Scheuermann's disease

Synonyms: juvenile kyphosis, Scheuermann's kyphosis

Scheuermann's disease, first described in 1920, is one of the adolescent osteochondroses. It is essentially osteochondrosis of the thoracic vertebral bodies which leads to wedge-shaped vertebrae[1] . It causes increased kyphosis, poor posture and backache. It is the most common cause of structural kyphosis in adolescents[2] [3] .

It is confusing that Scheuermann also described changes that can occur in the lumbar disc spaces - now called juvenile disc disorder. This is a quite separate condition.

There is a separate article giving an overview of Osteochondroses.

Pathophysiology

- Osteochondroses all involve a defect in ossification. In Scheuermann's disease there is a defect in the secondary ossification centres of the vertebral bodies. A few vertebral bodies may be involved or the whole thoracolumbar spine.
- The cause is unknown but hereditary factors are thought to play a part in the pathophysiology. However, no definite mode of inheritance or genetic marker has been established so far[4] .
- Other factors include a correlation between Scheuermann's disease and taller height, as well as an association with higher BMI and increased levels of growth hormone[5] . One study has suggested that the increased weight and height in those with Scheuermann's disease are not part of the pathogenetic mechanism, but suggests that these are secondary effects[6] .
- Mechanical factors and trauma have also been cited as possible causes; the aetiology is probably multi-factorial[1] .

Epidemiology

- Usually presents in children aged 13-16 years.
- The incidence is variously reported between 0.4% and 8% and some studies suggest it is more common in males, although other studies contradict this [1] .

Presentation [5]

History

- Poor posture with increased kyphosis (usually noticed by parents).
- Dull, non-radiating pain around the apex of the kyphosis.
- Neurological symptoms are rarely the presenting feature but there is an increased risk of disc degeneration and cord compression with age.

Examination

- It is normal to have a degree of kyphosis. Any degree of kyphosis of more than 50° is considered abnormal [7] .
- Kyphotic deformity may be progressive.
- Upper thoracic kyphosis is best seen in the forward flexed position; lower thoracic kyphosis may be seen at the thoracolumbar junction.
- There is reduced flexibility of the spine.
- There may be tenderness above and below the apex of the kyphosis.
- A third of patients will also have a mild scoliosis, spondylolysis of L5 or lumbosacral spondylolisthesis.
- You should carry out a neurological examination but neurological signs are rare.

Differential diagnosis

Includes:

- Postural kyphosis (known as postural round back deformity; kyphosis is flexible).
- Ankylosing spondylitis.
- Glucocorticoid-induced osteoporosis in Cushing's syndrome, which can lead to thoracic kyphosis.
- Skeletal dysplasia.
- Congenital kyphosis (failure of segmentation).
- Spinal tuberculosis (consider if there is atypical presentation).

Investigations

- X-ray of the spine is required for diagnosis of Sheuermann's disease, as it is defined by the appearance of hyperkyphosis and 'wedge-shaped vertebral bodies with anterior wedging of at least 5° or more in at least three adjacent vertebral bodies'. Anteroposterior (AP) and lateral views are required, in a standing position with arms raised by 90°. The whole thoracolumbar spine and pelvis should be imaged. Changes may also include [1] :
    - Narrow intervertebral disc spaces and lengthening of the vertebral bodies.
    - Irregular upper and lower vertebral endplates.
    - Schmorl's nodes - multiple herniations of the nucleus pulposus of the vertebral plates.
- MRI may also be necessary to exclude intraspinal abnormalities or cord compression at the apex of the kyphosis.

Management [5]

The first-line treatment is medical and includes rehabilitation and bracing. The earlier the start of treatment, the better the outcome, which highlights the importance of early diagnosis. Surgery is uncommon and must be limited to severe involvement after failure of conservative treatment [8] .

- For mild disease, avoidance of strenuous activity and weight-bearing may be all that is needed. Observation and X-ray follow-up are carried out.
- Physiotherapy may reduce pain, especially if the disease affects a short segment, but does not affect the progression of kyphosis.
- In more severe disease, if the bone is still immature, there is some flexibility in the kyphosis and the angulation is <65°, a spinal brace may be used. Braces may be advised for several years but although this can achieve improvement with reversal of wedging, this is partly lost in 30% of patients, even when used correctly.
- Surgery is usually only used if there is neurological deficit, uncontrolled pain, unacceptable cosmetic appearance or documented progression but is not indicated solely by the degree of kyphosis[2] .
- Cord decompression is carried out for neurological deficit and spinal fusion techniques are used for kyphosis correction and pain control.
- Over the period of a decade there has been an increase in spinal surgery for Scheuermann's disease, and a higher proportion of operations are in adults. Although there is a high risk of complications this is significantly less with new instrumentation techniques and an all-posterior approach[9] .

Complications

- Chronic back pain.
- Progressive and permanent deformity.
- Neurological deficit.
- Cardiorespiratory problems.

Prognosis

- Mild-to-moderate Scheuermann's disease rarely requires bracing or surgery.

- A Finnish study reporting on the 37-year follow-up of 49 people with Scheuermann's disease concluded that they have an increased risk of back pain and they are more likely to report difficulty carrying heavy loads and going upstairs without a rest. However, there was no correlation between the degree of their kyphosis and self-reported quality of life, quality of health or back pain [1] .
- Neurological and cardiorespiratory complication risks are very low.

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