THE NATURAL HISTORY OF SCOLIOSIS IN MYELODYSPLASIA

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Two hundred and fifty cases of myelodysplasia were reviewed in relation to spinal deformity. Approximately half of the children had, or were expected to develop, curves severe enough to need operations and only 10 per cent maintained completely undeformed spines. The most frequent deformity was scoliosis which could be subdivided into congenital and developmental types. The latter was of mixed aetiology, neuromuscular imbalance and asymmetry of the neural arch both contributing, while in some cases no causative factors could be identified. The best early indicator that developmental scoliosis was likely to appear was a high segmental level of both the neurological deficit and the neural arch defect. Deformity was very unlikely to start after the age of nine years.

In the last two decades infants with the more severe degrees of myelodysplasia have, as a result of energetic treatment, survived in large numbers until later childhood and beyond, and it has become progressively apparent in the older survivors that scoliosis, often severe, is a very common problem. In many instances after a childhood of multiple operations and much treatment, when a stable mechanical state appears to have been reached, progressive scoliosis develops and may convert a walker to a sitter, or a two-handed patient to a one-handed one. This paper considers the frequency and aetiology of such deformity, and discusses those features which might, at an early stage, be of prognostic value in relation to the ultimate onset of spinal curvature.

PATIENTS AND METHOD OF STUDY

A spina bifida clinic was established at the Birmingham Children's Hospital in 1966 in which children with all types of spina bifida and myelodysplasia are seen. The majority of patients arrive as neonates, transferred from nearby maternity units, but the more serious cases are referred from a wide surrounding area, so the material for this review is probably weighted towards the more severely disabled. Because of family movements some patients have inevitably first attended the clinic during the later stages of childhood and these have been balanced by others leaving the district. Nevertheless, the bulk of patients reviewed have been followed from the day of birth until the time of this review, which took place from July 1977 to June 1978. Patients with lesions entirely proximal to the first lumbar vertebra have been excluded since the neurological picture in these children is usually complicated by spasticity, and a further small number who did not attend for review and whose records were inadequate were also excluded, leaving a total of 250 cases for study.

Raycroft and Curtis (1972) divided spinal curves in myelodysplasia into congenital and developmental groups on the basis of whether or not the deformity was caused by malformation of the vertebral bodies; but some curves probably have a mixed aetiology in which developmental factors increase a congenital scoliosis, and within the purely developmental group multiple mechanisms may be at work. In this study congenital malformation of vertebral bodies, neurogenic imbalance of trunk control, malposition of spinal musculature, deformity of the hip and the occurrence of an idiopathic curve in association with myelodysplasia were all investigated as possible causes of deformity.

The following features were noted in every case: the type of spina bifida (sessile, meningocele, myelomeningocele); the levels and nature of the bony defect; the neurological levels, both sensory and motor, and whether partial or complete; the levels, laterality, the Cobb angle and degree of rotation of any lateral curve that was present, whether it was congenital or developmental, and in the latter case the age at which it was first recognised (in the early days of the clinic routine spinal radiographs were not taken); the presence of kyphosis or lordosis; the effect of spinal deformity on overall function; deformity and function of the hips and lower limbs; previous operations; functional level as a walker or sitter; hydrocephalus and its treatment; intelligence; the genito-urinary state; and any other abnormalities.

RESULTS

At review approximately one third of the patients were more than 10 years old, and boys slightly outnumbered girls. Of the 250 patients, 191 had a myelomeningocele, 46 a meningocele and nine a sessile lesion; in four patients the lesion was either a myelomeningocele or a meningocele, but the surgeon closing the lesion had been in doubt.

Table I. Incidence of spinal deformity

Deformity	All cases (*per cent*)	Over 10 years of age (*per cent*)
Scoliosis	54.0	82.5
Kyphosis	22.0	17.5
Lordosis	1.5	12.5
Undeformed	31.0	10.0

Incidence of deformity. Table I shows the incidence of all types of spinal deformity in the group as a whole, and separately in those over 10 years old. Only structural curves have been included; in a few instances where there was doubt whether a curve was structural or

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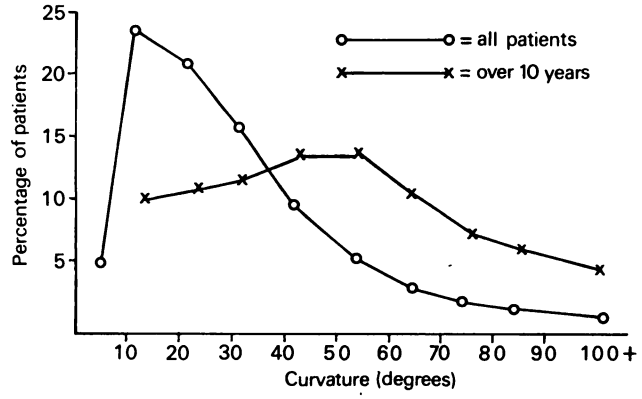

Fig. 1

Severity of lateral curvature measured by the Cobb angle: the shift to the right in those over 10 years of age shows the higher proportion of moderate and severe curves in older children.

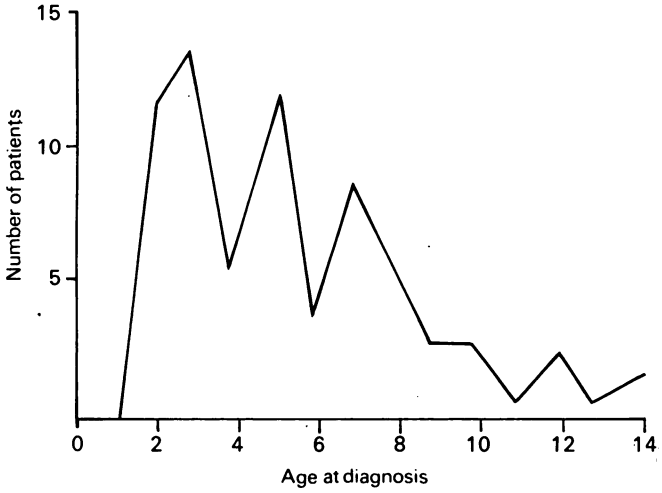

Fig. 2

Age at detection of scoliosis, excluding congenital curves.

postural it was recorded as undeformed. Scoliosis was much more common than other deformities and its incidence increased with age. Over the age of 10 years only one child in 10 was free of spinal deformity. Measurement of the lateral curvature showed that over the group as a whole mild curves predominated, but the severity increased with age and in the older group many curves were moderate or severe (Fig. 1). The progression of curvature was confirmed in those cases where serial standardised radiographs were available and few, if any, curves were static.

Age of onset. Figure 2 shows the age at which scoliosis was first observed, and in many cases it was clear from the magnitude of the curve that it had been present for some considerable time. The true age of onset was therefore undoubtedly younger than indicated in the graph, and there was probably only one genuine case of the onset of a curve after the age of nine years.

Congenital malformation. Figure 3 shows a simple example of congenital malformation of the vertebral bodies, causing a lateral curve; 38 per cent of lateral curves were of this type.

Neurogenic deficiency. A neurogenic contribution was considered to exist in all cases with any neurological anomaly above the first sacral segment, and in those showing neurological asymmetry at any level, even minor differences in power or sensation below the knee. In only two patients was it possible to demonstrate asymmetrical power of muscles acting on the spine in a manner which would cause the observed deformity. There were, however, 45 cases with asymmetric paresis at some level; the incidence of scoliosis in this group was only slightly higher than in the whole series, and the side of curvature bore no relation to the neural asymmetry. When the asymmetry was ignored and the total neurological deficit was expressed as a segmental level of paraplegia, there was a close relationship with the occurrence of scoliosis (Table II): the higher the level, the greater the incidence of lateral curves.

Muscle malposition. Muscle imbalance, though most often caused by change in muscle power, can also result from an abnormal position of muscles. Sharrard (1968) showed that in the most severe forms of neural arch deformity in spina bifida, where the two half-arches are hinged through 180 degrees to open up like the leaves of a book, the posterior spinal musculature is displaced anteriorly with the neural arch, and if innervated takes on an abnormal flexor activity which contributes to the progression of the kyphosis. In less severe deformities the halves of the neural arch project backwards and the

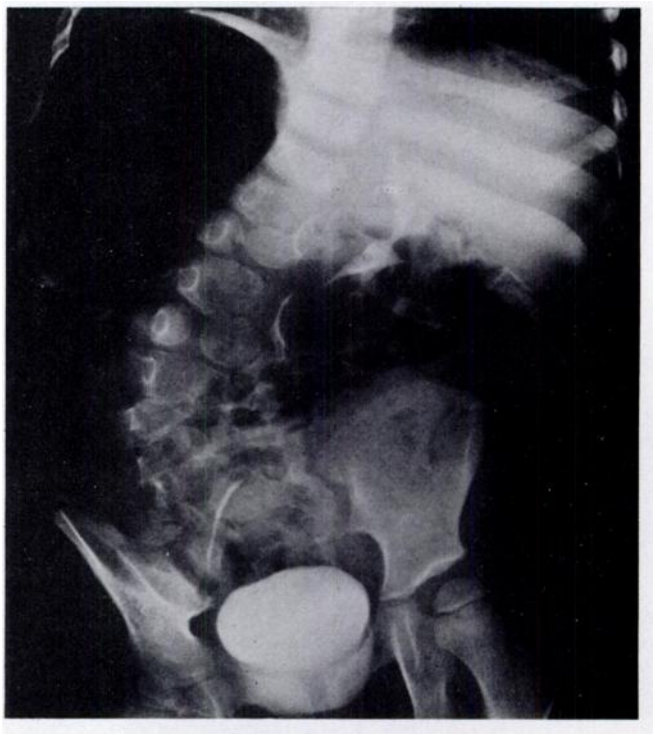

Fig. 3

Congenital scoliosis caused by malformation of the vertebral bodies. This patient had a myelomeningocoele affecting only the left side of the cord. Note also the gross renal anomaly.

musculature is displaced to the side (Fig. 4). If this displacement is asymmetric, and if the muscle is active, it may contribute to the onset, or at least to the laterality, of scoliosis.

The posteriorly projecting half-arches could be seen in many cases of spina bifida (Fig. 5), but it was not easy to demonstrate asymmetry of this malposition since once a curve appeared the accompanying rotation readily mimicked asymmetry of the arch, and, sometimes, in the very young prescoliotic spine the arches could not be clearly seen. Nevertheless, true asymmetry of the arch appeared to be present in 15 per cent of patients (Fig. 6). The upper level of arch defect was easier to see than asymmetry, and the incidence of scoliosis corresponded closely to this level, exactly as it did to the neurological level (Table II). As the neurological and bony defects tend to be closely linked, the relationship of scoliosis to the level of arch defect may be purely coincidental, but the possibility remains that the bony defect, through its effect on muscle action, contributes to the onset and laterality of deformity.

Deformity of the hip. Adduction deformity of one hip will impose a pelvic tilt, up on the affected side, to keep the lower limbs parallel, and an abduction contracture will similarly produce a pelvic tilt down on the affected side. The tilt is usually compensated by a lateral spinal curvature to preserve overall body alignment, and this curvature starts at the lowest mobile level, usually lumbosacral, but higher if the lower spinal segments are rigid, as may often be the case in myelodysplasia. The reverse also occurs when a scoliosis, extending down to the sacrum and thus causing a pelvic tilt, is compensated by adduction of the high hip and abduction of the low. In both situations the adducted hip, on the concave side of the spinal curve, is at risk of subluxation. Thus, deformity of the hip may cause scoliosis and vice versa; it is also possible that both deformities may be caused by

Table II. Incidence of scoliosis related to the upper levels of the neural deficit and the vertebral arch defect.

	Incidence of scoliosis (*per cent*)
Neurological level	
Above T12	83
L1 to low sacral	67
No demonstrable deficit	5
Bony arch defect	
Above T12	73
L1 to L4	44
L5 or lower	8

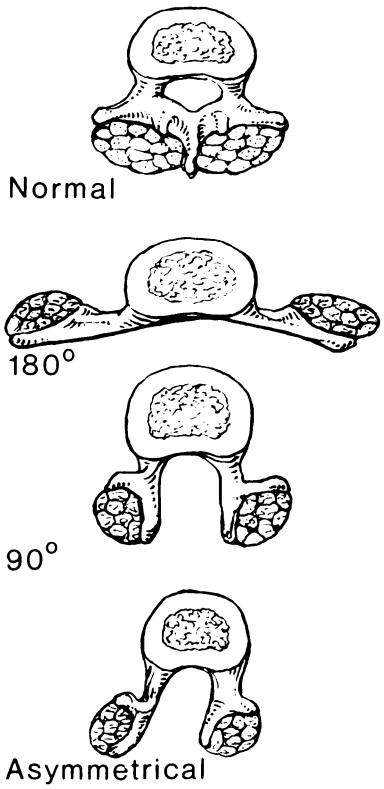

Fig. 4

Malpositions of the halves of the neural arch and associated musculature: hinged through 180 degrees, which is usually associated with kyphosis, symmetrical hinging through 90 degrees, and asymmetrical displacement which may contribute to scoliosis.

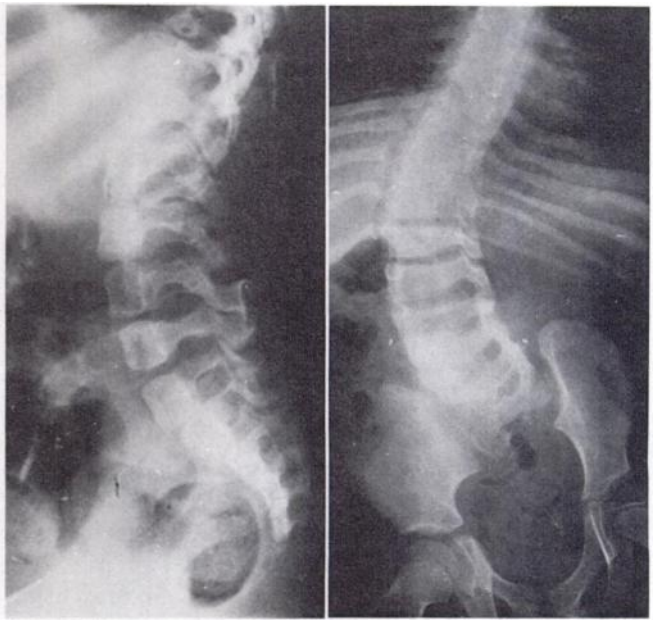

Fig. 5 Fig. 6

Figure 5—Posterior projection (and in this instance elongation) of the "half-arches", in myelomeningocele. Figure 6—Asymmetrical half-arches in myelomeningocele. The left halves of the neural arches project laterally, the right posteriorly. This appearance could be caused by true asymmetry or by vertebral rotation, or both. Clinically there was little or no rotation.

the same mechanism, for instance asymmetric activity of the iliopsoas.

An attempt was made to determine how often contracture or subluxation of the hip led to scoliosis, and if early treatment of the deformity of the hip would prevent the onset of spinal curvature, and the following facts emerged. In children without spinal deformity, 77 per cent had undeformed hips, 16 per cent had symmetrical deformity, and seven per cent had a unilateral hip deformity. In those with spinal curvature extending down to the sacrum or rigid lower lumbar spine, 40 per cent had undeformed hips, 33 per cent had symmetrical deformity, while 20 per cent had, or in the past had had surgical treatment for, an adduction deformity or subluxation of the hip on the concave side of the spinal curvature—the up-tilted side of the pelvis; surprisingly, the remaining seven per cent had had treatment for subluxation or adduction contracture of the hip on the convex side of the curve. In children with scoliosis not reaching to the rigid segments, the figures were intermediate 48 per cent having undeformed hips, 20 per cent symmetrical deformity, while in the remaining 32 per cent unilateral deformity was distributed equally between the convex and concave side of the curve. Thus, where deformity of the hip was asymmetrical and the curve extended down to the rigid segments of the spine, the well-known pattern of an adduction contracture or subluxation on the concave side, with or without an abduction contracture on the opposite side, was common; when analysis was restricted to curves over 30 degrees this picture was present in 40 per cent. The variability of the time of onset of deformity and of treatment precluded valid conclusions on the effect of the deformity of the hip in initiating or promoting spinal curvature. The effect of iliopsoas transfer (Sharrard 1964) on the incidence of spinal curvature was also analysed but there was nothing to suggest that this operation contributed either to the onset or prevention of scoliosis.

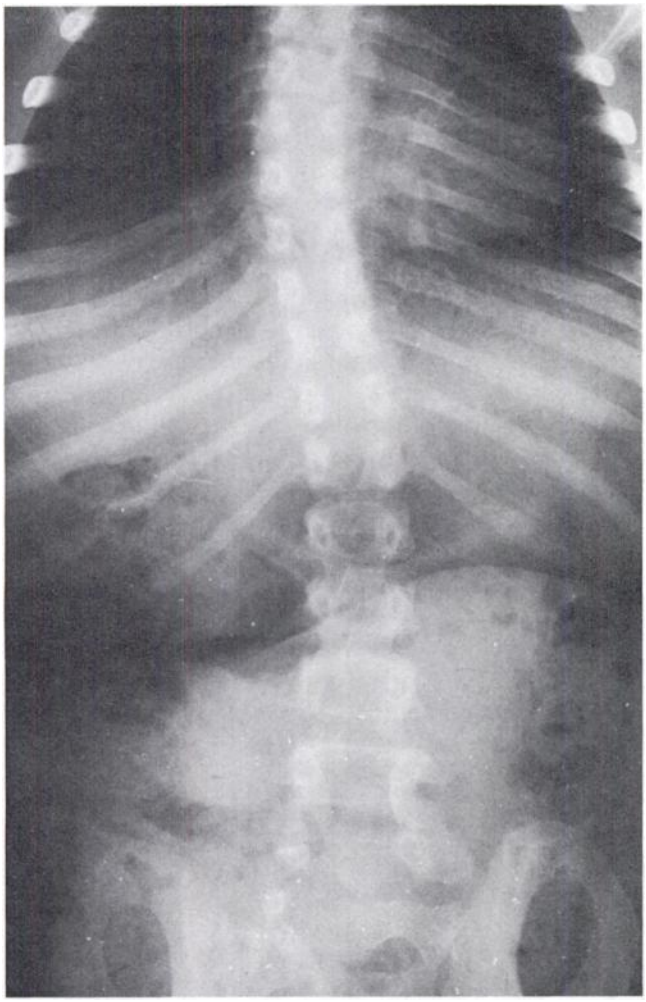

Fig. 7

Scoliosis of unknown cause in a child with myelomeningocele. The whole curve is cephalic to both neurological and bony defects and, if the abnormalities below the second lumbar level are ignored, the similarity to an idiopathic curve in an otherwise normal child is apparent.

Idiopathic curves. In 12 per cent of children with scoliosis no causative mechanism was apparent. The whole curve was proximal to both bony and neurological defects, and even when distal neurological changes were present there was no trace of asymmetry. These curves have therefore been called idiopathic: they are not identical with the curves of idiopathic scoliosis in otherwise normal children, but there are some remarkable similarities. Figure 7 shows a curve of this type, which would probably unhesitatingly be accepted as "idiopathic" if the defect in the neural arch were not obvious. Whereas in the series as a whole right and left curves were exactly equal in number, in the idiopathic group there was a preponderance (73 per cent) of right curves, as occurs in the common adolescent type of scoliosis. There were, however, equal numbers of boys and girls with idiopathic curves and the onset was usually well before adolescence.

Table III. Causes of scoliosis

	Per cent*
Congenital malformation	38
Neurological deficit	62
Asymmetric neural arch and associated muscle attachment	15
"Idiopathic" curve	12

*The percentages total more than 100 because the aetiology is multifactorial in some cases.

In each curve an attempt was made to identify the causative factors, and where no cause could be identified the curve was called idiopathic. The results are shown in Table III. It will be seen that many curves were thought to have a dual aetiology, most commonly a congenital malformation of vertebral bodies being associated with a neural deficit or asymmetric arches.

DISCUSSION

Causes of scoliosis in myelodysplasia. It is clear that several causes operate, sometimes acting together, and they fall into the three main groups of congenital malformation of vertebral bodies, inadequate neuromuscular control of the spine, and other factors unknown. To maintain the upright stance each vertebra is balanced on the one below by a combination of ligamentous support and muscular action. The muscles often traverse more than one segment and long distances may be spanned: they form a complex system of "guy-ropes", activated, self-adjusting, and computer-controlled. If there is a slight degree of asymmetric muscular weakness it is conceivable that adjustment could occur to prevent deformity, but if there is total muscle paralysis on one side then the system will bend to the active side. It is also apparent that if there is total bilateral paralysis over any section of the spine with overall loss of control, some very minor factor may determine the direction of the deformity. The onset of scoliosis and other spinal deformities may therefore be determined either by total loss or asymmetry of muscle action which may have resulted from nervous or muscular dysfunction, or from abnormal placement of muscles caused in the case of myelodysplasia by asymmetric deformity of the neural arch. In 12 per cent of the cases studied there was no demonstrable cause of scoliosis. It may be that other unknown factors are at work, but it is possible that these cases are really of neuromuscular origin. In spina bifida the spinal cord proximal to the main defect often shows quite extensive pathological changes (Emery 1972) and so does the brain. It is conceivable that these changes may be responsible for neuromuscular imbalance, not sufficiently severe to be demonstrated clinically, yet enough to fail under the very searching test of keeping the growing spine in balance.

Prognosis. Perhaps the most significant finding of this review is that by the age of 10 years 90 per cent of all children with myelodysplasia had some degree of spinal deformity and most of these (82.5 per cent) had scoliosis. Conversely, though longer follow-up is needed to confirm this, if a child reached the age of nine years without developing scoliosis he was unlikely to do so thereafter. In planning the long-term management of an infant with myelodysplasia it is useful to know if he is likely to escape major spinal deformity, and with this in mind many features were studied to assess their prognostic value. It has been shown that both the level of paraparesis and the level of neural arch defect were good guides, in that the higher the neurological or bony defect the greater the probability of spinal deformity. By studying the children over 10 years of age it became clear that if neither defect extended proximally beyond the fifth lumbar level, scoliosis was most unlikely to develop; if either reached to the fourth lumbar level there was a three in four chance of lateral curvature appearing; and if both defects reached this level the onset of scoliosis before 10 years of age was virtually certain. Not all curves, however, were bad enough to need surgical treatment and this study failed to reveal any reliable way of forecasting severe deformity, though there was a strong impression that both early onset, and fixed pelvic obliquity (whatever its aetiological relationship to lateral curvature might be) were bad omens for the progression of scoliosis. These prognostic considerations did not apply in congenital scoliosis which tended to progress, often severely, for there were so many different combinations of bony anomaly and neural deficit that reliable prognostic patterns could not be identified.

Treatment. It is not the object of this paper to consider treatment of spinal deformity. It is important, however, to remember that, in addition to scoliosis, kyphosis and lordosis are also significant problems in myelodysplasia and very often there is a combination of deformities, so treatment is then somewhat different from that of pure scoliosis. In all cases of major deformity, treatment has been operative or not at all, as no success has been achieved by bracing, except occasionally as a holding manoeuvre for an idiopathic curve. Selection of surgical treatment depends on the overall assessment of the child's problems; about 50 per cent will benefit from operation. This has usually taken the form of a Dwyer interbody fusion, though some have been treated by posterior fusion with a Harrington rod, and a few, usually the most extensive curves, have needed both Dwyer and Harrington procedures, with an interval of a few weeks. The decision to operate has often been difficult, but undoubtedly the worst mistakes have been to withhold or delay operation in a rapidly progressive curve.

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