Case Report

Dural ectasia: a likely cause of inadequate spinal anaesthesia in two parturients with Marfan’s syndrome


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We report two cases of Caesarean section in patients with Marfan’s syndrome where continuous subarachnoid anaesthesia failed to provide an adequate surgical block. This was possibly because of dural ectasia, which was confirmed by a computed tomography scan in both cases.

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Marfan’s syndrome is a genetic disorder of connective tissue characterized by significant cardiovascular and musculoskeletal complications.1 Anaesthetic management of parturients with Marfan’s syndrome can be challenging. We present two cases depicting similar erratic spread of intrathecal local anaesthetics, from two different institutions.

Case report 1

A 28-yr-old (G1P0) white female with Marfan’s syndrome and surgically corrected thoracolumbar kyphoscoliosis was admitted at 20 weeks’ gestation because of chest pain. A type A dissecting ascending aortic aneurysm was diagnosed and she was treated with β-blockers and discharged home after stabilization. At 30 weeks’ gestation she was readmitted for recurring chest pain. She was hospitalized until fetal lung maturity was confirmed at 34 weeks. The patient was then scheduled for an elective Caesarean section and 1 week later for aortic valve and ascending aorta replacement.

On physical examination she had evident Marfan’s syndrome body habitus: height 180 cm, weight 80 kg and arachnodactyly. Upon arrival at the operating theatre, arterial pressure was 140/80 mm Hg and heart rate was 60 beats min⁻¹. Airway examination was unremarkable. Her back revealed a scar extending from T1 to L3 from a previous kyphoscoliosis repair; she denied lumbar pain.

Continuous spinal anaesthesia was planned. Under standard non-invasive monitoring (ECG, arterial blood pressure and oxygen saturation), a 22 gauge spinal catheter over the needle (Spino-cath; B. Braun, Melsungen, Germany) was inserted in the left lateral position at L4–L5 interspace under sterile conditions (4 cm in the intrathecal space) by a midline approach. Aspiration of cerebrospinal fluid (CSF) was confirmed at all times. With the patient in the left lateral position, an initial bolus dose of plain bupivacaine 0.5%, 9 mg and fentanyl 20 μg was given, followed by plain bupivacaine 6 mg because of a bilateral low sensory level (T12). Twenty-five minutes later, after moving the patient from side to side in the supine wedged position with the knees flexed by a pillow, a third bolus dose of plain bupivacaine 6 mg was given. Despite the incremental dosing up to 21 mg of bupivacaine, the highest sensory level blocked was T10, as assessed by sharp pinprick. General anaesthesia was therefore induced using remifentanil 0.1–0.2 μg kg⁻¹ min⁻¹, propofol 2 mg kg⁻¹, succinylcholine and isoflurane in a mixture of oxygen–nitrous oxide. A male infant weighing 2120 g was delivered, with Apgar scores of 5, 7 and 9 at 1, 5 and 10 min respectively. Surgery lasted 50 min and the trachea was extubated at the end of the procedure, with excellent haemodynamic stability throughout the case, as previously described with a similar technique.2,3 On recovery, she had a sensory block at T12. She had no post dural puncture headache. A computed tomographic (CT) scan of her spine revealed an ectasic thecal sac (Fig. 1).

Eight days later, the aortic valve and ascending portion of the aorta were replaced. She had an uneventful postoperative course and was discharged home 6 days later.
Case report 2
A 21-yr-old (G1P0) white female with Marfan’s syndrome presented at 33 weeks’ gestation for elective Caesarean section. She had aortic root dilation of 5.8 cm and severe mitral valve regurgitation/prolapse treated with atenolol and digoxin. She also had severe thoracolumbar kyphoscoliosis. At the time of admission she was 165 cm tall, weighed 55 kg and had evident Marfan’s syndrome stigmata. She had a normal airway examination and no evidence of pulmonary compromise.

Continuous spinal anaesthesia technique was planned for the elective procedure. Preoperatively, left radial arterial and right internal jugular venous catheters were placed. An interventional neuroradiologist placed a spinal catheter at L5–S1 in the left lateral decubitus position, using CT fluoroscopic guidance with an 18 G pencil-point epidural needle (Special Sprotte®; Pajunk, Geisingen, Germany). The catheter was left 5 cm in the subarachnoid space. The procedure was uneventful and the images showed dural ectasia and severe kyphoscoliosis (Fig. 2). Plain bupivacaine (0.5%) 10 mg was administered incrementally via the spinal catheter with the patient in the wedged supine position. A patchy spread was noted by sensory testing using sharp pinprick. A further 7.5 mg of hyperbaric bupivacaine 0.75%, administered over the next 30 min, combined with moving the patient from side to side, led to only marginal improvement. The block remained patchy with a sensory block to T7 on the left and L1 on the right. Aspiration of CSF was confirmed at all times.

Because of the patchy nature of the spinal anaesthesia, this course of action was aborted and general anaesthesia was therefore administered using a rapid sequence induction with fentanyl 200 μg, propofol 70 mg and succinylcholine 100 mg. Anaesthesia was maintained using sevoflurane in oxygen–nitrous oxide mixture. The arterial pressure and pulse rate remained virtually unchanged from the preoperative values of 105/55 mmHg and 70 beats min⁻¹ respectively. A male infant was delivered with Apgar scores of 7 and 8 at 1 and 5 min respectively. At the end of surgery the trachea was extubated and the patient was transferred to the critical care unit. Intravenous ketorolac 30 mg and ondansetron 4 mg were given towards the end of surgery. Morphine 150 mg was administered via the spinal catheter and analgesia was maintained with patient-controlled spinal analgesia using a bolus of bupivacaine 2.5 mg with fentanyl 25 μg and a lockout period of 45 min. The catheter was removed 18 h after surgery. On day 3 the patient complained of headache and neck pain, but declined epidural blood patch for likely postdural puncture headache. She was discharged on postoperative day 5 and the headache resolved by day 9.

Over the following year she developed worsening of her cardiovascular symptoms and was scheduled to undergo aortic and mitral valve replacement along with replacement of the ascending aorta.

Discussion
To our knowledge, this is the first report describing unpredictable and inadequate spread of intrathecal local anaesthetics in patients with Marfan’s syndrome, presumably secondary to dural ectasia.

Marfan’s syndrome is an autosomal dominant disease resulting from a mutation in the fibrillin-1 gene, which is
located on chromosome 15. It is a disorder of fibrous connective tissue that shows striking pleiotropism (the control by a single gene of several distinct and seemingly unrelated phenotypic effects)\(^1\) and clinical variability that can affect both men and women of any race or ethnic origin. Its incidence is 1:5000 individuals and the clinical diagnosis relies on a combination of major and minor clinical signs.\(^1\)\(^4\)

The cardinal clinical features occur in three systems: skeletal, ocular and cardiovascular. The most common skeletal alterations are increased height, disproportionately long limbs and digits, anterior chest deformity (pectus excavatum), mild to moderate joint laxity, vertebral column deformity (kyphoscoliosis and thoracic lordosis), and a narrow, highly arched palate with crowding of the teeth.\(^5\)

The ocular changes include myopia, increased axial globe length, corneal flatness and subluxation of the lenses (ectopia lentis).\(^5\) Cardiovascular manifestations are most frequently mitral valve prolapse, mitral regurgitation, dilatation of the aortic root and aortic regurgitation, but the major life-threatening cardiovascular complications are aneurysm of the aorta and aortic dissection.\(^5\)

Widening or ballooning of the lumbosacral dural sac, dural ectasia, can be found in 63–92\% of patients with Marfan’s syndrome.\(^6\)\(^7\) It is considered one of the major manifestations of the syndrome, together with ectopia lentis and aortic dilatation/dissection.\(^8\) It can be associated with back pain, the intensity of which correlates with the dural volume.\(^9\) However, a high prevalence of dural ectasia (41\%) exists even in patients with Marfan’s syndrome without back pain.\(^9\)

Regional anaesthesia has been used previously with success in patients with this syndrome, both for analgesia during labour\(^10\) and Caesarean section.\(^10\)\(^--\)\(^14\) There are no reports, however, describing patchy, erratic and inadequate spread of intrathecal local anaesthetics in patients with this condition. Many factors that affect the extent of spread of spinal anaesthesia have been identified. One of the most important factors influencing block height in patients receiving spinal anaesthesia is the lumbosacral CSF volume, contributing to the variability in the spread of spinal block.\(^15\)\(^16\) The erratic spread of spinal anaesthesia in both of these cases was most likely the result of dural ectasia and the associated increase in CSF volume. In both cases confirmation of the diagnosis of dural ectasia was done with a CT scan. Another possible explanation for the patchy block was the severe degree of kyphoscoliosis in case 2. However, this seems unlikely since there are multiple reports of successful neuraxial anaesthesia performed on patients with spinal deformities\(^13\)\(^17\)\(^--\)\(^19\) and patients with corrected kyphoscoliosis and Harrington rods.\(^13\) On the other hand, a caudal direction of intrathecal catheters has been identified as a cause for delayed onset of spinal block and a requirement for more local anaesthetic to establish the block.\(^20\) However, this seems unlikely to be the cause of failure since, in both cases, we waited for an adequate period of time, we rolled the patients from side to side trying to achieve an even spread of the local anaesthetics, and administered a relatively large dose of bupivacaine. Similar doses were reported to be effective in achieving at least a T4 surgical block in 90\% of patients undergoing Caesarean section with a
continuous spinal anaesthesia technique using plain bupivacaine. Furthermore, in case 2 the catheter was inserted under CT fluoroscopic guidance and a cranial direction of the catheter was evident. Finally, confirmation of intrathecal position of the catheter in both cases was assessed by frequent aspiration of CSF.

Plain bupivacaine is hypobaric and redistributes to non-dependent areas. The upright position during spinal injection of plain bupivacaine promotes cephalad spread of the block. Positional changes also play a major role in promoting cephalad redistribution of plain bupivacaine, probably due to CSF dynamics associated with caval compression and epidural venous engorgement. In the cases presented, the spinal injections were made in the lateral and supine wedged positions and the patients were turned from side to side. It is unlikely that administration of the spinal anaesthetic in the sitting position would have overcome the diluting effect of CSF resulting from dural ectasia.

A continuous spinal anaesthesia was chosen instead of a single shot subarachnoid block in both cases, with the aim of incrementally inducing a surgical block, thus avoiding excessive haemodynamic instability. Furthermore, continuous spinal anaesthesia has been shown to be highly predictable in terms of level and quality of anaesthesia. The presence of the catheter would also provide excellent post-operative analgesia. A disadvantage of this technique, however, is a higher risk of postdural puncture headache. Since microcatheters are not currently FDA-approved in the USA, we attempted to decrease the risk of headache by using the Special 18 G Sprotte needle with a 20 G catheter in patient 2, since this needle might be associated with a lower risk of postdural puncture headache compared with the Tuohy needle.

We did not use an epidural technique because of concern about epidural space fibrosis in case 1 and concern about difficult insertion and position of the catheter in the presence of severe kyphoscoliosis in case two. In both cases we were reluctant to further increase the dose of bupivacaine for fear of potential neurological injury. While the incidence of ‘transient radicular irritation’ (TRI) is strikingly more common with intrathecal lidocaine compared with bupivacaine (plain or hyperbaric), there are cases where the only aetiologic factor for the TRI was bupivacaine.

Anaesthetic management in patients with Marfan’s syndrome with significant aortic root dilation or dissection includes β-blocker therapy in order to minimise sheer stress and increase in aortic wall tension, and maintenance of haemodynamic stability by careful titration of regional or general anaesthesia. Despite failure of the regional technique in both cases, haemodynamic stability was maintained and the general anaesthetic was uneventful. As an alternative to inducing general anaesthesia after failure of the regional technique, we could have attempted to supplement the patchy block using local anaesthetic infiltration. However, we felt that intraoperative pain, with the associated tachycardia and hypertension, was undesirable in our patients with cardiovascular compromise. Our general anaesthetic technique was effective in blunting these responses.

In summary, we present two cases of Caesarean section in patients with Marfan’s syndrome in whom continuous spinal anaesthesia failed to provide adequate anaesthetic block, most likely as a result of dural ectasia.

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