Syringobulbia: a surgical appraisal

David Morgan, Bernard Williams

Abstract

Syringobulbia is a term which has been clinically applied to brain stem symptoms or signs in patients with syringomyelia. Syringobulbia clefts are found on investigation or at necropsy caused by cutting outwards of the CSF under pressure from the fourth ventricle into the medulla. These should be differentiated from the ascending syringobulbia which may occur from upward impulsive fluid movements in a previously established syringomyelia. Clinical analysis of 54 patients suggests that bulbar features are most often found with neither of the above mechanisms but are due to the effects of pressure differences acting downward upon the hindbrain with consequent distortion of the cerebellum and brainstem, traction on cranial nerves or indentation of the brainstem by vascular loops. The commonest symptoms in the 54 patients were headache (35), vertigo (27), dysphonia or dysarthria (21), trigeminal paraesthesiae (27), dysphagia (24), diplopia (16), tinnitus (11), palatal palsy (11) and hypoglossal involvement (11). Careful attention to hydrocephalus is advisable before craniocervical surgery, but the decompression of the hindbrain and the correction of craniocervical pressure dissociation remains the mainstay of surgical treatment. The results of careful surgery are good, 45 of the 54 cases reported improvement. Most of the reported deterioration occurred in a few patients who did conspicuously badly.

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It is not uncommon for patients with syringomyelia or hindbrain herniation, to have dysfunction of the lower cranial nerve and brainstem. Such symptoms are often ascribed to "syringobulbia". Syringobulbia, however, is an uncommon lesion of the central nervous system; it may be defined as a pathological cavitation in the brain stem. There are several possibilities for the pathology and little agreement about what syringobulbia is.

We have reviewed patients with bulbar symptoms encountered in a neurosurgical practice expressing an interest in hindbrain herniation. The patients with adequate follow up have been selected from a database of patients with syringomyelia and related disorders.

Material and methods

Fifty four patients were selected because of hindbrain problems. Tumour cases have been excluded. All cases had clinical involvement of the brain stem and had craniocervical decompression (CVD) between 1962-88. Hindbrain hernia headache has not been used on its own as a selection criterion neither has syncope nor nystagmus. There were 34 females and 20 males. Forty four patients were available for review. Of the remainder, two had died of unrelated causes and 8 were untraceable or lived an excessive distance away. The age at diagnosis for females was 10-58, a mean of 37.3, and males 18-61, a mean of 44. The symptoms often started many years before diagnosis. The mean follow up period from the original operation was 14.2 years, range 3-39. Thirty four patients had either radiological or clinical evidence of syringomyelia, 20 cases had hindbrain features without syringomyelia.

Skull radiographs show basilar impression in 26 cases and CT scanning and myelography or MRI showed that 46 had tonsillar descent, 28 had a demonstrable syrinx in the spine. Two did not undergo CT scanning, myelography or MRI.

All patients had CVD. The technique included a small craniectomy decompressing the tonsils usually by removing the arachnoid and often by sucking away part of the tissue of the tonsils, and always stitching the dura back and leaving it widely open. Five patients had ventriculo-atrial shunting (VEA) following postoperative deterioration, one of these had a syringopleural shunt at the time of the CVD and two had late syringopleural shunting.

At review each patient completed a questionnaire and had a neurological and otolaryngological (ENT) examination.

CLINICAL FEATURES

Occipital headaches

Occipital headache was the commonest presenting symptom and occurred in 35 patients. In seven patients the headache persisted postoperatively, but was less. The headache was most characteristically a pounding pain usually posterior and bilateral, sometimes ascending to the vertex or present mostly unilaterally or in the neck. Typically this beat in time with the pulse, coming on two or three seconds after a strain, rising to a crescendo after a few more seconds and then dying away within half a minute. This is the "hindbrain hernia headache" stressed by Williams.¹⁻³ It may be tested for in the clinic by asking the patient to blow into a mouthpiece attached to a sphygmoman-

Department of
Otorhinolaryngology,
East Birmingham
Hospital,
Birmingham, UK
D Morgan

Midland Centre for
Neurosurgery and
Neurology, Holly Lane,
Smethwick, West
Midlands B67 7JX, UK
B Williams

Correspondence to:
Mr Bernard Williams

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ometer to a pressure of 50 mm Hg or so for five seconds and then suddenly relaxing. Observation of the patient usually makes the diagnosis but the patient may be asked to draw a graph of the pain.

Vértigo

Twenty seven subjects complained of intermittent vertigo. Ten patients were symptom free postoperatively, while the vertigo persisted in 9. In 2 cases, 1 and 2, persistent vertigo occurred postoperatively which was relieved following ventriculo-atrial shunting.

Voice disturbance

Twenty one patients had voice disturbance, either dysphonia due to cord palsy or in 5 of them, slurring of speech. Ten patients had confirmed vocal cord palsies. 1 of which was a bilateral abductor palsy and presented with stridor. Four unilateral and 1 bilateral recovered following operation. Only in 7 cases did the voice disturbance persist following CVD. However, 2 patients developed a cord palsy immediately postoperatively. Following ventriculo-atrial shunting the cord palsy completely recovered in one and in the other the symptoms resolved, although the cord remained immobile. Another patient developed a vocal cord palsy some years after operation.

Paraesthesiae

Of 27 patients with disordered trigeminal sensation, 1 of which was bilateral, 18 showed a complete resolution. In 8 the numbness persisted at least partially and in only 1 of these was there deterioration in symptoms. Only 1 case had trigeminal pain associated with paraesthesiae, this was the presenting symptom and was severe. This woman was completely cured by CVD, at which a loop of artery was found firmly pressed into the side of the medulla and was lifted out.

Dysphagia

Twenty four patients had pre-operative dysphagia, some had palatal regurgitation. In 18 cases the dysphagia resolved postoperatively. In most of the others there was some improvement but in one there was no change. Four patients developed postoperative dysphagia, 1 recovered following ventriculo-atrial shunting. One patient had cricopharyngeal myotomy with improvement.

Tinnitus

Eleven patients complained of tinnitus, 5 bilateral and 6 unilateral. Two patients with unilateral tinnitus became worse postoperatively. Out of the patients with bilateral tinnitus, 4 showed complete resolution while one persisted. Only one patient complained of persistent postoperative tinnitus, others said that it was present but unobtrusive.

Hearing difficulty

Of 9 patients complaining of hearing difficulty, all but one improved postoperatively; only one developed unilateral impairment postoperatively.

Nystagmus

Twenty six patients were noted to have nystagmus pre-operatively. Thirteen had rotatory nystagmus, which improved after CVD in nine, with marked amelioration of ocillopsia or dizziness in those who had symptoms. In only one case did severe rotatory nystagmus resolve postoperatively. Four patients developed nystagmus postoperatively, three of whom complained of ocillopsia, possibly contributing to difficulty in walking. Ocillopsia improved after VEA shunting in two of these and in none did it remain prominent.

Ptosis

Of 11 patients with ptosis in 5 it appeared to be due to a partial unilateral Horner's syndrome and in one a bilateral Horner's syndrome. In 2 cases unilateral Horner's syndrome persisted as did one with unilateral ptosis. Complete recovery occurred in 8 cases. A postoperative unilateral Horner's syndrome developed in one which resolved following VEA.

Diplopia

Sixteen patients complained of diplopia apparently due to abducent nerve palsies. 4 persisted postoperatively. In one of these, case 2, the paralysis was considerably worse after CVD and subsequent hydrocephalus but improved after VEA, although it remained a subjective problem. In 1 patient Myodil (Pantopaque) which had been used in the myelogram could be seen in the cavity of the syrinx in the immediate the post-myelogram period. Later radiographs showed that some of the contrast material was in the pons. This indicated that fluid had ascended from the cervical syrinx and the sixth nerve palsy may have been due to nuclear involvement.

Facial Nerve

Five patients had a pre-operative lower motor neuron (LMN) palsy of the facial muscles and 1 patient an upper motor neuron palsy. One of the patients with LMN features had a history of 6 episodes of facial palsy before the diagnosis was made. All but 1 LMN palsy recovered postoperatively. One patient had an acute herpes zoster of the left facial nerve a few days after discharge from hospital with temporary complete paralysis.

Palatal palsy

Eleven patients had a unilateral and 2 a bilateral palatal palsy associated with an absent gag reflex and analgesia of the palate. Six completely recovered including one with bilateral palsy and in the other bilateral case there was improvement in one side only, including restoration of the gag reflex.

One patient developed persistent palatal analgesia and an absent gag reflex immediately postoperatively, which resolved after VEA.

Accessory nerve palsy

Only 1 patient had a unilateral accessory nerve palsy, which recovered postoperatively.

Hypoglossal Palsy

Eight patients had pre-operative unilateral

Figure 1 Preoperation MRI scans of patient EM T2 weighted image (TE500, TR20) suggests flow void in the fourth ventricle indicating the energetic nature of pulsation.

Figure 2 Transverse MRI scan through the fourth ventricle of patient EM. T2 weighted to show flow void. This was taken at the same time as 3 & 4, after operation, at a time when hydrocephalus had advanced.

hypoglossal palsy, and 1 bilateral palsy. Two unilateral persisted postoperatively and 3 patients developed unilateral persistent hypoglossal palsy postoperatively.

SUBJECTIVE IMPROVEMENTS

Patients were asked to grade their clinical course postoperatively between "great improvement, some improvement, unchanged and worse". Great improvement was claimed by 31 patients, 14 reported some improvement, 5 were unchanged and 4 said they were worse. The subjective views correlated well with objective findings.

POSTOPERATIVE DETERIORATION

Five patients deteriorated considerably immediately postoperatively, at least two of them due to mismanagement, and they warrant further discussion. Most of the deterioration in the individual features described above occurred in these cases.

Case 1

A 34 year old female presented with syringomyelia and hoarseness due to a left cord palsy. She had marked basilar impression and arachnoiditis at the foramen magnum. After CVD combined with syringopleural drainage the patient was well for 6 days but then became suddenly worse. She became hoarse and developed total dysphagia. Her gait became considerably worse and she tended to fall over backwards and could not sit up on her own. She had severe nystagmus and oculocephalic reflex. There was acute urinary retention. Combined ventricular and intraspinal pressure studies showed that she had impacted her hindbrain more severely than pre-operatively. This result was probably contributed to because the syringopleural shunt which had been used was over draining the spinal compartment, probably including the spinal subarachnoid space. Hydrocephalus was present. The syringopleural drain was therefore removed and a Torkildsen's shunt from lateral ventricle to the upper spinal subarachnoid space was performed which resulted in slow improvement, eventually complete recovery of her dysphagia, oculocephalic reflex and urinary retention. The voice recovered only to its original state with persistence of the original cord palsy and some ataxia. Over subsequent years she has progressively deteriorated to become quadriplegic.

Case 2

This was a 40 year old woman who had suffered since the age of 19. She had headache, vertigo, diplopia, gait disturbance, dysphagia, dysphonia, and impaired mobility and hypoaesthesia of the hands. On examination she had weakness of abduction of both eyes, rotatory nystagmus and spasticity of all limbs. Investigation showed a basilar impression, hindbrain herniation but no syringomyelia or hydrocephalus. After CVD the patient was well for a few hours and then had a catastrophic decline. The CT scan showed a small haemorrhage at the site of the tonsillar removal and a sizeable haemorrhage around the front of the corpus callosum. She required tracheostomy and respiratory support and seemed likely to die.

The cause of the cerebral haemorrhages was unclear, it may have been related to platelet

Figure 3 Sagittal MRI scans (TE500, TR20) and adjacent to the midline to show the excavation of the floor of the fourth ventricle by clefts arising as a result of the combination of raised intraventricular pressure and probably low intraspinal pressure.

abnormalities. Angiography was negative. She did not have severe hydrocephalus at the time of her discharge back to another hospital where she convalesced for several weeks. Her gradual recovery was marred by the development of a hydrocephalus which was not acted upon for a further nine months. She did not actively deteriorate during this phase of the illness, but her recovery must have been delayed. Before VEA shunting she had total failure of abduction of both eyes, and all the symptoms with which she had presented were worse. The MRI scans showed a severe hydrocephalus with flow void in the third and the centre of the fourth, indicating marked pulsatile movement (fig 1 & 2). The fourth was particularly dilated and showed clefts in the sites normally associated with syringobulbia (fig 3 & 4). After VEA the patient's symptoms were all improved, but she did not recover to be better than before the CVD.

Case 3

A 68 year old woman complained of vertigo and had an ataxic gait with signs consistent with a syringomyelia. Investigations showed hydrocephalus and hindbrain herniation with some arachnoiditis. After CVD, she developed persistent vertigo, tinnitus, horizontal nystagmus, dysphasia, left vocal cord palsy, absent gag reflex, palatal palsy and unilateral hypoglossal palsy. She had severe ataxia and was

wheelchair bound thereafter. She refused further investigation or operation. It seems likely that hydrocephalus could have been shunted with good effect.

Case 4

A 38 year old woman, presented with occipital headache, nystagmus, facial numbness, tinnitus, giddiness, Horner's syndrome, dissociated sensory and tendon reflex loss and a Charcot's joint at the right shoulder. Investigations showed bilateral tonsillar descent with no arachnoiditis. Postoperatively she developed vertigo and a left sided Horner's syndrome, her syringomyelic signs and symptoms became worse. After initial but partial recovery she gradually deteriorated over many years. Re-investigation showed no hydrocephalus but there was a sizeable syrinx. A syringopleural shunt was performed 10 years later which led to the disappearance of the vertigo, tinnitus and Horner's syndrome and improvement in gait, although fine horizontal nystagmus persisted.

Case 5

A man aged 32 years, presented with stridor due to bilateral vocal cord abductor palsies in association with occipital headaches, tinnitus, dysphagia and rotational vertigo. He had basilar impression, assimilation of the atlas into the skull and congenital fusion of 3 cervical vertebrae with impression of the front of the brainstem by the odontoid peg (fig 5). After CVD he was acutely ill with a spastic triplegia, and was slow to recover consciousness, there was slight improvement in the bilateral vocal cord palsy eventually, but the patient had developed a bilateral hypoglossal palsy. Some spastic weakness persisted affecting the left foot severely. The right side of the tongue improved. Postoperative investigation showed a right anterior cerebral artery area infarction and deformation changes in the stem. The cause of this infarction is obscure. An anterior approach might have given a better result but this does not seem probable.

Discussion

The ideas expressed about syringobulbia in the literature are often obscure. Syringobulbia was reviewed by Jonesco Sisesti in 1932 and this work has been republished.⁴ He restricted his account to syringobulbia clefts and described such "syringobulbia" as normally associated with syringomyelia. Syringobulbia in isolation has been described.⁵⁻⁷

There is a long established tradition for regarding syringobulbia as a "developmental" lesion with the inference that it may be related to dysraphism. This attitude was taken by pathologists working in an era before modern imaging and surgery. More recently a hydrodynamic approach has proved helpful in understanding and managing syringomyelia, syringobulbia and related diseases. Nevertheless, perusal of the literature often reveals

Figure 4 Sequential MRI scans (TE500, TR20) through the mid-fourth ventricle a), through the lower part of the fourth ventricle to show the foramina of Luschka at b). Big fissures in the usual site for fourth ventricular clefts are shown on each side of c). Extension well down into the medulla is seen on d). Note the similarity of the clefts to those shown in figure 6. These clefts are widely opened, unlike those in the histological section.

findings that are difficult to explain on hydrodynamic grounds⁸ and odd syrinx cavities occasionally occur with no blockage of the CSF pathways to suggest an hydrodynamic cause. Some of these cases, however, are associated with dysraphic features. The majority of cases seen in clinical practice have a hydrodynamic component and this will be discussed.

Pathologists, surgeons and other clinicians differ in their concepts of syringobulbia. Pathologists concentrate on clefts running out of the fourth ventricle (as in fig 6). The lines of

structural weakness coincide with developmental planes and this has sometimes seemed to support the "developmental" school. It is proposed to call such fissures "syringobulbia clefts".

Surgeons are often more impressed by cavities running upwards from syrinx cavities in the cord (as in fig 7). These cavities may be found under the floor of the fourth ventricle and are readily displayed by MRI techniques. The fluid filling the syrinx in such cases may be not only CSF or fluid indistinguishable from CSF, but also sometimes proteinaceous fluid

Figure 5 CT scans after water soluble intrathecal contrast. Note the encroachment upon the foramen magnum by the malformed odontoid peg shown on the reformatted pictures (a). Angulation of the medullocervical junction was marked in this case but the impaction of the tonsils was thought to justify a posterior approach. The axial cuts b) appear to show the primacy of the hindbrain in deforming the medulla. note the deformity of the left side of the medulla and the proximity of the vertebral artery.

Figure 6 Histological section at the level of the hypoglossal and trigeminal nuclei to illustrate the traditional sites of clefts from the fourth ventricle. Note the destruction of the trigeminal nerve nucleus on the right of the picture and the way that the seepage of fluid reaches to the subarachnoid space. In another section there was a communication at that site.

from tumours. To differentiate them from syringobulbia clefts it is proposed to call cavities of this kind "ascending syringobulbia". It is possible to find combination of syringobulbic clefts and ascending syringomyelia with communication between the cavities as in fig 8.

Many clinicians, particularly before modern imaging, used the term syringobulbia to mean any symptom complex in association with syringomyelia which suggested bulbar involvement. Modern imaging and surgery indicate that syringobulbic clefts and ascending syringobulbia are less common than bulbar symptoms.

Pathogenesis of syringobulbia

Syringomyelia is not a disease, but is a condition with many possible causes. There are two main groups of cases, those in which the causative anomaly is at the foramen magnum level and those with a primarily spinal abnormality.

The cases with partial blockage of the CSF pathways at the level of the foramen magnum are the commonest and most important group. These may be called "hindbrain related" cases. Lesions at this level produce direct changes in the medulla, cerebellar tonsils, lower cranial nerves, posterior inferior cerebellar and vertebral arteries; this leads to difficulties in assessing the part played by intramedullary or pontine cavities.

The commonest lesion at the foramen magnum is downward herniation of the tonsils. Although commonly termed the Arnold-Chiari deformity, the non-eponymous term "hindbrain hernia" seems preferable. There is evidence that the main mechanism is the generation of pressure difference acting downwards across the foramen magnum, craniospinal pressure dissociation, or "suck". The cramming of hindbrain tissues into the foramen magnum has many effects. First, it maintains the morphology of the herniation and its valvular configuration, thus perpetuating the disease process. Second, it produces downward sliding and compression of the tissues. The dorsal regions of the cord move down relative to the front of the cord, the plane of weakness across the middle of the cord being accentuated by the actions of the dentate ligament. One side may slip down more than another and asymmetries and rotational deformities are common.

The most probable pathological explanation for ascending syringomyelia, the cavities occurring in the medulla and sometimes extending into the pons and beyond, is that upward movement of fluid occurs within the syringomyelia and this dissects along the tissue planes of least resistance, particularly into grey matter. Schliep¹⁰ has shown that extension of a syrinx from the cervical spine into the brain stem may occur. Such extension may be seen on investigation or at operation. Extension cephalad to the pons has been reported.^{8 10-12 18}

The cause of upward movement of fluid within syringomyelia cavities has been exten-

Figure 7 Unenhanced T1 weighted MRI scan to show ascending syringobulbia in a twelve year old boy with a spinal lipoma and no hindbrain herniation. The hindbrain area seems crowded because of the size of the syringobulbia. There were no bulbar symptoms in this case. The operation at the bottom end of the tethered cord, including a syringopleural shunt, produced resolution of the cavity and decompression at the hindbrain.

sively discussed by Williams.^{1 2 13-16} The movement of CSF in the spinal canal in response to sudden changes in pressure in the thoraco-abdominal cavities, is often violent, for example, after coughing. The pressure is transmitted from the trunk cavities through the epidural venous plexus producing a vigorous squeezing of the dura. When there is a sizeable cavity within the cord this provides a preferential flow pathway for CSF to ascend. Pulsatile compression of the dura and then the subarachnoid space followed by compression of the walls of a syringomyelic cord leads to upward movement of the intracord fluid. This phenomenon may be called "slosh" (fig 9). The movements of the fluid may be well seen on radiological studies and the changes in diameter of the cavities may be seen on static radiographs or CT scans. Patients may report symptoms such as upward advance of pain or sensory loss or neurological features such as diplopia experienced immediately after a cough or sneeze.^{14 15 17 18}

Benefits from operation, seem more likely to relate to reversible neurological mechanisms, such as traction upon nerves or compression of brain stem in the foramen magnum, than to

recovery of well formed glial lined cavities of syringobulbic clefts or ascending syringobulbia.

The clinical feature

A natural subdivision of brain stem features would be cranial nerve dysfunction and effects upon the stem itself. It is unclear, however, how this applies to complex symptoms such as nystagmus, vertigo and tinnitus.

Nevertheless, cases with correlations between the clinical state and the occurrence of clefts have been reported.^{1 4 6 19 20 23}

Figure 8 Unenhanced T1 weighted MRI scans of a patient with severe syringobulbia of mixed type, a) coronal and b) sagittal. There were three cavities in communication, one outside the fourth ventricle dorsal to the tonsils, the next one was the dilated bottom end of the fourth and the deepest cavity was deep within the medulla communicating with the main syringomyelia. The tissues were all thin and mobile which may account for the difficulty in obtaining a clear image.

Figure 9 The phase of upward movement of the CSF in response to a sudden rise in intracranial pressure. This gives rise to the most energetic phase of the slosh mechanism and the impulsive ascending surge of fluid may cut into the bulbo-pontine tissues as shown in (a). Note the way in which the hindbrain is lifted off the bone of the foramen magnum, the subarachnoid spaces are thus opened up to allow free movement of the CSF into the head. The phase of downward movement after a rise of intra-coelomic pressure is shown in (b). The close fit of the hindbrain in this part of the phenomenon provides the valvular mechanism and produces the suck phase of the phenomenon. The downward pressure of CSF across the foramen magnum in this part of the cycle of pressure changes causes cramming of the hindbrain into the upper spine, the overfolding of the medulla and traction on lower cranial nerves.

Cranial nerves

Involvement of the third and fourth nerves is uncommon but disordered eye movement can occur from a lesion of the posterior longitudinal fasciculus.²³ Twelve of our patients complained of diplopia on lateral gaze. One 1 case of combined facial and abducent nerve involvement was seen, despite the relationship of the seventh nerve fibres to the sixth nucleus. Only 2 had clinical evidence of an abducent palsy at follow up. The abducent involvement is thought by Schliep to be due to destruction of its fibres in the pons but involvement of the nucleus seemed likely in one of our cases and it is possible that traction on the nerves results from the downward movement of the brainstem.

Trigeminal nerve disorders are commonly reported but it is likely that the nerve itself is rarely directly affected.^{24 25} In one patient with a vascular loop trigeminal dysfunction was the only symptom; an uncommon event. In two cases reported by Lapresle and Metreau the presentation was of trigeminal type.²⁵ Trigeminal symptoms probably often originate from the spinal sensory nucleus in the upper cervical cord and the accompanying fibres which form the descending trigeminal root. These may initially be involved in syringomyelia, where the syrinx involves first the fibres representing the most caudal part of all three divisions, not

necessarily from ascending syringobulbia or a syringobulbia cleft. Extension of the syrinx upwards may cause dissociated sensory loss to progress in all three divisions, converging upon the tip of the nose and upper lip, due to the segmental pattern of facial sensory innervation.

Only 4 facial nerve palsies were seen, 3 of which were of lower motor neuron type. The significance of the relapsing case is unclear.

Deafness is an uncommon finding in syringobulbia according to Schliep,¹⁰ and Barnett *et al*²⁴ do not mention it. Five cases out of six in the present series improved following operation. Tinnitus was not associated with the deafness in these patients. Improvement was reported in 4 out of 6 cases. These symptoms may be due to traction and angulation of the nerves at the pons or at the porus acousticus. Alternatively the problem may lie in the brainstem.^{24 30}

Vagus dysfunction is common and may manifest as a breathing disorder^{4 31} vocal problems⁶ or swallowing difficulty.^{20 32} Others have reported improvement after operation.²¹

The tongue is frequently involved in syringobulbia, especially in the latter stages of the disease; inducing atrophy and fasciculation.^{10 22 24} Only 2 out of 6 palsies persisted following CVD.⁵ It may result from a syringobulbic cleft developing in the plane of

weakness down the middle of the brainstem. Bilateral Horner's syndrome was present in 1 patient who had bilateral lower cranial nerve involvement. Complete recovery occurred following decompression.

Nystagmus and ocillopsia are common accompaniments of hindbrain herniation.³³⁻³⁵ Improvement in nystagmus following operation has been reported.^{17 27 33-36} Nystagmus in our patients was more common than in some earlier series and improved less often after operation. Three patients developed nystagmus postoperatively. Vertigo commonly improved after operation.⁷ (10 of 15 patients.)

Occipital headache is common in syringomyelia and syringobulbia²⁶ and responds best to operation if there is a hindbrain hernia.^{5 27-29} Only 4 patients complained of occipital headaches at review.

Prognosis

Syringobulbia is sometimes rapidly progressive and may be responsible for death in patients with syringomyelia.³⁷ The survival of syringobulbic patients varies from 6 years^{4 32 36} to 27 years^{40 41} following the onset of brainstem signs. Brainstem dysfunction may result in swallowing and respiratory problems which threaten life.^{6 7 10 19 31 38 39} Sudden death is reported due to bleeding into a syringobulbic cleft.³⁹ It seems that syringobulbic clefts are more dangerous than the ascending syringobulbia. Jonesco Sisesti, however, considered that syringomyelia clefts are surprisingly benign.⁴

It is believed that prognosis is improved by operation with the objective of deflating hydrocephalus^{9 13 21 43 44} and then carefully correcting craniospinal pressure dissociation at the foramen magnum.^{13 24 27 28 38 43-45} We recommend that operation should include careful removal of the arachnoid and sometimes also of the tonsils. The dura must not be closed, lest suck be re-established, and a graft is not necessary.¹³ It is probable that part of the benefits of CVD are gained by the "depulsating" provided by a sizeable artificial cisterna magna.

Previous reports of the effect of CVD have usually concentrated on the syringomyelic symptoms or those from the hindbrain hernia such as headache.^{5 27} Banerjee and Millar²⁶ studied 8 patients with syringobulbia who had CVD. Six improved but there were no details of the pre- or postoperative signs or symptoms. Seaz *et al*²⁸ studied 60 operations on patients with Arnold-Chiari deformity, and showed that 65% improved. Over a longer period many of the patients with cord or brainstem cavities had a further deterioration. Hady-Djilani and Zander³⁶ reported a study of 12 patients for 12 years with cervicobulbar syringohydromyelia. They gave a qualitative assessment with few clinical details.

Conclusion

Of our 54 patients with features of bulbar involvement, commonly diagnosed as syringobulbia, 45 improved after operation. Deterioration after operation was uncommon; it occurred immediately postoperatively, but late hydrocephalus developed insidiously. Posterior fossa decompression improved lower cranial nerve dysfunction associated with syringomyelia and hindbrain herniation, usually ascribed to syringobulbia. Nevertheless, it remains unclear to what extent improvement is due to collapsing of cavities or clefts and the extent to which it may be attributable to mechanisms, such as, relieving compression and distortion of stem structures or traction upon nerves or vessels.

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The signs of Kernig and Brudzinski

"I have observed for a number of years in cases of meningitis a symptom which is apparently rarely recognised although it is, in my opinion, of significant practical value. I am referring to the occurrence of flexion contractures in the legs or occasionally also in the arms which become evident only after the patient sits up.

Disregarding for a moment the infrequent cases of acute meningitis without contractures (particularly purulent secondary meningitis), one may find, as is well known, in the vast majority of cases of tuberculous and epidemic spinal meningitis the classic more or less prominent stiffness of the neck and back. Only in some of these patients while they are lying supine, may contractures of the extremity muscles be noted . . . If one has the patient sit up on the edge of the bed, his legs dangling . . . the stiffness of the neck and back will ordinarily become much more severe and only now will a flexion contracture occur in the knee and occasionally also in the elbow joints. If one attempts to extend the patient's knees one will succeed only to an angle of approximately 135°. In cases in which this phenomenon is very pronounced, the angle may even remain at 90°."

Vladimir Mihailovich Kernig (1840-1917) first brought these observations to the medical public at a meeting of physicians in St Petersburg in 1882. On 29 December 1884 his work was published.¹ Born in Lepaia, Latvia, Kernig graduated in 1864, receiving a doctorate one year later. He obtained a post at the

Obuhovsk Hospital in St Petersburg, reaching the highest professional rank and continued his work until his retirement in 1911. Kernig's account followed one year after that of the often confused Lasègue's sign described in his thesis by Lasègue's pupil J-J Forst in 1881.²

Józef Brudzinski (1874-1917) was a Polish paediatrician. Dean of the University of Warsaw, he described several signs, but the one known in meningeal irritation was published in 1909.³

"I have noted a new sign in cases of meningitis: passive flexion of the neck causes the lower extremities to flex at the knees and the pelvis . . . The technique of examination is very simple. With the child in the supine position, the examiner flexes the neck of the child with the left hand while resting his right hand on the patient's chest to prevent it from rising. This examination is generally not difficult except in the very young who because of restlessness may not be able to maintain the lower extremities in extension. In such a case, the examiner should gently restrain the legs at the knees. To prevent errors, it is important to do repeated examinations."

JMS PEARCE

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