Disorders of the skeleton affecting the spine and rib cage fall broadly into the categories of scoliosis (lateral curvature of spine), kyphosis (backwards curvature), lordosis (forward curvature), and pectus (breast bone) abnormalities. The degree of lateral curvature in scoliosis is expressed by the Cobb angle, and is calculated from a standing X-ray as shown in Figure 1. There must also be a rotatory element.

The existing classification of scoliosis is via causation: congenital (seen at birth), neuropathic (caused by neurological or neuromuscular problems such as muscular dystrophies or polio), and syndromic (associated with other conditions, eg, Marfan and Klippel Feil syndrome). Each has particular consequences for breathing. However, by far the most common subtype is idiopathic scoliosis (75%), and the majority of these curves develop in adolescence. Idiopathic means the cause is not known.

Adolescent idiopathic scoliosis (AIS) is diagnosed between the ages of 10 and 18 years. In total there is an incidence of 3% for curves between 10° and 20° and 0.3% incidence for curves greater than 30°. The female to male ratio of curves greater than 30° exceeds 10:1.

Juvenile idiopathic scoliosis occurs in children aged 4 to 10 years and comprises 15% of scoliosis cases. It is more likely to progress than adolescent-onset curves, and can be associated with spinal cord conditions such as syringomyelia and Arnold Chiari syndrome.

Infantile scoliosis occurs after birth but before the age of four years and is relatively rare, with an incidence of 4% of scoliosis cases. In distinction to adolescent-onset and juvenile-onset scoliosis, it is more common in boys, and the curve is more likely to be to the left side of the chest, than right.

These early-onset curves, in particular, may also be linked with pulmonary hypoplasia (under-developed lung) and thoracic cage abnormalities, increasing the risk of long-term lung and heart complications. Congenital scoliosis often occurs as a result of failure of normal vertebral development during the fourth to sixth week of pregnancy. It is associated with congenital cardiac defects in approximately 10% of cases.

For scoliosis, these curves are related to underlying neurological or neuromuscular conditions such as cerebral palsy, muscular dystrophies, and spinal muscular atrophy. Curves, in contrast to idiopathic curves, can be more rapidly progressive and may advance after maturity. Furthermore, pulmonary function will be related to chest wall restriction but additionally by underlying respiratory muscle weakness if present.

Acquired structural scoliosis may occur as a result of thoracic surgery, traumatic onset spinal cord paralysis, a tumour affecting the thoracic spine, or radiotherapy to the spine if these conditions occur in childhood or adolescence, before spinal growth is complete.

How can scoliosis affect breathing?

The major effect of skeletal chest wall and neuromuscular disorders on pulmonary function is to cause a restriction in lung size, that is called a restrictive ventilatory defect. Any significant scoliosis or kyphosis results in a loss of height, so that arm span instead of height is required to predict normal lung volumes. In general, individuals who have a thoracic Cobb angle greater than 70° are subject to significant lung size limitation. In children with early-onset scoliosis, significant ventilatory limitation is unlikely if curve is...
Lung volumes

The simplest breathing tests measure the total volume of air breathed out in one second (forced expiratory volume, FEV1) and the total volume breathed out after a maximum breath in (forced vital capacity, FVC) – see Figure 2.

While both scoliosis and kyphosis diminish lung volumes, a lateral (sideways) curvature has a more profound effect on chest wall functioning. Restriction implies that both FEV1 and FVC are reduced in proportion, so the FEV1/FVC ratio remains normal. This distinguishes scoliosis from conditions such as asthma and chronic obstructive pulmonary disease where FEV1 is disproportionately reduced compared to FVC. A significant obstructive ventilatory defect is rare in adults with scoliosis and kyphosis, unless the individual has coexistent asthma, chronic obstructive pulmonary disease, or upper airway obstruction. However, in some patients with scoliosis and lordosis, bronchial torsion (twisting) or bronchial compression by adjacent vertebrae can occur. This is an important catch that measuring lung volumes can help us identify. If bronchial compression or torsion is suspected a computerised tomography (CT) scan of the chest can be done to help confirm this. The relationship between breathing impairment and the extent of curvature is complex and cannot be predicted accurately from the Cobb angle alone. The four underlying major determinants of a reduced FVC are the number of vertebrae involved in the curve, the higher the position of the curve, the Cobb angle, and the degree of loss of normal thoracic kyphosis. In paralytic scoliosis, lung volumes are reduced not only by chest wall restriction, but also by inspiratory muscle weakness.

Gas transfer or diffusing coefficient (KCO) is a measure of the ability of the lungs to transfer oxygen into the blood stream. This tends to be raised in scoliotic patients, because extra-thoracic compression squeezes more air than blood out of the lungs, thereby decreasing accessible alveolar volume. Importantly a low KCO value is suggestive of problems such as pulmonary hypertension (high pressure) in the pulmonary circulation or intrapulmonary disease, so should be investigated further.

Chest wall mechanics

‘Mechanics’ seems like an odd word to use when applied to breathing but is a physiological term describing how difficult it is to expand the chest and take breaths. ‘Compliance’ indicates how stretchy the chest wall is, and scoliosis can make it stiffer. Chest wall compliance (CCW) is an important determinant of lung volumes and the work of breathing. Individuals with a Cobb angle of less than 50° experience a minimal reduction in CCW, whereas CCW is likely to be significantly reduced if the Cobb angle is greater than 100°. A direct relationship between Cobb angle and CCW is not seen in patients who have neuromuscular disorders, as respiratory muscle weakness contributes independently to chest wall stiffness. Alteration in chest wall properties cannot solely be attributed to the mechanical condition of scoliosis, as a decrease in CCW has been found in patients affected by chronic respiratory muscle weakness in the absence of a scoliosis.

Lung compliance

Although lung expansion is compromised by chest wall properties, primary lung pathology is unusual in adult patients who have idiopathic scoliosis. However, lung compliance is reduced because small lungs are more difficult to expand. A simple analogy is blowing up a balloon – expanding it initially is much harder than increasing its volume once it has grown to the size of say, an orange. The lung is comprised of small air sacs (alveoli), and changes in pulmonary characteristics largely arise from an alteration in alveolar forces caused by chronic breathing at low lung volumes. In neuromuscular patients, collapse of some of the alveoli or larger lobes of the lung may complicate the picture as the respiratory muscles fail to expand all areas of lung. However, in early-onset scoliosis, failure of lung development (pulmonary hypoplasia) and undergrowth of the pulmonary vascular bed may occur. Diaphragm weakness and loss of lung ‘stretch’ may inhibit alveolar development in foetal and early life, causing further loss in lung volume and gas transfer ability. Recurrent pneumonia may occur in neuromuscular patients who have weakness of the swallowing muscles, or an ineffectual cough. Pulmonary scarring is also seen in patients who have old tuberculosis, and these individuals may have areas of lung damage that can lead to frequent infections (bronchiectasis). Cystic lung changes affect some individuals with neurofibromatosis or Marfan syndrome.

Breathing during sleep

During sleep we rest our muscles, including most of our breathing muscles, instead relying on our diaphragm. In addition our ‘drive’ to breathe from the brain is reduced, especially during rapid eye movement (REM) sleep, resulting in more gentle breathing at night, but in people with moderate and severe scoliosis this process can be magnified (nocturnal hypoventilation). In individuals with respiratory muscle weakness and chest wall restriction, nocturnal hypoventilation in REM sleep is seen when FVC falls below 60% predicted and tends to extend to non-REM sleep, ie, the whole night when VC is less than 40%. In a study of patients with nocturnal hypoventilation as a result of mixed respiratory muscle and chest wall disorders, 70% progressed to daytime ventilatory failure within 12 months and 90% within two years of the first appearance of symptomatic nocturnal hypoventilation. Nocturnal hypoventilation is therefore an indication to start
breathing support at night with nocturnal non-invasive ventilation (NIV).

How can you measure breathing?

Lung volumes should be measured as shown in Figure 2, or in a pulmonary function lab. Arterial blood gas measurement (O2 and CO2 level measured in an arterial blood sample), and assessment of respiratory muscle strength using mouth pressures are helpful, particularly in the group who has neuromuscular disease. Mouth pressures are measured by a simple inspiratory and expiratory test. Cough efficacy can also be measured by blowing into a mouthpiece device.

A fall in FVC greater than 15% predicted on lying in the supine position (flat on your back) indicates significant diaphragm weakness. A high daytime CO2 level is associated with an inspiratory mouth pressure less than 30% predicted. As well as inquiries about daytime breathlessness and exercise tolerance, individuals should be asked about symptoms of nocturnal hypoventilation (morning headache, poor sleep quality, frequent wakening from sleep, nocturnal breathlessness), and if any are present, the individual should be referred for monitoring of respiration during sleep - a sleep study carried out by Respiratory Medicine or Sleep Hospital departments. Simple overnight measurement of oxygen level (oximetry) can be done at home using a clip on the finger or ear. More detailed tests involving assessment of carbon dioxide level and respiratory pattern can be done in hospital overnight.

Who is at risk of breathing problems?

The good news is that the vast majority of individuals who have a thoracic spinal curve will not develop cardiorespiratory problems as most have small curves, that are more likely to be adolescent in onset, and therefore do not require long-term respiratory follow-up. But clearly, it is important to be able to identify the small minority at risk of problems so that appropriate monitoring and timely therapeutic intervention are carried out. Decades ago heart and lung failure were the primary cause of death in patients with severe idiopathic thoracic scoliosis. This is now hardly ever seen as treatments such as non-invasive ventilation are highly effective. Factors to look at to predict those at risk are firstly, age at onset of the scoliosis. In patients who developed cardiorespiratory problems attributable to their scoliosis, 90% had an early-onset curvature. A VC of 50% predicted is an important cut-off figure, as those with a VC less than 50% predicted at presentation are much more likely to develop respiratory decompensation than those who have larger lung volumes. In an historic untreated cohort with idiopathic scoliosis followed for 20 years, respiratory failure occurred in 25%, all of whom had a VC less than 45% predicted and a thoracic Cobb angle greater than 110°.

What can be done to help breathing?

For people with scoliosis and breathlessness as a result of chest wall restriction the first step is to see your GP who can measure your breathing or refer you to a respiratory medicine department to be assessed in more detail.

If there is a significant restrictive ventilatory defect and breathlessness is problematic, a pulmonary rehabilitation course can be helpful. This involves an exercise programme tailored to personal needs, coupled with advice on breathing control and healthy living. There is good evidence that pulmonary rehabilitation, particularly the exercise component, reduces breathlessness and enables the individual to walk further. The specific programme will take into account the scoliosis, and problems with any pain or truncal (torso) asymmetry. It is crucial to continue the exercise after the course ends, otherwise the benefits are lost. Your GP or respiratory physician can refer you to a pulmonary rehabilitation course either in the community or at a local hospital. Courses usually last around 6-8 weeks, with twice weekly attendance.

If the vital capacity is less than 50% predicted and/or there are symptoms of sleep disturbance such as waking with breathlessness or morning headaches, a sleep study may be carried out. In those with very small lungs this may show nocturnal hypoventilation during sleep.

How can I keep healthy?

Mostly this is common sense. Importantly don’t smoke, exercise regularly and keep to ideal body weight. Even putting on a little weight can make people with scoliosis breathless. Influenza vaccination is recommended in those over 65 years, or if you have an additional breathing problem such as asthma. If possible, keep an eye on your vitamin D level and take supplements if this is low. Many people are Vitamin D deficient so it could be argued that all scoliosis patients should take Vitamin D supplements. A bone densitometry scan is helpful in showing if individuals have osteoporosis, as this can be treated with bisphosphonate tablets +/- calcium, as advised by your GP. Upper respiratory tract infections are usually viral and settle with symptomatic measures such as paracetamol and rest. If symptoms don’t settle within a few days and are associated with production of green or yellow phlegm and increased breathlessness, then a check-up with your GP is advisable as an antibiotic might be required.