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REVIEW ARTICLE

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

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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.

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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

Table 1 Classification of spinal dysraphisms[5]

Open spinal dysraphisms	
Myelomeningocele	
Meningocele	
Hemimyelomeningocele	
Hemimyelocele	
Closed spinal dysraphisms	
With subcutaneous mass	Without subcutaneous mass
Lumbosacral	*Simple dysraphic states*
Lipomyelocele	Posterior spina bifida
Lipomyelomeningocele	Lipoma
Meningocele	• Intradural
Terminal myelocystocele	• Intramedullary
	• Filum terminale
Cervical	Tight filum terminale
Meningocele	Abnormally long spinal cord
Myelocystocele	Persistent terminal ventricle
Myelocele	
	Complex dysraphic states
	Dorsal enteric fistula
	Neurenteric cysts
	Split cord malformations
	• Diastematomyelia
	• Diplomyelia
	Dermal sinus
	Caudal regression syndrome
	Segmental spinal dysgenesis

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, 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 the spinal cord caudal to the thoracic or lumbar placode is 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 and 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 are 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 talipes equinovarus foot deformities, high arches, hammer toes and clawed feet.

Epidemiology

The prevalence of spinal dysraphisms ranges from 0.2 to 10 per 1000, with wide geographic variation, and it 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 10000 live births.[14] The majority were myelomeningocele and of these 79.9% 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,