DEVELOPMENT OF PATHOLOGICAL LUMBAR KYPHOSIS IN MYELOMENINGOCELE

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We analysed the cases of lumbar kyphosis in 151 (21%) of a series of 719 patients with myelomeningocele. Three different types were distinguished: paralytic, sharp-angled and congenital. In a cross-sectional and partly longitudinal study the size and magnitude of the kyphosis, the apex of the curve and the level of paralysis of each group were recorded and statistically analysed.

Paralytic kyphosis (less than 90° at birth) occurred in 44.4% and increased linearly during further development. Sharp-angled kyphosis (90° or more at birth) was present in 38.4% and also showed a linear progression. In both types, progression seemed to depend also on the level of paralysis. Congenital kyphosis occurred in 13.9% and we could find no significant factor which correlated with progression.

Lumbar kyphosis in myelomeningocele gives rise to a variety of clinical problems. Closure is more difficult (Brocklehurst 1976), and tension on the anaesthetic skin tends to cause ulceration with consequent meningitis (Christofersen and Brooks 1985). Abdominal compression leads to respiratory problems (Banta 1987) and paralysis causes stenosis of the ureters with infection of the bladder which jeopardises renal function. Mobilisation and orthotic treatment are extremely difficult (Heydemann and Gillespie 1987).

The reported incidence of lumbar kyphosis in myelomeningocele varies between 1% (Burney and Hamsa 1963) and 46% (Naik, Lendon and Barson 1978). Hoppenfeld (1967) described it as fixed, usually lumbar, and confined to the region of the spinal defect. Three different types have been described: paralytic, sharp-angled and congenital (Cotta, Parsch and Schulitz 1971; Raycroft and Curtis 1972; Sharrard and Drennan 1972; Banta and Hamada 1976; Brown 1978; Leatherman and Dickson 1978; Banta 1990; Lindseth 1990). The paralytic type has a nearly normal spinal curvature at birth, but due to muscle weakness the lumbar spine cannot be stabilised adequately and develops an increasing kyphosis (Raycroft and Curtis 1972; Lindseth 1990) (Fig. 1). The sharp-angled type has a typically rigid curvature of more than 90° at birth caused by the pathological position of the paravertebral muscles (Sharrard 1968; Drennan 1970; Lindseth 1990) (Fig. 2). The congenital type is primarily caused by vertebral anomalies such as an anterior defect of segmentation (Cotta et al 1971; Banta 1990) (Fig. 3).

It is widely agreed that in most cases deformity increases during childhood and that the increase in kyphosis is inevitable even after the completion of growth (Hoppenfeld 1967; Sharrard 1968, 1972, 1973, 1975; Hall and Bobeckho 1973; Banta and Hamada 1976; Sherk et al 1979; Mayfield 1981; Banta 1990). The deformity is more marked in patients with high neurological lesions (Mayfield 1981). Banta and Hamada (1976) reported an annual increase of 3° in paralytic kyphosis and of 8.3° in congenital kyphosis.

Our aim is to present a complete analysis of the incidence, occurrence, progression and possible predictive factors of lumbar kyphosis.

PATIENTS AND METHODS

Between January 1972 and May 1990 we treated 1466 patients at the Department of Paediatric Orthopaedics, University of Heidelberg of whom 719 had a lateral radiograph. In 151 (21%) a lumbar kyphosis was detected. Of these 87 (57.6%) were female and 64 (42.4%) male. The apex ranged from D12 to L5 with a maximum at L2 (Fig. 4) and the level of paralysis from D4 to L5 with a maximum in the lower thoracic region (Fig. 5). Five patients with paralytic kyphosis and one with sharp-angled kyphosis showed kyphoscoliotic deformity. In four cases the type of curvature could not be determined.

The first part of our study was a cross-sectional survey in which one radiograph per patient was included. In patients who had more than one radiograph one film was chosen at random. We studied age, gender, the size of the kyphotic curve in degrees, the apex of the curve, and the level of...
A patient with paralysis below D12 on the right side and below L1 on the left. Radiographs show paralytic kyphosis of 46° at the age of eight months (a), increasing to 60° at the age of nine years (b).

A patient with complete paralysis below D10. Radiographs show sharp-angled kyphosis of 94° at the age of two months (a) increasing to 145° at the age of three years (b).
paralysis. Regression analysis was applied to compare the adequacy of a linear and a logarithmic model. In the second part of the study we used longitudinal data.

**Statistics and data transformation.** To determine the correlation between age and the degree of kyphosis we used non-parametric and parametric procedures. The data were not normally distributed and we therefore first calculated the correlation according to Spearman ($r_s$) for each group and tested whether the coefficients differed significantly from zero. Then age and kyphosis were logarithmically transformed and regression models for both the original and the transformed data were calculated for each

A three-year-old boy with paralysis below L3. Radiographs show congenital kyphosis of 38° due to the anterior defect of segmentation between L2 and L4 (a). At 11 years of age it is unchanged (b).
group. We compared the linear and the logarithmic models obtained for each group. The linear model fitted better in all three groups as judged by the size of the Pearson correlation. Hence we calculated the 95% confidence interval of the mean progression rate per year. We then added an extra independent variable, level of paralysis, as a potential predictor of progression. The results of the equations obtained by the cross-sectional analysis were compared with the longitudinally estimated progression per year. Of 146 patients 76 had at least two radiographs. In one patient with sharp-angled kyphosis the curve in the second film could not be estimated exactly and therefore this case was not used for longitudinal analysis. Based on the data of the remaining 75 patients the mean progression per year with a 95% confidence interval was calculated as:

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\text{progression (°)/year} = \frac{\text{kyphosis 2(°) - kyphosis 1(°)}}{\text{age 2 (years) - age 1 (years)}}
\]

This was then compared with the mean progression rate per year obtained by cross-sectional analysis.

RESULTS

Table I shows the relationship between kyphosis and age for each group. Spearman’s correlation was significant for the lumbar and sharp-angled groups, but not for the congenital group. The gradient of the best-fitting linear equation gives the estimated progression (in degrees) to be expected annually, being 3.4° for the lumbar group and 3.8° for the sharp-angled group.

The first and the second films of those patients who had more than one radiograph were then analysed. The number of patients showing increased rather than decreased kyphosis were compared by the sign test. In the paralytic group kyphosis increased in 20 patients and decreased in eight (p < 0.04). In the sharp-angled group it increased in 24 patients, decreased in five and was unchanged in three (p < 0.001) and in the congenital group it increased in all 15 patients (p < 0.001).

In studying the level of paralysis as a further potential predictor for the development of kyphosis we initially transformed our data as follows:

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\begin{align*}
D4 & = 1234567 \\
D5 & = 89 \\
D6 & = 1 \\
D7 & = 0 \\
D8 & = 1 \\
D9 & = 1 \\
D10 & = 2 \\
D11 & = 1 \\
D12 & = 3 \\
D13 & = 1 \\
D14 & = 4
\end{align*}
\]

We then applied multiple regression with age and level of paralysis as predictors and the kyphosis curve as a dependent variable. Table II shows the results for the first two groups. Kyphosis appeared to increase by 3.2° and 4.2° annually for the lumbar and the sharp-angled groups, respectively. It decreased with the level of paralysis by 3.8° and 4.0° per level in the two groups. The results of the procedure for congenital kyphosis gave no significant predictor.

We also validated our regression models by calculating the progression rate per year for each individual who had been examined at least twice (Figs 6 to 8). The results are given in Table III and show that progression of the kyphos-
sis should be expected in all three groups. In the paralytic and sharp-angled cases it seems to be influenced mainly by the age of the patient and the level of paralysis. For these predictors in neither case does the 95% confidence interval include 0. The age of the patient seems to have a linear influence in both groups. For the paralytic group the estimated yearly progression ranges from 1.7° to 5.1°, and for the sharp-angled group from 2° to 5.6°. The more cranial the level of paralysis the faster the progression in both groups. In the congenital group neither age nor level of paralysis showed a statistically significant correlation with the kyphosis angle. It is of interest, however, that in the patients who could be examined longitudinally (n = 15) individual progression took place.

**DISCUSSION**

The incidence of pathological lumbar kyphosis in the 719 patients in our series was found to be 21%. In spite of the large number of cases our findings cannot claim absolute precision because the number of patients who were excluded influences the accuracy of this figure. This was discussed by Barson (1965) who concluded that his finding of an incidence of 27.5% was too low. We feel, however, that this conclusion is questionable. A suspicion of kyphosis is a clinical diagnosis which requires additional lateral radiographs to verify the presence of the spinal deformity. Hence, our findings probably overestimate the real incidence which would be about 10% rather than 20% if none of the patients without lateral radiographs had kyphosis.

Female patients seem to be slightly more often affected by kyphosis in myelomeningocele although this difference was not statistically significant. This finding is also seen in scoliosis in myelomeningocele. Carstens, Vetter and Niehhard (1990) suggested that there was a link to the hormonal status which loosens the surrounding soft tissues and destabilises the spine.

In all patients high-level paraplegia is the main reason for developing a spinal deformity and three types of kyphosis can be distinguished: paralytic, sharp-angled and congenital. Banta and Hamada (1976) found kyphoscoliosis to be a separate special entity. In our patients there were six cases of kyphoscoliosis. According to our criteria we classified them as the paralytic type in five and as the sharp-angled type in one and we did not therefore consider this deformity separately.

*Paralytic deformity* causes no complications at birth because of the small size of the child. Any progression, and its known consequences, is therefore of major importance for the patient’s future. We found that progression could be expected in all cases. In the paralytic group both linear and logarithmic statistical models fitted our data relatively well,
but the former was better and hence more appropriate. Thus, a yearly progression in a range between 1.7° and 5.1° should be expected. The development of kyphosis was influenced by the level of paralysis; the more cranial the level of neurological deficit the faster the development of the spinal deformity. The mechanism appears to be muscular instability with lack of dorsal bony structures and is similar to that for paralytic scoliosis. Surgery is indicated in cases of progression.

*Sharp-angled kyphosis* seems to be caused by a pathological anterior displacement of the psoas and erector spine muscles which act as perverted lumbar flexors (Drennan 1970). Because of its marked size at the time of birth and its significant kyphosis causes substantial impairment and early surgical treatment is indicated. We found a positive correlation between the size of the curvature and the age of the patient. As in the paralytic cases a linear statistical model seems appropriate. These kyphoses are typically extensive. According to our definition the minimum angle of deformation is 90° and may increase up to 180° when the chest rests on the iliac crests. A yearly progression in a range between 2° and 5.6° should be expected. As in the paralytic cases the level of paralysis seems to have some effect.

In the congenital group cross-sectional analysis showed no statistically significant variables, with no correlation between age and kyphosis or between the level of paralysis and kyphosis. In all patients who were examined longitudinally, however, individual progression occurred. Progress is therefore unpredictable. In our patients we have seen slow as well as fast deterioration which is not dependent on time.

In order to validate our data we compared our cross-sectional analysis with the longitudinal data. This showed a marked overlap of the range of values. It seemed, however, that progression was more rapid in patients who were seen at least twice, compared with those examined only once. This suggests that the more severely affected patients seek medical help more often.

The increase in kyphosis is synonymous with the deterioration in the patient’s condition. Few authors have discussed the progression of kyphosis in myelomeningocele and only Banta and Hamada (1976) give figures. They found paralytic kyphosis to increase by 3° per year and our results confirm this figure. For congenital kyphosis they give a figure of 8.3° per year, but we could not confirm this.

We compared our findings on kyphosis on this with studies on scoliosis in myelomeningocele. Carstens et al (1990) found that the development and progression of scoliosis are mainly influenced by the level of paralysis and the age of the patient. According to Müller, Nordwall and Oden (1994) the rate of progression depends mainly on the degree of the curvature and the age of the patient. Walking capacity seems to be a possible predictive factor for the progression of scoliosis. According to our results the paralytic and the sharp-angled groups were mainly influenced by the age of the patient and the level of paralysis. For these two types therefore the factors which affect prognosis are common to both scoliosis and kyphosis.

Spinal deformities are major problems in the care and treatment of patients with myelomeningocele. An incidence of at least 10% to 21% indicates that pathological lumbar kyphosis is quite common and we recommend that all patients should be examined for this deformity.

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