

ABSENT SUPERFICIAL ABDOMINAL REFLEXES IN CHILDREN WITH SCOLIOSIS

AN EARLY INDICATOR OF SYRINGOMYELIA

HAMID G. ZADEH, SAMIR A. SAKKA, MICHAEL P. POWELL, MIN H. MEHTA

From the Royal National Orthopaedic Hospital Trust, Stanmore, England

We describe 12 children with idiopathic scoliosis who had a persistent absent superficial abdominal reflex (SAR) on routine neurological examination. MRI showed syringomyelia to be present in ten. The average age at detection of the scoliosis was 4.3 years and at diagnosis of syringomyelia 6.6 years.

In all ten children the SAR was consistently absent on the same side as the convexity of the curve. In two it was the only abnormal neurological sign. An absent SAR in patients with scoliosis is an indication for investigation for underlying syringomyelia.

In the children with syringomyelia, six had thoracic and four thoracolumbar curves. The clinical features differed in the two groups. Patients with thoracic curves were generally asymptomatic. Their neurological signs were subtle and none had any motor signs. By contrast, patients with thoracolumbar curves had symptoms and neurological signs. Abnormal gait was present in all four patients with thoracolumbar curves. In three this was due to considerable motor weakness.

In eight children syringomyelia was associated with a Chiari-I malformation. In seven the syrinx was treated surgically by decompression of the foramen magnum.

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The term 'syringomyelia' derives from the Greek words for 'tube' and 'marrow' and was first described by Ollivier in 1827.

The presenting features are diverse (Williams 1979). In children or adolescents, scoliosis is found in over 50% of cases (Tashiro et al 1987; Gurr, Taylor and Stobo 1988;

Burwell et al 1992; Williams 1992). Arai et al (1993), in a comprehensive study, reported that 4.0% of patients with scoliosis with curves larger than 20° had syringomyelia. New imaging techniques and improved clinical awareness have identified more patients with idiopathic scoliosis who have syringomyelia (Nohria and Oakes 1990). It is progressive and early diagnosis and treatment are therefore paramount (Williams 1992).

Our aim was to indicate the clinical features of importance in the early detection of syringomyelia with special reference to the superficial abdominal reflex (SAR) and to report our experience of the use of corrective plaster jackets and decompression of the foramen magnum in the management of these patients.

PATIENTS AND METHODS

Among patients with scoliosis referred to our unit are children with infantile or juvenile idiopathic scoliosis. Routine clinical assessment includes a detailed neurological examination and recording of the tendon reflexes, the plantar response and SAR. It has been our policy since 1985 to suspect syringomyelia when an absent SAR was detected, even if there were no other neurological signs.

Between 1986 and 1993 we observed 12 such children with an abnormal SAR. MRI showed syringomyelia to be present in ten. In the other two no intracranial or intraspinal lesions were demonstrated. There were six girls and four boys. Their average age at the detection of the scoliosis was 4.3 years (1 year 8 months to 6 years 7 months) and at presentation to our unit for the first time 5.8 years (2 years 5 months to 7 years 3 months). The time of diagnosis of syringomyelia was 6.6 years (4 years 11 months to 11 years 9 months). The average follow-up was 4.6 years (1 year 3 months to 8 years 8 months).

RESULTS

Six children had thoracic and four thoracolumbar curves. The clinical features differed in these two groups. Those with thoracic curves and syringomyelia were asymptomatic and had few abnormal physical signs. None had motor signs, but four had abnormal sensation over the trunk. By contrast, all four patients with thoracolumbar curves had

H. G. Zadeh, FRCS, Orthopaedic Registrar
S. A. Sakka, FRCS, Coxen Fellow
M. P. Powell, FRCS, Consultant Neurosurgeon
M. H. Mehta, FRCS, Consultant Orthopaedic Surgeon
Scoliosis Unit, Royal National Orthopaedic Hospital Trust, Brockley Hill, Stanmore, Middlesex HA7 4LP, UK.

Correspondence should be sent to Mr S. A. Sakka.

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symptoms and obvious neurological signs. An abnormal gait was common in all four and in three this was due to considerable motor weakness. Abnormal sensation was noted in three patients. In both groups abnormal reflexes were present (Table I).

In all ten children the SAR was absent on the same side as the convexity of the curve. That on the side opposite to the convexity of the curve was more variable and was present in three patients but only partially in two. In one patient partial recovery was noted after surgical decompression of the syrinx.

The average Cobb angle at first presentation was 34° and at the latest follow-up examination 38°. Orthopaedic treatment of the scoliosis consisted of the application of serial corrective plaster jackets or removable braces. General anaesthesia was necessary to apply plaster jackets in younger children. The brace was used during periods of slow growth (as judged by growth charts), when the scoliosis had been nearly corrected or during warm summer months.

MRI findings in eight patients showed the syrinx to be associated with a Chiari-I malformation (Fig. 1). In one, it was associated with tethering of the conus to S2 and an intraspinal lipoma, and in another it was multiloculated.

In nine children the syrinx has been managed surgically. Seven had decompression of the foramen magnum (performed by MPP). Two other children were operated on at different neurosurgical units. One had insertion of a syngoarachnoid shunt and the other lumbar laminectomy and syringoperitoneal shunting. For the child with a multiloculated syrinx there was no satisfactory neurosurgical procedure.

The average follow-up of the seven children who had decompression of the foramen magnum was 3.4 years (11 months to 6 years 6 months). All had MRI at 9 to 12 months after the operation to assess the degree of decompression (Fig. 2). This showed that two required further surgery; in one a syringopleural shunt was inserted and in the other a C1 arch division was performed. Further MRI in both patients confirmed satisfactory decompression of the syrinxes. Obvious neurological improvement has been noted in five patients, and in two who had mild sensory deficits no further deterioration has occurred.

After combined orthopaedic and neurosurgical treatment the scoliosis improved in three patients, stabilised in three and progressed in four. None of the children has had surgical correction of their scoliosis, but in some this may be necessary in the future.

DISCUSSION

The key to the early diagnosis of syringomyelia in scoliosis is a high index of suspicion and a thorough neurological examination (Huebert and MacKinnon 1969). A number of authors have reported that all affected patients have abnormal neurological signs (Depotter et al 1987; Phillips, Hen-

singer and Kling 1990; Lena et al 1992; Arai et al 1993). We agree with this observation, although in two of our ten patients the only abnormal neurological finding was an absent SAR. Lewonowski, King and Nelson (1992) reported that in 26 patients with idiopathic scoliosis under 11 years of age without neurological signs, MRI showed abnormal intraspinal pathology with Chiari-I malformation in five. They did not, however, record the SAR.

The SAR is a part of routine neurological examination and was first described by Rosenbach (1876). Lonnum (1956) reported that this reflex is present in the newborn and infants and Madonick (1957) stated that it may be absent in over 10% of normal individuals less than 50 years of age. Clinical work by Lonnum (1956) and EMG studies by Teasdall and Magladery (1959) showed that it is essentially a spinal reflex which can be modified by activity from higher centres in the CNS via the pyramidal tracts. An abnormal SAR in scoliosis associated with syringomyelia has been mentioned by a number of authors (Mehta 1992; Arai et al 1993; Charry et al 1994). In our experience an absent SAR on the same side as the convexity of the spinal curve is a consistent and early physical sign in the pathological evolution of this disorder. In some patients this may be the only abnormal neurological sign and precedes the development of other such signs.

In young children detailed neurological examination is often difficult. Routine MRI in children with scoliosis may be desirable but it is impractical for two main reasons. First, MRI is costly and not readily available to all specialist units and secondly young children do not tolerate it well and general anaesthesia is often necessary to obtain good-quality images. An absent SAR is therefore a reliable and useful indicator for selecting those children likely to have syringomyelia.

Radiological features which suggest syringomyelia include an increase in the width and depth of the cervical canal, bony abnormalities at the craniovertebral junction, diastematomyelia, and occipitalisation of the atlas (McRae and Standen 1966; Williams 1979). They are often difficult to detect and are generally observed retrospectively.

MRI findings in the Chiari-I malformation include herniation of the cerebellar tonsils by more than 5 mm, reduction in the subarachnoid space both anterior to the brain stem and posteroinferior to the cerebellum, descent of the brain stem, syringobulbia and cervicomedullary kinking (Pillay et al 1991) (Fig. 1).

The primary and the most important step in the successful treatment of scoliosis with syringomyelia is early surgical decompression of the syrinx. There are many different operative techniques and the ideal procedure is somewhat controversial (Madsen, Green and Bowen 1995). In patients with Chiari-I malformation the pathogenesis of syringomyelia appears to be due to the phenomenon of cranio-spinal pressure dissociation sometimes described as 'suck' (Williams 1969, 1980). Clinical and experimental work has shown that after episodes of raised thoracoabdominal pres-

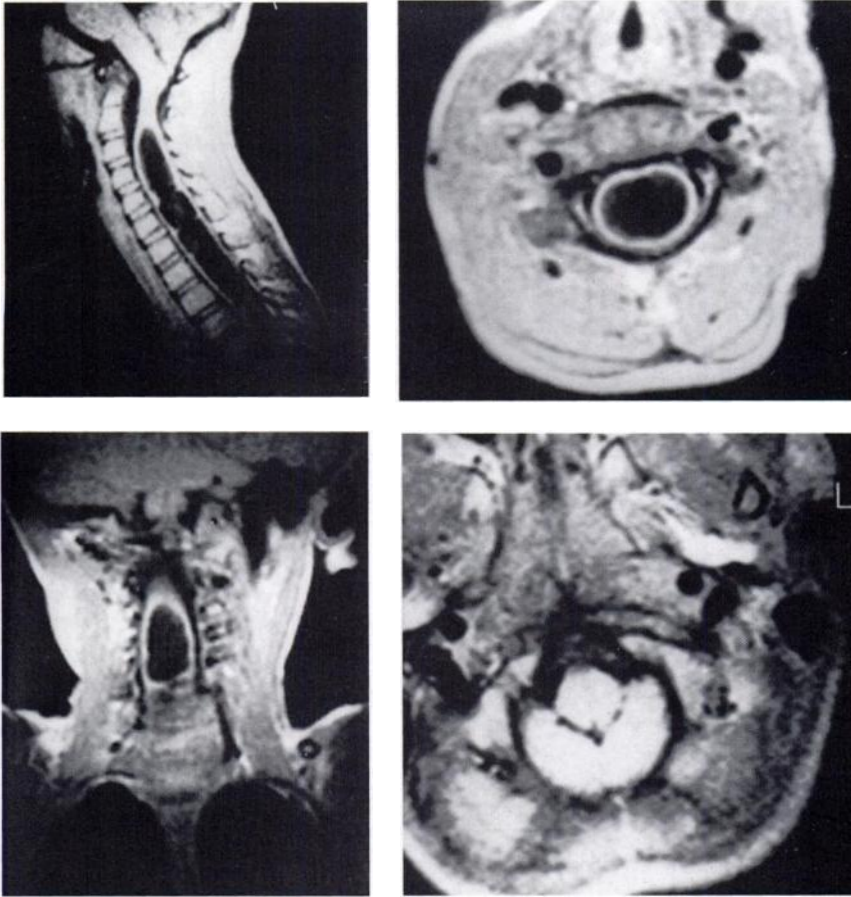


Fig. 1

MRI of the spine and hind brain before decompression showing a large syringomyelia and Chiari-I malformation.

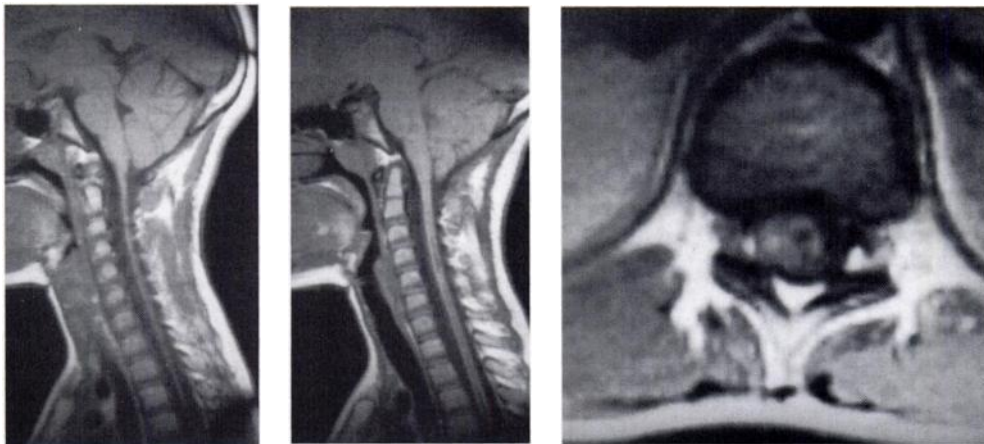


Fig. 2

MRI of the spine after decompression showing total collapse of the syrinx.

sure the herniated hindbrain or cerebellar tonsils behave as a valve and block the normal redistribution of the pressure between the subarachnoid space of the cranium and the spinal cord. The resultant pressure difference drives the CSF from the high-pressure ventricular system into the central canal of the cord which develops into the 'syrinx'. The basic principle of hindbrain decompression is to

reverse this phenomenon and to release the neural tissue impacted in the foramen magnum.

In 1965 Gardner first performed decompression surgery in which the foramen magnum was enlarged posteriorly, the lamina of the upper cervical vertebrae was removed, the dura was opened widely and the floor of the fourth ventricle was incised. The communication between the syrinx and

Table I. Details of ten children with scoliosis and associated syringomyelia

| Case | 1 | 2 | 3 | 4 | 5 | 6 | 7 | 8 | 9 | 10 |
|--|--|------------------------------|---|--|---|---|------------------------------|-----------------------|------------------------------|--------------------------|
| Sex | M | F | M | M | F | F | F | F | F | M |
| Age (yr mth) | 7 7 | 12 5 | 8 10 | 13 8 | 12 5 | 10 2 | 10 10 | 6 2 | 8 1 | 14 6 |
| Age at detection of scoliosis (yr mth) | 5 9 | 5 | 3 6 | 2 | 4 | 5 3 | 5 8 | 1 8 | 6 7 | 3 6 |
| Age at diagnosis of syringomyelia (yr mth) | 6 3 | 11 9 | 6 6 | 7 | 6 2 | 6 3 | 7 2 | 4 11 | 7 11 | 8 7 |
| Type of curve | Right thoracolumbar | Left thoracolumbar | Left thoracolumbar | Right thoracolumbar | Right thoracolumbar | Left thoracolumbar | Left thoracolumbar | Right thoracolumbar | Right thoracolumbar | Left thoracolumbar |
| Symptoms | Abnormal gait, Right hemiparesis | Abnormal gait | Abnormal gait | Abnormal gait | Abnormal gait, Right hemiparesis, Enuresis, Headaches | Asymptomatic | Asymptomatic | Asymptomatic | Asymptomatic | Asymptomatic |
| Abnormal sensory signs | Yes | Yes | No | Yes | Yes | Yes | Yes | No | Yes | No |
| Abnormal motor signs | Yes | No | Yes | Yes | No | No | No | No | No | No |
| Superficial abdominal reflex | -/+ | +/- | +/- | -/+ | -/+ | -/- | -/- | -/- | -/+ | +/- |
| Other abnormal reflexes | Absent right cremasteric reflex, Up going plantars | Absent left ankle reflex | Absent left cremasteric reflex | Up going plantars, brisk lower limb reflexes | None | Absent right quadriceps and biceps reflexes | Up going plantars | None | None | None |
| Initial Cobb angle (degrees) | 30 | 24 | 28 | 38 | 25 | 38 | 31 | 62 | 38 | 24 |
| Latest Cobb angle (degrees) | 33 | 10 | 41 | 61 | 27 | 48 | 22 | 52 | 60 | 26 |
| MRI findings other than the syrinx | Chiari I | Chiari I | Tethering of conus to S2 and intraspinal lipoma | Chiari I | Chiari I | Chiari I | Chiari I | Multiloculated syrinx | Chiari I | Chiari I |
| Operative procedure | Foramen magnum decompression | Foramen magnum decompression | Lumbar laminectomy and syringoarthachnoid shunt | Foramen magnum decompression and fascia lata graft | Foramen magnum decompression | Foramen magnum decompression | Foramen magnum decompression | None | Foramen magnum decompression | Syringoarthachnoid shunt |
| Revision surgery | - | - | - | Syringopleural shunt | - | C1 arch division | - | - | - | - |
| Postoperative neurological progress | Improved | Improved | Improved | Improved | No change | No change | Improved | - | Improved | Improved |

the fourth ventricle was closed by a plug of muscle in the obex. Since then a number of authors have expressed reservations regarding the use of an obex plug and advocated modifications to the original procedure. Williams (1978) avoided the use of an obex plug and created an artificial cisterna magna by incision and suture of the dura and the arachnoid. Matsumoto and Symon (1989) reported a higher mortality and complication rate with Gardner's operation and recommended craniovertebral decompression and syringoperitoneal shunting. Logue and Edwards (1981) also observed a higher complication rate with Gardner's operation and preferred a simple posterior decompression leaving the dura open but preserving the arachnoid membrane. Syringostomy is reserved for selected cases.

In foramen magnum decompression the cerebellar tonsil herniation is decompressed by a combination of C1 laminectomy and enlargement of the foramen magnum, without opening the dural sac. In our first patient the dura was opened and a fascia lata graft inserted. Despite neurological improvement, this patient had an inadequate decompression of the syrinx and later required a second operation to insert a syringopleural shunt. In the next six operations the dura was left intact. In one patient MRI revealed the remnants of the lamina of C1 which appeared to be the cause of incomplete decompression of the syrinx. Further surgery was required in the form of C1 arch division. Overall, our short-term results appear to be satisfactory. The latest MRI in all seven patients confirmed complete collapse of the syrinx. None of our patients has deteriorated and five have shown considerable neurological improvement.

In idiopathic infantile or childhood scoliosis a plaster jacket is used to counteract the effect of scoliosis and the child's natural growth is used to correct the residual spinal and ribcage deformities (Mehta 1984). For scoliosis asso-

ciated with syringomyelia, however, the results are less predictable.

We have observed that a plaster jacket can control the spinal curve, but due to the underlying neurological pathology the deformity can rapidly deteriorate if this treatment is discontinued. We also found that the use of a brace alone is inadequate. For advanced curves, the plaster jacket can control the deformity until adolescence, when surgical correction may be undertaken. For small curves, early surgical decompression of the syrinx and a plaster jacket can control or halt the progression of the curve until skeletal maturity and avoid the need for the surgical correction of the scoliosis. We believe that a plaster jacket in combination with surgical decompression of the syringomyelia slows down or halts the progression of the spinal deformity and is useful in improving the cosmetic appearance of the ribcage deformity.

Conclusions

- 1) Syringomyelia is an important cause of scoliosis in children and diagnosis relies on a high index of suspicion and a thorough neurological examination.
- 2) Neurological signs in patients who present with thoracic curves are often subtle.
- 3) An absent SAR on the same side as the convexity of the curve is an early and sensitive indicator of underlying syringomyelia. Sometimes this may be the only abnormal neurological sign.
- 4) Early surgical decompression of the syrinx is associated with recovery or stabilisation of the neurological deficit and reduction of the rate of progression of the scoliosis.
- 5) Preliminary results after decompression of the foramen magnum are favourable.

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