Review article

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REVIEW ARTICLE

Spinal dysraphisms in the parturient– implications for perioperative anaesthetic care and labour analgesia

C. J. Murphy,[a] E. Stanley,[b] E. Kavanagh,[b,d] P. E. Lenane,[b,c] C. L. McCaul[a,b,d]

[a]*The Rotunda Hospital, Parnell Square, Dublin, Ireland*

[b]*Mater Misericordiae University Hospital, Dublin, Ireland*

[c]*Children's University Hospital, Temple Street, Dublin, Ireland*

[d]*School of Medicine and Medical Sciences, University College Dublin, Belfield, Dublin, Ireland*

Short title: Spinal dysraphisms

Correspondence to: Dr Conan McCaul, Consultant Anaesthetist, The Rotunda Hospital, Parnell Square, Dublin 1, Ireland.

E-mail address: cmccaul@rotunda.ie

ABSTRACT

Anaesthetists may encounter parturients with a spectrum of anatomical and functional abnormalities secondary to spinal dysraphisms which are among the most common neurodevelopmental anomalies. These range from surgically corrected open dysraphisms to previously undiagnosed closed dysraphisms. Both bony and neural structures may be abnormal. In true bony spina bifida, which occurs in up to 50% of the population, failure of fusion of the vertebral arch is seen and neural structures are normal. Ninety percent of such cases are confined to the sacrum. In open dysraphisms, sensory preservation is variable and may be present even in those with grossly impaired motor function. Both epidural and spinal blockade have been described for labour analgesia and operative anaesthesia in selected cases but higher failure and complication rates are reported. Clinical assessment should be performed on an outpatient basis to assess neurological function, evaluate central nervous system shunts and determine latex allergy status. Magnetic resonance imagining is recommended to clarify anatomical abnormalities and to identify levels at which neuraxial techniques can be performed. Of particular concern when performing neuraxial blockade is the possibility of a low-lying spinal cord or conus medullaris and spinal cord tethering. Previous corrective de-tethering surgery frequently does not result in ascent of the conus and re-tethering may be asymptomatic. Ultrasound is not sufficiently validated at the point of care to reliably detect low-lying cords. Epidurals should be performed at anatomically normal levels but spread of local anaesthetic may be impaired by previous surgery.

Introduction

Spinal dysraphism refers to an extremely heterogeneous group of disorders of the vertebral arches, spinal cord and meningeal layers which have multiple implications for the provision of peripartum anaesthetic care.[1] It encompasses a range of conditions that have been described as spina bifida aperta, cystica, manifesta and occult spinal dysraphisms. Analysis of reports in the anaesthetic literature show that neuraxial blocks are possible in select cases but challenging with a relatively high incidence of failure and complications for both epidural and spinal techniques. This review aims to identify issues relevant to labour ward analgesia and operative anaesthesia.

Classification

Interpretation of the existing literature is rendered difficult by inconsistent definitions and variable use of terminology, which have caused confusion since the first descriptions of spina bifida were published.[2-4] Unfortunately, there is to date no universally agreed classification of spina bifida and its variants. The recently proposed classification by Tortoni-Donati uses a combination of clinical and radiological assessment(Table 1).[5] Clinical assessment determines whether a mass is present and whether the overlying skin is intact. Accordingly, lesions are classified as open or closed spinal dysraphism, with or without a mass. Masses are either simple or complex. Radiological investigations determine the nature of the lesion and associated anatomical abnormalities. This classification supersedes previous systems, which used the terms spina bifida aperta, cystica and occulta. The term spina bifida occulta is a particular source of confusion as it has been used to describe a spectrum of conditions, which range from isolated bony abnormalities identified on x-ray, to cases in which a spinal dysraphism is present but has gone undiagnosed. True spina bifida occulta affects the vertebral arches only and overlying skin is normal with no visible abnormalities.[6] The vast majority of these involve the sacrum only and less than 10% involve L5. Using this definition, a patient with either radiologically diagnosed spinal dysraphism, symptomatic or not, does not fulfil the criteria for spinal bifida occulta. For the purposes of this review, we have used the original classification used by the authors of papers describing their clinical experience of cases.

Open spinal dysraphisms

In open spinal dysraphisms, the malformed segment of spinal cord (placode) and meningeal layers are not covered by skin and are open to the environment. In all cases the bony vertebral arch is deficient and the placode and covering meningeal membranes protrude. Four types exist; myelomeningocele, meningocele, myelocele, hemimyelomeningocele and hemimyelocele. Of these myelomeningocele is by far the most common. In myelomeningocele, the neural placode is elevated above skin level by the expanded subarachnoid space while in myeloceles, the placode is flush with the surrounding skin. In most myelomeningoceles, the placode is terminal i.e. at the caudal end of the spinal cord but segmental variants have been described in which spinal cord caudal to the thoracic or lumbar placode are normally formed. In hemimyelomeningoceles and hemimyeloceles, the lesion affects one side of a split spinal cord. Open spinal dysraphisms are always associated with a Chiari II malformation which is variable in severity.[5]

Closed spinal dysraphisms

In closed spinal dysraphisms, overlying skin is present but the spinal cord and associated structures are abnormal. Where a mass is present, it most commonly occurs in the lumbosacral area above the natal cleft. In this area, the majority of masses are lipomatous and are associated with dural defects.[5] The lipoma typically has a subcutaneous portion which extends into the spinal canal through the spina bifida defect and tethers the spinal cord.[5] The range of anatomical variations in closed spinal dysraphisms is wide and encompasses all developmental abnormalities in the midline of the back. These include a low-lying spinal cord and conus medullaris, tethered spinal cord, split cord, lipomata (including lipomyelomeningocele). Rarer conditions include terminal myelocystoceles and neurenteric cysts. In some cases patients do not report any symptoms and go undiagnosed into adulthood. Approximately 70% of patients with closed spinals dysraphisms have abnormal skin overlying the lesion but these skin abnormalities are not universally present. While their presence should increase the clinical suspicion of underlying dysraphism, they are not pathognomonic (Fig. 1) (Table 2).[7,8] Clinical manifestations of closed spinal dysraphism is usually secondary to tethering of the filum terminale and is known as tethered cord syndrome (TCS). Symptoms of TCS include urinary frequency and incontinence and non-dermatomal back and lower limb pain.[9] Back pain secondary to TCS is typically worse when the spine is flexed and alleviated when extended.[10] Signs include limb, buttock and foot asymmetry, pes cavus and tallies equinovarus foot deformities, high arches, hammer toes and clawed feet.

Epidemiology

The prevalence of spinal dysraphisms ranges from 0.2–10 per 1000 with wide geographic variation and is among the most common birth defects.[11-13] In the recently published USA National Birth Defects Prevention Study, the combined prevalence of myelomeningocele, meningocele, myelocele, lipomyelomeningocele and lipomeningocele was 3.06 per 10 000 live births.[14] The majority were myelomeningocele and of these 79.9 % of were lumbar, 11% sacral, 8.4% thoracic and 0.8% cervical.[14] In another study, the anatomic level of the lesion was T12 or lower in 83.3% of open and 84.1% of closed spinal dysraphisms.[1] Neurological impairment, manifest as motor and sensory dysfunction, absent reflexes, sphincter dysfunction, hydrocephalus

and Chiari II malformations were more common with higher lesions and those that were classified as "open" at birth.[1]

Isolated bony abnormalities commonly known as spina bifida occulta are much more common with a reported incidence ranging from 1.2–50% depending on the definition used.[15,16] The majority of these abnormalities are vertebral arch defects in the sacrum, with 80% occurring at S1, 10% at S1-2, 8.4% at L5 and 0.2% at L5-S1. The clinical significance of such findings in asymptomatic patients is disputed and is largely considered to be a variant of normal.

Reproductive health in parturients with spinal dysraphisms

Fertility is thought to be normal in patients with open spinal dysraphisms.[17] In the Dutch ASPINE study, over 46% of patients with open spinal dysraphisms and shunted hydrocephalus were reported to be sexually active.[18] Successful pregnancies have been reported in multiple patients with spinal dysraphisms including its more severe forms.[19-24,25] In Arata et al.'s series of 23 pregnancies in 17 women, 12 were delivered by caesarean section.[20] Hypertensive disorders occurred in six of the 23 pregnancies.[20] Premature delivery is also common, secondary to cephalopelvic disproportion and postoperative complications reported to be common requiring prolonged inpatient stays.[17, 20] Caesarean section is more common in wheelchair-bound compared to independently mobile patients.[20] In Sterling et al.'s series of 32 spinal cord injured patients, (69% secondary to neural tube defects) caesarean section was performed in 60% for indications such as failure to progress, pelvic instability or contracture, fetal distress and concerns regarding pushing in the presence of a CSF shunt.[25] Urinary tract infections occurred during pregnancy in 68% of cases and recurrent urinary tract infection was particularly common in those who self-catheterised. Pyelonephritis developed in 11%.

Current practice

Clinicians providing care for patients with spinal dysraphisms have opted not to place neuraxial blocks in those with a history of spinal fusion,[26] tethered cord,[26,27] previous dural puncture,[26] existing neurological deficit[28] and in the absence of appropriate diagnostic imaging.[29] Such clinical decisions are not strongly evidence based and are unlikely to be universally accepted and many of the clinical cases described below have at least one of the above contraindications. In true bony spina bifida, the ligamentum flavum is malformed or absent adjacent to the bony deficit

and avoidance of needle placement at the level of the lesion is recommended because of increased risk of dural puncture.[30-32] Pian Smith and Leffert suggest that spinals and epidurals are contraindicated in patients with a tethered cord and preserved neurological function.[33] It has, however, also been suggested that in the presence of severe compromise of extremity and sphincter function, a low-lying tethered cord should not interfere with spinal needle placement.[32] This approach is not reasonable if partial neurological function is present as residual neural placode function may maintain intact local spinal reflexes governing bladder and bowel function even in the absence of cortical control.[34] Other authorities do not feel that neuraxial blocks are contraindicated but if epidural blocks are used they should be placed proximal to the deficit or surgical scar.[31] Spinal anaesthesia for caesarean section has been advocated using normal local anaesthetic volumes.

Anaesthetic care in cases reports

Despite the relatively high prevalence of spinal dysraphisms, the number of cases in the English literature in which detailed descriptions of peripartum analgesia and anaesthesia is small consisting of 16 case reports, three cases series and a registry with a cumulative total of 84 patients.[3,20,27,29,35-50] There are more limited descriptions in meeting abstracts of an additional 55 patients.[26,28,51]

Overall, epidural techniques were used in 52 cases. Reported complications in these cases included asymmetric block (n=1),[49] dural puncture (n=3),[3,45,49] excessive block height (n=1),[49] suboptimal analgesia (n=7),[48,49] rapid onset block (n=1),[26,48] spinal catheter migration (n=1),[39] pain on needle placement (n=1),[45] increased number of attempts (n=2),[28] difficulty locating the epidural space (n=1),[20] and post-procedural neurological deficit (n=1).[52] Spinal or combined spinal-epidural (CSE) anaesthesia was used in 15 cases.[20,26,39,41,42,44,49] One of these cases involved the use of a T7-8 spinal catheter and another, a T10-11 single-shot injection.[41,47] Reported complications included multiple attempts (n=1),[41] block failure and suboptimal block height (n=4),[26,43,44,49] asymmetric block (n=1),[39] difficulty locating the subarachnoid space (n=1),[20] rapid block regression (n=2),[41,43] paraesthesia on injection (n=2),[44] postoperative sciatic distribution pain with full recovery (n=1).[44] Paravertebral blockade was used in two cases one at T11-12 and the other at L2.[40, 42] In neither case, was complete analgesia achieved.

Of the 41 cases clearly described as either spina bifida cystica, spina bifida aperta, myelomeningocele, meningocele or occult spinal dysraphism, spinal or CSE anaesthesia was used in 10 cases and epidural in 17.[3,20,26,28,39,41,43,47-49,53]Slow onset block with rapid regression was reported in one case of spinal anaesthesia.[43] In the sole case where a CSE technique was used, an asymmetric block with accidental intrathecal catheter placement was described.[39] Analgesia was successfully attained for labour using an intrathecal catheter at T7[41] and a single-shot technique at T10-11[47] in patients with pre-existing neurological deficits. The remaining cases were described as uncomplicated but reports do not detail needle placement or medication use. In 21 cases where an epidural was used, incomplete analgesia occurred in nine, accidental dural puncture in one, complete analgesia after a test dose in one. In nine epidurals, no complications were recorded and analgesia was satisfactory. In a case reported by Ahmad et al., postoperative foot drop occurred after low lumbar spinal needle placement for caesarean section. A low terminating spinal cord was discovered on postoperative magnetic resonance imagining (MRI).[54]

Previous back surgery, neurological deficits including neurogenic bladder, tethered cord (symptomatic and asymptomatic), limb symptoms and undiagnosed dysraphism were present in 10 of 62 patients described as having spina bifida occulta.[26,27,35,37,42,44,52] In these cases a spinal technique was used in five, CSE in none and epidural analgesia in a further 14. Where spinal anaesthesia was used, block failure requiring conversion to general anaesthesia (GA) occurred in one case.[49] Paraesthesia on injection with an L3-4 spinal attempt occurred in another patient on two separate injections.[44] This patient developed a temporary neurological deficit and on subsequent MRI was discovered to have a low conus medullaris.[44] Two of 14 epidurals were complicated. In one, radicular pain occurred on needle placement caused by difficulty in determining the vertebral level and a high needle placement.[45] This patient developed a post dural puncture headache. In the other case, a neurological deficit occurred after an uncomplicated L3-4 epidural and an MRI showed previously undiagnosed spinal dysraphism.[49]

In cases managed with GA, failed intubation with supraglottic airway rescue is described in one case and awake intubation in two.[36, 37] Hee and Metias reported a single case in which a parturient with repaired spina bifida cystica required GA for caesarean section but refused upper limb intravenous access, cricoid pressure and mask application and was induced with

intramuscular ketamine 10 mg/kg.[55] In Aratas et al.'s series of obstetric patients, GA was used in 91% of caesarean sections.[20]

In the non-obstetric literature Wood and Jacka reported a subarachnoid hematoma and paraplegia after spinal anaesthesia in a patient with asymptomatic spina bifida occulta with low lying spinal cord.[56] Cooper and Sethna reported the uneventful use of perioperative epidural anaesthesia for elective paediatric surgery in three patients with repaired closed spinal dysraphism. In all cases the level of insertion was above the level of repair.[57]

Risks of neuraxial anaesthesia

Other than the cases detailed above, a review of existing publications in the anaesthetic literature, which details 602 reports of neurological injury out of a total of 5 304 115 cases, does not identify spina bifida in any form as a risk factor in any individual case. In related work, Sharpe et al. recently reported the use of neuraxial anaesthesia is a series of eight patients with spinal cord injuries of unspecified origin of whom six had complete injury and two incomplete injuries. Neuraxial blocks were used in all patients without subsequent neurological deterioration.[58]

Surgical repair and implications for neuraxial anaesthesia

Anaesthetists typically encounter patients with repaired lesions, usually performed in infancy. Neurosurgical repair attempts to protect neural elements, provide a seal over the dura and prevent future cord tethering. Filum terminale release is often performed. Placode reconstruction or preservation may be attempted followed by dural mobilisation and closure over the neural elements. Dissection and closure of five layers is typically performed. The pia and arachnoid layers are dissected away from their junction with the neural placode and sutured together in the midline. Similarly, the freed dural edges are approximated and closed in the midline where possible. Where dural tissue is inadequate, the defect may be patched with fascial flaps and more rarely cadaveric dura, bovine pericardium or colloidal collagen.[34] The paravertebral muscles and associated fascia are closed in the midline where possible. Latissimus dorsi flaps are sometimes used and may be reinforced by mobilised lumbosacral fascia. Bony defects are not repaired but are sometimes osteotomised to facilitate closure.

After surgical repair, the epidural space may is unlikely to be normal and can be non-existent. Identification of the vertebral level at the site of the defect is not possible and the

ligamentum flavum is not present. Consequently, the epidural space cannot be located using a loss-of-resistance technique at the level of a repair. In patients who have undergone surgical de-tethering of the spinal cord, re-tethering (which may be asymptomatic) is common, particularly in those with a posterior dural attachment before surgery.[59,60] In MRI studies of patients who had previously undergone neurosurgery for occult spinal dysraphism and myelomeningocele closure, anterior migration of the conus or the cord or filum complex was observed only in a minority in the prone position.[59,61] In contrast normal patients demonstrate anterior movement of the tip of the conus medullaris (mean 6.3 mm) in the hip flexed lateral position compared to supine.[62] In the presence of cord tethering or re-tethering, the caudal neural elements are more likely to be posteriorly placed within the spinal canal with a reduced safety margin for neuraxial needle placement and possibly more vulnerable to direct needle trauma. Furthermore, previous de-tethering does not automatically result in elevation of the conus within the spinal canal. Kim et al. reported ventral movement of the conus in 44% of subjects imaged, a mean of 2.5 years after surgical sectioning of the filum terminale.[63] The cauda equina, which is vulnerable to injury from neuraxial approaches, is frequently low and remains so despite corrective surgery in many cases.[63,64]

Issues in peripartum care

Intrapartum pain perception

Sensory deficit is extremely variable in patients with open spinal dysraphisms and some degree of sensory preservation is seen even in patients with profound motor deficits.[65] Oakeshott et al. found preserved perineal sensation in 33% of patients with spinal dysraphisms who had undergone early surgical closure.[65] Sensory abnormalities may be asymmetrical and isolated sensory sparing in the perineum has been described in a number of patients with lumbar and thoracic sensory levels.[66] Motor and sensory levels may also be unequal.[18] Patients with complete lesions above T10 may not perceive contraction pain and are at risk of unattended birth.[25] Parturients with impaired sacral sensation may not experience the added pain of the second stage of labour but experience pain in the first stage if lumbar sensory function is even partially preserved.[31] T4 levels of anaesthesia are required for caesarean section to cover peritoneal stimulation and therefore consideration must be given to the need to achieve this level of block even in those with relatively proximal neurological levels.

Associated Neurological Conditions

Hydrocephalus secondary to Chiari malformation is common in patients with open spinal dysraphisms and is treated with central nervous system shunts. Shunts are almost universal in patients with thoracic level and in 70% of lumbar myelomeningoceles.[13,18]The most common shunt is a catheter running from the ventricles to the peritoneum (ventriculo-peritoneal) but may also be ventriculo-atrial or ventriculo-pleural. Shunt malfunction has been described in a number of patients during pregnancy and has been attributed to increased total body water, increased cerebrospinal fluid (CSF) volume and increased intra-abdominal pressure as a consequence of uterine enlargement.[67] Acute shunt malfunction was reported 12 hours after caesarean section by Hwang et al. and was attributed to an obstructing blood clot from the surgical field.[68] Liakos et al. reported seven shunt revisions in 138 pregnancies and a further 15 cases of transiently increased intracranial pressure not requiring surgical intervention.[69] None of the six myelomeningocele related shunt patients in this series developed shunt malfunction but one had a shunt infection at 30 weeks of gestation. Symptoms of shunt malfunction include headache and visual disturbances. Clinical signs include confusion, amnesia, pupillary abnormalities, seizures and altered level of consciousness. It is important to distinguish headaches due to hydrocephalus from other causes of headaches such as migraine or preeclampsia. Magnetic resonance imaging is recommended as the investigation of choice. It is important to note that it is not an appropriate choice of investigation in women, who have a metallic component to their shunts. Even in patients who have not had shunts, raised intracranial pressure which may have previously been undiagnosed may occur.[70]

The presence of a shunt is not a contraindication to either spinal or epidural anaesthesia. In the largest series published to date, Bradley et al. described the use of epidural analgesia in 13 of 41 vaginal deliveries and 11 of 22 caesarean sections in patients with shunts.[71] Spinal anaesthesia was used in two caesarean sections, the remaining nine were performed under GA. Block failures occurred in four cases. Numerous case reports exist of the performance of spinal anaesthesia in parturients who are neurologically stable with shunts in place. There is an argument to be made that the loss of CSF using a small gauge needle is a safer alternative than laryngoscopy, especially in the case of a potentially difficult airway where increased intracranial pressure could occur.

Autonomic dysfunction may occur in patients with spinal cord injury but is unusual in patients with lesions below T6.[58] Manifestations of autonomic dysfunction include hypertension, headache, blurred vision, piloerection, diaphoresis, palpitations, tachycardia or bradycardia.[72] Epilepsy, chronic pain, spasticity and visual impairment have been reported in 9%, 25%, 13% and 8%, respectively, of those with spina bifida aperta and hydrocephalus.[18]

Genitourinary tract

Renal dysfunction is common in patients with more severe spinal dysraphisms and most common in those with neuropathic bladders and bladder diversions and may require transplantation.[17,20] Renal function may further deteriorate during pregnancy secondary to hydronephrosis and conduit obstruction.[73] Patients with spinal dysraphisms, who have neurogenic bladders may undergo a variety of urological surgical procedures which aim to maintain continence which include ileal conduits, clam cystoplasties, urethral slings and bladder neck reconstructions.[17] These may make the performance of a caesarean section more complex and require urological expertise. Urinary tract infections are common during pregnancy particularly among those who practice self-intermittent catheterisation and are a frequent cause of hospitalisation during pregnancy. Urinary tract infections are also associated with premature labour.

Latex allergy

The reported incidence of latex allergy in patients with spina bifida is high with some older studies reporting rates of up to 35%.[74] More recently much lower incidences have been attributed to the adoption of latex free environment for patients from their first hospital exposure, and for this group, the incidence of latex allergy is similar to that of the normal population. This avoidance of latex also appears to prevent the development of allergy to other common allergens.[75]

Skeletal abnormalities

Talipes equinivarus (clubfoot), contractures, hip dislocation, scoliosis and kyphosis are common in patients with myelomeningoceles and corrective surgery is common.[76,77] Scoliosis is reported in up to 69% of patients.[76] It is most common in those with thoracic lesions and less frequent in lumbar lesions.[18] Many patients with scoliosis will have undergone surgical correction, which

makes the performance of neuraxial anaesthesia technically difficult. Scoliosis repair may include deliberate spinal cord transection.[78] Severe kyphoscoliosis has also been associated with restrictive lung disease and cardiorespiratory failure.[79] Closed spinal dysraphism can present with limb asymmetry, high arches, hammering and clawing of the toes.

Mortality outside of pregnancy

Oakeshott et al.'s longitudinal cohort study of 117 patients born with open spina bifida and who underwent back closure as infants identified a mortality rate of 10 times the UK national average between the ages of 5 and 35 years.[80] Survival was lowest in those with sensory levels above T11 and the most common causes of death were pulmonary embolus, acute hydrocephalus, epilepsy and urinary tract sepsis.

Preoperative assessment

Recommendations

Parturients with a history of spinal dysraphism should be seen on an outpatient basis well in advance of delivery.[31,49] A full sensory and motor examination should be performed and documented. The extent of the anatomical level of the bony defect should be established by clinical examination and diagnostic imaging as required. The presence of a central nervous system shunt should be ascertained and its function determined. Latex allergy status should be determined.

Neurologic Assessment

Functional neurological motor impairment can be graded according to ability to ambulate. This may be useful in projecting the need for caesarean section, as non-ambulant patients are less likely to deliver vaginally. A sensory examination is performed and the lowest completely unimpaired dermatome level on both sides measured with sensitivity to pin-prick and light touch is identified. [81]The neurological level is defined as the most caudal level at which sensory and motor examination in normal on both sides.

Imaging studies

Anatomical abnormalities in patients with actual or suspected spinal dysraphisms cannot be assumed on the basis of clinical examination and appropriate imaging is required. Correspondence between the anatomic level of the bony defect and functional level of impairment is variable and impairment can be higher than the anatomic level in up to 48% and lower in 14% of patients.[13] Existing imaging studies may be useful in anatomical assessment of the lesion and identification of normal anatomical levels as potential areas for neuraxial needle placement. An MRI scan is the study of choice and can be performed in pregnancy if required. X-rays of the lumbar spine yield limited information and are not recommended during pregnancy. Magnetic resonance imaging allows determination of the level of termination of the conus medullaris, the presence of tethering, assessment of CSF cistern volume and the presence of masses e.g. lipoma or syrinx (Figs.2-7). It should also be used to identify normal anatomical levels with an intact ligamentum flavum. When a clinical suspicion of occult spinal dysraphism exists, MRI should be considered especially in the presence of urinary symptoms, sensory or motor abnormalities,[43] previous back surgery, back pain, limb deformity and midline cutaneous abnormalities.[56] It has been suggested that patients with suspicious midline skin markings who are not symptomatic do not benefit from MRI. It should, however, be borne in mind that many of the cases in the literature described as asymptomatic had abnormal clinical signs and the absence of symptoms is not a reliable predictor of disease absence.[26,27] Magnetic resonance imaging may also be useful if previous surgery has occurred, as de-tethering may not result in a change in the position of the conus.

Ultrasonic evaluation of the spine is of established value in neonatology as a screening tool for detection of spinal dysraphisms.[82-84] Ultrasonic findings with repaired myelomeningocele in a paediatric population have shown that usable images were obtained in 80% of cases. Concordance between ultrasonography and MRI was seen in 82% of cases regarding the level of the distal end of the cord, in 59% of cases regarding the position of the cord in the canal, in 63% regarding the presence of hydromyelia, in 96% regarding cord duplication, in 16% regarding adhesions, in 37% regarding intradural mass and in 83% regarding dural sac measurements.[85] The relevance of these data to anaesthetic practice is limited as the technique is heavily dependent on operator expertise and bony windows. Ultrasonic spine evaluation is rapidly evolving as a useful tool in obstetric anaesthesia but its use to guide neuraxial blockade in spinal dysraphisms has not been described or validated.[86, 87]

Based on the clinical and radiological assessment, labour options can be discussed with patients providing realistic expectations of analgesic outcomes.[49] Neuraxial techniques can be used in selected cases. Needle placement through lesions or scars is not recommended. Epidurals should be performed at anatomically normal levels with an intact ligamentum flavum. Analgesia may, however, be incomplete if the epidural space has been altered by corrective surgery and supplementary distal nerve blocks may be required in the second stage. Reduced bolus doses of epidural medication are recommended in those with abnormal anatomy. No recommendations can be given regarding paravertebral blockade, spinal catheter techniques, epidural opioids or additional epidural catheters placed below the level of the lesion as clinical experience with these approaches is extremely limited.

General anaesthesia has been frequently used for caesarean section in patients with spinal dysraphism and is usually uneventful.[20] In a small number of cases airway problems, unrelated to the spinal dysraphisms have been reported. Succinylcholine may trigger hyperkalaemia in the presence of neuropathy or myopathy but has been safely used in many cases of myelomeningocele outside of pregnancy.[88] In patients with kyphoscoliosis, reduced lung volumes are associated with more rapid oxyhaemoglobin desaturation and short tracheal length predisposes to right main bronchus intubation.[79]

Conclusion

Neuraxial techniques can be used in select patients with spinal dysraphisms. Considerable variation is encountered in both anatomy and sensory perception. Both spinal and epidural techniques have been successfully used but overall success rates are lower than in the normal population. Imaging studies are recommended in order to understand individual patient anatomy and identify appropriate levels for needle placement.

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Figure Legends

Fig. 1 Panel A. Lumbosacral hypertrichosis. Panel B. Frontal abdominal radiograph showing dextroscoliosis and absence of posterior elements at L5 (arrow). Panel C. CT image showing bony bar at L3 and diastematomyelia. Panel D. CT image showing spina bifida at L5. This is a non-pregnant patient whose images are used for illustrative purposes.

Fig. 2 T2 weighted sagittal image demonstrating an enlarged lumbar cistern (white arrow) and a low-lying cord to the level of the L4 vertebral body (grey arrow). Incidental degenerative disc at L3-4.

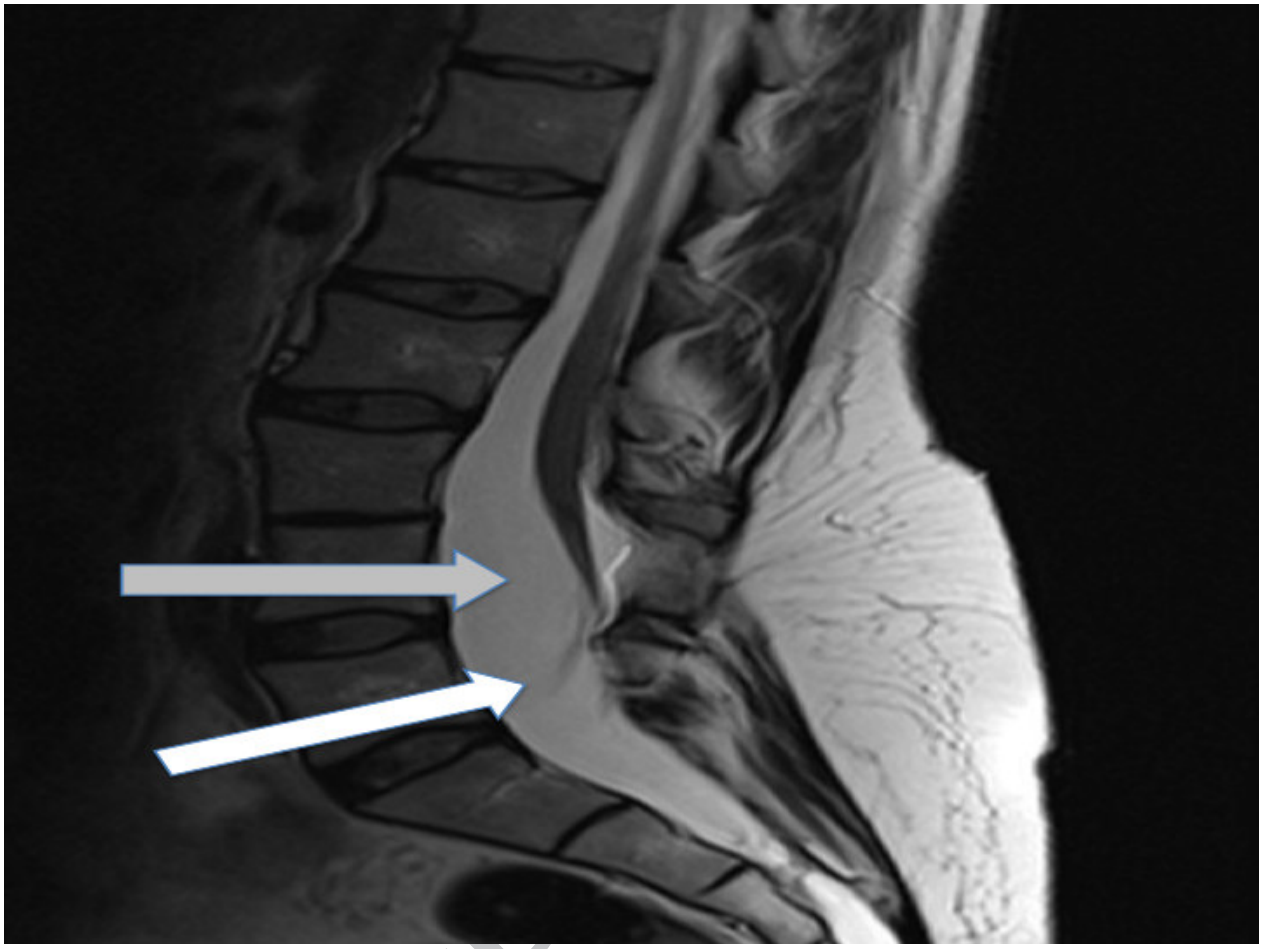
Fig. 3 T2 weighted axial image at S1 level demonstrating an enlarged lumbar cistern with a low-lying cord and a dorsal dermal sinus.

Fig 4 T2 weighted sagittal image showing a moderately enlarged lumbar cistern with a well circumscribed intramedullary lesion consistent with an intramedullary lipoma (arrow) at the level of the conus.

Fig.5 A. T2 weighted sagittal image show a moderately enlarged lumbar cistern with a T2 hyperintense well circumscribed intramedullary lesion consistent with an intramedullary lipoma at the level of the conus. B. T2 weighted axial image shows diastematomyelia of the cord just above the level of the previously lipoma at L1.

Fig. 6 T2 weighted sagittal image showing hydrosyringomyelia of the distal cord at the level of L1 and cord tethering at S1.

Fig. 7 A. T2 weighted axial image showing hydrosyringomyelia of the distal cord at the level of L1 (arrow). B. Diastematomyelia distal to the syrinx (arrows).



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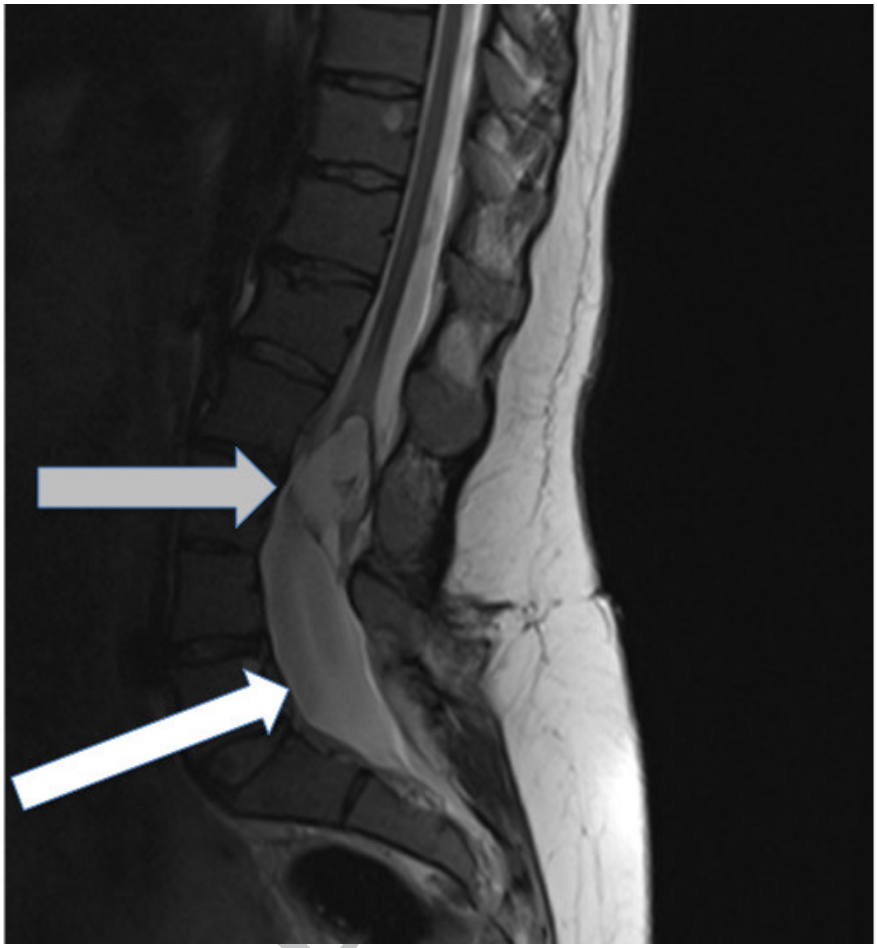

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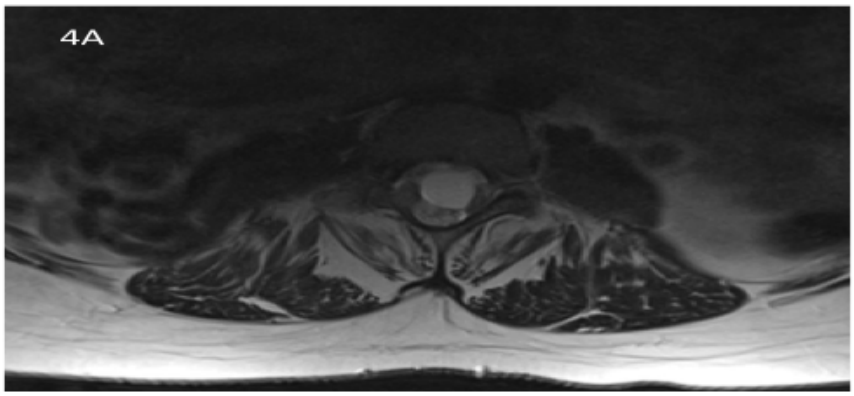

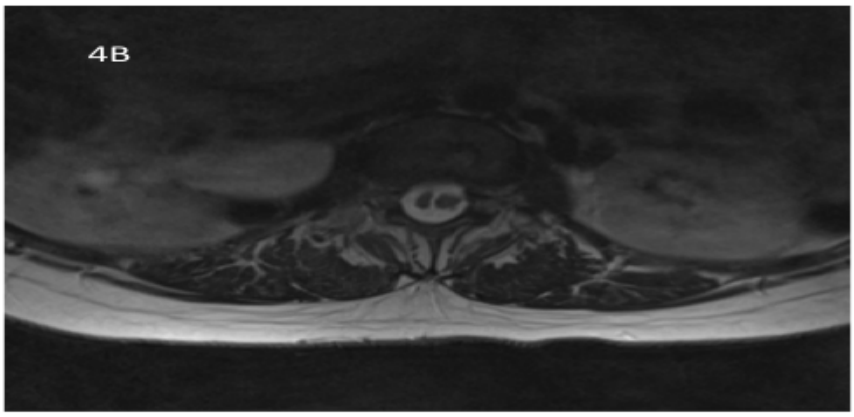

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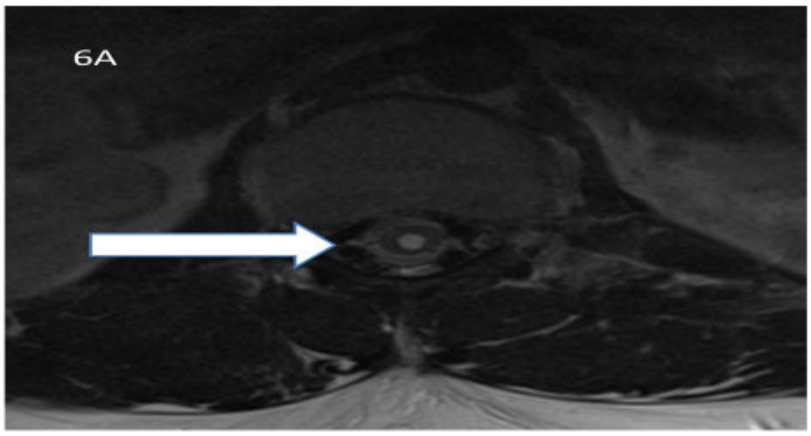

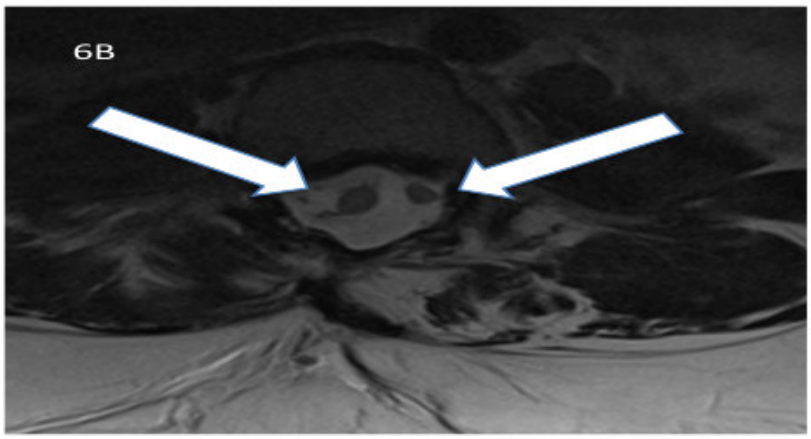

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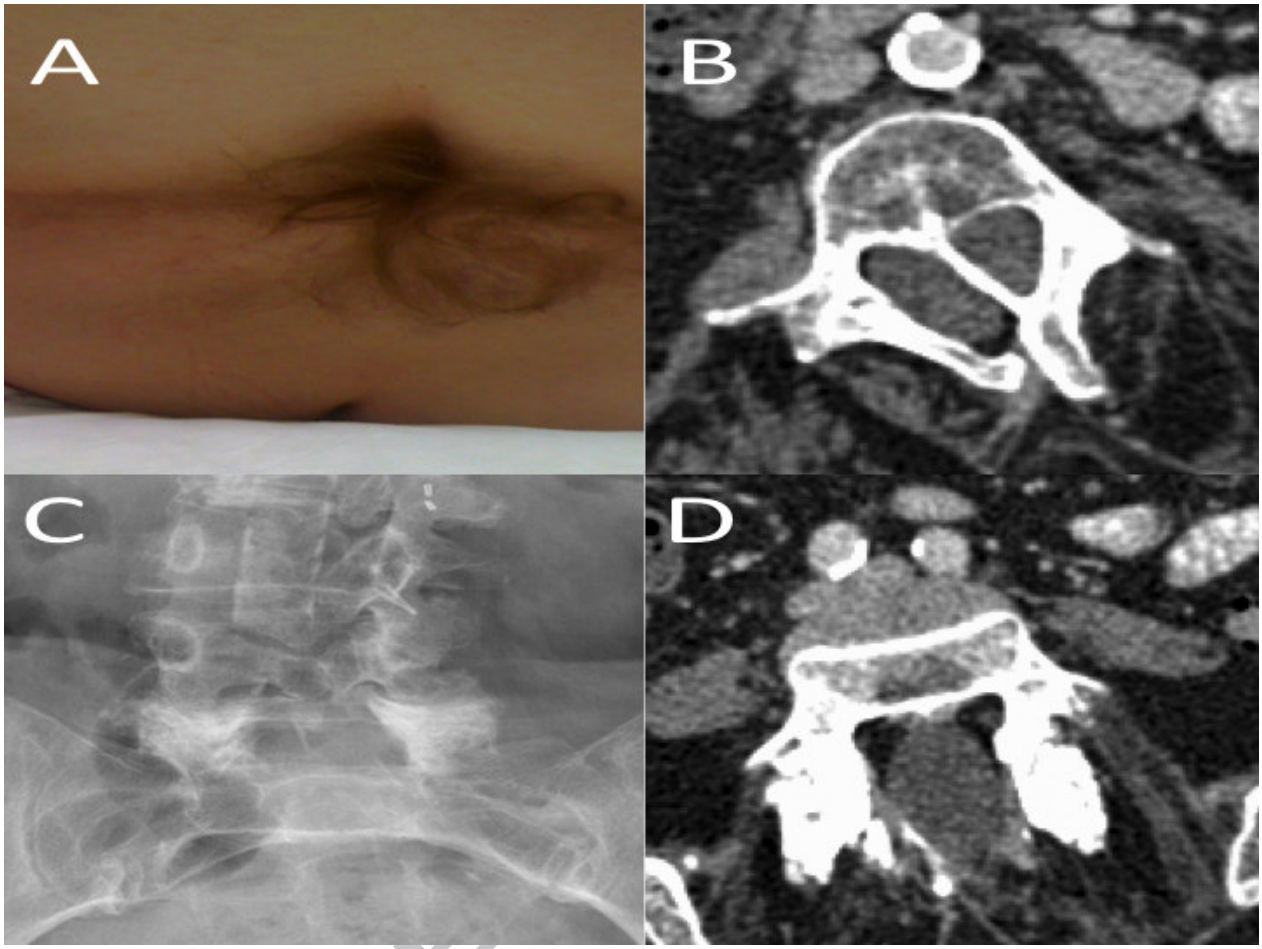

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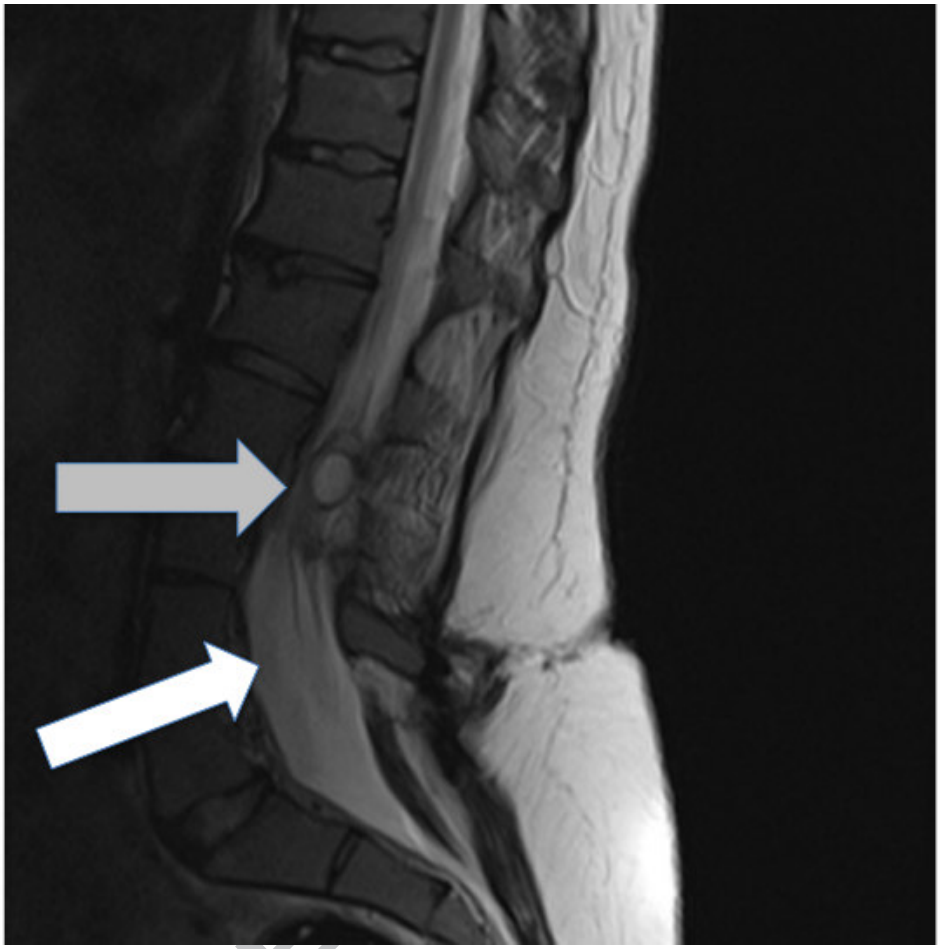

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660 **Table 1 Classification of spinal dysraphisms[5]**

Open Spinal Dysraphisms		
Myelomeningocele		
Meningocele		
Hemimyelomeningocele		
Hemimyelocele		
Closed Spinal Dysraphisms		
With Subcutaneous Mass		Without Subcutaneous Mass
Lumbosacral	*Cervical*	*Simple Dysraphic States*
Lipomyelocele	Meningocele	Posterior spina bBifida
Lipomyelomeningocele	Myelocystocele	Lipoma
Meningocele	Myelocele	• Intradural
Terminal myelocystocele		• Intramedullary
		• Filum terminale
		Tight filum terminale
		Abormally long spinal cord
		Persistent terminal ventricle
		Complex Dysraphic States
		Dorsal enteric fistula
		Neurenteric cysts
		Split cord malformations
		• Diastematomyelia
		• Diplomyelia
		Dermal sinus
		Caudal regression syndrome
		Segmental spinal dysgenesis

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662 **Table 2 Clinical manifestations of spinal dysraphisms**

Cutaneous[89]	**Urological**[60, 90]	**Neuro-orthopedic**[60, 91]
High Index of Suspicion	Incontinence	Talipes equinovarus
Hypertrichosis	Recurrent UTI	Pes cavus
Dimples		High arches
• Large		Hammer Toes
• >2.5 cm from anal margin		Clawed feet
Acrochordrons		**Asymmetry**
Pseudo-tail		Buttock
True tail		Leg
Haemangiomas		Foot
Aplasia Cutis/ Scar		**Symptoms**
Dermoid Sinus or cyst		Non dermatomal back pain
		Numbness
Low Index of Suspicion		Weakness
Telangectasia		
Capillary malformation (port wine stain)		
Hyperpigmentation		
Melanocytic nevi		
Teratomas		

663 UTI: urinary tract infection

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Highlights

- Spinal dysraphisms are among the most common neurodevelopmental anomalies.
- Bony and neural structures may be anatomically abnormal.
- Neuraxial blocks are possible in selected patients.
- The incidence of complications and failure is relatively high.
- Magnetic resonance imaging can clarify anatomical abnormalities and assist decision making regarding neuraxial techniques.