SYRINGOMYELIA AND SCOLIOSIS

H. T. HUEBERT and W. B. MACKINNON, WINNIPEG, CANADA

From the Shriners’ Hospital for Crippled Children, Winnipeg and the Orthopaedic Department, University of Manitoba

Pathological cavitation of the spinal cord was first called "syringomyelia" by Ollivier in 1837. He did not distinguish between cysts and central canal dilation, because the canal was not recognised as a normal structure until 1859. The first known description of the clinical signs was that of Duchenne (1872), though he called it progressive muscular atrophy. Schultze (1882) correlated the pathology of syringomyelia with the clinical syndrome.

Syringomyelia is a chronic, slowly progressive degeneration of the spinal cord and medulla. Pathologically there is cavitation and gliosis within the substance of the cord. Clinically there is sensory dissociation with pain and temperature sensation involved, but not touch, at the level of the lesion, and weakness and wasting of the muscles of the involved segments. Impaired long tract function may be found distally. The cervical spine is most commonly affected, though the cystic lesions often extend proximally into the medulla and distally as far as the lumbo-sacral area.

<table>
<thead>
<tr>
<th>TABLE I</th>
<th>SYRINGOMYELIA AND SCOLIOSIS</th>
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</thead>
<tbody>
<tr>
<td>Total number of patients with syringomyelia</td>
<td>43</td>
</tr>
<tr>
<td>Patients with scoliosis</td>
<td>27</td>
</tr>
<tr>
<td>Mild scoliosis (under 25 degrees)</td>
<td>15</td>
</tr>
<tr>
<td>Moderate scoliosis (25-50 degrees)</td>
<td>5</td>
</tr>
<tr>
<td>Marked scoliosis (over 50 degrees)</td>
<td>7</td>
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<tr>
<th>TABLE II</th>
<th>CHARACTERISTICS OF SCOLIOTIC CURVES</th>
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<tbody>
<tr>
<td>Site of scoliosis</td>
<td>Thoracic</td>
</tr>
<tr>
<td></td>
<td>Thoraco-lumbar</td>
</tr>
<tr>
<td></td>
<td>Lumbar</td>
</tr>
<tr>
<td>Type of curve</td>
<td>Single primary</td>
</tr>
<tr>
<td></td>
<td>Double primary</td>
</tr>
</tbody>
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The diagnosis of syringomyelia presents a problem. Other conditions such as cord tumours may mimic the disease. Whereas a radiologically enlarged cord together with an enlarged spinal canal is often found in syringomyelia, Ford (1960) believed that the cord might be flattened and atrophic in some cases. The final diagnosis is dependent on the total general impression obtained, taking into account the clinical, radiological and operative findings.

Syringomyelia is often associated with other abnormalities such as Arnold-Chiari deformity, basilar impression, Klippel-Feil syndrome, spina bifida and cervical ribs. Gardner.
and Collis (1960) and Gardner (1964) believed that the skeletal changes common to syringomyelia, diastematomyelia and meningocele suggested a common origin. On the other hand Finlayson (1962) found in his seventy-five cases of syringomyelia that the incidence of spina bifida occulta was the same as that of a control group (10.5 per cent).

Scoliosis has been described in cases of syringomyelia, but there is diversity of opinion about its significance. Moe (1967) encountered several cases. Finlayson considered scoliosis a frequent early finding, and Ford found that syringomyelia in children often presents a severe and rapidly progressive scoliosis. Perret (1963) considered that it occurs eventually in most cases of syringomyelia. McIlroy and Richardson (1965) found scoliosis in thirty-three out of seventy-five cases of syringomyelia. They did not mention the magnitude of the curves, nor whether treatment was necessary. Woods and Pimenta (1944) noted five cases of scoliosis among twenty patients with syringomyelia.

A review of the records of four Winnipeg hospitals and of all the Shriners' hospitals revealed forty-three well documented cases of syringomyelia. All pertinent radiographs were reviewed. Twenty-six patients were female, seventeen male. The average age at the onset of symptoms was 21.4 years, varying from soon after birth to fifty-four years. Thirty patients had had cervical myelography, which confirmed the diagnosis in nineteen. Eleven had had laminectomy and three had had spinal fusion (two for scoliosis and one for instability of the cervical spine). Involved segments varied from the medulla down to the distal end of the cord. Five patients had associated abnormalities of the skeleton or central nervous system.

Of the forty-three patients studied twenty-seven (63 per cent) had evidence of scoliosis. Fifteen had mild curves (0–25 degrees by Cobb measurement), five had moderate (25–50 degrees) while seven had severe curves (over 50 degrees) (Table I). Twenty-one curves were confined to the thoracic region, three were thoraco-lumbar and three were lumbar. There were twenty-five single primary curves and two double primary curves (Table II).

No published report of attempts to treat scoliosis associated with syringomyelia has been found. Two such cases were seen in Winnipeg and illustrated some of the problems that may be encountered.

**CASE REPORTS**

**Case I**—A girl was first seen at the Shriners' Hospital in Winnipeg at the age of thirteen. She had noticed chest and back deformity for four months. The parents had noted abnormalities of the face, tongue, left arm and leg almost since birth. On examination there was obvious scoliosis convex to the right. There were minimal right facial weakness and atrophy of the right side of the tongue. The left arm and leg were slightly weaker than the right; reflexes in the left arm were decreased and there was diminished perception of pinprick in the second to fourth cervical dermatomes. Radiographs showed thoraco-lumbar scoliosis convex to the right from T.5 to L.2, the curve measuring 69 degrees. Myelography showed the cervical cord to be enlarged from the upper cervical region down to T.1 (Fig. 1). Syringomyelia with syringobulbia was diagnosed. Because of the severity of the scoliosis treatment was begun using a Milwaukee brace, but the curve increased to 86 degrees (Fig. 2). At operation Harrington rods were inserted from T.4 to L.1 and the spine was fused. Thereafter a plaster jacket was worn for eight months. A year after operation the correction was maintained at 63 degrees (Fig. 3). When last seen three years after operation the patient had no complaints; the curve measured 64 degrees.
Comment—It is of interest that the underlying disease was, in retrospect, noted from early infancy, yet the presenting symptom was finally the scoliosis. Treatment of this patient was begun with caution because of lack of previous experience with this disease. Greater correction of the scoliosis could have been achieved at the time of operation, but was not attempted because of the risk of damage to the abnormal spinal cord.

Case 1—Figure 2—Radiograph showing the thoraco-lumbar scoliosis of 86 degrees (Cobb measurement). Figure 3—Showing correction obtained one year after spinal fusion with insertion of Harrington rods.

Case 2—A girl first complained of backache and leg pain at the age of nine years. Scoliosis was noted at the age of twelve and increased during the next five years. It was centred in the lumbar area, was convex to the right and measured 61 degrees. At laminectomy (T.11 to L.2) yellow fluid was aspirated from the distended cord. The symptoms improved, but some weakness of the legs and pain in both feet persisted. At the age of thirty she again had increasing backache with paraesthesias in the legs. She could not differentiate heat from cold on the right side of the body and had diminished pinprick in the same area. The curve now measured 74 degrees. Operation for insertion of Harrington rods and spinal fusion was advised. At operation dense scar tissue was encountered. A cyst which had become adherent to the fibrous tissue ruptured during the dissection and a large amount of fluid escaped. It was thought unwise to use Harrington rods because of the cerebrospinal fluid leak, so a Hibbs' facet fusion was done. After operation the patient was paraplegic below the twelfth thoracic segment, that is, the level of the cyst. Two weeks later a corrective Risser jacket was applied despite the paralysis. Four weeks after the operation the patient died suddenly from pulmonary embolism. At necropsy the cord distal to the site of operation was softened and largely replaced by brownish gelatinous material. The diagnosis of syringomyelia was confirmed. Comment—This patient illustrates the hazards of treatment when the spinal cord is diseased. The danger was, of course, compounded by the previous laminectomy.

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DISCUSSION

In patients with syringomyelia, scoliosis is probably a skeletal change secondary to the pathology in the spinal cord. The cystic lesions in this disease are most commonly situated just dorsal to the central canal. Expansion of the cyst damages the neighbouring nervous structures; this expansion is usually asymmetrical. The function of pain and temperature fibres is lost very early in the disease because of their vulnerable position as they cross the midline near the central canal (Fig. 5).

With lateral expansion of the cyst the medial nuclear group of cells is frequently affected. From the position and extent of these columns it has been supposed that they innervate the muscles of the trunk (Strong and Elwyn 1964). The ventro-medial nuclei form a column of cells of the anterior horn and extend from the first cervical to the sacral segments. The nucleus of the hypoglossal nerve in the medulla seems to be a continuation of this column. The dorso-medial nuclei are present in the sixth and seventh cervical segments, then continue from the second thoracic to the first lumbar area (Fig. 4). The proximity of these tracts to the central part of the cord could explain the early development of scoliosis—due to damage done by an asymmetrically expanded cyst (Fig. 6).

From this series of cases it is difficult to ascertain the exact natural history of scoliosis in syringomyelia, though a general trend is noted (Figs. 7 and 8). The more severe curves tended to occur in the patients who developed symptoms early in life. The incidence of
scoliosis in patients who developed definite symptoms of syringomyelia at the age of sixteen or earlier was 82 per cent, while those first noting symptoms after sixteen years had an incidence of 48 per cent only (Fig. 9). Presumably scoliosis will be more severe if the pathological changes in the cord develop while the spine is still growing. The exceptions may be due to the difficulty in obtaining an accurate history and in ascertaining just when weakness of the trunk muscles began. Asymmetry of cyst development, size of the cyst, as well as the time of development probably account for the presence or absence of scoliosis and for the severity of the curvature when it occurs.

Correction of scoliosis in patients with pathological changes in the spinal cord should be approached with greater care than usual. Fragility of the cord associated with cysts tends to
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make it prone to damage with sudden changes in configuration of the bony canal. Laminectomy, with resultant removal of the bony protection posteriorly, makes the cord especially vulnerable to injury.

SUMMARY AND CONCLUSIONS

1. In this analysis of forty-three patients with syringomyelia, twenty-seven (63 per cent) had scoliosis. This association is probably due to the early involvement of the ventro-medial and dorso-medial nuclei of the spinal cord by expanding lesions.
2. The literature makes no reference to the treatment of scoliosis associated with syringomyelia. Two cases are presented of attempts to correct this scoliosis— one because of increasing deformity, the other for increasing backache.
3. Due to the presence of cystic lesions characteristic of syringomyelia, corrective operative treatment of scoliosis may present an added risk.
4. Because of the high incidence of scoliosis in patients with syringomyelia, any patient with syringomyelia should be examined for evidence of neurological deficit.

We should like to thank Dr David McQueen for allowing us to use one of his patients as a case presentation. We are also indebted to the Shriner's hospitals and the Winnipeg hospitals for the use of their records and radiographs, as well as to the Children's Hospital of Winnipeg for their help with the radiographs and other illustrations.

REFERENCES