

# LABOR ANESTHESIA IN MARFAN SYNDROME, AN INTERESTING CASE

Tobias Robinson, MD; Rebecca Evans, MD; Bridget Marroquin, MD  
Department of Anesthesiology , University of Vermont Medical Center, Burlington, Vermont

## Introduction

Marfan syndrome, named after Antoine-Bernard Marfan who in 1896 described young 5-year-old Gabrielle’s arachnodactyly, is considered a rare hereditary connective tissue disorder with a prevalence of 1 in 5,000 people worldwide.[1] It carries many potentially morbid medical complications including aortic root dilation, aortic dissection, ectopic lentis, tall stature with arachnodactyly, scoliosis or kyphosis of the spine, pectus excavatum or carinatum, and dural ectasia. [3]

Dural ectasia, as defined by De Paepe et al, is a dilation of the dural sac anywhere along the spinal column often accompanied by thinning of the lamina, cortex, and pedicles of the vertebral bodies, and can be found in 63% to 92% of individuals with Marfan syndrome.[2,3,6] These are often overlooked as part of Marfan syndrome but can influence anesthetic management dramatically, such as unpredictable intrathecal anesthetics, inadvertent dural puncture, fecal and urinary incontinence, and pain in the lower back, buttocks, and lower extremities.

## Case

A 34 year old G2P1001 female at 38 weeks and 6 days presented for induction of labor in the setting of gestational hypertension, with a known diagnosis of Marfan syndrome (FBN1 32 mutation). She has a history of prior vaginal delivery with epidural labor analgesia, mild-moderate mitral regurgitation, aortic root dilation, premature ventricular contractions, and mild dextroscoliosis of the thoracolumbar spine.

Epidural from prior labor was placed using a 17g Touhy at the L3-4 level in a single attempt. She had incomplete coverage of her left hip and elected to have the epidural replaced. A combined spinal-epidural was done with 17g Touhy needle and accompanying 27g Whitacre. Labor analgesic goals were accomplished as indicated by absence of painful contractions. However, she continued to endorse lumbosacral pain despite repeated boluses from the PCEA. The infusate was changed from 0.0625% bupivacaine-2% fentanyl solution to a more concentrated 0.125% bupivacaine-2% fentanyl solution. She delivered several hours later with minimal pain.

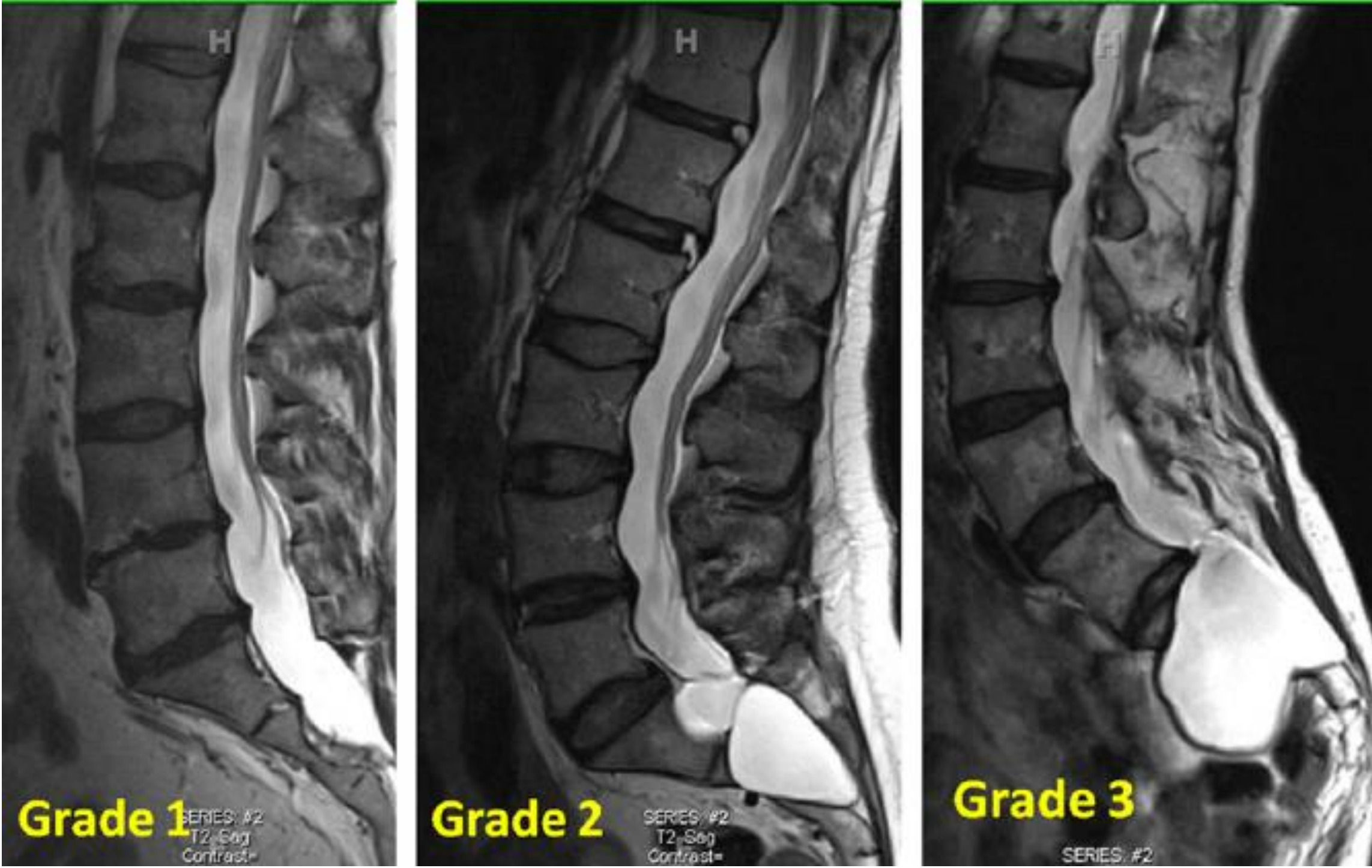
During her presentation for this case, a labor epidural was requested. Using ultrasound for assessment of anatomic landmarks, an epidural was placed on first attempt at the L3-4 level using a 17g Touhy. No paresthesias were noted during placement of catheter. Assessment of epidural at 60 minutes demonstrated no discernable level of analgesia despite repeat boluses of 0.125% bupivacaine. Decision was made to replace the epidural. The second attempt at L3-4 level resulted in accidental dural puncture. Catheter was threaded into the intrathecal space and continuous spinal catheter was used for analgesic control. However, follow up over the next several hours revealed no discernable analgesic level. She revealed that the spinal catheter had never given her any pain relief.

## Case Continued

Following discussions with the patient, the catheter was removed and a 3<sup>rd</sup> attempt was made to replace the epidural. A combined spinal-epidural was attempted at the L4-5 level. Ultrasound guidance was once again used to evaluate level and midline approach, litmus paper identified fluid return as CSF. An intrathecal dose of 1ml 0.25% bupivacaine with 50mcg of fentanyl was given. The catheter threaded easily into the epidural space and was confirmed by a negative test dose. The patient continued to have significant pain at 20 minutes following placement, so an epidural bolus dose of 7ml of 0.25% bupivacaine was administered and achieved a therapeutic T10 dermatomal level of analgesia. She delivered unassisted one hour later and the epidural catheter was removed

The following day she developed a postural headache consistent with a post dural puncture headache. After failing conservative management we elected for a sphenopalatine ganglion block given her neuraxial anatomy and difficulty with epidural anesthesia. She got little relief from the block and elected to continue with conservative management. She was discharged home the following day.

## MRI illustration of a Dural Ectasia Grading Scale



- The grading system of Fattori et al.[9]
- **Grade 0, Normal – Regular and rectilinear dural sac**
  - **Grade 1, Mild – Bulging of the dural sac and lack of epidural fat**
  - **Grade 2, Moderate – Bulging of the dural sac and lack of epidural fat at the level of the posterior wall of 2+ vertebral bodies, and presence of large radicular cysts**
  - **Grade 3, Severe – Presence of anterior sacral meningocele**

## Discussion

Parturients with Marfan syndrome are high risk due to aortic dilation and risk of rupture and/or dissection. Therefore, neuraxial anesthetic management during labor is essential for safety, especially with regard to hemodynamic perturbations. Labor epidurals are widely used, functionally to decrease pain while also reducing systemic blood pressure through peripheral vasodilation, improving the stress on the aorta. Additionally, these patients pose increased risk during epidural placement for unintentional dural puncture and scoliosis causing difficult needle placement. Though epidural anesthesia is beneficial, thorough preoperative evaluation, including MRI should be considered given the increased risks and challenges they pose.

As discussed by Yang et.al and Lacassie et.al, dural ectasias often result in unpredictable spinal anesthesia, require variable local anesthetic doses, and have high failure rates. Though this patient did not have a formal diagnosis with imaging demonstrating a dural ectasia, the high prevalence in Marfan syndrome and her unpredictable response to intrathecal anesthetics, makes this diagnosis likely. She was administered spinal anesthetic doses that reliably produce a measurable level of analgesia in patients with normal spinal anatomy. Over the course of her labor, she received intrathecal doses ranging from 3 ml of 0.0625% bupivacaine with 6mcg Fentanyl to 1.5 ml of 0.25% bupivacaine (1.875 mg to 3.75 mg) with modest analgesic effects. Following the final combined spinal epidural where the spinal portion had no measurable effect, she was given a total of 7ml of 0.25% bupivacaine epidurally in divided aliquots, which lead to analgesia with a dermatomal level up to T10 bilaterally, as measured with ice. Ultimately, the epidural functioned exceptionally well and the patient was able to deliver a healthy baby with minimal discomfort.

## References

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