Scoliosis in myelomeningocele

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Prevalence, type, and magnitude of scoliosis were studied in 163 patients with myelomeningocele. A scoliosis was diagnosed in 143 of them, congenital in 15 percent of the cases and developmental in the remaining patients. The severity of scoliosis increased with age and was more severe the higher the level of the neurologic deficit. The direction of the curves was correlated with pelvic obliquity, but not with hip dislocation. Although some patients with low level anomalies may develop severe scoliosis, patients with levels above L3 run a considerably higher risk in this respect. The radiographic baseline examination of the spine carried out in the newborn seems to permit a reasonable prognosis in regard to future scoliosis. Gross syringohydromyelia caused or contributed to scoliosis in 5 percent of the cases. However, less extensive syringohydromyelia and Chiari malformations due to abnormal neuromuscular control also promote the development of scoliosis in these patients.

In patients with myelomeningocele, the incidence of scoliosis is reported to vary between 9 and 88 percent depending on age and the neurologic level of the lesion (Raycroft and Curtis 1972, Shurtleff et al. 1976, Piggott 1980). Two main origins of scoliosis are recognized – congenital, a small number, and developmental, the majority. Patients belonging to the former group are born with vertebral anomalies in addition to those anomalies characteristic of myelomeningocele. In the developmental type, inadequate neuromuscular control results in scoliosis (Raycroft and Curtis 1972). Further, idiopathic curves with no apparent causative mechanism are on record (Piggott 1980); these are, however, sometimes associated with central nervous system abnormalities, e.g., syringohydromelia and Chiari malformations (Hall et al. 1976, Samuelsson et al. 1987).

We have attempted to identify correlations between prevalence, type, location, and degree of scoliosis, on the one hand, and age, gender, radiographic and neurologic levels of the lesion, pelvic obliquity, and hip dislocation, on the other. We also wanted to correlate scoliosis with manifestations of brain stem and/or spinal cord lesions cranial to the myelomeningocele.

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Patients and methods

Totally, 196 patients with myelomeningocele were treated between 1952 and 1985 at the Karolinska Hospital, Stockholm, or at Örebro Medical Center Hospital, Örebro, Sweden.

Eighty-five males and 78 females, mean age 15 (2–40) years, underwent a clinical and radiographic follow-up examination between January 1, 1984, and December 31, 1985. Of the remaining 33 subjects lost for study, 11 were dead, 17 had moved to other parts of Sweden, and 5 were untraceable. Review of the case histories and available radiographs revealed no differences between this subgroup and the main series as regards age and levels of lesions.

Radiographic level of myelomeningocele. The upper level of the vertebral anomaly related to the myelomeningocele was radiographically deter-

mined. For this purpose, radiographs obtained before 6 years of age were reassessed. Special attention was paid to defects and deformities of the neural arches, increase in interpedicular distance, and distortions and deviations of the pedicular bases (Samuelsson and Eklöf 1987). Radiographs from previous examinations and those obtained at the follow-up were also assessed for congenital vertebral anomalies in addition to those typical of myelomeningocele.

Radiographic assessment of scoliosis, pelvic obliquity, and hip dislocation. Scoliosis was defined as a curve of 5° or more. It was measured according to Cobb (1948) on anteroposterior radiographs obtained in an unsupported sitting position. Due to the lack of relevant radiographs, 2 patients were omitted from this appraisal. Thoracic curves had their apex between T2 and T11, thoracolumbar curves at T12 or L1, and lumbar curves between L2 and L4. The same radiographs were used for evaluating pelvic obliquity according to Osebold et al. (1982) by measuring the angle formed by the intercrest line and the horizontal line. Assessment proved feasible in 131 patients, whereas 32 were excluded because available radiographs were unsuitable for appraisal. Radiographs of the pelvis, also obtained at the follow-up, were screened for dislocation of the hips.

Magnetic resonance imaging (MRI). The brain and spinal cord were examined with MRI in 35 patients using a 1.0 Tesla (T) superconducting unit (Siemens, Magnetom) operating at 0.35 T or 0.5 T. Selected for this examination were patients with developmental scoliosis and/or symptoms and signs suggestive of lesions in the brain stem or spinal cord at levels above the myelomeningocele.

Assessment of neurologic level and deficiencies. Using the criteria set up by Lindseth (1976), the level of the motor deficit was established, permitting identification of six neurosegmental levels. Thus, the motor level deficit was defined as the lowest level, on the better side, at which the patient was able to perform an antigravity grade 3 movement through the available range of joint mobility. In addition, sensory disturbances and upper or lower motor neuron paralysis above the myelomeningocele were recorded.

Statistical methods. Differences between measurements of scoliosis stratified according to age, gender, radiographic and neurologic level, and location of the curvature were analyzed with the Spearman rank correlation, Kruskal-Wallis test, and Mann-Whitney *U*-test. The relationship between curve pattern, pelvic obliquity, and hip dislocation was tested by chi-square analysis. The 5 percent level of significance was accepted.

Results

Age, gender, and prevalence of scoliosis. Scoliosis was found in 77 males and 66 females. Fifty-two patients had also developed compensatory curves. If age and motor level are disregarded, this implies an 89 percent prevalence of scoliosis. However, the age dependence was reflected by the fact that only 59 percent of all the patients below aged 6 years presented with scoliosis. This rate increased to 97 percent in the age-interval 11–16 years (Table 1).

Congenital scoliosis. All 21 patients with congenital scoliosis had vertebral anomalies besides those characteristic of myelomeningocele. There was a broad spectrum of malformations. Single or, more frequently, multiple hemivertebrae with or without neural arch abnormalities and fusions of vertebral bodies occurred. In 11 patients such anomalies were found at discontinuous levels. Patients belonging to this category had, on an average, higher lesions independent of whether the assessment of the level was radiographic or neurologic. Their curvatures were generally more severe.

Prevalence of scoliosis related to radiographic and neurologic levels. The radiographic level of the lesion was thoracic in 15 percent of the cases, lumbar in 69 percent, and sacral in 16 percent. The

Table 1. Occurrence and degree of scoliosis in 143 patients with myelomeningocele

Age	Total number	With scoliosis (fraction)	Degrees of scoliosis Mean, median (90 percent range)
0–6	22	13 (0.59)	20, 11 (5–60)
6–11	28	24 (0.85)	26, 21 (5–90)
11–16	35	34 (0.97)	32, 24 (5–108)
16–	78	72 (0.92)	34, 20 (5–148)

Table 2. Mean degrees, median degrees, and percent of patients with scoliosis according to radiographic level. N 141

Level	Total number	With scoliosis (fraction)	Degrees of scoliosis Mean, median (90 percent range)
Thoracic	24	24 (1)	54, 48 (5–108)
L1–L2	20	19 (0.95)	38, 36 (5–84)
L3	19	17 (0.90)	32, 20 (9–84)
L4	40	34 (0.85)	28, 20 (5–94)
L5	33	29 (0.88)	23, 15 (5–61)
Sacral	25	18 (0.72)	11, 9 (5–20)

Table 3. Mean degrees, median degrees, and percent of patients with scoliosis according to neurologic level. N 143

Level	Total number	With scoliosis (fraction)	Degrees of scoliosis Mean, median (90 percent range)
Thoracic	35	35 (1)	52, 48 (7–107)
L1–L2	5	5 (1)	58, 36 (9–130)
L3	26	26 (1)	28, 20 (5–61)
L4	32	26 (0.81)	30, 20 (7–100)
L5	20	17 (0.85)	14, 11 (5–23)
Sacral	45	34 (0.75)	16, 10 (5–47)

frequency and severity of scoliosis were worse in patients with upper lesions (Table 2). The neurologic level was thoracic in 22 percent of the patients, lumbar in 51 percent, and sacral in 27 percent. Eleven patients with no scoliosis had neurologic sacral lesions (Table 3).

Curve pattern. There was no sex difference as regards the site or direction of scoliosis. In general, thoracic curves were more pronounced than those with a thoracolumbar of lumbar location.

Pelvic obliquity and hip dislocation versus scoliosis. Of 131 patients with radiographs permitting assessment, 103 had an oblique pelvis and 28 a horizontal one. The left side was high in 55 cases and the right side in 48. The mean amplitude of pelvic obliquity was 8°, the median 5° (0°-39°). In 46 patients with an obliquity of 5° or more, 22 had a high right hemipelvis and 24 a high left hemipelvis. In 36 of them the convexity of the scoliosis was away from the high side of the pelvis. Thirty patients had scoliosis and dislocated hips – 10 of them bilaterally. There was no correlation between the side of the dislocation and the direction of the scoliosis.

Brain stem and spinal cord lesions. Eighteen patients has neurologic deficits rostral to the myelomeningocele. Fifteen of them were examined by MRI. Ten of these patients had signs of cranial nerve deficits. Syringohydromelia in a widened or bulging spinal cord was shown in 5 of them. Another 20 patients, without neurologic deficits above the myelomeningocele, underwent MRI. Nine of them had less extensive syrinxes in atrophic or normal-sized spinal cords. All 35 patients had Chiari malformations of varying extent (Samuelsson et al. 1987).

Discussion

The distribution of patients according to radiographic and neurologic levels (Tables 2 and 3) conforms with the findings of Menelaus (1980). However, only 3 percent of our patients belonged to the L1–L2 motor level, as compared with approximately 20 percent of the cases reported by Raycroft and Curtis (1972). The divergence of findings reported in the literature may partly be explained by the lack of common criteria for classification of neurologic deficits. In our opinion the method of Lindseth (1976) used in this investigation is superior, and it permits reclassification for comparison with findings obtained according to Sharrard (1964) or Hoffer et al. (1973).

Congenital scoliosis. Our 15 percent rate of congenital scoliosis is lower than in most previous reports. Raycroft and Curtis (1972) reported an incidence of 21 percent and Piggott (1980) 38 percent. This discrepancy might be accounted for by differences in age at the time of assessment. In contrast to previous reports, our evaluation was based on radiographs obtained before 6 years of age. It was, therefore, possible to separate true congenital curvatures from developmental scoliosis.

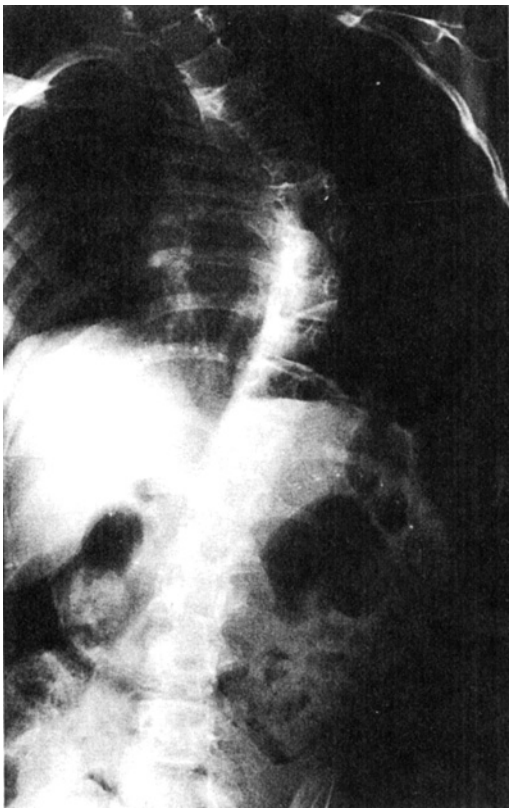
Scoliosis related to age and level of lesion. Both the prevalence and the severity of scoliosis increase with age and higher neurologic level. In our patients aged 20 years or older, the prevalence of scoliosis exceeding 30° was higher than the percentage reported by Shurtleff et al. (1976). This feature was particularly obvious in patients with thoracic or sacral lesions. Although patients with low lesions may develop a severe scoliosis, our results indicate a preponderance of such curves in

patients with levels higher than L3. Eleven patients without scoliosis had sacral lesions. Out of the remaining 7, all except 1 patient had neurologic levels below L1–L2. The agreement demonstrated between radiographic and neurologic assessments of the level of the lesion makes it possible to use the baseline radiographic examination as an indicator of the likely degree of future scoliosis (Samuelsson and Eklöf 1987).

Scoliosis, pelvic obliquity, and hip dislocation. Raycroft and Curtis (1972) found an elevated pelvis, ipsilateral luxation or subluxation of the hip, and lumbar scoliosis to the contralateral side in all the cases of moderate or severe developmental scoliosis. We could confirm the observation that the convexity of the scoliosis was opposite to the the elevated side of the pelvis. However, there was no correlation between the direction of scoliosis and side of hip dislocation. Independent of the side of dislocation, the left hemipelvis tended to be higher twice as frequently as the right hemipelvis. These findings accord with the observations of Lonstein and Beck (1986) in their study of patients with cerebral palsy. Our results seem to exclude infrapelvic factors, such as hip dislocation, as a probable cause of scoliosis also in myelomeningocele.

Scoliosis and syringomyelia. Including cases of idiopathic scoliosis (Piggott 1980), 85 percent of all our curves were developmental. An idiopathic scoliosis may originate as an inborn or acquired neurologic abnormality associated with myelomeningocele. In these patients, syringohydromyelia and/or Chiari malformations have been demonstrated, both at necropsies (Peach 1965, Emery 1972, Gilbert et al. 1986) and by means of neuroradiologic methods (Hall et al. 1976, Venes et al. 1986, Samuelsson et al. 1987).

There is an established relation between idiopathic syringomyelia and scoliosis (Huebert and MacKinnon 1969, Williams 1979, Baker and Dove 1983). Previous reports indicate that spinal cord cavities may also cause scoliosis in myelomeningocele patients (Hall et al. 1976, Samuelsson et



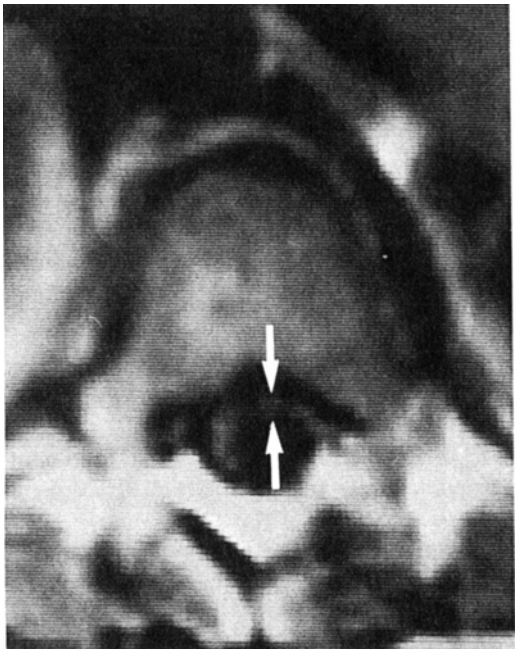


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Figure 1. A 10-year-old girl with a radiographic level of the myelomeningocele at L5.
A. Thoracic scoliosis.
B. Thoracic, axial MR image demonstrating syringohydromyelia. Rim of spinal cord between arrows.

al. 1987). In association with myelomeningocele the prevalence of syringohydromyelia has been estimated to be as high as 40 percent, but the majority of cases probably remain clinically occult.

Six of our patients had neurologic deficits clinically ascribable to syringohydromyelia. Five of them underwent MRI, which revealed large spinal cord cavities. This indicates a prevalence of gross syringohydromyelia causing or contributing to scoliosis in at least 5 percent of the cases. One may assume that the more frequent, less extensive, and clinically asymptomatic cavities may also affect spinal stability, thus contributing to the development of scoliosis. These small cavities are probably the origin of the gross, clinically manifest syringohydromyelia.

In patients with myelomeningocele, syringohydromyelia is commonly located at the cervicothoracic junction (Samuelsson et al. 1987). This location is in common with idiopathic syringomyelia, which is often associated with high thoracic scoliosis. In myelomeningocele patients, similar thoracic curves associated with low radiologic levels may indicate syringohydromyelia, especially if the corresponding neurologic level is high (Figure 1).

Brain stem dysfunction and scoliosis. During periods of vertebral growth, patients with adolescent idiopathic scoliosis have an equilibrium dysfunction at the brain stem. As a consequence, it has been postulated that scoliosis may develop secondary to a disturbance of the central mechanism of the postural reflex system (Nachemson and Sahlstrand 1977, Sahlstrand et al. 1979, Yamada et al. 1984). Dreitakis (1984) described 8 patients with congenital brain stem abnormalities associated with scoliosis. Riley and Swift (1979) reported on kyphoscoliosis in association with congenital horizontal gaze palsy, probably due to lesions in the pontine reticular formation. Recently, Wyatt et al. (1986) found pathologic vibratory responses that were due to a malfunctioning of the posterior column pathways in patients with adolescent idiopathic scoliosis. These facts indicate that lesions of the brain stem, by causing neuromuscular disturbances, contributed to the development of scoliosis.

Brain stem deformation due to Chiari malformations seems to be invariably associated with myelomeningocele (Samuelsson et al. 1987). These patients also show symptoms and signs of cranial nerve deficits (Sieben et al. 1971), abnormal brain stem auditory-evoked potentials (Holliday et al. 1985), and eye-movement disorders of central etiology (Cogan 1968). In addition, aplasia of cranial nerve nuclei have been recorded in neuropathologic studies (Gilbert et al. 1986). Due to the spinal cord lesion, the postural reflexes are abnormal. All the patients examined with MRI had deformation of the brain stem, and 10 of them had symptoms and signs of cranial nerve deficits. In patients with myelomeningocele, Chiari malformations may, apart from syringohydromyelia, in a yet unknown way, lead to scoliosis.

References

Baker A S, Dove J. Progressive scoliosis as the first presenting sign of syringomyelia. Report of a case. J Bone Joint Surg (Br) 1983;65(4):472–3.

Cobb J R. Outline for the study of scoliosis. In: Instructional Course Lectures, The American Academy of Orthopedic Surgeons, Edwards, Ann Arbor 1948;5:261–75.

Cogan D G. Down beat nystagmus. Arch Ophthalmol 1968;80(6):757–68.

Dretakis E K. Scoliosis associated with congenital brain stem abnormalities. A report of eight cases. Int Orthop 1984;8(1):37–46.

Emery J L. The back lesions, lipomas and dermoids. In: The American Academy of Orthopaedic Surgeons Symposium on Myelomeningocele, C.V. Mosby, St Louis 1972:41–9.

Gilbert J N, Jones K L, Rorke L B, Chernoff G F, James H E. Central nervous system anomalies associated with meningomyelocele, hydrocephalus and the Arnold Chiari malformation: reappraisal of theories regarding the pathogenesis of posterior neural tube closure defects. Neurosurgery 1986;18(5):559–64.

Hall P V, Lindseth R E, Campbell R L, Kalsbeck J E. Myelodysplasia and develomental scoliosis. A manifestation of syringomelia. Spine 1976;1:48–56.

Holliday P O, Pillsbury D, Kelly D L Jr, Dillard R. Brain stem auditory evoked potentials in Arnold Chiari malformation: possible prognostic value and

changes with surgical decompression. Neurosurgery 1985;16(1):48–53.

Hoffer M M, Feiwell E, Perry R, Perry J, Bonnett C. Functional ambulation in patients with myelomeningocele. J Bone Joint Surg (Am) 1973;55(1):137–48.

Huebert H T, MacKinnon W B. Syringomyelia and scoliosis. J Bone Joint Surg (Br) 1969;51(2):338–43.

Lindseth R E. Treatment of the lower extremity in children paralyzed by myelomeningocele (birth to 18 months). In: Instructional Course Lectures, The American Academy of Orthopedic Surgeons, C.V Mosby, St Louis 1976;25:76–82.

Lonstein J E, Beck K. Hip dislocation and subluxation in cerebral palsy. J Pediatr Orthop 1986;6(5):521–6.

Menelaus M. The spine. In: The Orthopedic Management of Spina Bifida Cystica. Churchill-Livingstone, Edinburgh 1980:152–87.

Nachemson A L, Sahlstrand T. Etiologic factors in adolescent idiopathic scoliosis. Spine 1977;2(3):176–84.

Osebold W R, Mayfield J K, Winter R B, Moe J H. Surgical treatment of paralytic scoliosis associated with myelomeningocele. J Bone Joint Surg (Am) 1982;64(6):841–56.

Peach B. Arnold Chiari malformation. Anatomic features of 20 cases. Arch Neurol 1965;12:613–21.

Piggott H. The natural history of scoliosis in myelodysplasia. J Bone Joint Surg (Br) 1980;62(1):54–8.

Raycroft J F, Curtis B H. Spinal curvature in myelomeningocele. Natural history and etiology. In: The American Academy of Orthopedic Surgeons Symposium on Myelomeningocele. C.V. Mosby, St Louis 1972: 186–201.

Riley E, Swift M. Congenital horizontal gaze palsy and kyphoscoliosis in two brothers. J Med Genet 1979; 16(4):314–6.

Sahlstrand T, Petruson B, Örtengren R. Vestibulospinal reflex activity in patients with adolescent idiopathic scoliosis. Postural effects during caloric labyrinthine stimulation recorded by stabilometry. Acta Orthop Scand 1979;50(3):275–81.

Samuelsson L, Bergström K, Thuomas K A, Hemmingsson A, Wallensten R. MR imaging of syringohydromyelia and Chiari malformations in myelomeningocele patients with scoliosis. AJNR 1987;8(3):539–46.

Samuelsson L. Eklöf O. The value of conventional radiography as supplement to the neurological assessment in myelomeningocele. Acta Radiol (Diagn) (Stockh) 1987;28(5):615–9.

Sharrard W J W. The segmental innervation of the lower limb muscles in man. Ann R Coll Surg 1964;35:106–22.

Shurtleff D B, Goiney R, Gordon L H, Livermore N. Myelodysplasia: the natural history of kyphosis and scoliosis. A preliminary report. Dev Med Child Neurol 1976;(Suppl 37):126–33.

Sieben R L, Hamida M B, Shulman K. Multiple cranial nerve deficits associated with the Arnold Chiari malformation. Neurology 1971;21(7):673–81.

Venes J L, Black K L, Latack J T. Preoperative evaluation and surgical management of the Arnold-Chiari II malformation. J Neurosurg 1986;64(3):363–70.

Williams B. Orthopaedic features in the presentation of syringomyelia. J Bone Joint Surg (Br) 1979;61(3): 314–23.

Wyatt M P, Barrack R L, Mubarak S J, Whitecloud T S, Burke S W. Vibratory response in idiopathic scoliosis. J Bone Joint Surg (Br) 1986;68(5):714–8.

Yamada K, Yamamoto H, Nakagawa Y, Tezuka A, Tamura T, Kawata S. Etiology of idiopathic scoliosis. Clin Orthop 1984;(184):50–7.

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