

Syringomyelia and its surgical treatment— an analysis of 75 patients

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SUMMARY A consecutive series of 75 patients with syringomyelia is presented, all of whom were treated by cranio-vertebral operations. Attention is drawn to the difficulty in assessing the results of treatment but 56 were stabilised or showed modest improvement after surgery. Occluding the central canal appeared to have no greater influence on the progression of the disease than did simple decompression and did have a higher incidence of complications. Upper motor neurone weakness, joint position sense and central neck pain are the features most likely to improve and it is concluded that relieving the medullary compression resulting from a Chiari type 1 malformation, rather than influencing the syrinx, is the means by which this may occur. Simple decompression with preservation of the arachnoid membrane, combined with syringostomy in certain cases, is recommended.

The condition of syringomyelia has been recognised as a nosological entity for over 150 years,¹ and operative treatment was carried out as early as 1891,² but even at the present time, the place of surgery in its management and the results that may be expected are still uncertain despite many publications on this subject. The factors responsible for this state of affairs are numerous and comprise the rarity of the disease, the lack of a uniform classification, the multiplicity of signs from random and asymmetric involvement of the tracts of the spinal cord and medulla oblongata and, in many patients, its long duration so that premature conclusions have tended to be drawn from too short a post-operative follow-up. Again, in many reports the results have been graded by such categories as “excellent,” “good” or “poor” which, despite definition, has little meaning in a condition with so much variation in the response of individual neurological modalities to surgical treatment, some of which may show improvement and others deterioration. The occasional occurrence (although not in this series) of a patient,

usually young and with a short history, who makes a surprising and near full recovery has tended to have had an exaggerated influence on clinical impression and has distorted the overall picture of the response to surgery.

As a result of these several factors a clear idea of which neurological aspects can be expected to improve with operation, which cases can be expected to benefit, which patients are unsuitable for operation and what are the acceptable risks of surgery has not emerged.

We have attempted in this presentation to correct some of these deficiencies by restricting our analysis to 75 consecutive patients who presented with the commonest variety of syringomyelia and who have been extensively investigated clinically and radiologically and have been reviewed many times during a follow-up of mean duration of five years.

Case material

CLASSIFICATION OF PATHOLOGICAL ANATOMY

We have modified Barnett's classification of syringomyelia³ but retained the distinction between two main types:

1 *Syringomyelia due to primary dilatation of the central canal of the spinal cord.* This is the more common type sometimes referred to as communicating syringomyelia.⁴ It is almost always associated with

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an abnormality of the foramen magnum, usually the Chiari type 1 malformation (90% in this series), and is due to dilatation of the central canal (simple syringomyelia) although fluid often transgresses the bounds of this canal forming irregular channels penetrating both outwards and vertically in the cord parenchyma (syringomyelia).

2 *Syringomyelia without primary involvement of the central canal.* This may be due to spinal trauma or spinal arachnoiditis. In this variety the cyst arises in the cord substance without primary communication with the fourth ventricle or subarachnoid space.

The natural history of these two varieties differs as does the method of surgical treatment. We have, therefore, excluded from consideration all the Type 2 cases and report a series of 75 patients whom we believe conform to the more common Type 1, due primarily to dilatation of the central canal.

The 75 cases can be divided into three groups:

1 Syringomyelia with Chiari type 1 malformation: 68 cases (syringomyelia proven: 58 cases, syringomyelia strongly suspected on clinical evidence: 10 cases).

2 Syringomyelia associated with other lesions of the foramen magnum: 4 cases.

3 Syringomyelia without anomaly at the foramen magnum: 3 cases.

Chiari malformation and its association with syringomyelia

The term Chiari I malformation is the first of the four categories of hind brain anomaly described by Chiari⁵ and in essence comprises a herniation of the *tonsils* of the cerebellum through the foramen magnum with symptoms presenting in adult life and which is only rarely associated with spina bifida. This is in contrast to the Chiari II malformation which is a herniation of the *vermis* of the cerebellum largely presenting in infants and children and usually associated with spina bifida at some level in the spine and often with hydrocephalus. There are surprisingly few exceptions to these two generalisations. In both types a downward displacement of the medulla oblongata through the foramen magnum may also occur. Varieties III and IV of the Chiari classification are not concerned with syringomyelia. All 68 cases reported here with the cerebellar herniation were of the Chiari I variety and there was no case of spina bifida.

The neurological symptoms and signs in syringomyelia with the Chiari malformation derive from a permutation and combination of three mechanical factors:

1 Direct compression by the tonsils of the fibre tracts and nuclei in the medulla and upper segments of the cervical cord within the foramen magnum and upper cervical canal.

2 Herniation downwards of the brain stem which results not only in stretch of the cranial nerves from as high as CN 5 down to CN 12, but also permits higher segments of the medulla to be subjected to

pressure by the tonsils at the foramen magnum.

3 Distortion and disruption of the long tracts in the spinal cord by the central cyst itself.

These abnormalities of the nervous structures are sometimes associated with congenital cranio-vertebral anomalies (32% of this group) which may suggest the diagnosis, but the disconcerting feature is the large number of patients who have normal skull radiographs.

We have treated a total of 122 cases of the Chiari I malformation by operative decompression. In the 68 cases reported here the anomaly was associated with syringomyelia but in the other 54 patients there was no evidence of a syrinx (these cases will be the subject of a later publication). The appearance at operation of the tonsils was similar in cases with and without syringomyelia.

The frequent association of syringomyelia with the Chiari I anomaly established by Gardner⁷ has been the subject of much ingenious speculation, notably by Gardner himself, who suggested that the abnormal hind brain anatomy causes a disturbance of cerebrospinal fluid (CSF) dynamics whereby the pulsating ventricular fluid tends to be diverted by the obstructing tonsils down a patent central canal in the spinal cord to form the syrinx. Gardner devised an operation to block the mouth of the canal and by tonsillar decompression to restore normal hydrodynamic flow and this procedure was carried out on our patients until five years ago. Williams⁸ suggested a modification to this theory, but still proposed that the syrinx fills from the fourth ventricle. This "hydrodynamic theory" has come under criticism from Banerji and Millar⁹ and Aboulker¹⁰ who give several cogent arguments against it and also from ourselves. Our views mainly rest on the basis of a random collection of 19 patients who had died with syringomyelia (which was not necessarily the cause of death) in whom the central canal leading from the fourth ventricle to the cyst was shown on serial microscopical section to be totally occluded or very narrow and incapable of transmitting fluid in a pulsatile fashion. (These cases will be published later.)

It may be therefore, that the relationship of the malformation and syringomyelia is not a causal one and we now regard them as associated congenital lesions each capable of inflicting neurological damage. The important implication of this view is that tonsillar decompression may relieve those symptoms due to medullary and upper cervical cord compression, but will not influence those arising independently from the syrinx, although it is possible that widening the foramen magnum may have a favourable influence on *progression* of the syrinx by restoring the normal cranio-spinal hydrodynamics.

Clinical manifestations

GROUP 1A

In this group of 58 cases all had the Chiari malformation with syringomyelia established by un-

equivocal radiological evidence or confirmation at operation or both. The average age at time of diagnosis was 45 years, the male to female ratio 27:31 and the average length of history 7.8 years. The age of onset ranged from birth (from the observation of a weak arm) to 56 years, an average of 37.2 years. The length of history was very variable: some patients were seriously crippled within two years, others were still able to get about after 20 years. The onset of pain and temperature loss was sudden in four cases although the symptoms in these patients subsequently progressed slowly. A rapid deterioration sometimes occurred during the course of a slow progression following a cough or sneeze or straining. It is tempting to think that these sudden changes may be related to rupture of the ependymal wall of the cyst and extrusion of fluid out into the substance of the cord.

The striking feature was the vast range of variables in the neurological picture.

Pain This was a common symptom occurring in 31 cases and two types were distinguishable. (1) A sub-occipital and lower central neck pain, particularly provoked by coughing or sneezing occurred in eight patients and was probably due to a compression or stretch of the upper cervical roots by the Chiari malformation itself. This view is supported by the fact that this particular symptom has been seen to occur commonly in patients harbouring only the Chiari malformation without an associated syringomyelia (series to be published later.) One patient complaining solely of increasing neck pain had radiological and operative evidence of a syrinx but no abnormal neurological signs. (2) In 23 cases the pain had an aching or burning quality and tended to be lateralised in the face, neck, limbs or trunk and occurred within the area of pain and temperature impairment, sometimes at the upper or lower border. In one case the pain moved ahead of the advancing level of sensory impairment.

Neurological signs

(a) *Sensory change* The characteristic reduction of pain and temperature sensation occurred in 54 cases, usually to a variable degree on both sides of the body, particularly affecting the head, upper limbs and often of a cape or half cape distribution. However, there were numerous variations on this basic pattern. One patient had a unilateral area of suspended dissociated loss as the only abnormal sign: in another it was confined to a single dermatome and in a third patient localised to one ear. Four patients had "root entry zone syndromes" in which all modalities of sensation were impaired in

one arm only. No deficit of pain or temperature occurred in four patients. Light touch was usually well preserved and, if involved, was confined to the most densely affected area of pain and temperature loss. Joint position sense was impaired, often lost completely in 44 patients (in three without accompanying ataxia) affecting the arms alone in 11, the legs in 11 and both arms and legs in 22. It should be mentioned that testing the sensation in toes and ankles often showed it to be normal which tended to obscure the presence of gross reduction of proximal proprioception particularly in the hip joints and which was responsible for the ataxia. In the upper limbs the affected joints usually lay within the area of cutaneous sensory reduction but was noted in isolation in one case. In the legs, however, joint position sense was frequently lost without any local cutaneous sensory impairment. No case had evidence of Charcot's arthropathy.

(b) *Cerebellar dysfunction* Apart from nystagmus which was present in 32 patients (rotatory in 15), the signs of cerebellar involvement were uncommon in this series. Fifty patients had evidence of limb incoordination or ataxia and in 41 of these, as noted above, it was explicable on the basis of proprioceptive loss. In six other patients the ataxia was best explained by gross wasting and weakness of the arms in three and by severe weakness and spasticity of the legs in the remainder. In only three of the 50 cases was there definite evidence of cerebellar dysfunction without significant weakness or proprioceptive impairment. Owing to its infrequent presence in this group of patients cerebellar dysfunction in the section on "results" is grouped under the heading of "Motor Disability", a designation which therefore comprises weakness, spasticity and cerebellar deficit.

(c) *Motor function* The upper limbs showed the characteristic weakness, wasting, hypotonia and reduction or absence of reflexes in most patients. However, exceptions were commonplace: in nine the upper limb tendon reflexes were all preserved and in two patients tone in the upper limbs was increased with appropriate reflex changes, indicating an upper rather than a lower motor neurone lesion. This could be explained either by a high extension of the cyst or by the Chiari malformation compression. Two other patients had lower motor neurone signs affecting the C5 myotomes but with spasticity in the hands which was also present in the legs. In the lower limbs there was always evidence of a pyramidal lesion, even in the two cases of lumbar syringomyelia, although one had both upper and lower motor neurone signs in the legs.

(d) *Sphincter disturbance* Bladder symptomatology was a late complication and occurred in only five patients. The symptoms were incontinence in one, urgency in two, hesitancy in one and frequency in one. Impotence was a prominent symptom in only two individuals.

(e) *Scoliosis* This occurred in 23 patients and was not confined to the younger age group or to those with a long history.

(f) *Lumbar syringomyelia* In two patients physical signs were restricted to the lower limbs. One of them presented combined upper and lower motor neurone signs and loss of all modalities of sensation in one leg, the upper limbs being normal apart from some reduction of tendon reflexes. The other had bilateral loss of pain sensation up to the waist and developed a unilateral spastic foot-drop, the upper limbs being normal.

(g) "*Syringobulbia*" Eighteen patients had one or more features conventionally ascribed to syringobulbia. Seven of these had mild difficulty of swallowing with, in three, definite impairment of pharyngeal sensation; one had asymptomatic unilateral palatal weakness. Eight had nystagmus (five rotary, three vertical) and three had marked fibrillation of the tongue.

GROUP 1B

All these patients (10 cases), except one described below, exhibited classical signs similar to the majority in Group 1A as would be inferred from the fact that the diagnosis rested on clinical findings alone, oil or air myelography or both being normal in all. Two had bulbar signs (a depressed gag reflex in one, difficulty in swallowing in the other).

The absolute confirmation of the presence of a syrinx may be difficult. As already mentioned, the Chiari malformation by itself causes a wide variety of symptoms and signs. However, the findings of muscle wasting, absent or reduced arm reflexes and particularly a suspended area of impaired pain and temperature sensation has led to the diagnosis of syringomyelia even when delineation of the spinal cord by oil or air has been normal and where no cord enlargement had been seen at operation.

In one patient in this group the neurological signs did not conform to the classical pattern.

Case history

Eight months before surgical treatment this, 49-year-old male patient developed urgency of defaecation, followed two years later by weakness of the left foot. The main physical signs were increased arm reflexes on the left side, a left

foot-drop and some weakness of the left hamstrings and glutei. The left ankle reflex was reduced but the plantar response was equivocal. Fasciculation was seen in both calves and feet. Sensation was normal everywhere. Nerve action potentials in the left lateral popliteal nerve were reduced. Myelography showed a characteristic Chiari malformation but no cord swelling. Although there were mild upper motor neurone signs in the left limbs, there was also unequivocal evidence of a lower motor neurone disturbance in the legs which led to a diagnosis of an associated syringomyelia even though this was not confirmed radiologically.

GROUP 2

There were only four examples of obstruction at the foramen magnum other than by Chiari malformation. The lesions were dural cystic fibroma, localised arachnoiditis obstructing the foramen magnum associated with atlanto-occipital fusion, simple cerebellar cyst, and basilar invagination. All these lesions were probably congenital. Indeed, the rarity of an acquired lesion at the foramen magnum such as a meningioma or neurofibroma is a striking feature.

It is not certain that the proven syrinx in these cases was due to dilatation of the central canal, but they were included tentatively in this series as there was no evidence of other disease known to cause the purely intramedullary type of syringomyelia such as spinal arachnoiditis or trauma. The clinical signs in these cases all conformed to the classical pattern.

GROUP 3

Three patients (4% of the total number) had no indication of the presence of a Chiari malformation or other obstruction at the foramen magnum. There was no evidence of spinal arachnoiditis, tumour or trauma and in each case at exploration the presence of a syrinx containing clear, colourless fluid was confirmed. The clinical signs in these three were typical of a cavitating lesion of the spinal cord.

Radiological diagnosis (75 cases)

Plain radiographs

These were abnormal in a total of 24 patients (32%) (table 1). It is emphasised that plain radiographs were within the normal range in 68% of case.

Oil myelography

Lumbar myelography was undertaken in all patients (table 2), except for two early in the

Table 1 *Abnormalities on plain radiographs*

Wide cervical canal	14
Atlanto-axial fusion or apposition	9
Basilar invagination	2
Bifid C1	1
Bifid C6	1
Extra bone between clivus and dens	1
Fusion C2-3	1
Asymmetrical posterior fossa	1

Six patients had more than one abnormality.

Table 2 *Confirmation of syringomyelia by myelography*

	Groups			
	1A	1B	2	3
<i>Myodil</i>				
Syrinx not demonstrated	7	8*	0	1
Syrinx demonstrated	51	0	4	1
<i>Air</i>				
Syrinx not demonstrated	2	3	0	0
Syrinx demonstrated	24	0	0	2

* Ventricular introduction in two other cases was normal.

series who were investigated by Myodil injected into a lateral ventricle. It should be noted that myelography failed to demonstrate a dilated cord in seven patients who were later confirmed to have syringomyelia by air myelography or by operative findings, as well as in eight who were diagnosed as having a syrinx on clinical grounds. The foramen magnum was delineated by screening contrast in both prone and supine positions. There was no case in Group 1 showing a complete block and in none did prone screening show significant abnormality. In most cases the Chiari malformation was shown in the lateral view with associated narrowing of the cisterna magna, but oblique views revealed the degree of tonsillar protrusion more accurately. In cases of arachnoiditis supine screening usually disclosed a complete posterior obstruction with an irregular contour causing Myodil to flow anterior to the cord (six patients with the Chiari malformation also had a considerable degree of local arachnoiditis).

In Group 2 the abnormalities were distinguished radiologically from the Chiari malformation. In the case of the cerebellar cyst, the myelogram suggested that the tonsils were prolapsed but further investigation disclosed a posterior fossa mass. In this case Myodil entered the fourth ventricle and passed straight down into the syrinx which was the only occasion in this series in which this feature was observed.

In Group 3 the tonsils were demonstrated by Myodil or air studies to be above the foramen magnum in all three cases. Myelography failed to reveal the syrinx in one, although it was later confirmed by operation.

Air myelography (31 cases)

This investigation has the advantage that it may show dynamic changes in the size of a cord for when a flaccid cyst is surrounded by air the fluid in it will fall to its most dependent part, causing the cord to expand or to shrink according to whether the patient is tipped head-up or head-down. (Ellertson¹¹ claims that cystic tumours are more tense and in contrast change very little if at all with posture). However, the "collapsing cord" is not always demonstrable and in two cases later proven to have syringomyelia the examination was essentially normal.

In two patients myelography caused temporary but significant neurological deterioration; one following the use of Myodil and the other following air. Our present practice is first to use metrizamide rather than oil and to employ air myelography only if the water soluble contrast fails to establish the presence of a syrinx in cases where the diagnosis is obscure. Demonstration of a Chiari malformation makes it virtually certain that any enlargement of the spinal cord or the presence of characteristic signs of central cord disease (severe muscle wasting, absence of tendon reflexes, dissociated sensory loss to pain and temperature) is caused by syringomyelia rather than a tumour. When a Chiari malformation cannot be demonstrated, these features could be due to solid or cystic spinal cord tumour and air myelography may then be useful in establishing a diagnosis.

Vertebral arteriography

This has not been used routinely though the position of the tonsils may be demonstrated in some cases by the low course taken by the tonsillar branches of the posterior inferior cerebellar artery; however, these vessels are seen to be low running as a frequent and chance finding in normal individuals.

Computed tomography (CT)

Before 1973 the size of ventricles was not routinely investigated in the absence of clinical evidence of hydrocephalus. In those patients of Group 1A who had had further investigation, the ventricles were judged to be normal in size in 12 and mildly or moderately dilated in six. Since 1973 routine CT scans before operation have demonstrated normal ventricles in eight and mild to moderate hydrocephalus in two. In some, the Chiari malformation was shown. Other recent cases, not included here, have been investigated by CT scanning of the spinal cord; this sometimes, but not reliably, demonstrated the presence of a syrinx.

Operation

GROUPS 1A AND 1B

(1) *Decompression of the cerebellar tonsils*
(All 68 Group 1 Cases)

As a result of Gardner's ideas and its modifications, most surgical procedures have been directed to the cranio-vertebral abnormality.^{4 7 10 12-15} In the earlier cases of this series, the arachnoid was incised and the opening of the central canal into the fourth ventricle plugged with a piece of muscle (Gardner's operation). This has now been abandoned on two counts; firstly, because of a high incidence of complications, albeit usually temporary, and secondly, the observation that in those cases where the canal was not plugged because of difficulty in exposing the fourth ventricle, the final results were in no way inferior (table 3). More importantly; as we stated earlier, we now have considerable doubt that the mechanism described by Gardner explains the development and progression of so called "communicating syringomyelia." Our present practice is to carry out decompression of the foramen magnum so as to relieve the tonsillar impaction. This is usually performed in the prone position and involves the removal of the lower two-thirds of the squama of the occipital bone, the posterior margin of foramen magnum extending far laterally (bearing in mind the entry of the vertebral arteries), and the laminae of the first, second and sometimes third cervical vertebrae to reach below the level of the herniated tonsils. Every effort is made to keep the arachnoid

intact to avoid any blood entering the subarachnoid space. Any neural abnormality can be seen through the arachnoid. The dura is then left open.

(2) *Syringostomy*

(14 cases, 7 primary combined with cranio-cervical decompression, 7 secondary)

Syringostomy has been employed for over 50 years.^{16 19} It has been adopted in this series of cervico-occipital decompressions as part of the primary procedure only when the cyst was revealed at the time of decompression and was enclosed only in a very thin layer of spinal cord tissue. A small incision in the arachnoid and pia mater is made and a silastic tube passed from the sub-arachnoid space into the syrinx and secured to the pia mater with a stitch. If these findings were not present, syringostomy may be considered later as a secondary procedure if the patient deteriorates thereafter, in which case it is usually performed in the lower cervical or thoracic zone.

(3) *Terminal syringostomy*

(1 Case) This is a procedure recently introduced by Gardner and colleagues¹⁷ and based on the assumption that the dilated central canal extends into the filum terminale and that the syrinx may be drained by dividing the filum. It has merit of safety but we are doubtful that the canal is usually patent as low as the filum.

(4) *Ventriculo-caval CSF diversion*

This has not been used as a primary procedure because it is not clear to us why insertion of a ventriculo-caval shunt as recommended by Krayenbuhl and Bernini¹⁸ could have an influence where the ventricles are normal in size, which is the usual situation. Another factor, previously mentioned, is that any communication between the fourth ventricle and the syrinx is usually obstructed or narrowed so that reduction of ventricular CSF pulsation is irrelevant. Where there is a definite hydrocephalus or where it develops as a complication of foraminal decompression, a CSF diversionary procedure may be needed and was carried out for this reason on two cases in this series.

Table 3 Results of operations in groups 1, 2 and 3

	Groups					Total
	1A	1B	1A+ B	2	3	
<i>Gardner's operation</i>						
Number	26	4	30	2	3	35
Improved	9*	0	9	1	0	10
Unchanged	11	3	14	0	0	14
Worse	6	1	7	1	3	11
Follow-up (yr)	7	8	7	6	7	
<i>Decompression</i>						
Number	25	6	31	2	0	33
Improved	9*	2*	11	0	0	11
Unchanged	13	3	16	0	0	16
Worse	3	1	4	2	0	6
Follow-up (yr)	3½	4	3½	11	—	—
* One patient in each of these groups subsequently deteriorated again.						
<i>Decompression and primary syringostomy (Group 1A)</i>		<i>Secondary syringostomy (All Groups)</i>				
Number	7					7
Improved	1					2
Unchanged	3					4
Worse	2					1
Died	1					0
Follow-up (yr)	3					3

GROUP 2

The operative findings and the procedures in these four cases are described in table 4.

GROUP 3

In these patients (table 4) the foramen magnum was explored in the face of normal radiological findings in the hope that a band, not discernible

Table 4 Results of operations in patients with craniovertebral anomalies other than the Chiari malformation (Group 2), or having no significant craniovertebral lesion (Group 3)

Group 2 (Sex, age)	Associated lesion	Primary procedure	Result	Secondary procedure	Result
M39	Dural cystic fibroma	Cranio-vertebral decompression. Excision of dural nodule	Progressive deterioration JPS, UMN, ST Grade: 3-4 FU: 4.5 yr	—	—
M33	Cranio-vertebral arachnoiditis. Atlanto-axial fusion	Cranio-vertebral decompression	Progressive deterioration in power (?LMN) Grade: 3 FU: 6 yr	(Died of carcinoma of bronchus) Thoracic syringostomy	Stable for 5 yr
M20	Cerebellar cyst	Cranio-vertebral decompression. Partial excision of cyst.	Improved (UMN) Grade: 3 FU: 8.5 yr	—	—
F38	Basilar invagination	Cranio-vertebral decompression. Syringostomy	Stable for 8 yr then deteriorated in gait (JPS)	1. Thoracic syringostomy 2. Filum terminale syringostomy	Stable for 7 yr then Grade: 4 Improved (UMN) for 1 month only Grade: 4 FU: 1½ yr
Group 3 M33	None	Cranio-vertebral decompression. Posterior fossa exploration	Slow deterioration in gait (UMN, JPS) Grade: 4 FU: 7 yr	—	—
M21	Membrane partially occluding foramen of Magendie	Cranio-vertebral decompression	Progressive deterioration in gait (UMN, ST) Grade: 3-4 FU: 8 yr	Thoracic syringostomy	Stable for 2 yr
M35	Tonsils fused together	Cranio-vertebral decompression	Progressive deterioration in gait (UMN) Grade: 3-4 FU: 6 yr	Thoracic syringostomy	Stable for 2 yr

by x-ray examination, might be funnelling CSF into the opening of the central canal. The tonsils were confirmed to be well above the foramen magnum level in all cases. Minor abnormalities were seen in two. In one a membrane 6 mm in vertical extent only partially obstructing the foramen of Magendie was incised and the central canal plugged with muscle. In the second case the anatomy was normal except that the tonsils proved difficult to separate in order to expose the foramen. Clear CSF was aspirated from the cord. In the third the anatomy of the hind brain was entirely normal, clear CSF being aspirated from the cord and a syringostomy performed.

Results

The difficulty in assessing response to treatment derives from the number of neurological modalities involved, often to a varying degree which frequently respond in disparate fashion. All factors were considered individually and it was found that the main beneficial influence was on the function

of the pyramidal tracts and posterior columns, but in contrast to the generally good results obtained by decompression of the Chiari malformation in the absence of syringomyelia these cases showed only modest gains. No patient was followed for less than one year and most cases were examined annually. The length of follow-up was determined by the date of the last examination.

GROUPS 1A AND 1B

One patient died post-operatively. He had a five year history of progressive limb dysfunction and difficulty in swallowing and was finally admitted to hospital with quadriplegia and respiratory paralysis. He failed to improve following decompression of a Chiari malformation and syringostomy which was carried out as a last resort. He died 10 weeks later. Necropsy confirmed a large syrinx with extension into the medulla (syringobulbia) but only a microscopically small communication with the fourth ventricle via the extremely narrow upper end of the central canal. There were four late deaths due to carcinoma in two, myocardial in-

fraction in one and bronchopneumonia in one (respectively 4, 5, 10 and 11 years after the operation).

Pain

Central neck pain and cough headache usually responded well. In one patient this symptom had resolved spontaneously five years before surgery, but in six of the other seven the pain was completely relieved. However, in only two of these was there also improvement in other modalities, respectively gait, leg spasticity and joint position sense in one and gait and spasticity in the other. In one of the remaining four cases there was increased weakness of the arms and deterioration of gait due to spasticity (tables 5A and 6).

Of the 23 patients with, usually, lateralised pain in the face, trunk or limbs, one had ceased to suffer pain before surgery. In the remaining 22 the results were disappointing compared with those of neck pain. In five pain was still markedly improved after an average interval of four years four months, but in another four it improved only temporarily for less than one year. Pain was largely unchanged in the remaining 13 with an average follow-up of five years four months. In the long term, therefore, 17 out of the 22 failed to obtain persistent relief. Among the five who

showed improvement this was exhibited in three cases in other signs, gait, use of hands, spasticity and joint position sense in one, gait and spasticity in another and power in right arm spinothalamic sensation and leg spasticity in the third patient (table 5A).

Motor disability (tables 5 and 6)

Disability in limb function is the most obtrusive and restrictive neurological deficit for these patients. In many cases it was owing to a combination of a pyramidal disturbance with reduction of joint position sense with, in a few cases, a cerebellar deficit and it proved difficult to quantify the importance of each modality. For this reason an assessment was made of overall motor ability when comparing improvement using the following grades:

- 1 Minimal symptoms.
- 2 Mild symptoms or disability not sufficient to impair livelihood or former activity.
- 3 Some limitation of function but capable of independent life.
- 4 Limited function requiring aid.
- 5 Total dependence on others.

Twenty-one Group 1 patients (20 Group 1A, one Group 1B) noted some improvement. In 18 this was related in varying degree to recovery in power

Table 5A Patients showing improvement after craniovertebral surgery (Groups 1A and 1B (21))

Case no	Group 1A Sex, age	Symptoms improved	Signs improved	Grade	Operation	Follow-up (yr)
1	M17	Sensation right arm. Able to write	JPS (Right arm)	3-2	G	8
2	M44	Gait no longer spastic	UMN	3-2	D	3.75
3	M26	Foot drop	UMN, LMN, ST	3-1	D	1.5
4	F66	Gait. Able to go out un- accompanied. Pain (C)	UMN, JPS	3-2	G	10
5	M52	Gait	UMN, JPS	4-3	G	10
6	F37	Gait	UMN, JPS	3-2	G	8
7	F57	Gait. Power left arm	UMN, JPS	3-2	G	9
8	M58	Gait. Pain (C)	UMN	3-2	D	3.5
9	F54	Gait. Use of hands. Pain (L)	UMN, JPS	3	D	6
10	F48	Gait	UMN	3	D	17
11	M61	Gait. Pain (L)	UMN	2	D	1
12	F56	Gait	UMN, ST	4	G	5
13	F48	Gait. Able to stand from sitting position	UMN, JPS	3	D	6
14	M38	Power of hands	UMN, LMN, ST	3	G	9
15	M44	Power right hand. Pain (L). Handwriting. Sensation right cheek	UMN, ST	3	D	4
16	F38	Gait	JPS	4	G	10
17	M57	Power left arm and leg	UMN	3	D	2
18	M49	Foot drop	UMN	3-2	D	5.25
19	F30	Gait	UMN	3*	D	6
20	F42	Use of hands	UMN, JPS	3*	G	9
<i>Group 1B</i>						
21	M39	Gait	JPS	3-2†	D	7.5/6-75

G—Gardner's operation; D—Decompression; *—Delayed deterioration. Thoracic syringostomy; †—Delayed deterioration at 7 years; Pain (C)—Central neck pain relieved; Pain (L)—Lateralised dysaesthetic pain relieved; UMN—Upper motor neurone signs; LMN—Lower motor neurone signs; ST—Pain and temperature sensation; JPS—Joint position sense.

Table 5B Modalities improved after surgery

Motor function	Joint position sense	Pain sensation and temperature	Number of cases
7	7	—	7
7	—	—	7
4	—	4	4
—	3	—	3
18	10	4	21

and reduction of spasticity (in the lower limbs) in conjunction with improvement in joint position sense in seven cases and pain sensation in four (tables 5A and B). In three others joint position sense improved as the only modality. The function in the upper limbs improved in seven cases and in three this was in association with that of the legs.

With regard to grades, nine patients improved one grade and one two grades. In the remaining 11 the improvement in symptoms was mild and often difficult to quantify in terms of distance walked or speed in writing, but was supported by definite improvement in physical signs although overall it was insufficient to warrant a change in grade (tables 5A and B).

Sensory change

Cutaneous sensory loss to pain and temperature improved in only four patients. Loss of these two modalities apart from the well recognised liability to burns and scalds from which the patient learns early on to take avoiding action did not produce much disability. Although the upper and lower levels of sensory reduction can be charted fairly accurately and any change in the way of ascent or descent noted, alteration of quality of sensation is much more difficult both for patient and examiner to assess, particularly when comparing

results at intervals of one or more years. Moreover many patients in the first few days or weeks postoperatively claimed that sensation had improved (or that the limbs were stronger) but at review from three months onwards this subjective recovery had often disappeared and it is only the real and persistent changes that are recorded in tables 5A and B. It is this common but short-lived, mainly subjective, response to surgery that has frequently led to extravagant claims of surgical benefit and has tended to make true comparison of results between different series so difficult.

The extent of the real improvement in our four patients, however, is limited: (1) hypalgesia on the trunk changed to normal sensation in four segments on the right and two segments on the left (2) patchy loss in segments C2 to D12 right and left returned to normal (3) the quality of appreciation of pin prick improved significantly, but not to normal, in CN5+2 cervical dermatomes on the right (4) cold sensation returned in two lumbar segments.

Light touch sensation was uninfluenced by operation and has not been included in table 5.

Muscle wasting

No improvement in muscle wasting in the upper limbs was noted and in no case did lost reflexes recover.

Syringobulbia

In only three out of 18 patients did the features of syringobulbia improve. Two out of seven patients showed improvement in swallowing difficulties and one out of two noticed an improvement in voice. Nystagmus was abolished in two, un-

Table 6 Patients showing deterioration after craniovertebral surgery (Groups 1A and 1B (13))

Case no	Sex, age	Symptoms worse	Signs worse	Grade	Operation	Follow-up (yr)
<i>Group 1A</i>						
1	F61	Gait	UMN	4	G	4
2	F52	Gait	Nystagmus	4	G	9
3	F41	Gait	UMN	3-5	G	6
4	M32	Gait and use of arms	UMN	3-5	G	6
5	M58	Use of left hand, gait	JPS	4	D	10
6	M66	Ataxia, diplopia, vertigo	Nystagmus	4	D	1
7	F39	Power in hands	UMN	3	S	3
8	M35	Use of hands, gait	UMN, LMN, ST	2-3	G	7
9	M30	Use of left arm, gait, swallowing	UMN, Ataxia	3-4	S	4
10	M20	Gait	UMN	3	D	3
11	F50	Gait	UMN	2-4	G	8
<i>Group 1B</i>						
12	M58	Arms weaker gait*	UMN, ?LMN	4	G	6
13	M58	Gait	UMN, JPS	3-4	D	5

G—Gardner's operation; D—Decompression; S—Decompression and previous syringostomy; *—Central neck pain relieved; UMN—Upper motor neurone signs; LMN—Lower motor neurone signs; ST—Pain and temperature sensation; JPS—Joint position sense.

changed in five and worse in three and in one case developed for the first time after operation. Marked fibrillation of the tongue was unchanged in three patients. Several of the patients had more than one bulbar symptom. As already mentioned the mechanism of bulbar improvement may be either compression by the tonsils (when decompression might be expected to relieve the symptoms) or extension of the syrinx into the medulla when decompression is unlikely to help.

ANALYSIS OF RESULTS

GROUP 1A AND 1B

The twenty-one cases showing improvement have already been described (table 5A); an analysis of the actual neurological modalities influenced is shown in table 5B. No patient showed improvement in all three modalities, namely motor function, joint position and pain and temperature and no patient returned to normal. In thirty-three patients the progressive deterioration that had brought them to surgery ceased after operation and their symptoms and signs remained static, that is stabilised, throughout their period of follow-up, an average of 5 years and 4 months. In twelve of these cases the duration of their observation after operation was longer than their preceding history of symptoms and we believe that the previous relentless progress has probably been permanently arrested. However, it must be accepted that late and even very late deterioration may be seen as the duration of follow-up increases.

Thirteen patients were worse (table 6), four having deteriorated one grade and two two grades and seven not sufficiently to alter the grade. One of these patients has deteriorated very slightly in ten years and it is clear that the previous rapid progress has been arrested to a large extent. In three grossly disabled patients the operation resulted in immediate deterioration of gait which thereafter

continued slowly to get worse. Another case deteriorated when a ventriculo-caval shunt inserted to control hydrocephalus after operation blocked to cause profound quadriplegia with only partial recovery. Nine other patients showed delayed slow relentless deterioration of limb function despite surgery (four after Gardner's operation, three after simple decompression of the cerebellar tonsils and two after decompression and primary syringostomy.)

Comparison of the operative procedures showed similar results following simple decompression and Gardner's operation (table 3) but the complication rate was higher in the latter (table 7).

Relapse

In three patients improvement was only temporary, 5, 6 and 7 years respectively; the first two of these patients underwent secondary syringostomy and the third patient remains under observation.

Secondary syringostomy

Syringostomy has been carried out as a second procedure in three Group 1 cases (table 3): in two of these the primary procedure has been temporarily successful and syringostomy has given further improvement.

GROUP 2

This is a heterogenous group and the results are given in table 4. Only one patient improved. The patient with basilar invagination is of interest as she improved following terminal syringostomy as a third surgical intervention.

GROUP 3

It is hardly surprising that these three patients received no benefit from posterior fossa surgery as no compressive lesions were found. Two have subsequently received syringostomies (table 4).

Table 7 Complications of surgery

Operation	Patients with complications	Pleocytosis/pyrexia	Subarachnoid haemorrhage	Kyphosis	Hydrocephalus	Meningocoele	Confusion	Neurology worse	Wound infection
Gardner's operation	15 (30)	6	2	1	2	2	3	4	
Simple decompression	4 (31)								
a) Arachnoid opened	3 (7)	1				1		2	
b) Arachnoid intact	1 (24)								1
Simple decompression and Syringostomy	0 (6)								

*Some patients had more than one complication.

SUMMARY OF 75 CASES

Improved 22 (21—Group 1A and B; 1—Group 2 (case 3)).

Stabilised 33 (33—Group 1A and B).

Worse 19 (13—Group 1A and B; 6—Groups 2 and 3).

Died 1.

The chances of cure or improvement, expressed as a percentage, for certain symptoms and neurological modalities are as follows:

74 cases

Central suboccipital and neck pain—	6 of 7 cases 87%
Lateralised dysaesthetic pain—	5 of 22 cases 23%
Motor function—	22 of 74 cases 30%
Joint position sense—	10 of 44 cases 23%
Spino-thalamic sensation—	4 of 70 cases 5.6%

Discussion and recommendations

It is recognised from these modest results that the ideal operation for syringomyelia has not been found. All the reported procedures, comprising suboccipital and cervical spinal decompression with or without high cervical syringostomy, insertion of a tube leading from the fourth ventricle to the cervical subarachnoid space, ventriculocaval deviation of CSF (in the presence of normal sized ventricles), and syringostomy and filum terminale syringostomy all seem to influence the condition for a time in some patients, the ventriculocaval shunt probably being the least effective.

To us it appears that suboccipital decompression by relieving the pressure of the Chiari malformation as well as influencing, in some way, the progression of the syrinx is the best primary procedure, with syringostomy employed as an additional measure.

Our results show that with longer follow-up the original hopes and expectations of surgical treatment based on Gardner's hypothesis have not been achieved. However, foramen magnum decompression does have some influence on the condition as 74% of this series were either improved in some aspects or stabilised throughout the period of follow-up with some degree of persisting deficit, but even this outcome was gratefully accepted by most patients when faced with inexorable neurological deterioration. However, with a longer follow-up it is possible that more cases may show a late relapse. In our hands, leaving the arachnoid intact, foramen magnum decompression is a safe procedure. Patients with neck pain or cough headache and those whose disability appears mainly to be related to pyramidal and posterior column func-

tion may be expected to do best; indeed, this means any patient where it is thought that the Chiari malformation is the main cause of symptoms.

Surgery is best avoided in those with severe disability such as walking with two sticks, or chair bound or bed bound particularly with poor respiratory reserve; but even then surgery may be undertaken as a calculated risk where deterioration is rapid or where central neck pain is very obtrusive. In all the patients in this series operation has been proposed only in the presence of demonstrable continuing deterioration. Patients in a quiescent phase have been left alone; there seems to be no place for "prophylactic" surgery, because any reappearance of deterioration in symptomatology can always be dealt with as soon as it arises. The risk of an acute incident of deterioration forestalling surgery, although possible, is so small that it does not seem to us to justify the indiscriminate application of a major surgical procedure, with potential complications, to all static patients particularly as it would be impossible to assess any beneficial effect.

Syringostomy

The final question remains as to whether foramen magnum decompression is preferable to primary syringostomy. Although the first syringostomy was performed by Elsberg as long ago as 1913¹⁹ it has been difficult to answer this question precisely for the reasons stated in the beginning of this communication. The article by Love and Olafson²⁰ probably offers the best comparison although their results are graded simply as "excellent" (showing subjective or objective improvement), "good" (status unchanged) and "poor" (progressive deterioration) and the longest follow-up is only ten years. In forty cases subjected to surgery, 33 were available for follow-up (of which only three were shown to have a Chiari malformation) with the following results:

Excellent 10 cases (follow-up 1–10 years)—30%

Good 13 cases (follow-up 1–4 years)—40%

Poor 10 cases (follow-up 1–6 years)—30%

The surgery in all three groups was similar and comprised syringotomy (simple incision of cyst), or syringostomy using retained wicks or drainage tubes ranging from polythene tubes to tantalum wire, which latter was stated to give the best results. As far as it is possible to compare this series with ours, the results are not dissimilar and it would seem that both operations are effective to some extent, though for different reasons, foramen magnum decompression by relieving the effect on tonsillar compression and syringostomy by drain-

ing the syrinx directly. In the individual case it may be difficult to determine which factor is more important clinically and it would seem logical to treat both where possible. Our present view is that foramen magnum decompression is used as the first procedure combined with primary cervical syringostomy where the cyst is easily accessible.

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