

# Spinal dysraphic anomalies; classification, presentation and management

Dominic N P Thompson

## Abstract

Spinal dysraphism comprises an array of congenital anomalies of spinal cord development. Each of the dysraphic disorders can result in neurological, orthopaedic and urological dysfunction. This potentially confusing array of conditions is best understood from the embryological perspective. Whilst not all of these conditions are managed in the same way similar principles govern the initial investigation, multidisciplinary evaluation and long term follow up of these cases.

**Keywords** dermal sinus track; lipomyelomeningocele; myelomeningocele; spina bifida; spinal dysraphism; tethered cord syndrome

## Introduction

A number of aberrations of spinal cord development, some common some exceedingly rare are embraced by the term spinal dysraphism. However a bewildering array of diagnostic terms, syndromes and synonyms surrounds the subject of spinal dysraphism, to the extent that patients are commonly inappropriately labelled and doctors and medical staff incompletely informed. Even amongst “specialists” and in the contemporary medical literature there are many terms that are used inconsistently, interchangeably and on occasion incorrectly. It is therefore an impossible task to provide an overview of the subject that would withstand scrutiny from all interested parties. It is hoped however that the account that follows, with its basis in simple undergraduate embryology will provide a framework within which the dysraphic disorders might be better understood.

## Embryological basis of spinal dysraphism

### A. Normal spinal cord development

A rudimentary knowledge of the development of the spinal cord is essential to begin to understand the dysraphic pathologies and with this in mind three stages of spinal cord development need to be examined.

**Gastrulation:** the very early embryo comprises an inner and outer cell mass, a bilaminar plate. The primitive streak, a midline groove that extends from cranial to caudal on the dorsal aspect of the embryo is the first indication of the rostro-caudal polarity of

the embryo. Cells migrate toward and through the region of the primitive streak creating a middle layer, thus transforming the structure from bilaminar to trilaminar. This process, establishing the three germ cell layers of ectoderm, mesoderm and endoderm is referred to as gastrulation, and forms the basis of the embryo from which all tissues and organs will derive. Opposite the ectoderm (future nervous system and skin) is the amniotic cavity, opposite the endoderm (future alimentary canal) is the yolk sac. The middle lamina, the mesoderm will be the origin of the musculoskeletal system. As the embryonic disc enlarges during gastrulation the primitive streak regresses in a caudad direction leaving in its wake a midline tubular structure, the notochord. For a short period of time the notochord provides temporary communication between the yolk sac and amniotic cavity via the neurenteric canal.

**Primary neurulation:** the ectodermal layer will give rise to the central nervous system and the integument (skin and its adnexae). Under the influence of the underlying notochord two longitudinal folds develop in the ectoderm, the lips of these folds will meet in the midline and then “zip up” in a rostral and caudal direction establishing the tubular structure of the central nervous system. It is this process that is dependent on the activity of folate dependent enzymes. The open ends of the neural tube, anterior and posterior neuropores are last to close at approximately 24 and 27 days post-fertilization. In fact it is now known that the closing of the tube occurs at a number of “closure points” thus there are in essence more than “two neuropores”. This folding of the ectoderm establishes the tubular primordium of the central nervous system and is known as primary neurulation, the structure so formed will undergo a complex series of folds at its rostral end to form the brain, the caudal part of the neural tube retains its tubular form in the form of the spinal cord with its slender central canal. Once the folds of the ectoderm have met, the neural tube separates from the rest of the ectoderm (that destined to be skin) in a process referred to as dysjunction, in this way that part of the ectoderm destined to be neural, the neuroectoderm becomes distinct from that which will form the skin, the cutaneous ectoderm.

The spinal cord thus formed only extends however to the second sacral segment, the terminal spinal cord, conus medullaris, filum terminale and elements of the cauda equina form by a third process.

**Secondary neurulation:** in the tail bud of the embryo is a mass of pluripotent cells the caudal cell mass, this will form the lower end of the spinal cord and will also contribute to the lower gastrointestinal and genitourinary tracts. Cells within the caudal cell mass coalesce around a central lumen and will join the lower end of the neural tube that was formed by primary neurulation thus completing the spinal cord. Initially the spinal cord extends into the tail of the embryo, however with growth and regression of the tail bud much of this terminal component of the spinal cord gradually involutes, what remains constitutes the conus of the spinal cord and the filum terminale.

### B. Abnormalities of spinal cord development

The term spinal dysraphism refers not to a specific condition but to a group of congenital conditions each of which has its

*Dominic N P Thompson MBBS Bsc FRCS (SN) is a Consultant Paediatric Neurosurgeon in the Department of Neurosurgery, Great Ormond Street Hospital for Children NHS Trust, Great Ormond Street, London WC1N 3JH, UK. Conflicts of interest: none.*

### Classification of the dysraphic disorders according to presumed embryological origin

Process	Gastrulation	Primary neurulation	Dysjunction	Secondary neurulation
Features	Establishment of the three germ layers	Formation of the neural tube	Separation of the neural and cutaneous ectoderm	Formation of the terminal spinal cord, conus and filum
Dysraphic condition	Split cord malformations Neurenteric cysts Anterior meningocele	<b>“Open” dysraphism</b> Myelomeningocele Meningocele	<b>Incomplete dysjunction</b> - Dermal sinus track <b>Premature dysjunction</b> - Lipomas (dorsal type)	Lipomas (caudal, transitional) Thickened filum Myelocystocele

**Table 1**

presumed origin at different points in spinal cord development (Table 1).

**Disorders of gastrulation:** signalling from the notocord instigates the process of folding of the ectoderm. It has been postulated that under some circumstances the notochord is divided or duplicated resulting in the formation of a “double” or split spinal cord, **split cord malformations (SCM)**. The two hemi spinal cords may occupy separate dural sheaths with an intervening spur of bone (type I SCM or diastematomyelia) or may share a common dural tube with little more than a fibrous band between the two (type II SCM or diplomyelia).

As noted above, during gastrulation there is a period when a temporary communication exists across the embryo in the form of the neurenteric canal, at this time there is a connection between the ectoderm and endoderm. The canal usually disappears though may persist with cyst formation somewhere between the endodermal derivatives and the spinal cord, so called **neurenteric or enterogenous cysts**.

The period of gastrulation establishes the primitive germ layers and so it is not uncommon to see congenital spinal disorders such as hemivertebrae and segmentation anomalies accompanying these dysraphic conditions.

**Disorders of primary neurulation:** defects of primary neurulation result from failure of the neural tube to close or from abnormal separation (dysjunction) of the skin ectoderm from the neural ectoderm. Failure of neural tube closure results in open neural tube defects, the posterior spinal elements are deficient and the neural tissue, the placode is exposed. This is termed **myelocele or myelomeningocele**, the terms are used interchangeably although strictly a myelocele is situated flush with the surrounding skin whilst in a myelomeningocele the placode is thrust proud of the back due to the accumulation of cerebrospinal fluid beneath (Figure 1).

Separation of the neural tissue from the surrounding cutaneous ectoderm (dysjunction) may fail to complete and there may persist a track of tissue that connects the terminal spinal cord to a punctum in the overlying skin, this is known as a **dermal sinus track**. By contrast it has been suggested that the process of separation may occur prematurely, before the neural tube has closed over, this exposes the open placode to the underlying mesoderm and permits the formation of a **spinal lipoma or lipomyelocele**. In this condition the terminal spinal

cord does not float freely within the CSF filled dural tube but is attached by a pedicle of fat to the subcutaneous fat.

**Disorders of secondary neurulation:** many of the dysraphic conditions affect the termination of the spinal cord and so must have their origins in secondary neurulation. The most terminal component of the spinal cord may fail to involute and remain thick and infiltrated with fat, **thickened or fatty filum terminale**. It is likely that many of the more complex forms of spinal lipoma have their origins in secondary neurulation as involvement of the conus is common. There are a number of more severe conditions affecting the entire caudal cell mass and its derivatives which as a result are associated with major congenital anomalies of the hindgut and lower urinary tract. These are not infrequently associated with more widespread congenital anomalies described



**Figure 1** Myelomeningocele. The meninges and spinal cord are exposed through a skin defect in the lumbar region.

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