syringomyelia, particularly in the presence of a co-existing Chiari malformation, as there are case reports describing the onset of signs and symptoms up to 2 weeks after dural puncture.20,21 Whatever the mode of anaesthesia, these patients require close postoperative observation, particularly if there is syringobulbia, autonomic neuropathy or recently progressive disease, when sudden apnoea or cardiac arrest may be more likely.

The decision to perform a Caesarean section in this case was an interdisciplinary one, with the full agreement of the woman concerned. In the absence of existing guidance in the literature, others may have opted to permit labour. In this event, we would recommend early siting of an extradural catheter and strict avoidance of straining during the second stage.

References


