



Inadequate Neuraxial Anaesthesia in Marfan's Syndrome

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Background

Marfan's syndrome is an autosomal dominant disorder of connective tissue, associated with an abnormal gene coding for fibrillin-1. Fibrillin-1 is a connective protein that provides support for extracellular structures. It also normally binds to transforming growth factor beta (TGF- β), a protein which has a negative impact on vascular smooth muscle and the extracellular matrix.

It can affect several different organ systems, including the heart, eyes, skeletal system and lungs. It is known to cause dural sac ectasia in 63-92% of affected adults¹. Dural sac ectasia can lead to inadequacy of spinal anaesthesia² due to the increased volume of the caudal dural sac.

Case Report

We analysed ten years of our databases at St. Mary's Hospital, Manchester for patients with Marfan's syndrome who had undergone anaesthesia. We retrieved the anaesthetic records of 11 parturients with Marfan's syndrome who had undergone a total of 14 Caesarean sections under regional anaesthesia. We looked for evidence of an inadequate regional block which we defined as: requirement for a second spinal, conversion to a GA or additional perioperative analgesia required.

Of the 14 procedures (7 spinals and 7 combined spinal-epidurals(CSE), 7 patients (50%) developed a block adequate for Caesarean section. 7 anaesthetics (50%) were deemed inadequate, requiring further anaesthesia or analgesia.

Of the 7 inadequate blocks, the mean initial spinal dose of 0.5% hyperbaric bupivacaine was 2.8ml (ranging from 1.5-4ml). It is worth noting that the mean dose has been skewed downwards by one low dose CSE with an initial starting dose of 1.5ml of heavy marcaine. Six received intrathecal opiates. 5 of the 7 procedures were performed under CSE and 2 under spinal. 4 of the 5 patients that had a CSE received epidural local anaesthetic but despite this all four required additional measures (1 GA, 1 repeat spinal, 2 intravenous opiates).

Interestingly, one patient who received an inadequate spinal had had a recent MRI of her lumbar spine showing no evidence of dural ectasia which suggests that MRI appearances of the lumbar spine may not be useful in predicting response to spinal anaesthesia.

Details of Cases with an Adequate Block for LSCS

Case Number	Method of RA	Initial spinal dose (heavy marcaine and diamorphine unless stated)	CSE epidural used?	Repeat Spinal	Supplementary Analgesia	Convert to GA
1	CSE	3.5+300		No	No	No
2	Spinal	3.2+150mcg morphine and 20mcg fentanyl		No	No	No
3	Spinal	3.0+300		No	No	No
4	CSE	2.0+300	10ml of chirocaine	No	No	No
5	Spinal	3.0+300	No	No	No	No
6	Spinal	2.6+300		No	No	No
7	Spinal	3.0+300		No	No	No

Details of Cases with an Inadequate Block for LSCS

Case Number	Method of RA	Initial spinal dose (heavy marcaine and diamorphine unless stated)	CSE epidural used?	Repeat Spinal	Supplementary Analgesia	Convert to GA
1	CSE	1.5	5ml 0.5% bupiz	No	Alfentanil 250mcg	No
2	CSE	2.5+300	5ml 2% lignocaine and 10ml 0.5% chirocaine	No	GA	Yes
3	Spinal	4.0+400		Yes - 2ml	100mcg fentanyl, paracetamol	No
4	CSE	2.8+300	No	No	Alfentanil 250mcg	No
5	Spinal	2.6+300		No	Alfentanil 1.4mg, N2O	No
6	CSE	3.4+300	23ml chirocaine in divided doses	No	5mg diamorphine, 500mcg alfentanil	No
7	CSE	2.8+300	10ml of bupivacaine 0.5%, 10ml of 2% lignocaine with bicarb and adrenaline	Yes - 1ml	Yes - 1mg alfentanil and N2O	No

Discussion

This case series suggests that patients with Marfan's syndrome have a higher incidence of inadequate block with subarachnoid anaesthesia. We do not have figures at our hospital but our rate of 50% of blocks being inadequate is a factor of 10 above the Royal College of Anaesthetists accepted incidence for inadequate block.

Attempts to compensate for the possibility of dural ectasia with a higher dose of local anaesthetic in the spinal, or by placing a CSE are not successful in all cases. The former approach may also be relatively contraindicated if there are concerns about cardiovascular pathology as it may lead to a deleterious hypotensive response. Radiological appearances of the dural sac may not correlate with clinical response to neuraxial anaesthesia. The preoperative discussion with the patient should highlight the higher risk of inadequate block with increased need for supplemental analgesia and conversion to general anaesthesia.



Magnetic Resonance Image of a Marfan's Patient with Dural Ectasia³

References

1. A. Altman et al, Dural Ectasia as Presenting Symptom of Marfan Syndrome, IMAJ 2008;10:194-195
2. Laccasie HJ, Millar S, Leithe LG, et al. Dural ectasia: a likely cause of inadequate spinal anaesthesia in two parturients with Marfan's syndrome. Br J Anaesth 2005;94:500-4.
3. <http://radiopaedia.org/images/1578406> Accessed 11th March 2013

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