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ORIGINAL ARTICLE

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Can hindbrain decompression for syringomyelia lead to regression of scoliosis?

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Abstract Scoliosis in childhood develops secondary to syringomyelia in some children. The existing literature does not provide a clear answer as to whether surgical treatment of the syrinx can allow subsequent improvement of the spinal deformity, thus preventing the need for scoliosis surgery. This series comprised 16 patients with syringomyelia who presented with significant scoliosis in the absence of major neurological deficit. All underwent a hindbrain decompression, and follow-up ranged from 1 to 6 years (mean 2.5 years). Subsequent deformity surgery was necessary in eight cases, but the scoliosis was seen to improve or arrest its progression in six (37.5%). Improvement was found to be statistically more likely in children of younger age at the time of syrinx surgery and in those with left thoracic curves. Improvement occurred in 71.4% of those under the age of 10 at the time of hindbrain decompression.

Key words Syringomyelia · Scoliosis · Hindbrain decompression

Introduction

Syringomyelia is a well-recognised, though uncommon, cause of scoliosis in children. Although many such children will present with neurological signs and symptoms, modern imaging techniques may reveal an underlying syrinx as the cause of a scoliosis otherwise thought to be idiopathic [1, 9, 14]. Cases of syringomyelia that present with major neurological features in addition to any spinal deformity have conventionally been treated by addressing the neurosurgical problem initially with subsequent treatment of the associated deformity.

The development of magnetic resonance imaging (MRI) has led to increased spinal imaging in cases of apparent idiopathic scoliosis, with the resultant diagnosis of syringomyelia in association with little or no neurological deficit. This has led some authors to explore the role of surgical treatment of the syrinx and to examine its effect on curve progression. However, no clear answer regarding the efficacy of such an approach has developed. This may be partly due to the fact that previous studies have contained small cohorts of patients consisting of cases with varying degrees of neurological deficit and in whom the syringomyelia was treated by a variety of surgical methods [2–4, 7, 8, 11, 12, 16]. Within those series, the number of patients who actually underwent some type of syrinx surgery ranged from only 6 to 21 (mean 12). Due to such small numbers with differing degrees of neurological deficit, age, curve severity and types of surgical treatment it is difficult to deduce whether the treatment of the syrinx itself can reverse the deformity process in any or some cases.

This study presents the preliminary results of a carefully defined population with syringomyelia, who presented with scoliosis and who each underwent an identical surgical procedure for the treatment of their syrinx.

Materials and methods

As the numbers of patients with syringomyelia who actually present with scoliosis as their initial symptom is comparatively small, one author (D.S.) scrutinised the operative records of three large neurosurgical departments from 1990 onwards. All cases undergoing hindbrain decompression (HBD) for syringomyelia who were aged under 17 years at the time of surgery were identified. All

cases of scoliosis seen in the associated deformity surgery department were also screened to identify any further patients with syringomyelia. Twenty-eight patients whose presentation was with spinal deformity were identified. Of these, 12 were excluded for the following reasons. Four patients found in the deformity departments had only very small syrinxes, which were not treated surgically. In two patients, deformity surgery had been performed prior to the diagnosis of syringomyelia being made. In a further six patients there were other associated conditions of spinal dysraphism. The remaining 16 patients form the cohort group for this study.

All 16 patients presented with scoliosis as the initial symptom. Syringomyelia was diagnosed in all by MRI. Careful neurological examination revealed subtle neurological signs such as loss of abdominal reflexes in ten patients, and two patients had minimal upper limb weakness. After gaining careful informed consent from the child and the parents, all underwent a standard HBD of their Arnold-Chiari malformation as described by Williams [17].

At the time of HBD, the ages of the patients ranged from 7 to 16 years (mean 11 years). All of the patients had an Arnold-Chiari malformation and syringomyelia. The syrinx affected the cervical spine in all cases and also the brainstem in two. The syrinx extended over 5–21 vertebral segments (mean 14 segments). The pattern of the curvature showed the scoliosis to be left thoracic in eight; right thoracic in four; and a double curve in the remaining four cases. At the time of HBD, the severity of the deformity ranged from 15° to 67° (mean 41°) as measured by their Cobb angle.

The rate of curve progression was assessed on serial anteroposterior spinal films taken during follow-up and, where possible, prior to the HBD. The end points of the study were the final Cobb angle at either latest follow-up or at the time of deformity surgery. The decision that deformity surgery was necessary was made by the scoliosis surgeon purely on the grounds of the rate of curve progression or the severity of the deformity. The duration of follow-up was from 1 to 6 years (mean 2.5 years) in patients who did not require deformity surgery, and from 1 to 5 years (mean 2.5 years) in those who did eventually require deformity surgery. Children who have not yet reached skeletal maturity remain under careful follow-up.

Statistical analysis was by the Anova method, chi-squared or Student's or paired *t*-test as appropriate.

Results

The clinical data of those patients who did not require deformity surgery are shown in Table 1. Table 2 shows the data for those who did require scoliosis surgery. Eight of the 16 patients eventually underwent deformity surgery. The deformity improved in five patients, stabilised in one and progressed to a degree that did not require deformity surgery in the remaining two patients, who did not require scoliosis surgery. The amount of improvement ranged from 9° to 19° (mean 11.6°). The magnitude of the scoliosis as assessed by the Cobb angle at the time of HBD had no bearing on the need for deformity surgery (Table 3).

Table 4 displays the data regarding the rate of curve progression, when known, both prior to and following

Table 1 Clinical data in cases not requiring scoliosis surgery (*HBD* hindbrain decompression, *FU* follow-up, *RTh* right thoracic, *LTh* left thoracic)

Sex	Age at HBD (years)	Scoliosis type	Curve at HBD (°)	Curve at FU (°)	FU (years)
F	7	RTh	40	30	3
M	10	RTh	15	25	6
M	11	LTh	40	53	2
F	9	LTh	51	40	1
F	16	LTh	40	30	2
M	9	LTh	40	40	2
M	9	LTh	40	30	3
M	9	LTh	32	13	1

Table 2 Clinical data in cases requiring scoliosis surgery

Sex	Age at HBD (years)	Scoliosis type	Curve at HBD (°)	Curve at surgery (°)	FU (years)
F	8	Double	28	60	5
M	7	RTh	45	60	3
F	14	Double	45	68	1
F	15	Double	67	90	1
M	16	RTh	40	58	2
M	11	Double	42	50	1
M	12	LTh	52	65	3
M	14	LTh	45	51	4

Table 3 Effect of curve size at HBD and the need for deformity surgery (P = n.s. (Student's t-test)

	Cobb angle at HBD	Mean Cobb angle
No deformity surgery	15°–51°	37°
Deformity surgery	28°–67°	46°

Table 4 Rate of curve progression before and after HBD

Age at HBD	Curve type	Rate before HBD (degrees/year)[a]	Rate after HBD (degrees/year)[a]	Scoliosis surgery (yes/no)
7	RTh	2	–3	N
10	RTh	1.5	2	N
11	LTh	9	6	N
9	LTh	0	–19	N
16	LTh	0	–7	N
9	LTh	0	–8	N
9	LTh	7	–3	N
9	LTh	Not known		
8	Double	4	4	Y
7	RTh	6	9	Y
14	Double	18	14	Y
15	Double	12	17	Y
16	RTh	5	20	Y
11	Double	7	8	Y
12	LTh	16	6	Y
14	LTh	Not known		

[a] Figures extrapolated to full year if final radiograph was not at complete year end

Table 5 The effect of age on curve progression

	Curve at HBD	Improvement
Age at HBD < 10 years	28°–51°	5/7 (71.4%)
Age at HBD > 10 years	15°–67°	1/9 (11.1%)
	P = n.s. (t-test)	$P < 0.05$ (χ^2)

HBD. Of the eight left thoracic curves, five showed improvement following HBD. Only one of the four right thoracic and none of the four double curves showed improvement. Where it was possible to compare the rate of progression of the curve both before and following HBD, the improvement in those with a left thoracic curve was highly significant ($P < 0.009$; paired t-test). Of those children who ultimately required scoliosis surgery, the curve progressed in all cases, but the rate of progression diminished in two cases. However, as their curves were respectively 45° and 52° at the time of HBD, they still required surgical correction of their deformity despite the reduced rate of curve progression.

In ten of the 16 cases it was possible to assess the degree of syrinx collapse on postoperative MRI scans using the method described by Grant et al. [6]. The degree of syrinx collapse ranged from 10 to 90% (mean 57%), but this had no significant effect on the need for deformity surgery. The sex of the patient had no significant effect either.

The rate of curve progression following HBD was analysed by linear regression analysis using sex, age, curve type and the size of the curve at HBD as variables. The only factor that showed a significant effect was age ($P = 0.045$; ANOVA). Table 5 shows the effect of age on curve progression in those below or above the age of 10 years at the time of HBD. Those under the age of 10 years showed a significantly greater incidence of curve improvement ($P < 0.05$; χ^2).

Discussion

Scoliosis in the growing child does not necessarily progress, and in some cases may improve spontaneously. Soucacos et al. [15] examined 839 cases with idiopathic scoliosis of at least 10° and found progression in 14.7% and improvement in 27.4%. Similarly, Lonstein and Carlson [10] followed 727 children with idiopathic scoliosis with curves of less than 30° and found progression in 78.8% of children with initial curves of between 20° and 30°.

Clearly it is possible that the rate of progression of scoliosis in a population of children with syringomyelia may be dissimilar to that in patients with idiopathic scoliosis. Charry et al. [2] reported on 25 patients with scoliosis secondary to syringomyelia. Thirteen of those patients were observed without treatment of their syrinx, and in 12 of them the scoliosis was seen to progress, particularly in the more mature patients.

Previous studies [2–4, 7, 8, 11, 12, 16] have attempted to analyse the effect of the treatment of syringomyelia on scoliosis progression. However, these studies report only small numbers of treated patients, who frequently had other associated dysraphic states and who were treated by a variety of neurosurgical procedures. They also frequently presented with significant neurological deficit in addition to the spinal deformity.

Charry et al. [2] reported nine patients within their series who underwent HBD. The mean age of this group was only 8 years, with five patients being aged 5 or less. At the time of HBD, their curves ranged from 15° to 44°, with a mean of 34°. In seven of the nine cases, the deformity either stabilised or improved by an average of 21° following HBD.

Tomlinson et al. [16] described six patients with syringomyelia in the absence of other dysraphic conditions. Five had scoliosis, though it was the presenting symptom in only two. Five of the cases were treated by a syrinx shunting procedure, and in three cases the deformity either stabilised or improved. No information was given about the magnitude of the curvature, which makes the assessment of the significance of this study uncertain. Their ages ranged from 6 to 15 years, with a mean of 9 years.

Farley et al. [4] reported 28 patients, in whom a syrinx shunting procedure was performed in 21. Their ages ranged from 1 to 16 years, with a mean of 10 years, and in twelve of the patients the deformity was less than 30° at the time of surgery. They found no difference in curve progression either between the operated and non-operated groups or by age, sex, or the type of curve. The discrepancy between their results and our own may be related to their choice of surgical treatment of the syrinx, which leaves the primary mechanism of the cause of the syrinx (the tonsillar herniation) untreated.

Muhonen et al. [11] described 11 patients with Chiari malformations and deformity, of whom eight had syringomyelia. Their cases showed a high incidence of neurological abnormality. They underwent HBD with a shunting procedure in all, with obex plugging in some cases. Their preoperative deformity ranged from 15° to 54°, with a mean of 29°, and their mean age was 10 years. Only one of their patients showed progression of their curve.

Ghanem et al. [5] reported seven cases of syringomyelia with scoliosis treated by HBD with occipitocervical fusion. Only one case showed an improvement in the deformity and five cases deteriorated, though all had a preoperative curve greater than 40°.

The current series reports 16 patients with syringomyelia whose first presentation was with scoliosis in the absence of major neurological deficit and in whom the syrinx was treated by a standardised surgical procedure. Associated dysraphic states were excluded. The mean fol-

low-up from the time of HBD was 2.5 years. All cases showed significant degrees of spinal deformity, with the mean curvature at the time of HBD being 41°.

The degree of spinal deformity was found to improve or stabilise in 6 of the 16 cases (37.5%). It is interesting to note that the magnitude of the deformity at the time of HBD had no bearing on the subsequent progress of the deformity. Neither the sex of the patient nor the degree of syrinx collapse were significant factors. However, this study suggests that, if HBD is performed before the age of 10 years, there exists a significant chance of deformity improvement, with 71.4% of the curves showing improvement. In addition, the presence of a left thoracic curve was associated with a significantly greater chance of improvement. Although the degree of improvement of the deformity in the six cases was small (range 9°–19°; mean 11.6°), previous authors have accepted a change of only 5° to represent a clinically significant change [15].

As in all previous studies, examination of the effect of HBD for syringomyelia on the progress of scoliosis is hampered by the relatively small numbers of cases suitable for inclusion. This study of a well-defined population with long-term follow-up does suggest that early intervention by means of HBD may lead to improvement in the severity of scoliosis associated with syringomyelia in younger children, particularly in association with a left thoracic curve.

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