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Syringomyelia: cyst measurement by magnetic resonance imaging and comparison with symptoms, signs and disability

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SUMMARY The severity and distribution of symptoms and signs in patients with syringomyelia is considered to be dependent on the longitudinal and transverse dimensions of the syrinx and it is thought that clinical examination can identify the extent of the cyst. Magnetic resonance imaging has made the anatomical localisation of intramedullary spinal lesions more exact and probably more specific than previous methods of investigation. Syrinx length, diameters, cyst:cord and cord:canal ratios have been studied in 12 patients with syringomyelia to assess whether the dimensions of the syrinx relate to the clinical findings. The length of syrinx appeared to be related to cyst diameter, cyst:cord and cord:canal ratios. Patients with a small syrinx tended to have a small cyst diameter, and small cyst:cord and cord: canal ratios. No significant relationship was found between muscle wasting or weakness, distribution of sensory loss, degree of disability or distress and the dimensions of the syrinx. These findings should be borne in mind when surgical management is being considered.

Syringomyelia predominantly affects the cervical region but cavities may extend cranially, producing syringobulbia, or caudally involving the thoracic and even lumbo-sacral segments.[1] Several neurological textbooks[2–4] state that the specific signs depend on the anatomy and dimensions of the cyst, in particular its cross sectional location and length determine the extent of dermatome involvement. This would be in keeping with the theory that cyst enlargement in the transverse or longitudinal dimensions causes direct damage to central cord structures, and also provides the rational basis for cyst draining or decompressive operations.

The presumed dimensions of the syrinx, as well as the severity of its clinical effects and consequent disability, are important factors in decisions about management. Magnetic resonance imaging (MRI) provides an accurate non-invasive method of imaging intrinsic cord lesions and has made the localisation and evaluation of cystic intramedullary spinal lesions more sensitive and specific.[5] We have studied patients with unoperated syringomyelia to discover if the transverse and longitudinal dimensions of their syrinxes, or the ratio of cyst:cord diameters related directly to their symptoms, clinical signs or disability. The relationship between the diameter of the cord to canal at the level of widest cyst diameter was also measured to assess if extrinsic pressure or a narrow canal diameter might result in more extensive or severe disease.

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Patients and methods

Patients

Twelve patients with syringomyelia were studied. There were eight females and four males. Their ages ranged from 22–58 years, with a mean age of 42 years. Mean age at the time of the first symptom was 31 years and length of history ranged from 2 to 35 years. Eleven patients did not have evidence of any preceding cause. One patient (case 7) had suffered a subluxation of her 6th cervical vertebrae in an accident 4 years previously.

In three cases, the diagnosis was made fortuitously. One patient presented with neck ache and cough headache and MRI of the posterior fossa was requested; this revealed a tonsillar herniation and an extensive syrinx. A second, with a presumptive clinical diagnosis of multiple sclerosis, had cranial MRI carried out and this demonstrated the upper end of cervical syrinx. The third patient had evidence of hyrocephalus on cranial CT examination and this was confirmed and in addition, syringomyelia was demonstrated by MRI.

Patients were asked about their presenting symptoms, the speed of onset, and the rate and degree of progression. Specific features assessed included neck pain, arm pain, paraesthesia or numbness in the limbs, limb weakness or bladder disturbance. Clinical examination was performed within 2 weeks of MRI in all cases. The presence of wasting and weakness, and the distribution of sensory loss was recorded.

To assess the severity of the patients disability and distress, a questionnaire was completed. This took into account the activities of daily living, mobility and the degree of reliance on other people, as well as a measure of the patient's subjective distress. The mobility of the patient was scored in points 0: able to go outdoors with no difficulty; 1: able to go outdoors but only with difficulty; 2: not able to go outdoors except with help, 3: bedridden/chairbound.

If patients were unable to care for themselves and required help in (a) washing, (b) dressing, (c) feeding, (d) toileting, a further two points were given for each. The total score for disability could therefore range from 0–11. Patients were asked to place a cross on a unmarked 10 cm line to represent the degree of overall distress their disorder causes them. This line was then subdivided into five segments of 2 cm each and graded from 1–5 depending on increasing severity of subjective distress. The degree of combined disability and distress could therefore score a maximum of 16 points. Eight patients underwent operation and in each the MRI diagnosis was confirmed.

Imaging

Magnetic resonance imaging of the posterior fossa and spine was performed in sagittal and selected axial planes using a Picker vista 1100 0·15T resistive magnet operating at 6·38 MHz. Four slice T1 weighted sequences such as the Inversion Recovery (IR) 780/400/40 and short echo time (TE); short repeat time (TR) Spin Echo (SE) 512/40 were used routinely with the standard head coil (fig 1) and purpose built spine coils (fig 2). Using 8 mm contiguous slices, 4 averages and a 256^2 high quality images were obtained in 8·7 minutes. T2 weighted sequences such as SE 2200/80 or SE 1100/80 were used at the radiologist's discretion.

To define the full extent of a syrinx from the brain stem to the conus required three sets of sequences and took approximately one hour. The SE sequences produced the best signal/noise ratio and spatial resolution on our imager but the highest tissue contrast between CSF and cord was produced by the IR sequence (fig 3).

The presence and number of cysts, if multiple, were recorded. Cyst site and length in relation to the vertebral bodies was recorded and cyst diameters, cyst:cord and cord:canal ratios measured with electronic calipers, after enlarging the area of interest by × 4 on the viewing console. Cervical cord diameter was measured through the cyst at its broadest diameter. The measurement of the cervical canal diameter was taken at the same level to include the signal

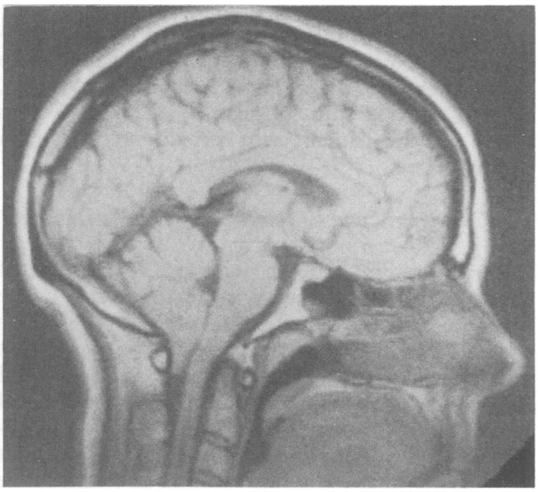

Fig 1 *SE 512/40 sagittal head scan demonstrating "peg like" tonsillar herniation and the upper end of a syrinx.*

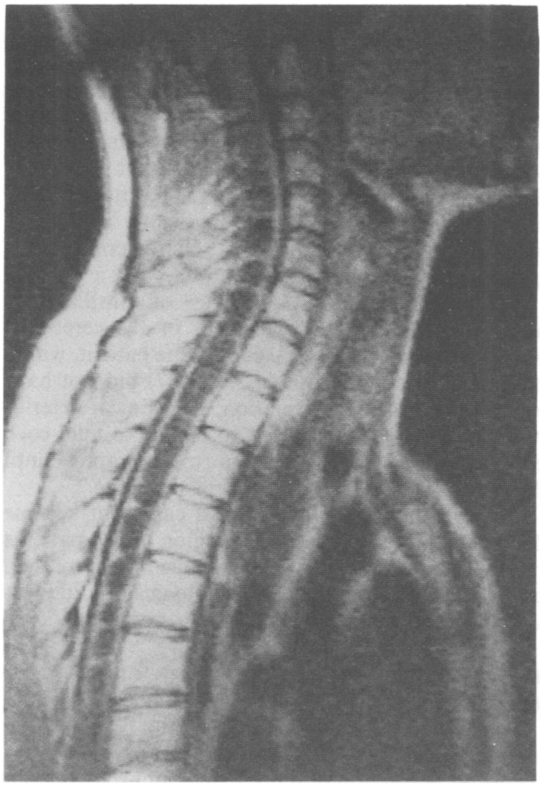

Fig 2 *SE 512/40 sagittal scan using local spinal cord, demonstrating a large cervico-thoracic syrinx.*

void representing cortical bone. The presence of bulbar extension, ventricular connection or tonsillar herniation was noted. Arnold Chiari type 1 malformation was reported when there was evidence of displacement of the cerebellar tonsils below a line drawn between the cortical lip of the anterior and posterior margins of the foramen magnum on a midline sagittal image but without displacement of the 4th ventricles. All measurements were reported independently, without access to the clinical information.

Results

Clinical and radiological characteristics

The onset of symptoms was so slow in nine patients that they were not able to say accurately when the symptoms started, but three patients described the onset symptoms as occurring fairly quickly over a period of approximately one week. Seven patients described a slowly progressive worsening of symptoms over some years and five had experienced a stepwise deterioration. Neck or arm ache was the presenting symptom in eight patients and was a symptom at the time of MRI in 10 patients. Four patients complained of numbness in the upper limbs and four complained of weakness in the upper limbs as an initial presenting complaint and at the time of investigation these symptoms occurred in nine and seven patients, respectively. At MRI five patients complained of unilateral sensory symptoms in the upper limbs. Nine patients described stiffness or unsteadiness when walking. Eleven of the 12 patients felt that severity of the disease had worsened with time, and five had noticed a stepwise progression, frequently associated with neck pain, which often had characteristics suggestive of nerve root pain.

Ten patients had MRI evidence of tonsillar herniation with caudal displacement of the cerebellar tonsils well into the cervical canal. One patient, with a post traumatic syrinx (table 1, case 7) did not have evidence of tonsillar herniation; her clinical deterioration was initially suspected to be due to mild cord compression demonstrated on myelography until three cysts were found on MRI of the spine.

The syrinx was central in most cases but became dorsally situated in the cervical region in two patients (table 1, cases 4 and 12). Two patients had more than one cyst in the cord and this radiological finding was varified at operation.

RELATIONSHIP BETWEEN MRI AND CLINICAL FINDINGS

(A) Tonsillar herniation and neck and arm pain

Tonsillar herniation of more than 3 mm was found in 10 patients (table 2). In the remaining two patients the cerebellar tonsils lay 3 mm above the foramen magnum, in one case associated with a posterior fossa arachnoid cyst. Ten of the 12 patients complained of occipital headache or neck ache. These included both patients without imaging evidence of tonsillar herniation, three of five patients with tonsillar herniation, less than 10 mm and all five patients with herniation greater than 10 mm. Arm pain was present in three patients with tonsillar herniation less than 10 mm and two patients with herniation greater than 10 mm.

Fig 3 *IR 780/400/40 axial scan through C2/3 vertebral level showing a large central syrinx.*

(B) Cyst length and its relationship to symptoms and signs

The relationship between wasting, distribution of weakness and sensory signs, is shown in table 1. The upper limit of the syrinx in all patients was C2 or higher. In three cases, imaging evidence of a syrinx was confined to the high cervical region (C2–C3) and in the others the syrinx extended from the cervical cord caudally to a vertebral level of T8–T10. Five patients had facial numbness and the MRI upper limit of the syrinx was C2 in three and C1 in two. Muscle wasting in the upper limbs was present in four of the nine cases with cervico-thoracic syringomyelia and in

Table 1 *Relationships between syrinx dimensions and clinical signs*

	Cyst length in Vertebral Body Lengths	*Cyst:cord ratio*	*Wasting*	*Weakness*	*Sensory loss*
1	C2	0·2	C8–T1 (B)	C7–T1 (B)	C6–T1 (B)
2	C2	0·33	—	C4–T1 (R) P (B)	C5–S4 (R)
3	C2 C3	0·22	—	C6–T1 (B) P (B)	T5–T6 (B)
4	C2–T8	0·22	—	C5–T1 (R)	V3–C6 (R) T2–T10 (R)
5	C2–T8	0·88	—	P (B)	T6–S4 (R) T7–S4 (L)
6	C2–T9	0·33	—	—	V2–C2 (L) C6–S4 (L) T6–S4 (R)
7	C2–C6 C6–C7 C7–T10	0·82	C6–C8 (B)	C5–T1 (B) P(B)	C7–T1 (L) C6–T1 (R) T4–S4 (B)
8	C2–T10	0·33	C5 (R)	C4–C6 (R) P (R)	C4–T1 (R)
9	C2–T10	0·73	—	—	V3–C5 (L) T7–T9 (L)
10	C1–T10	0·43	C4–C6 (B)	C4–C6 (B)	V3–C6 (B)
11	C1–T10	0·73	C8–T1 (L)	C5–T1 (L) P (B)	V2–S4 (L)
12	MED.–T11	0·80	—	C6–T1 (L) P (B)	C2–C5 (R) C2–L3 (L)

P = Pyramidal lower limb findings, B = Bilateral, R = Right, L = Left.

one of the three cases where only the cervical cord appeared involved. There was no relationship between cyst length and muscle weakness in the upper limbs or upper motor neuron findings in the legs.

Sacral sensory loss occurred in four cases with cervico-thoracic syrinx and one case with a cervical syrinx. Thus a consistent relationship could not be identified between the clinical features and the length of the cyst (fig 4).

(C) Cervical cyst:cord ratio and its relationship to symptoms and signs

The diameters of the cervical canal, cervical cord and syrinx at its broadest level were measured and results are shown in table 2. Every patient had a syrinx that involved the cervical region but cervical cyst:cord ratios ranged from 0·2–0·88. For analysis, the patients were separated into two groups; six had cord:canal ratios less than 0·4 and six ratios greater than this. In both groups, five patients had neck pain and four arm pain. Of four patients who complained of facial numbness, three had cervical cyst:cord ratio greater than 0·4 and four of seven patients who complained of arm weakness had ratios greater than 0·4. Equal numbers of both groups complained of paraesthsiae in arms or legs and there was no appreciable difference in numbers of patients complaining of leg weakness and stiffness or bladder disturbance.

The distribution of sensory loss, weakness and wasting is shown in table 1. There was no relationship between the cervical cyst:cord ratio and the extent of sensory involvement or the presence of sacral sparing.

Table 2 *MRI syrinx dimensions*

Case no	*Cyst length (In VB'S)*	*Tonsillar herniation (mm)*	*Cyst diameter (mm)*	*Cervical Cyst:cord Ratio*	*Thoracic Cyst:cord Ratio*	*Cervical Cyst:Canal Ratio*
1	1	23	2	0·2	—	0·43
2	1	5	2	0·33	—	0·55
3	2	4	2	0·22	—	0·56
4	7	8	2	0·22	0·16	0·82
5	7	17	8	0·88	0·88	1·0
6	8	15	3	0·33	0·63	0·85
7	9	—	9	0·82	0·82	0·73
8	9	—	3	0·33	0·75	0·85
9	9	16	8	0·73	0·73	0·85
10	10	3	6	0·73	0·83	0·875
11	10	6	8	0·43	0·80	0·85
12	12	13	8	0·80	0·46	0·83

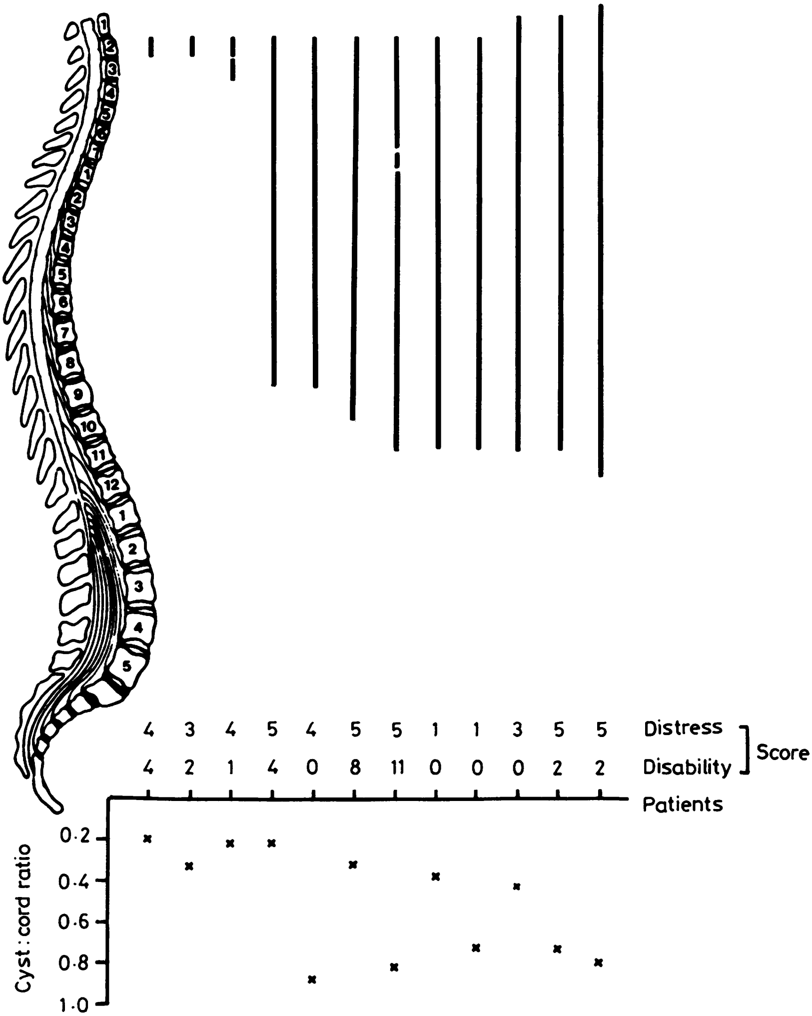

Fig 4 *Relationship of cyst length and cyst:cord ratio to distress and disability score.*

Two patients with cyst:cord ratios of less than 0·4 had muscle wasting in the upper limbs compared with three patients with a ratio of greater than 0·4. Five of the six patients with a ratio less than 0·4 had upper limb weakness and 4 had pyramidal weakness of the legs. Four patients with a ratio greater than 0·4 had upper limb weakness and five had pyramidal weakness in the lower limbs. Therefore, there was not an obvious relationship between the ratio of the diameter of the syrinx to that of the cervical cord and the patients symptoms or distribution of signs.

Similar comparisons were made between the diameters of the cord and the cervical canal. Six patients had a cervical cord:canal ratio of less than 0·85 and six a ratio of 0·85 or more (table 2). There was no evidence of any association between a high ratio (>0·85) and either the presence of wasting, or the presence and distribution of sensory signs or weakness.

Of four patients with a disability score of 4 or over, three had a cyst:cord ratio of less than 0·4 and two had a cord:canal ratio of less than 0·85. There was no obvious relationship between distress score and cyst:cord or cord:canal ratio (fig 4). Three of six patients with a combined disability and distress score of 7 or over had cyst:cord ratios less than 0·4 and three had cord:canal ratios less than 0·85. An association could not be found between the degree of disability or subjective distress and either the cervical cyst diameter or the ratio of cyst:cord or cord:canal.

(D) Thoracic cyst cord ratio and its relationship to symptoms and signs

Nine patients had imaging evidence of extension of the syrinx to involve the thoracic cord, with cyst:cord, ratios ranging from 0·16–0·88. Cyst:cord ratio was greater than 0·5 in six patients and the distribution of symptoms and signs in these cases was compared with the remaining six patients. Only one patient in either group complained of paraesthesia or numbness on the trunk or lower limbs and one in each group complained of bladder disturbance. Upper motor neuron distribution, lower limb weakness was present in only four of the six patients with a ratio greater than 0·5 and in three patients with a ratio $<0{\cdot}5$ in the other group. Diminution of pain sensation on the trunk or legs occurred in four patients with a cyst:cord ratio of greater than 0·5, three patients with a ratio of less than 0·5, and in two patients without imaging evidence of the syrinx extending to involve the thoracic region.

Scolosis was present in one case in each of these three groups. None of the patients had muscle wasting in the lower limbs. Four patients with cyst:cord ratios greater than 0·5 had disability and distress scores of less than 5, while all patients with ratios less than 0·5 had scores of 5 or over.

Despite the predilection for signs of syringomyelia to be most evident in the upper limbs, where present, the thoracic cyst:cord ratio was larger in four patients, similar in three and smaller in two patients than the cyst:cord ratio in the cervical region.

Lower motor neuron signs were not found. The "classical" findings of dissociated sensory loss and lower motor neuron findings in the upper limbs with hyperreflexia in the lower limbs were noted in only 50% of cases. In summary, it was not possible in any case to estimate accurately from any clinical features the radiological extent of the cavity as shown by MRI.

Discussion

In the past much emphasis has been placed on the anatomy of the syrinx as an explanation for the clinical effects,[2–4] and as an index of the indications and likely response to operation.[6] Clearly these views need to be revised. Our results suggest that there is no direct relationship between the imaging dimensions of a syrinx and the associated neurological findings and, therefore, the anatomy of the syrinx alone cannot provide a satisfactory explanation for the pattern of clinical signs, nor the severity of the disability they cause.

The predilection for wasting, weakness and dissociated sensory loss to affect the C6–T1 dermatomes, is well known, but the reason for this is not understood. In seven of the nine cases with a thoracic extension, the diameter of the syrinx in the thoracic cord was at least as large as that in the cervical area. The present study shows that the explanation can not be that the cervical region is the site of maximum cavitation. Patients with a normal or an atrophic spinal cord lesion and a cord:canal ratio within the accepted normal limits, were just as severely affected as those with an expanded cord and cord:canal ratios greater than 0·85. This view is supported by the finding of other authors using myelography with computed tomography.[7–8] It is not surprising that there is no relationship between diameter of syrinx and distribution and severity of signs. Central cord ischaemia may be associated with cord atrophy, and if the syrinx produces sufficient pressure, it may result in local ischaemia and atrophy.

Bulbar extension of syrinx is thought to be present in approximately 25% of cases at necropsy.[9] One patient in this study had MRI evidence of syringobulbia and yet five of the 12 patients had trigeminal sensory involvement. A possible explanation for this apparent discrepancy may be that the extension into the medulla or pons may be so small as to be beyond the resolution of the MRI or that trigeminal sensory involvement has been a result of the damage to the spinal trigeminal tract or nucleus which can extend to a caudal vertebral level of C3.[10–12] Therefore, it is not possible to predict accurately the MRI upper limit of the syrinx using this sign.

Asymptomatic patients with extensive syringomyelia are well recorded and of interest because they had a change in dimension of the spinal cord without clinical manifestations. Patients may not present with the classical findings of syringomyelia, and this may lead to incorrect diagnosis. Only 50% of the cases in this study presented with classical signs of syringomylia and the diagnosis was fortuitously made in three cases.

There was, in general, a direct relationship between the different dimensions of the cyst. Thus cysts of a small length tended to have a smaller cyst diameter and small cyst:cord and cord:canal ratio. If it were to be established, as suggested by Gebarski,[6] that only large cysts respond to shunting, MRI can provide the information necessary to allow the neurosurgeon to decide which patients may benefit from this procedure. Failure of surgical treatment may be due to failure of the operation to reduce the syrinx[13] and successful decompression can be assessed postoperatively and follow up scans performed if clinically indicated.

Magnetic resonance imaging is a non invasive, sensitive technique which has a specificity at least as high as contrast assisted myleography.[14] It has the advantages of absence of bone artefact and will provide a direct image of the spinal cord in several planes. It is the investigation of choice in patients suspected of having syringomyelia.

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