# Scoliosis: a review

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## Abstract

The normal spine has a straight profile when seen from behind. Scoliosis occurs when this profile is deformed by a lateral curvature which may appear in one or more segments. This curvature is associated with rotation and wedging of the vertebrae. In addition to the spinal curvature there may be prominence of the thorax and or shoulder blade unilaterally or asymmetry of the shoulders and pelvis. The majority of cases present in adolescent females with no obvious cause. Spinal deformity presenting in younger children is much more likely to have an identifiable cause. Treatment and management of scoliosis is aimed at identifying those curves which are at risk of progression, monitoring and treating if necessary. This article details the different types of scoliosis, risks of progression and principles of assessment and management.

**Keywords** investigation; natural history; risk factors; scoliosis; surgery

## Introduction

Scoliosis is a structural three-dimensional deformity of the spine defined by a lateral curvature of more than 10 degrees. The development and progression of scoliosis is related to growth and is accelerated at the periods of growth spurts. Scoliosis may be structural or non-structural. The latter can be caused by conditions such as lower limb disorders resulting in limb length discrepancy or hip dysplasia, limb deficiency syndromes and herniated discs in the older child. This type of spinal deformity is managed by treating the primary cause.

Scoliosis can also be classified by cause into idiopathic or secondary. Idiopathic scoliosis is further classified into infantile, juvenile and adolescent types or early and late onset. Scoliosis can also be secondary to congenital disorders, neuromuscular conditions, tumours, trauma or syndromic conditions. They are also classified according to the side of occurrence into left or right sided curves.

The prevalence of scoliosis in the UK has been established in a survey of 10,000 children in Edinburgh as 1.3/1000 in those under 8 years and 1.8/1000 in those over 8 years of age. In the former group the incidence was similar in both sexes but in the latter group girls outnumbered boys by almost 3 to 1. These figures are similar across the world with a few exceptions.

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A scoliosis has a primary curve and a secondary curve. Each curve has a convex side and a concave side. The primary curve is usually stiffer. The curve site is classified according to the location of the apical vertebra in the coronal plane – cervical, thoracic or lumbar. A junctional curve (cervicothoracic, thoracolumbar or lumbosacral) has the apical vertebra at the interface between these two areas. The apical vertebra is the most rotated vertebra in the curve. The end vertebrae are the most cephalad and caudal vertebrae whose superior and inferior surfaces respectively, tilt maximally towards the concavity of the curve. The curve size is assessed by measurement of the Cobb angle (Figure 1) which is usually described in the direction of the concavity. This is done by drawing lines perpendicular to the transverse axes of the upper and lower end vertebrae. Where these lines intersect is the Cobb angle. The Cobb angle is useful is assessing the initial curve, charting the increasing magnitude of the curves and also in helping to decide when operative intervention may be most beneficial to the child. Previously this was done using a device called the Cobbometer but can now be done with computer software. The curves can be further classified according to their apical location as indicated below:

- Cervical C1-C6
- Cervicothoracic C7-T1
- Thoracic T2-T12
- Thoracolumbar T12-L1
- Lumbar L2-L4
- Lumbosacral L5-S1

The successful treatment of scoliosis to a large extent depends on a good understanding of the natural history of the curve and the consequences of not treating it. This must be tempered by an appreciation of there being no single natural history but rather a multitude of natural histories depending on the type of curve.

**Box 1**: What is scoliosis and who gets it?

- Scoliosis is a three dimensional deformity involving lateral curvature and rotation
- F:M Prevalence more than over 8 years
- Curve site dependent on location of apical vertebra

## Natural history of scoliosis

Scoliosis is known to present with increased frequency in siblings and in children of patients with scoliosis. At initial presentation, the primary focus usually lies in distinguishing between idiopathic and non-idiopathic causes. As expected non-idiopathic causes tend to present earlier, progress much more rapidly and can present with neurological symptoms.

The most common presentation is deformity noticed either by the patients themselves, their immediate family or friends either at home or in social surroundings. The deformity noticed could be either of the spine/prominence of the ribs or asymmetry of the pelvis/shoulders. Breast asymmetry may be noticed by the adolescent female patient.

In a majority of patients pain is not a usually a reason for presentation but is noted as back pain or rib prominence pain.





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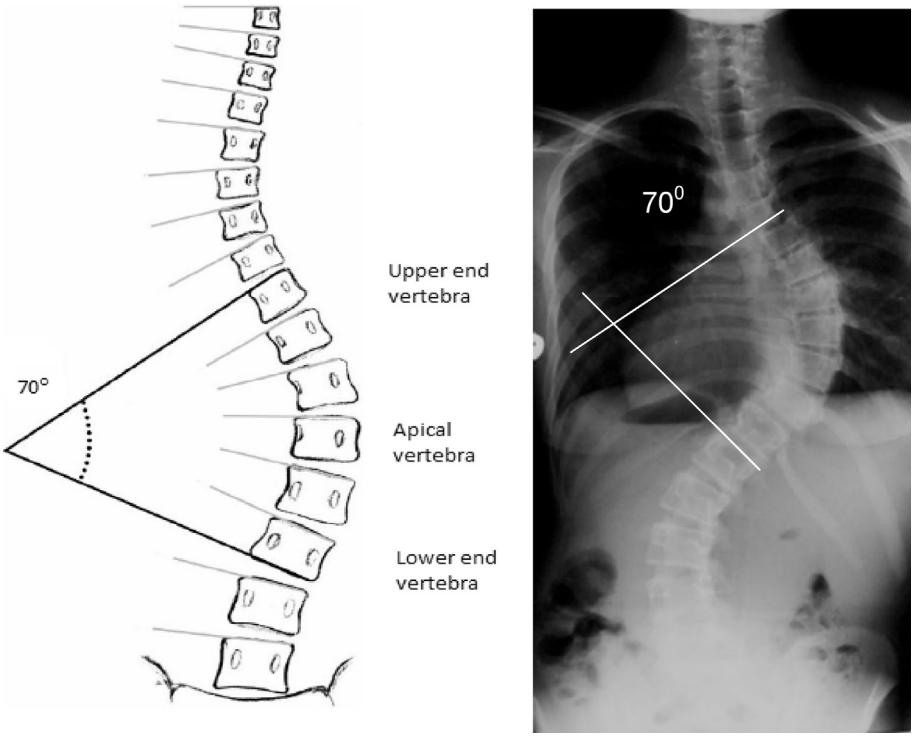


**Figure 1** Cobb angle. The two 'end' vertebrae are the last vertebrae whose end plates are tilted into the concavity. The Cobb angle is measured between a line drawn along the upper end plate of the superior vertebra and another along the lower end plate of the inferior vertebra. Rotation may be estimated from pedicle asymmetry, the more marked the asymmetry the greater the vertebral rotation (normally maximal at the apex).

The presence of pain should prompt the clinician to rule out spinal infection particularly if fever is also present. Night pain in one constant area is unusual and should prompt imaging to rule out a spinal tumour. Other important features to keep in mind are balance and gait disturbance, bowel and bladder control or lack of and any other neurological deficit. These should alert the examiner to look for other pathological causes including tumours (both intra and extradural) and other central causes (e.g. syringomyelia).

Curve progression needs to be carefully monitored. The risk of curve progression depends on timing of growth spurts and also on the amount of growth remaining in each individual patient. In younger children (particularly those less than 10 years of age) lung volume growth may be impaired and untreated severe or progressive scoliosis can lead to or worsen chronic restrictive respiratory disease.

The perception of the patient regarding their appearance is also an important factor in management planning as dissatisfaction with appearance could lead to psychological impairment in the future.

The long term outcomes of scoliosis are mainly based on observational studies which include heterogeneous groups of patients. This calls into question the conclusions drawn from these populations. It is generally felt that curves over 90 degrees are associated with an increased risk of mortality and morbidity. In congenital or early onset curves these consequences can be devastating if not treated early. Adult scoliosis can either develop de novo usually secondary to degenerative disease or result from untreated or missed adolescent deformity. The aim of treatment in all cases is to prevent curve progression. The different types of scoliosis are discussed below:

*Congenital curves*: the anomalies causing congenital scoliosis are present at birth but may take many years to become evident. Genes from the *HOX* group are responsible for many cases of congenital scoliosis.

These are classified into failure of formation, failure of segmentation and mixed groups. They are further sub-divided into anterior formation failure, posterior formation failure, lateral formation failure, anterolateral formation failure and anterocentral formation failure depending on site of pathology (Figure 2). The classification provides a guide to the natural history of these curves. As a general principle, failures of segmentation have a more benign prognosis whilst failures of formation and mixed anomalies have a poorer prognosis. Anomalies at junctional areas such as the thoracolumbar junction also tend to fare worse. Generally, the worst curves occur with a unilateral unsegmented bar with a contralateral fully segmented hemivertebra. Other factors include the type of anomaly, site of anomaly and age of onset. These curves should be referred early for specialist opinion. These anomalies are commonly associated with neuroaxis, cardiac and urogenital anomalies and are increasingly identified during prenatal ultrasound examinations. Congenital scoliosis ranges from the trivial to catastrophic depending on the capacity to progress (and in kyphosis, the capacity to cause spinal cord compression and paralysis). Children diagnosed with congenital scoliosis should undergo Magnetic Resonance imaging (MRI) of spinal cord and brain as approximately 20% are associated with abnormalities




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