Paediatric spinal conditions

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Abstract

Children with spinal problems present to a wide range of healthcare providers. These providers include the emergency department, their GPs, physiotherapy and the paediatric medical and surgical clinics (including orthopaedics). They may present with a variety of symptoms, but the common complaints are: back pain, leg pain or a change in back shape (spinal deformity). Some children may experience a combination of these problems. A systematic approach to the history and examination with knowledge of the common spinal conditions in children will allow you to select the best investigations. This will maximize the chance of making the correct diagnosis and providing appropriate care. This article aims to give an overview, appropriate for surgeons in the early years of their training.

Keywords Disc prolapse; kyphosis; sciatica; scoliosis; spinal infection; spinal injury; spine; trauma; tumour

Assessment

The majority of patients presenting for non-deformity-related complaints present with pain as their main symptom. A careful history is vital to help direct further investigation and assessment. A general pain history should first be sought with details of site of pain, radiation, exacerbating and relieving factors (e.g. NSAIDs) and character. Night pain is an important factor; the presence of night pain is often associated with significant pathology. It is helpful to distinguish, where possible, between central bony spine pain and more lateral muscular pain, and establish the presence or absence of associated leg pain. If the child has leg pain, you will want to try and establish whether this fits for a dermatome, and if so, which dermatome? Consider other diagnoses, for example, could this be a hip problem?

Enquiries should be made to establish the presence of any associated neurological disturbance – bladder or bowel dysfunction or leg pain/weakness. Additional questions should focus on fevers/malaise/systemic upset (discitis), recent trauma, significant sporting activity (spondylolysis), other joint pain, family history (juvenile arthritis), significant recent weight loss or malignant disease (neoplastic disease). These symptoms do not confirm or exclude any of these diagnoses but will help to guide further assessment.

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For patients complaining of a change in back shape, you should establish what was first noticed, when, and by whom. Ask how the back has changed over time, and ask what the child and family thinks about it. It is relevant to ask about birth history, motor milestones and family history.

Measurement of height and weight (plotted on an appropriate growth chart) should happen in every children's clinic. This will provide a historical record of growth, but predicting future growth can also be helpful when treating spinal problems. Past growth provides a clue, as does parental height and (for girls) the timing of menarche. Formal assessment of skeletal age can be done by looking at x-rays of the hand a wrist or the Risser grading of the iliac apophysis if the pelvis is included (as it should be) on whole spine imaging.

Children will need to undress down to their undergarments (asking them to come prepared with shorts or a swimming costume can help them feel more comfortable during the examination).

Clinical examination will include an inspection, looking for deformity, shoulder balance, waist crease symmetry and any associated signs such as spinal hairy patches (spinal dysraphism) or café au lait patches (neurofibromatosis).

Adams' forward bend test involves asking the child to lean forwards from the waist to touch their toes with their knees straight. This makes any rotational deformity of the trunk (and underlying scoliosis) more obvious. Spine range of movement should also be assessed along with careful assessment of leg length and pelvic obliquity. A full neurological examination should also be undertaken, including assessment of abdominal reflexes. Beighton's score should be recorded if there is any suspicion of hyperlaxity – conditions such as Marfan's and Ehlers-Danlos can be associated with spinal deformity.

For spinal deformity, the initial imaging is a standardized posterior-anterior (PA) and lateral whole spine radiograph. There is probably little use in un-standardized X-rays prior to being seen in a specialist clinic, as this can lead to increased X-ray dose. This is a particular worry for children with spinal deformity, as they will invariably have a number of different radiographs during their childhood.

Radiographs allow identification of spinal deformity, quantification using the Cobb angle method shown below and allows monitoring of the deformity over time. We may identify underlying vertebral abnormalities such as hemi-vertebrae or fusions that have caused the deformity. The disadvantage of plain film assessment is that it gives only a two-dimensional assessment of a three-dimensional (3D) deformity.

Topographical scanning such as image and scanner interface specification (ISIS) scans are useful to monitor spinal deformity over time by plotting body surface shape (from photographs) to produce a height map of the posterior chest wall. Using topography reduces the radiation dose to patients under long-term follow-up. Some X-ray machines can capture simultaneous antero-posterior (AP) and lateral views and produce a calculated approximation of 3D shape. The cost of these machines remains just above the reach of most of the NHS.

MRI scanning is the first investigation of choice for most patients presenting with back pain or leg pain. It is sensitive and specific for infection and tumours. It shows disc pathology well, and gives a clear view of the neural structures. Plain X-rays may

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be helpful for identifying spondylolysis, but MRI is not so good at this. So, if there is still a clinical concern that there may be a relevant spondylolysis after a negative MRI, you may wish to consider a radio-isotope scan, such as single-photon emission computed tomography (SPECT). SPECT is also good at confirming or excluding osteoid osteoma.

CT scanning should be used sparingly due to the large dose of radiation to a young patient, with the main indication being demarcation of bony anatomy in patients with suspected segmentation anomalies or spondylolysis.

Paediatric deformity

A spinal deformity is a condition in which the spine is not the 'normal shape' because of illness, injury or because it has grown wrongly, that is, the spine is misshapen or malformed. Deformity may be scoliosis (coronal plane), kyphosis (sagittal plane), or a combination of the two.

Deformity may present at any age in childhood. It may be an isolated presentation or may be associated with other conditions such as metabolic disease, neuromuscular disease (e.g. Duchenne's, cerebral palsy), chromosomal disorders or associated with other abnormalities such as in VACTERL (vertebral anomalies, anal atresia, cardiac defects, tracheoesophageal fistula and/or oesophageal atresia, renal and radial anomalies and limb defects).

Patients with spinal deformities present in a number of ways, influenced by the presence or absence of associated conditions.

Children with idiopathic deformity typically present when either they or a parent or healthcare worker notice a change in their spinal shape. This may manifest itself as shoulder asymmetry, waist asymmetry, a rib hump or change in standing or sitting balance. Back pain may or may not be an associated feature at the time of presentation, and it is unusual for idiopathic deformity to significantly limit physical activity. Patients with conditions that may be associated with spinal deformity are often referred from other healthcare specialists for specialist opinion.

Scoliosis

This is a 3D deformity of the spine, characterized by a curvature in the coronal plane which measures 10° or more on standing radiographs, measured using the method described by Cobb (Figure 1). A curve is considered structural if it does not fully correct with lying down, or correction of an underlying factor such as leg length discrepancy. Although defined as a coronal plane abnormality, abnormalities of sagittal spinal profile and rotation are also seen. Adams' forward bend test is a useful test and emphasizes the rotational component of a structural curve.

Scoliosis classification

Scoliosis can be classified according to both the age of onset, and the aetiology.

We differentiate between early-onset and late-onset scoliosis. Early-onset scoliosis will affect lung development and impact on lung function in the long term. Late-onset scoliosis is unlikely to impact on measures of lung function until the curve exceeds 90°. Various ages have been suggested as a cut off between the two groups; the arguments revolve around the timing of lung development. The Scoliosis Research Society has settled on the age of 0–9 years and 10 years or older.

The identified causes of scoliosis may be congenital, neuromuscular, or syndromic. Many cases of scoliosis have no identifiable cause and said to be 'idiopathic'.

Historically, idiopathic scoliosis was subdivided into infantile (0–3 years of age), juvenile (4–9 years of age) or adolescent (10 years or older). It is common to see the term 'infantile' used interchangeably with early-onset idiopathic, and the term 'adolescent' used interchangeably with late-onset idiopathic.

Curves are described according to location in the spine (thoracic, thoracolumbar or lumbar etc.) and side (named for the side of the convexity).

Approximately 70% of children with spinal deformity present with a late-onset idiopathic scoliosis.

Early-onset idiopathic scoliosis

Early-onset idiopathic scoliosis usually presents with a left-sided curve, and is more commonly seen in male patients. Many of these curves will resolve spontaneously by 5 years of age, but is it important they are treated correctly and followed up closely due to potential issues with alveolar development. Curves greater than 20° may be associated with neural axis abnormalities and need MRI scanning under general anaesthetic.

Outcomes in this group fall into two groups. Approximately 80–90% of these curves will resolve, but of those that do not there is a risk of rapid curve progression with potential for significant cardiorespiratory compromise. Cobb angle greater than 30° and rib vertebral angle difference (RVAD) greater than 20° (Figure 2) are both associated with a high risk of curve progression. Curves with Cobb less than 30° or RVAD less than 20° can be observed with regular follow up and imaging. Curves greater than this can be treated with serial casting in the first instance (Mehta casting). This requires serial casting over several months, and often years. Casts are worn continuously and changed every 3 months or so, with a few days out of cast periodically for skin care. Treatment is resource intensive for patient, parents and healthcare staff.

Curves which progress, despite cast treatment, may require surgical treatment. One of the goals is to maximize growth, particularly in the thoracic spine to maximize lung development. This may be achieved with a 'growing rod' spinal instrumentation system. Growing rod treatment traditionally requires a return to theatre at regular intervals for rod lengthening, which interferes with schooling. Alternatively, MAGEC (*Mag*netic *E*xpansion *C*ontrol) rods may be used. This system allows lengthening via an actuator to drive a mechanism contained in the rod, avoiding repeat surgeries. This is currently approved by NICE for NHS use, but evaluation of this technology is ongoing.

Late-onset idiopathic scoliosis

The cause of idiopathic scoliosis is un-known, but there are a number of theories regarding the cause late-onset idiopathic scoliosis; genetics, melatonin signalling pathway abnormalities and calmodulin dysfunction have all been suggested to play a role, There is an established genetic association with an 11% incidence of scoliosis in first-degree relatives, although concordance in monozygotic twins can be as low as 1%.[1]

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