

PROGRESSIVE NONINFECTIOUS ANTERIOR VERTEBRAL FUSION

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We reviewed 26 patients with progressive spinal kyphosis due to anterior fusion between the vertebrae. No patient had back pain or any neurological defect.

The kyphosis appeared to be progressive until the fusion had included all of the disc. Progression was faster during the adolescent growth-spurt. Kyphosis increased with the number of discs involved, from one to six, and the extent of fusion within each disc. In six of the nine cases treated by spinal bracing, progression of the kyphosis was arrested.

Progressive noninfectious anterior vertebral fusion (AVF) is a rare disease of uncertain aetiology, which was first described fully by Mosenthal (1931). The first change is narrowing of the anterior disc space with approximation of the anterior corners of the vertebral bodies. This may involve one or more disc spaces (Fig. 1). Anterior fusion of the vertebrae extends posteriorly in involved spaces, eventually involving the whole of the body of the vertebra (Fig. 2).

A total of 80 cases, including our series, have now been reported; 60% were female.

We have reviewed 26 cases seen in Copenhagen from 1947 to 1988.

PATIENTS AND METHODS

Of the 35 cases recorded since 1947 at Copenhagen University Hospital and the former Orthopaedic Hospital of Copenhagen, we were able to review the notes and radiographs of 26 patients. Eight were seen on only one occasion. Their mean age at first consultation was 20.7 years (4 days to 84 years) and mean follow-up for the 18 patients kept under review was 13.3 years (1 to 25). None of the patients had presented with pain or neurological

symptoms, but some patients developed dorsal pain during follow-up. No patient was operated upon, but nine were treated by spinal bracing.

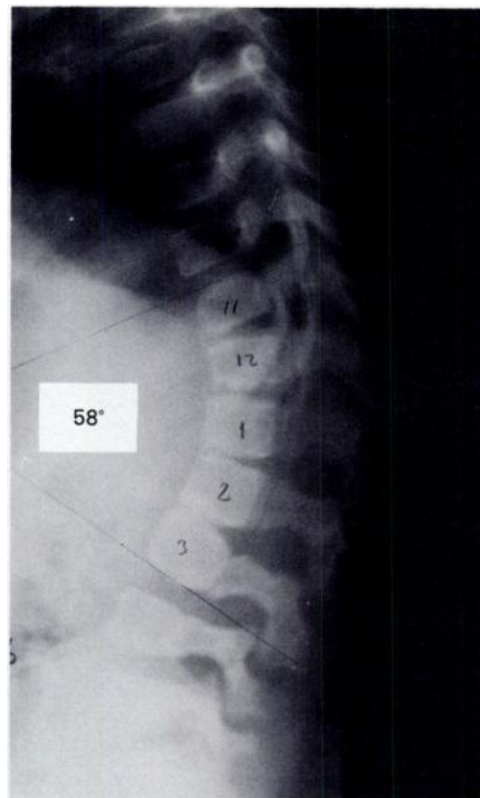


Fig. 1

Lateral radiograph of a one-year-old girl showing a 58° kyphosis due to early disease in four disc spaces at the thoracolumbar junction.

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We recorded the number of vertebrae involved, their vertical height, the extent of anterior fusion, the angle and the progression of kyphosis.

RESULTS

Ten patients had involvement of one disc, five patients had two discs affected, four had changes at three levels

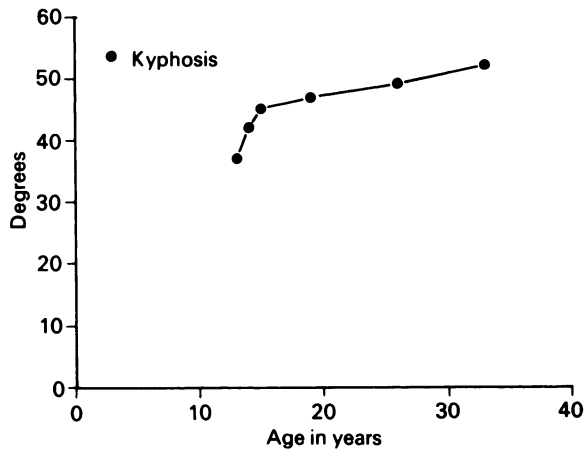


Fig. 2a

Case 1. Changes in angle of kyphosis during 21 years follow-up.

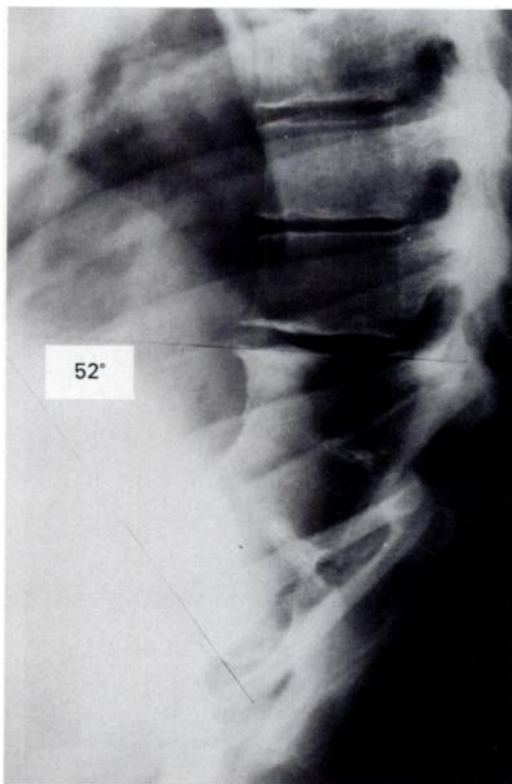


Fig. 2b

Case 1. Radiograph at 33 years of age, showing 75% fusion of the discs.

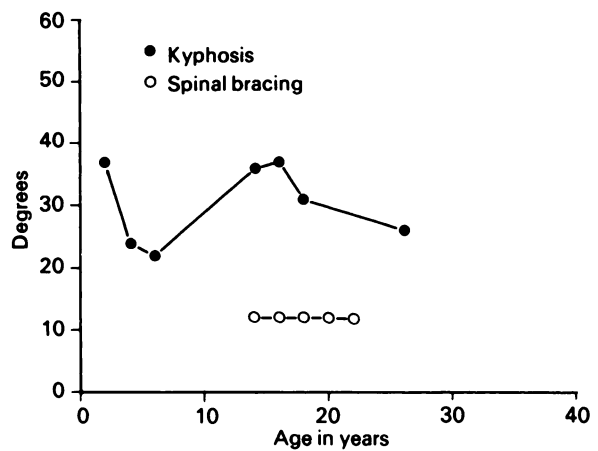


Fig. 3a

Case 2. Changes in the angle of kyphosis during follow-up, showing the influence of spinal bracing.

and four at four. Five discs were affected in two patients and six in one.

The changes were in the thoracic spine in 12, at the thoracolumbar junction in another 12, and in the lumbar region in two. The extent of intervertebral disc space fusion was estimated as 0%, 25%, 50% or 100%. The most extensive degree of fusion was recorded in patients with more than one disc space involved.

At the first consultation the mean kyphosis was 25.5° (-3° to 78°). At the end of follow-up, the mean kyphosis was 37.7° (3° to 120°). As shown in the case histories, some reduction in severity or decrease of the kyphosis was seen in six of the cases treated by spinal bracing.

ILLUSTRATIVE CASE REPORTS

Case 1. Typical development. A boy was first seen at the age of 12; two disc spaces were involved. No clinical records were available but radiographic follow-up spanned 21 years (Fig. 2a). There was rapid progression of kyphosis during adolescence, but some slower deterioration has continued into adult life. At the latest review, at aged 33 years, there was 75% fusion of the discs (Fig. 2b).

Case 2. Early development and bracing. A one-year-old girl was referred with kyphoscoliosis, and treated in a plaster bed. Radiographs at two years of age showed typical involvement of three discs at the thoracolumbar junction. No further treatment was given until the patient was 14 years of age, when spinal bracing was started (Fig. 3a). With this treatment, the kyphosis decreased from 37° to 26°. At 23 years of age she stopped wearing her corset because she was pregnant (Figs 3b and 3c).

Case 3. Spinal bracing. A ten-year-old girl with involvement of one disc was treated with a Boston brace for five years. During this time the kyphosis decreased and then became stable (Fig. 4).

Case 4. Spinal bracing. A 15-year-old girl with involve-

Case 2. Radiographs at 16 and 23 years of age.

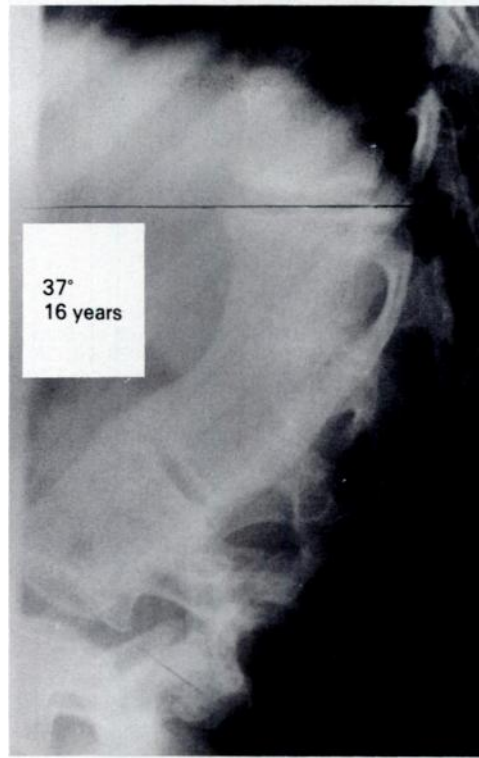


Fig. 3b



Fig. 3c

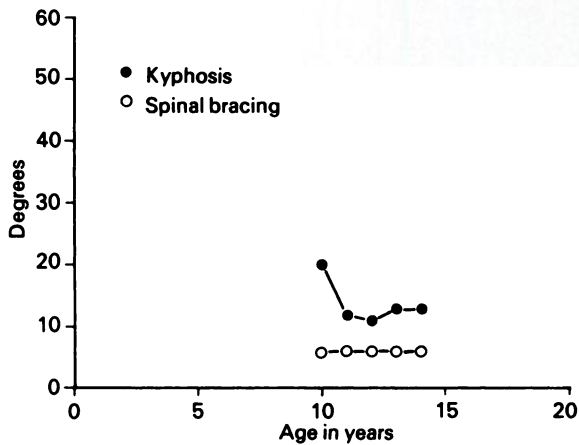


Fig. 4

Case 3. Changes in kyphosis during a period of spinal bracing.

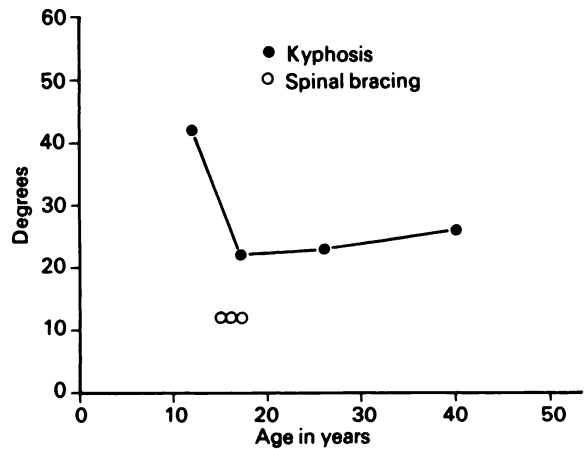


Fig. 5

Case 4. Changes in kyphosis during and after a period of spinal bracing.

ment of one disc was treated in a plaster bed and then for three years in a brace with good effect (Fig. 5).

DISCUSSION

Anterior noninfectious vertebral fusion seems to be unique in its radiological presentation and course, resembling no other disease of the spine. Some authors consider that it is related to Scheuermann's disease (Lindemann-Kiel 1931; Knutsson 1949), but others have

implicated infection or trauma (Hueck 1930; Lindemann-Kiel 1935). Morin et al (1985) attempted, but failed, to reproduce the changes by means of hypoxia in mouse embryos, while Smith et al (1986) compared it with changes induced by thalidomide and found some similar-

ities in the late stages. We have found no connection with these causes, and think that it is probably a congenital disease. Kharrat and Dubousset (1980) report two instances of familial involvement: in one family a grandmother, her daughter and two grandchildren were affected, and in the other a brother and sister both had the condition.

Because of the chronic nature of the disease, very long follow-up is necessary to assess patterns of progression. In our series, increase in kyphosis was related to the number of vertebrae involved and the extent of fusion of the discs. We found no correlation with the height of the vertebral column. None of our patients had any other significant bony malformations.

Conclusions. Progressive anterior vertebral fusion can sometimes be diagnosed from radiographs taken shortly after birth because of the characteristic anterior defect in the involved vertebrae (see Fig. 1). The resultant kyphosis tends to progress rapidly during adolescence and more slowly during adult life. Anterior fusion of disc spaces progresses backwards until the bodies of the vertebrae are fully united.

Patients usually present with a progressive kyphosis with no significant pain or neurological symptoms. Spinal

bracing, appears to reduce and sometimes even reverse the kyphotic deformity. Treatment and review should continue throughout adolescence, and into early adult life, though once there is 100% fusion of all the involved disc spaces, the deformity does not appear to alter.

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