Pragmatic approach to neuraxial anesthesia in obstetric patients with disorders of the vertebral column, spinal cord and neuromuscular system

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ABSTRACT

Neuraxial anesthesia provides optimal labor analgesia and cesarean delivery anesthesia. Obstetric patients with disorders of the vertebral column, spinal cord and neuromuscular system present unique challenges to the anesthesiologist. Potential concerns include mechanical interference, patient injury and the need for imaging. Unfortunately, the existing literature regarding neuraxial anesthesia in these patients is largely limited to case series and rare retrospective studies. The lack of practice guidance may lead to unwarranted fear of patient harm and subsequent avoidance of neuraxial anesthesia for cesarean delivery or neuraxial analgesia for labor, with additional risks of exposure to general anesthesia. In this narrative review, we use available evidence to recommend a framework when considering neuraxial anesthesia for an obstetrical patient with neuraxial pathology.

BACKGROUND

Disorders of the vertebral column, spinal cord and neuromuscular system, such as degenerative disease, scoliosis or simple closed spinal dysraphisms with a gap in a single vertebral body, are common, affecting more than 40% of women of childbearing age.[1–3] These and the rarer conditions, such as cysts, tumors and vascular lesions, are highly relevant to the anesthesiologist as neuraxial analgesia and anesthesia are the preferred methods for labor analgesia and obstetric surgical intervention. Questions often arise as to whether the neuraxial procedure could harm these patients and whether these techniques will be effective. Unfortunately, published data to guide practice are often limited in scope or quality, and clinical practices may be excessively cautionary. This review reacquaints the provider with the relevant anatomy and physiology, synthesizes and summarizes the available data (table 1), and provides guidance to facilitate the rational use of neuraxial anesthesia (figure 1).

Anatomy of the lumbar spine

The lumbar spine consists of five vertebrae (L1–L5) that are connected in sequence by intervertebral discs and facet joints formed by the inferior and superior articular processes of the adjacent vertebrae. The anterior and posterior vertebral elements connect by the pedicles bilaterally. The interlaminar space is bound by the inferior margin of the vertebrae above and the superior margin of the vertebrae below the interspace (figure 2). Normal pregnancy-induced changes include engorged epidural veins with displacement of the dura and cerebrospinal fluid (CSF), resulting in decreased lumbar CSF volume.[4] Administration of epidural medication further compresses the dural sac, transiently increasing the pressure in the epidural space and the intracranial pressure. The optimal neuraxial technique depends on the clinical context and the specific patient's anatomic considerations. For example, the epidural space may be scarred or obliterated, but the intrathecal space may still be accessible. In general, if a spinal anesthetic or lumbar puncture is contraindicated due a patient's anatomy or physiology, then an epidural procedure is also contraindicated since inadvertent dural puncture is always a risk.

Use of imaging in planning for neuraxial procedures

In addition to a careful history and physical examination, prior imaging studies may be used to assess the technical feasibility of a neuraxial procedure, identify favorable interspaces and avoid known lesions. Relevant imaging modalities include standard X-rays, MRI, CT scanning and ultrasound. MRI (without gadolinium) and ultrasound are generally preferred during pregnancy to avoid ionizing radiation exposure to the developing fetus.

Preprocedure neuraxial ultrasound may assist the anesthesiologist in identifying midline, specific interspaces and select pathologies. Anesthesiologists inaccurately identify specific lumbar interspaces when using surface landmarks, with estimates ranging from one space below to four spaces above the actual location.[5] While ultrasound is also imperfect, the margin of error in one study was at most one space above or below the intended target.[6] A recent meta-analysis confirmed a high correlation between ultrasound-measured depth to ligamentum flavum and actual needle insertion depth.[7] Randomized controlled trials in healthy obstetric patients also demonstrate fewer needle passes and improved analgesia with preprocedure ultrasound compared with surface landmarks alone.[6–9]

Expert consultation

The care of patients with complex neurological disease is enhanced by an interdisciplinary approach with neurologists, pain specialists and surgeons to help interpret the impact of or need for additional imaging and the nuances of the disease

Table 1 Summary of disorders of the vertebral column, spinal cord and neuromuscular system and consequences for neuraxial anesthesia

Pathology	Implications for neuraxial anesthesia	Evidence	References
Disorders of the vertebral column			
Degenerative spine disease	► Back pain is common; typically safe to proceed with neuraxial anesthesia. ► Severe symptoms or new neurological deficits merits immediate preprocedure neurological workup. ► If disc causes central canal stenosis, seek alternative interspace. ► Patient positioning during labor and delivery is key.	Case reports	Chow *et al*, 2008[11] Simon *et al*, 1989[10] Vercauteren *et al*, 2011[2]
Scoliosis	► May be more technically challenging but frequently successful. ► Consider neuraxial ultrasound to elucidate curve and identify optimal interspace.	Case reports and series, retrospective cohort study	Bowens *et al*, 2013[27] Butwick and Carvalho, 2007[17] Carlson *et al*, 1978[12] Chan *et al*, 2017[24] Ko and Leffert, 2009[1] Li *et al*, 2019[29] McLeod *et al*, 2005[25] Moran and Johnson, 1990[13]
Prior spine surgery	► Simple discectomies or microdiscectomies rarely compromise epidural space. ► Spinal fusion with or without Harrington Rods distorts or obliterates those interlaminar spaces. – Successful spinal and epidural anesthetics can be achieved but may be more technically challenging.	Case reports and series, prospective matched case–control studies	Bauchat *et al*, 2015[40] Crosby and Halpern, 1989[30] Daley *et al*, 1990[32] Hebl *et al*, 2010[31] Hwang *et al*, 2009[140] Lee *et al*, 1995 Kardash *et al*, 1993[35] Majeed *et al*, 2017[36]Okutomi *et al*, 2006[38] Perlas, 2010[33] Silva and Popat, 1994[37]
Spinal dysraphisms	► 'Closed' lesions with no signs of lumbosacral neurologic deficits are typically safe for neuraxial anesthesia. ► 'Open' lesions or evidence of lumbosacral neurological deficits requires additional investigation.	Case reports and series	Ahmad *et al*, 2006[50] Kuczkowski and Zuniga, 2007[52] McGregor *et al*, 2013[54] Murphy *et al*, 2015[3] Palmer and Baysinger, 2019[51] Vaagenes and Fjaerestad, 1981[48]
Spinal cord stimulator	► Typically safe for neuraxial anesthesia as long as no mechanical interference with the device.	Case reports	Patel *et al*, 2014[63] Saxena and Eljamel, 2009[61] Segal, 1999[60] Sommerfield *et al*, 2010[62]
Dorsal root ganglion stimulator	► Thought to be safe for neuraxial anesthesia as long as no mechanical interference with the device.	No available literature	
Disorders of the spinal cord and nerve roots			
Multiple sclerosis	► Prior concerns for exacerbation of multiple sclerosis with spinal anesthesia have been unsubstantiated. ► Increased risk of postpartum relapse irrespective of anesthetic use or type ► Candidate for neuraxial anesthesia.	Case reports and series, retrospective cohort studies, prospective cohort studies, systematic review	Bader *et al*, 1988[88] Confavreaux *et al*, 1998[85] Lavie *et al*, 2019[90] Lee *et al*, 2009[99] Leigh *et al*, 1990[98] Pastò *et el*, 2012[97] Vukusic *et al*, 2004[100] Wang and Sinatra, 1999[94]
Spinal cord injury	► Early neuraxial anesthetic recommended to prevent autonomic dysreflexia for lesions at T6 or above. ► Concern for further injury with neuraxial anesthesia unsubstantiated.	Case reports and series	Agostoni *et al*, 2000[79] Burns and Clark, 2004[70] Crosby *et al*, 1992[69] Jadhav and Brooks, 2004[76] Owen et al., 1994[73]
Neurofibromatosis	► Neurofibromas are associated with neural sheath tumors originating at the nerve route. – Spine tumors are morecommon in NF-2 than NF-1 but can occur in either. ► Pregnancy can accelerate tumor growth. ► Neurology consultation and review of radiology studies can aid in decisions regarding neuraxial anesthesia.	Case reports	Dounas *et al*, 1995[107] Esler *et al*, 2001[104] Galvan nd Hofkamp, 2018[108] Spiegel *et al*, 2005[109]
Spinal cord vascular malformations	► Occur rarely in women of childbearing age. ► AVF - superficial on spinal cord. ► AVM - intermedullary. ► Rarely at the lumbar level. ► Of greatest concern are large lesions or those with vascular congestion. ► Otherwise, may likely proceed with neuraxial anesthesia after neurology consultation.	Case reports	Abut *et al*, 2006[117] Eldridge *et al*, 1998[111] El Shobary *et al*, 2009[118] Gemma *et al*, 1994[115] Warner *et al*, 1987[113]
Tarlov cysts	► Lesions are typically sacral. ► If not symptomatic, likely not to interfere with neuraxial anesthetic. ► Theoretical cause of 'failed spinal', not substantiated.	Case reports	Fettes *et al*, 2009[57] Hoppe and Popham, 2007[58] Pfund *et al*, 2018[59]
Neuromuscular and peripheral nerve disorders			
Myasthenia gravis	► Early neuraxial anesthetic recommended to prevent myasthenic crisis and exposure to general anesthesia	Case reports and series	Almeida *et al*, 2010[138] Jaleel et al., 2018[139]
Amyotrophic lateral sclerosis	► No high or medium quality evidence for contraindication to neuraxial anesthesia. ► Baseline respiratory insufficiency requires increased vigilance.	Case reports	Jack and Sanderson, 1998[124] Kock-Cordeiro *et al*, 2018[123]

Continued

Table 1 Continued

Disorders of the vertebral column			
Pathology	Implications for neuraxial anesthesia	Evidence	References
Guillan-Barre syndrome	► Risk of worsening symptoms/signs postpartum regardless of anesthetic technique. ► No contraindication to epidural or spinal anesthetics.	Case reports	Alici *et al*, 2005[126] Kocabas *et al*, 2007[127] Mangar *et al*, 2013[136] McGrady, 1987[132] Vassiliev *et al*, 2001[129]

AVF, arteriovenous fistula; AVM, arteriovenous malformation.

in the context of pregnancy and planned neuraxial anesthesia. Collecting the key patient data in advance of hospital admission, minimizing the use of specialty-specific jargon, and formulating defined clinical consult questions will greatly facilitate the conversation. For example, for a patient with a structural lesion: is there an obstruction to the needle/catheter entry, path or target? If so, is it safe and feasible to navigate around it?

DISORDERS OF THE VERTEBRAL COLUMN AND SPINAL CORD

Degenerative spine disease

Common degenerative lumbar spine disorders in women of childbearing age include intervertebral disc herniation, facet arthropathy, spondylolisthesis, pars interarticular stress fracture and intervertebral disc degeneration. Low back pain with or without pelvic pain is common in obstetric patients, typically increasing as the pregnancy progresses. If the pain is severe or associated with new lower extremity, bowel or bladder neurological deficits, then immediate neurological consultation and work-up is indicated prior to a neuraxial procedure.

In general, a lumbar degenerative spine disorder without central canal stenosis should not impact the feasibility or safety of neuraxial anesthesia, although high-quality randomized controlled trials are lacking. Conversely, disc herniation causing central canal stenosis may result in markedly decreased cerebrospinal fluid (CSF) around the cauda equina and impair access to the corresponding intrathecal space (figure 3A). Needle placement at an alternative interspace with ample surrounding CSF is preferable (figure 3A). Despite case reports of patients developing cauda equina syndrome from lumbar disc herniation after vaginal and cesarean delivery with epidural anesthesia,[10 11] changes in body mechanics during pregnancy and delivery are more likely than the neuraxial procedure to be the cause.[2] It is prudent to remind patients with epidural analgesia to pay extra attention to positioning during labor as they may be less aware of body positions that exacerbate their symptoms.

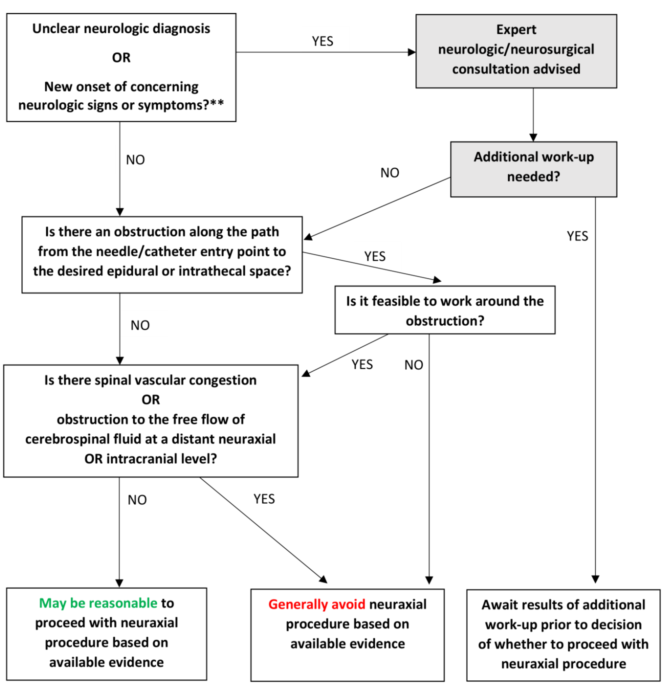

Figure 1 Decision aid for evaluating the safety of neuraxial procedures in obstetric patients with disorders of the vertebral column, spinal cord or neuromuscular system.*,+
*Assumes no other contraindication to neuraxial anesthesia. +The decision of whether to proceed with a neuraxial procedure occurs in a clinical context. This decision aid does not define a legal standard of care or replace the care or judgment of the responsible medical professional. **Concerning neurological symptoms include, but are not limited to, severe headache, new-onset extremity weakness or numbness, or bowel/bladder changes beyond mild incontinence commonly observed in pregnancy.

Scoliosis

Scoliosis is present in roughly 2% of the population, primarily in women.[1] The defect severity is estimated by the rate of progression and the greatest angle measured from the top of the involved superior vertebra to the bottom of the inferior vertebra (Cobb angle). Scoliosis may be treated with conservative measures (eg, observation, bracing) or surgical intervention.

Individual case reports and series historically document multiple procedure attempts to achieve effective neuraxial anesthesia[12–15] and technical complications (eg, inadequate analgesia, intravascular placement and accidental dural puncture) in parturients with scoliosis.[16–23] A narrative review of published case reports found similar success rates of epidural and spinal anesthetics (80% and 73%, respectively) in uncorrected scoliosis patients.[1] More recently, a retrospective cohort study of parturients with adolescent idiopathic scoliosis reported successful

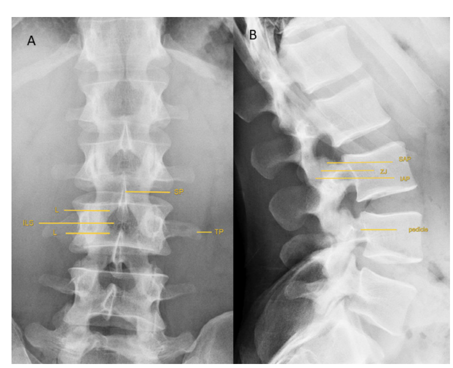

Figure 2 Plain X-ray of lumbar spine. (A) Anterior–posterior view; (B) lateral view. IAP, inferior articular process; ILS, interlaminar space; L, lamina; SAP, superior articular process; SP, spinous process; TP, transverse process; ZJ, zygapophyseal joint.

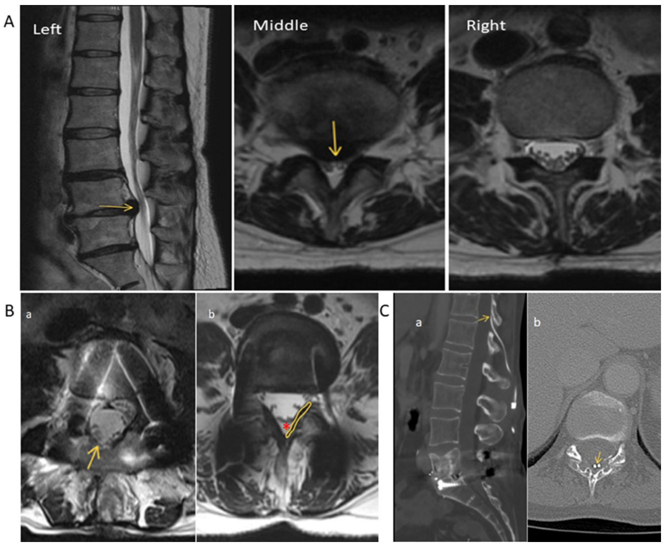

Figure 3 Images of spinal pathology relevant to neuraxial anesthesia. (A) MRI of lumbar intervertebral disc herniation. Left: a T2-weighted MR sagittal image depicting an intervertebral disc herniation at the L4/5 level (yellow arrow) indenting the dural SAC, causing severe central canal stenosis. Middle: a T2-weighted axial view image at L4/5 interlaminar level depicting the same L4/5 intervertebral disc herniation (yellow arrow) causing severe central canal stenosis. Right: a T2-weighted axial view image at L3/4 interlaminar level. (B) MR images of two patients with posterior instrumented spinal fusion through level of hardware. (a) Ligament flavum absent; (b*)* Intact ligament flavum (left side ligament outlined) and epidural fat (asterisk). (C) CT images of a patient with spinal cord stimulator. (a) Sagittal view SCS lead entering the dorsal epidural space at T12/L1 level (yellow arrow). (b) Axial view dual SCS leads at T12 level (yellow arrow). SCS, spinal cord stimulator.

spinal anesthesia in 99% of the cases, including women with prior spinal fusion.[24]

Preprocedure ultrasound can facilitate identification of interspaces that are least affected by rotation and curvature and provide a trajectory for the spinal or epidural needle.[25–28] In the transverse view, the ultrasound probe can be angled slightly cephalad to mimic the usual needle entry position and then repositioned to place the spinous process in midline. The probe is then further angled (by rocking or rotating) until the articular processes are roughly symmetric in a horizontal plane. If the probe is now visually to the left or right of midline, or asymmetric on the back, then the spine is likely curved and/or rotated at that interspace. The needle trajectory should be adjusted accordingly. A spinal procedure with a small gauge needle and no catheter may be technically easier to perform than an epidural procedure. Dual epidural catheters have been used to improve spread of epidural medication in patients with severe scoliosis and unilateral block, although there is a theoretical risk of damaging the in situ catheter by the second epidural needle with this technique.[1 29]

Prior spine surgery

Prior spine surgery can dramatically change the neuraxial anatomy, obscure palpable surface landmarks and alter the 'loss of resistance' on entering the epidural space.[30] Fibrosis in the epidural space may make threading the epidural catheter difficult[31] and limit the spread of local anesthetic leading to patchy or unilateral epidural blocks.[32] Review of the operative notes and MR images can help determine if there is likely to be an intact ligamentum flavum.[33] Choosing an interspace distinct from the surgical site, if feasible, is advised to ensure successful catheter insertion and anesthetic spread.[34]

In general, simple discectomies or microdiscectomies do not compromise the ligamentum flavum or epidural space. Laminectomies may involve partial or full thickness removal of the ligament flavum which may compromise the loss of resistance technique. Figure 3B shows the MR images of two patients with history of posterior instrumented spinal fusions: the first has a surgically removed ligamentum flavum whereas the second has an intact ligamentum flavum. Performing a 'blind' epidural procedure in the second patient may be more successful than in the first as the anesthesia provider can still rely on the 'loss of resistance' when the needle is advanced through the ligamentum flavum.

Harrington rod instrumentation is a common scoliosis correction surgery that entails posterior spinal fusion of the curve and one spinal segment above and below, resulting in distortion or obliteration of the involved interlaminar spaces.[35] Numerous case reports and series describe successful spinal[28 35 36] combined spinal-epidural (CSE)[26 37] and intrathecal catheter[38 39] anesthetics in these patients although multiple attempts are often reported.[30 32] A recent prospective, case–control study in parturients with surgically corrected scoliosis found an 88% success rate of CSE or epidural anesthesia.[40] However, the mean time required for procedure completion was 41% longer (6.5 vs 4.6 min) in prior surgery group, with a greater number of needle redirections, attempted interspaces and need to switch to a more experienced provider. The 12% failure rate was significantly lower than a previous estimate of 50% in this patient population.[2]

Anterior scoliosis corrective surgery or vertebral body tethering is a new minimally invasive thoracoscopic technique wherein screws are placed across the vertebrae at each level under tension by a flexible tether to modulate growth and straighten the spine. There are no reports to guide subsequent neuraxial procedures in these patients.

Spinal dysraphisms

Spinal dysraphisms include the broad spectrum of congenital defects with incomplete or abnormal neural tube closure, most often in the sacral spine. They are characterized as being 'open' or 'closed' based on whether overlying skin is present.[41] Spinal dysraphisms can be particularly vexing for anesthesiologists due to complex and varied terminology.[3] Whereas 'spina bifida occulta' rarely has major implications for neuraxial anesthesia, 'occult spinal dysraphism' may introduce additional risk.[42]

Spina bifida occulta, the simplest closed spinal dysraphism, is present in 10%–20% of the population.[43 44] These patients have a gap in one or more vertebrae with normal overlying skin and underlying neural tissues. They are typically asymptomatic, and these lesions are often found incidentally on imaging for other indications. Other closed spinal dysraphisms include cutaneous masses, tethered spinal cord and low-lying filum terminale. Open spinal dysraphisms include meningocele, where the meninges surrounding the spinal cord protrude from the defect, and myelomeningocele, where both the meninges and spinal cord are contained within the protruding sac with associated neurologic deficits. More than 90% of patients with meningomyelocele also have a severe form of Chiari malformation, with cerebellar tonsillar herniation through the foramen magnum and potentially non-communicating hydrocephalus due to obstruction of CSF flow. Spina bifida cystica is a less precise term applied to open or closed lesions with associated neurological abnormalities.

Risk stratification of these complex lesions is facilitated by a few basic considerations. For patients with a simple closed spinal dysraphism (eg, spina bifida occulta) without lumbosacral deficits or other indications of cord abnormalities (see below), it is typically reasonable to proceed with the neuraxial procedure.[3] We do not routinely recommend imaging of these patients. Some reports recommend avoiding the level of the bony defect to minimize the inadvertent dural puncture risk due to ligamentum flavum abnormalities, but this remains a theoretical concern.[45–47] Sacral analgesia may be compromised if the neuraxial procedure is done at high lumbar or low thoracic interspaces to avoid the lesion.[48]

Indications of spinal dysraphism associated with spinal cord abnormalities often include cutaneous, urinary, and orthopedic anomalies (eg, hypertrichosis, large dimple or cyst midline in low lumbar region; incontinence or atonic bladder).[49] Of particular concern is spinal cord tethering, wherein the most distal part of the spinal cord, the conus medullaris, is abnormally attached ('tethered') to the bony spine, ligaments and/or other tissues. Spinal cord tethering can be seen radiologically by a low-lying conus medullaris attached to a thickened (more than 2 mm) filum terminale extension of the conus. Tethered cord syndrome includes pain, bowel/bladder dysfunction and lower extremity symptoms that are exacerbated with flexion and can result in permanent neurological deficits. These anomalies may require one or more cord 'release' procedure(s), which may not be successful.[49] A low-lying or tethered spinal cord is likely more susceptible to needle injury during a spinal procedure or an unintentional dural puncture during an epidural procedure and is generally considered to be an impediment to neuraxial procedures.[50 51] In these cases, an MRI can illustrate the anatomy and help guide decision making (figure 1).[3]

Despite the concern that obstetric patients with spina bifida may experience a greater frequency of block failure or complication,[47 52 53] a review of 84 patients including 41 with complex spinal dysraphism documents success in 80% of cases: 15 patients underwent spinal or CSE procedures and 52 patients underwent epidural procedures without serious complications. The most common issues were suboptimal analgesia or block height.[3] One report describes a complex case of a woman with partial sacral agenesis, urinary retention and colostomy who had a successful CSE anesthetic at the L4/5 interspace for cesarean delivery.[54] A prior CT myelogram showed an anterior meningocele with the tip of the conus medullaris at L2, without tethering.

Tarlov cysts

Tarlov perineural cysts are CSF filled sacs arising at the junction of the posterior nerve roots and the dorsal root ganglion (DRG). Present in 5%–10% of the population, they are often small, sacral and asymptomatic.[55] Larger or ruptured cysts may cause radicular symptoms including hip, leg or foot pain, perineal pain, paresthesia, or bowel or bladder dysfunction.[56] The impact of a Tarlov cyst on candidacy for neuraxial anesthesia is likely negligible. Injection of spinal medication directly into a Tarlov cyst has been postulated to cause failed spinal anesthesia,[57] although MRIs in suspected cases have not confirmed this hypothesis.[58] One recent report describes successful spinal anesthesia in a parturient with a symptomatic cyst, despite her repeatedly needing CT-guided drainage antepartum.[59] In a symptomatic patient, medication in the epidural space could potentially increase nerve compression.

Spinal cord stimulators

Spinal cord stimulators (SCS) are increasingly used to manage refractory chronic neck, back and limb pain conditions including complex regional pain syndrome, postlaminectomy syndrome and painful peripheral neuropathy. SCS components include electrical leads, an optional extension and a pulse generator. For neck and upper extremity pain, SCS leads are placed in the cervical posterior epidural space up to the C2 level depending on the area affected. For low back and lower extremity pain, SCS leads are placed at mid-thoracic levels accessed through the mid to high lumbar levels. The pulse generator is typically implanted in the upper buttock area subcutaneously. The leads are then connected with a pulse generator through a subcutaneous tunnel. Cervical leads may require an extension to reach the generator.

While the safety and effects of SCS during pregnancy have not been systematically studied, case reports document successful SCS therapy throughout pregnancy.[60 61] It is imperative to review the surgical documentation and previous imaging studies to ascertain the location and course of the SCS leads and connecting wires. Spinal anesthesia can be feasible if there is no hardware or other obstruction to reaching the dura and intrathecal space.[62 63] Although there is no literature on use of continuous epidural infusion in parturients with indwelling SCS, epidural labor analgesia should also be feasible. Accessing the epidural space at a level well below where the SCS leads enter is advantageous to avoid the needle and catheter contacting the leads, keeping in mind that the average height of the human lumbar vertebrae and intervertebral disc complex is 3–4 cm.[64] Figure 3C illustrates the SCS leads entering the dorsal epidural space at T12/L1 level. Access to the epidural or intrathecal space should be attained below this level, ideally with the help of preprocedure ultrasound. In addition to direct damage to the SCS hardware, potential risks of neuraxial anesthesia in patients with indwelling SCS include migration of SCS lead(s), contamination and infection, and possible alteration in electrical behavior of the SCS with electro-conductive epidural medication.

DRG stimulators

In the last decade, devices that stimulate the DRG have shown similar and sometimes superior efficacy to traditional SCS for treating chronic pain.[65 66] The DRG stimulator components are similar to SCS: electrical leads, an optional extension, and a pulse generator. The electrical leads are threaded into the epidural space and then into the intervertebral foramen, in which the DRG lies. To treat back and lower limb pain, the leads are typically threaded to one or more lumbar DRGs. There is no current literature describing neuraxial anesthesia in patients with an indwelling DRG stimulator. However, management is similar to management of neuraxial anesthesia with an SCS: providers should avoid accessing the neuraxial space at the level of DRG leads and follow strict aseptic techniques. Since the leads are near the lumbar DRG, it is advisable to turn off the DRG stimulator if a lumbar epidural infusion is used.

Chronic pain patients, with and without indwelling stimulators, represent an extremely complicated subset of parturients and predelivery consultation with the patient and her pain physician would likely be advantageous. Coexisting diseases to consider include chronic opioid use, buprenorphine management, pain catastrophizing and hyperalgesia.

SPINAL CORD AND NERVE ROOT DISORDERS

Spinal cord injury

Advances in rehabilitation and reproductive technology have led to an increasing number of pregnant women with spinal cord injury (SCI). Injuries are defined by permanent or temporary change in neurologic function, the level at which injury occurs, and whether the SCI is complete or incomplete. Physiological perception of labor and delivery depends primarily on the injury level: above T10 eliminates the sensation of uterine contractions; above T11/T12 eliminates the sensation of cervical dilation; and above the lumbosacral plexus eliminates sensations in the perineum and vagina.[67]

Autonomic dysreflexia is a dangerous complication of SCI occurring in over 85% of patients with SCI at T6 or above, and characterized by uninhibited sympathetic response to noxious stimuli below the level of the SCI. The absence of perceived pain does not prevent autonomic dysreflexia.[68] Stimulation at lower spinal segments tends to lead to more autonomic disturbance. Triggering stimuli during labor and delivery include bladder distension, vaginal exams, episiotomy and laceration repairs.[69] Responses range from asymptomatic hypertension to hypertensive crisis resulting in encephalopathy, hemorrhagic stroke or cardiac arrest.[68] While most peripartum episodes occur in patients with a prior history of autonomic dysreflexia, it may occur for the first time in a parturient. Fundal massage, intermittent uterine contractions, and bowel or bladder distension have been reported to trigger autonomic dysreflexia up to 5 days post partum.[70–72]

Neuraxial anesthesia should not cause further cord injury in SCI patients with stable neurological function.[68] While randomized controlled trials are lacking, several case series suggest that epidural[73–76] or CSE[77–79] anesthetics are effective and essential to prevent autonomic dysreflexia in at-risk SCI patients. As thromboprophylaxis or systemic anticoagulation may be used, guidelines governing the timing of neuraxial procedures should be considered.[80 81] It can be difficult to precisely define a satisfactory block in an insensate parturient. In some cases, the resulting block 'level' may be identified where previously spastic limbs become flaccid.[68]

For SCI above T6 with sympathetic dysfunction, spinal anesthesia could lead to severe hypotension and shock from the inability to vasoconstrict the splanchnic bed (innervated by T5–L2) or increase heart rate via cardioaccelerator fibers (innervated by T1–T4). A small series of 6 SCI patients receiving spinal anesthetics demonstrated cardiovascular stability secondary to baseline low sympathetic tone.[82] Epidural anesthetics are generally well tolerated even in patients with orthostatic hypotension, with appropriate intravascular volume expansion prior to the procedure.

Multiple sclerosis

Multiple sclerosis (MS) is the most common among the chronic demyelinating diseases that include neuromyelitis optica (Devic's disease) and idiopathic inflammatory demyelinating disease. The course of MS may be either relapsing-remitting (80%–90%) or progressive (10%–20%). Central nervous system lesions result in neurologic sequelae ranging from mild lower extremity weakness and/or sensory changes to respiratory failure with diaphragmatic paralysis.[83 84] Stimuli including heat, infection and emotional stress may trigger disease relapse or progression. MS disproportionately affects women of childbearing age and is highly relevant to obstetric practice.[85] Most relapses occur postpartum, as the disease is often quiescent during the antepartum period. As such, the anesthetic may be falsely implicated in the relapse.[85]

There are concerns about the safety of neuraxial anesthesia for MS patients that some authors believe are unsubstantiated. Isolated case reports from the mid-1900s raised concern for the development of permanent neurological deficits resulting from spinal anesthesia in MS patients.[86 87] In the frequently cited 1988 retrospective cohort study by Bader *et al*, 18 parturients with MS who received epidural anesthesia had no significant difference in relapse incidence compared with those who had received local techniques (eg, pudendal block). No patients received spinal anesthetics. The authors highlighted that three parturients who relapsed received higher concentrations of epidural local anesthetic (bupivacaine, 2% lidocaine) and speculated that 'theoretically, epidural anesthesia may be less of a risk than spinal anesthesia because the concentration of local anesthetic in the CSF, and thus, the spinal cord is higher following subarachnoid block.[88] Notably, some patients with relapse also received concentrated epidural local anesthetic (chloroprocaine 3%). Subsequent case reports and retrospective cohort studies suggest no difference in relapse with spinal, epidural or CSE anesthetics in parturients with MS.[89–99] Two prospective cohort studies and a systematic review also found no difference in relapse rate or disease progression in parturients with MS with and without neuraxial analgesia.[85 100 101] Nonetheless, the American Society of Regional Anesthesia and Pain Medicine concluded that definitive evidence supporting the use of neuraxial anesthesia in MS patients with stable neurological symptoms is lacking and the decision to perform neuraxial blockade should include a careful discussion with the patient about risks and benefits (class II).[102] Curiously, the associated text repeats the unsupported recommendation that local anesthetic doses and concentrations be limited and that epidural anesthesia is considered safer than spinal anesthesia.

In summary, the best available evidence suggests that neuraxial anesthesia (spinal, epidural, or a combined technique) is a reasonable option for women with MS without other contraindications, and there is no evidence that the disease impacts the success of the neuraxial anesthetic.[91 92 94–96 98 99 103]

Neurofibromatosis

The neurofibromatoses, NF1, NF2 and Schwannomatosis, are inherited disorders of benign peripheral nerve sheath tumor proliferation. Some investigators have reported accelerated tumor growth during pregnancy.[104 105] Patients with NF1 often have scoliosis and a variable burden of neurofibromas throughout the body, including along the spinal nerve roots in up to 35% of patients.[106] Most of these neurofibromas are small, slow growing and arise laterally in the intervertebral foramen. As such, these patients do not routinely undergo surveillance MRI of the spine. Rarely, large tumors can extend towards midline and cause pain or lower extremity symptoms and potentially obstruct access to the epidural space during neuraxial procedures. NF2, a rarer disease than NF1, features vestibular schwannomas, meningiomas and spinal tumors (including ependymomas, schwannomas and meningiomas) in as many as 90% of affected patients. These patients typically undergo routine gadolinium-enhanced brain and spine MRIs every 1–2 years for surveillance. Schwannomatosis is clinically very similar to NF2 but is genetically distinct.

The safety of a neuraxial anesthesia in NF patients depends on their burden of disease as revealed by imaging. In general, NF1 patients with a low disease burden, normal neurological examinations, and no lumbar or adjacent spine lesions on a previous MRI are likely to be reasonable candidates for neuraxial procedures. They may not require additional imaging. Needle entry should avoid cutaneous lesions to limit bleeding. In contrast, NF patients with large nerve root tumors of the lumbar or adjacent spine with or without intracranial tumors with mass effect (eg, vestibular schwannomas or meningiomas in NF2) are typically not candidates for neuraxial procedures. The former is problematic due to mechanical obstruction, whereas the latter poses a risk of cerebellar tonsillar herniation from a dural puncture. Of note, MRIs performed during pregnancy are typically without gadolinium and are likely to be suboptimal, particularly in detecting small spine lesions. All NF patients with new or worsening neurologic symptoms (eg, persistent headache, hearing loss, extremity numbness/weakness) warrant a formal neurologic evaluation prior to undertaking a neuraxial procedure (figure 1).

Several case reports document successful epidural procedures in NF1 and NF2 patients.[104 107–109] Most patients had prior brain and lumbosacral spine MRIs. One case reported an epidural hematoma with cauda equina syndrome in a patient with NF1 after inadvertent dural puncture. However, follow-up urgent MRI revealed no spine neurofibromas and a small, inconsequential hematoma, highlighting that NF was unrelated to the outcome.[104]

Spinal cord vascular malformations

Spinal cord arteriovenous malformations (AVMs) are rare in pregnant women. When the transition from the artery and the vein is direct, these malformations are called AV fistulas (AVFs).[110] AVFs typically occur superficially on the spinal cord. In 'nidus' AVMs, which originate in the spinal cord parenchyma, the artery and veins are connected through a network of abnormal vasculature. Indolent symptoms include the slow onset of back pain with gait disturbance, lower extremity spasticity, sensory changes and bladder or bowel symptoms. Neurological deterioration stems from venous congestion, hemorrhage, redistribution of blood supply or rarely mass effect.

Spontaneous spinal AVM rupture causing paraplegia has been reported during pregnancy.[111 112] The physiological changes of pregnancy including compression of venous outflow by the gravid uterus, labor and Valsalva during delivery can lead to venous congestion within the AVM. This can then cause spinal cord edema, ischemia and potentially even AVM rupture. There are also case reports of undiagnosed spinal AVMs in predominantly non-obstetric patients who presented with catastrophic neurologic outcomes after neuraxial procedures.[113–117]

There are few data to guide screening and neuraxial anesthetic management of parturients with known or suspected spinal AVMs.[118–120] Most publications discuss hereditary hemorrhagic telangiectasia (HHT), a rare autosomal dominant disease characterized by AVMs in the lungs, gastrointestinal tract and more rarely, in the brain or spine (<1%). If present, spine lesions are seldom at the lumbar level. In the International Guidelines for the Diagnosis and Management of HHT, there was insufficient evidence to recommend general screening for spinal AVMs,[120] but if screening is performed, then angiography (or a sagittal T2 MRI scan of the spine) is suggested. Successful cases of spinal, epidural and CSE anesthesia have been reported in parturients with HHT.[111 118 121] Most reported cases document negative spine imaging as a prerequisite for proceeding with neuraxial anesthesia. In general, the anatomic location of AVMs makes direct trauma with the needle during a neuraxial procedure unlikely. However, a large spinal AVM at a distant site could theoretically be compromised by marked changes the A-V transmural pressures (eg, due to intracranial hypotension from a large volume CSF leak) or decreases in spinal cord perfusion (eg, maternal systemic hypotension or increasing lumbar CSF pressure by loading the epidural space), increasing the risk of the neuraxial procedure (figure 1).

SELECT NEUROMUSCULAR DISORDERS

Although neuromuscular disorders are rare in pregnancy, familiarity with some general principles can facilitate anesthesia care. Several of these disorders predispose patients to respiratory insufficiency, aspiration, and sensitivity to neuromuscular blocking agents.[122] Neuraxial anesthesia, particularly titratable techniques such as epidural or CSE anesthesia paired with non-invasive respiratory adjuncts (eg, bilevel positive airway pressure, BiPAP) should be considered as viable or preferred options.[122–132]

Amyotrophic lateral sclerosis (ALS) is a rare disorder of upper and lower motor neurons of the spinal cord leading to progressive disability and death. Patients exhibit hyperreflexia, spasticity, and weakness due to dorsal motor neuron degeneration and muscle denervation. Reports of pregnant women with ALS are limited and therefore innovative solutions are warranted. One parturient with ALS manifesting with bulbar symptoms, lower extremity weakness, and progressive respiratory insufficiency acutely decompensated in the setting of a viral infection and required BiPAP. There was hesitation to intubate her for her cesarean delivery out of concern that she would fail to extubate. Instead, she was successfully managed with a CSE and BiPAP.[123] Another report documents a parturient with severe, progressive ALS who tolerated epidural anesthesia for cesarean delivery despite her baseline respiratory insufficiency.[124]

Guillain-Barre syndrome (GBS) is the most common acute peripheral neuropathy.[125] Whereas these patients have an increased risk of autonomic dysfunction and sensitivity to muscle relaxants including hyperkalemia with succinylcholine during general anesthesia, several case reports support the safety and efficacy of epidural, spinal, and CSE anesthesia.[126–132] The postpartum onset of GBS has been reported in patients after neuraxial anesthetics,[133 134] although these cases are confounded by infections which can trigger GBS. As with MS, some authors suggest reducing the concentration or dose of local anesthetic to minimize additional damage to affected nerves although there is no high-quality evidence to support this recommendation.[135] Mangar *et al* reviewed eight published cases and their primarily non-obstetric patients who developed the acute onset of GBS after neuraxial anesthesia, finding 'no direct link between neuraxial anesthesia and GBS can be confirmed.[136]

Myasthenia gravis (MG), a chronic autoimmune disease of antibodies targeting postsynaptic nicotinic acetylcholine receptors at the neuromuscular junction, is twice as common in women and has peak incidence during reproductive age. Intermittent exacerbations, often with acute respiratory failure, are common. In pregnancy, 30%–40% patients have symptomatic improvement, 30%–40% have unchanged disease and 20%–30% have worsened symptoms.[122] General anesthesia is complicated by potentiation of neuromuscular blockade and relative contraindication of acetylcholinesterase inhibitors. In contrast, neuraxial labor analgesia can help to mitigate labor-induced stress and fatigue, myasthenic crisis and acute respiratory failure.[137] The largest case series describes 17 MG patients, primarily with favorable outcomes: six stable pregnant patients and three patients with exacerbations or new antepartum symptoms received epidural anesthesia without complication. The last received a spinal anesthetic associated with severe dyspnea requiring mechanical ventilation.[138] A separate report describes an asymptomatic patient who successfully received a spinal anesthetic for cesarean delivery.[139]

CONCLUSIONS

In summary, despite a paucity of high-quality evidence, anesthesiologists and their parturients with neurologic disease of the vertebral column, spine and neuromuscular system must weigh the risks and benefits of neuraxial analgesia and anesthesia. Using a pragmatic approach that highlights whether there is a physical obstruction to the needle or catheter trajectory, or a distant spinal or intracranial lesion that could contribute to morbidity, allows shared decision-making around the available anesthetic options (figure 1). If neuraxial anesthesia or analgesia are deemed to have an excessive risk or if the patient wants to avoid neuraxial intervention, other forms of labor pain management (eg, intravenous analgesia) or general anesthesia for cesarean delivery, if needed, can be viable alternatives for labor and delivery.

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