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Chronic herniation of the hindbrain

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Summary

Herniation of the hindbrain occurs when the lowest parts of the cerebellum and sometimes part of the medulla are moved downwards through the foramen magnum, a pressure difference acting across the foramen magnum moulding the tissues into a plug. It is suggested that the clinical course in both adults and babies with spina bifida may be explained by the hindbrain hernia acting as a valve.

The term 'Chiari Type I deformity' is commonly used for an abnormality in which the tonsils and lowermost parts of the cerebellar hemispheres are prolapsed through a normal foramen magnum. Acute herniation may occur as a result of space-occupying lesions. Chronic herniation may be morphologically identical although it tends to be more severe. Sometimes it will produce few symptoms which often may be delayed so that the original causative lesion may not be apparent. Causes include bone softening, tumour, or previous meningitis. Birth injury is probably the commonest cause of the deformity, which presents clinically in adults.

In infants with severe forms of spina bifida a hindbrain herniation is present. This abnormality may be called 'Chiari Type II deformity' or Arnold–Chiari deformity and is an intrauterine abnormality in which the fourth ventricle and medulla are grotesquely herniated before they are properly developed and the foramen magnum is enlarged.

The commonest clinical presentation of Chiari Type I deformity is syringomyelia, which is usually not diagnosed until adult life. Other presentations include syringobulbia, headache, oscillopsia, attacks of giddiness, lower cranial nerve palsies, and ataxia. Particularly characteristic are cough headache and cough syncope. Syringomyelia and syringobulbia in particular may be irreversible by the time they are diagnosed. Nevertheless, surgical decompression may be successful in relieving symptoms of headache, cough syncope, and long-tract compression; most cases of syringomyelia show some improvement and in others progression of the disease is arrested. Operative techniques for hindbrain herniation are discussed.

Chiari Type II deformity is probably responsible for the progression of hydrocephalus after birth in the majority of babies with spina bifida. Measurement of pressure in the cerebrospinal fluid above and below the foramen magnum shows that intermittent pressure difference is commonly present at times of neurological deterioration. Surgical decompression of the hernia in adults allows correction of the valvular effect, which may be monitored by pressure measurements. In babies the associated hydrocephalus is usually so gross that it requires separate treatment, but pressure monitoring may be of value in assessing the state of the disease.

Introduction

In discussing herniation of the hindbrain two age groups must be considered. Herniation in infants with spina bifida is more gross than the herniation formed in adults and is often labelled a 'malformation'. The resemblances between the infantile and adult forms have sometimes

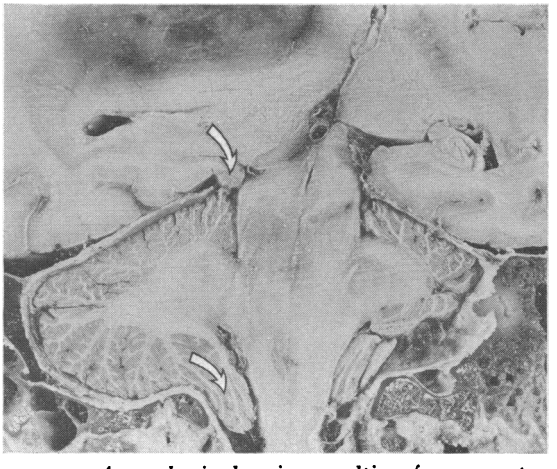

FIG. 1 *Acute brain hernias resulting from a supratentorial glioma (above). There is a hernia of the uncus (top arrow) pushing a part of the temporal lobe through the incisura and a hindbrain hernia (bottom arrow) due to the downward displacement from the tumour.*

Based on an Arris and Gale Lecture given in Birmingham on 27th February 1980

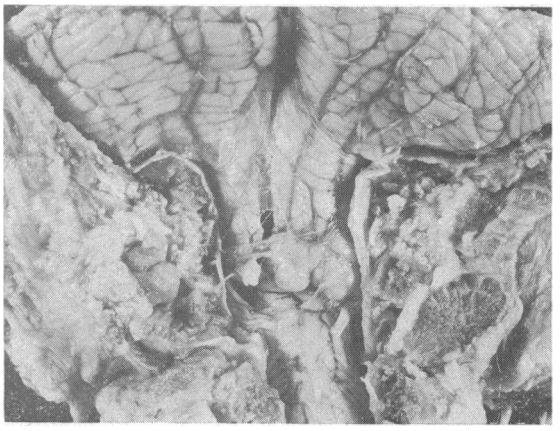

FIG. 2 *Acute hindbrain hernia dissected from behind in a patient who died of an untreated cerebellar haemangioblastoma (Chiari Type I).*

led to the assumption that the adult chronic herniation is also a 'malformation'.

Interest in the hindbrain hernias of adults has been stimulated by the realisation that syringomyelia in adults is usually accompanied by hindbrain hernia, that the commonest chronic presentation of herniation in adults is syringomyelia, and that operations on the posterior fossa may produce remarkable benefit *(1)*.

Both Eric Newton, who gave his Hunterian Lecture 11 years ago, and I were introduced to the subject by Mr Jack Small, of the Midland Centre for Neurosurgery and Neurology, who has built up a large personal series, thoroughly investigated and documented, which has provided clinical material.

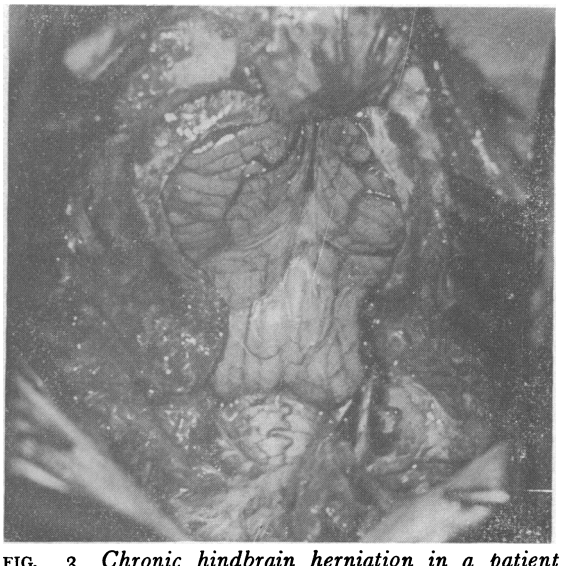

FIG. 3 *Chronic hindbrain herniation in a patient who presented with cough headache 27 years after precipitate labour. The limited normal operative exposure to decompress the tonsils is shown. Note that there is no morphological difference between this case and Fig. 2 apart from the fixation shrinkage in the patient who died contrasted with the tight impaction during life.*

Herniation of the lower part of the cerebellum through the foramen magnum is the simplest form of hindbrain herniation and is commonly caused by space-occupying lesions (Figs 1 and 2). There is little difference between the tonsils engaged in the foramen magnum as the result of an acute event such as a tumour and the tonsils which are deeply engaged in the upper part of the spinal canal as a result of a long-standing herniation (Fig. 3). In the latter case the moulding is commonly greater and the tonsils may extend downwards as far as the upper border of the axis. The patients with the most severe herniation are those in whom the deformities have come on during fetal development. In newborn babies with spina bifida the herniation of the cerebellum may extend down as far as

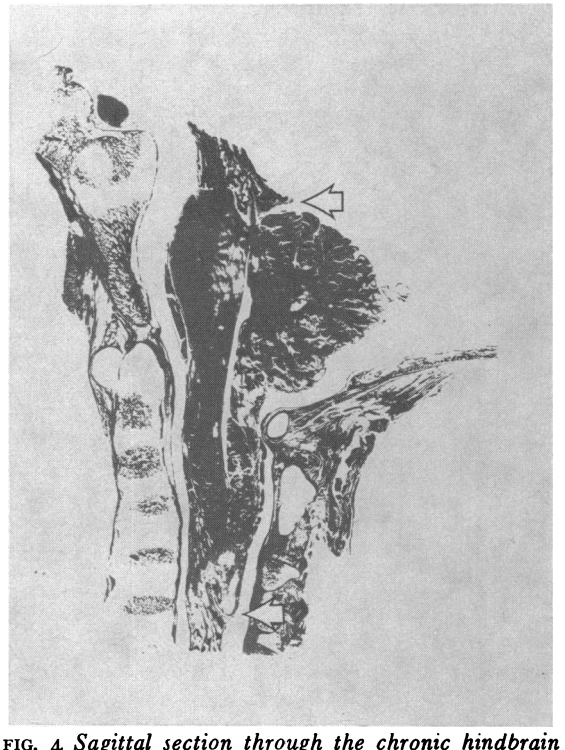

FIG. 4 *Sagittal section through the chronic hindbrain hernia associated with severe meningocele (Chiari Type II). The upper arrow shows the moulding of the midbrain by the cerebral hemispheres and the lower arrow shows the overfolding of the medulla. The fourth ventricle and lower parts of the cerebellum are elongated, deformed, and herniated down to the level of the body of C6.*

the thoracic spine; appearances as shown in Figure 4 are common.

The deformities of adult life are so different from those in infants that Chiari divided the cerebellar herniation into two types and this division remains convenient for discussion. The term Arnold–Chiari malformation has passed into common usage although it lacks a precise definition. Chiari's classification seems preferable; his Type I occurs in adults and Chiari Type II refers to the severe deformities found in infants with spina bifida.

Clinical course

ADULTS

Clinical presentation may be divided into two groups, syringomyelia and posterior fossa symptomatology. Many patients have both; some patients with brainstem features suggesting a posterior fossa presentation may have syringobulbia. Both kinds of patient may have long-tract signs from compression of the upper spinal cord, particularly of the posterior columns, by the herniated cerebellar tonsils.

Syringomyelia Syringomyelia is a well-recognised symptom complex and identifies a useful subdivision of patients with chronic hindbrain hernia for such purposes as enquiry about aetiological factors or correlations with changes in the ventricles. There are of course other causes of syringomyelia such as spinal tumours or cystic cavities above a paraplegia and therefore it is necessary to note that 'syringomyelia' in this paper refers only to cases associated with chronic hindbrain herniation. This type, sometimes called 'communciating syringomyelia', accounts for the biggest group of patients presenting with clinical evidence of cord cavities and it is also the commonest and most severe presentation of chronic adult hindbrain hernia.

When syringomyelia presents with dissociated sensory loss or a Charcot joint it is readily recognised. The earliest symptom, however, is usually pain, which commonly takes the patient to an orthopaedic surgeon or a physician. The stigmata of syringomyelia and a review of the presenting features, including a discussion on the origins of the pain, have been presented elsewhere *(2)*.

Posterior fossa presentation Posterior fossa presentation may be of several kinds. Headaches, particularly occipital or nuchal, may be exacerbated by coughing; in these cases surgical treatment is likely to be successful. Many patients have headache related to exertion which is overshadowed by other complaints. Acutely raised intracranial pressure with papilloedema may come on in the course of chronic hindbrain herniation, sometimes precipitated by investigation, but patients with papilloedema and headache usually have space-occupying lesions.

Cranial nerve symptoms, particularly deafness and diplopia, are common and patients sometimes complain of oscillopsia. More common is nystagmus, often with vertical or rotatory components. Ataxia is most troublesome when it affects stance and gait.

Attacks of unconsciousness may occur; syncope produced by coughing, laughing, or some other form of straining is the characteristic finding. It comes on a few seconds after the strain and the patient usually recovers in a minute or so. Death may occur from lower cranial nerve dysfunction, dysphagia and associated respiratory distress, or syncope which passes into respiratory arrest. Occasionally in patients presenting with cough headache alone the symptom may remit, either spontaneously or following an event such as a sudden jerk or a pneumoencephalogram. It is probable that most cases of posterior fossa presentation progress to death if not treated.

INFANTS

In newborn children with spina bifida the neurological and orthopaedic deficits associated with the spinal lesion dominate the presentation. Of those severely affected, with the spinal cord everted on to the surface, around 95% will develop progressive hydrocephalus. The children are usually born normally because the head is not enlarged and there is little external evidence of hydrocephalus. There are some external signs: the ears seem low on the head, the forehead may protrude, and the occiput seems large and overhangs the back of the neck. The internal morphology of the cerebrospinal fluid (CSF) pathways is grotesquely deformed in such infants. The lateral ventricles are large, unequal, and fused in the middle, and the occipital horns are blended in with the enlarged bodies. The frontal horns and temporal horns appear to be in competition with each other for space, and a patient with large frontal horns may have small temporal horns and vice versa. The third ventricle is small and deformed, having been compressed from side to side by the enlarging lateral ventricles. The massa intermedia is large and may obliterate part of the third ventricle. The foramina of Monro are elongated and slit-like from front to back as a result of compression. The aqueduct is usually narrowed from compression by the lateral ventricles or the suprapineal recess and may be

elongated as the fourth ventricle is displaced downwards through the foramen magnum. The fourth ventricle and lowest part of the cerebellum are sometimes so far herniated through the enlarged foramen as to be unrecognisable.

The choroid plexus is concealed in a mass of enlarged veins and such structures as the cerebellar tonsils have not been formed. The foramina of Magendie and Luschka are deep in the cervical canal and not identifiable.

The cortical mantle may show microgyria, the cortex having become corrugated and compressed owing to the hydrocephalus. Many morphological changes may be due to hydrocephalus and those that cause narrowing of the CSF pathways may in turn aggravate it; thus the hydrocephalus may become progressive and irreversible. Clinically these babies are compensated at birth and the external circumference of the head may be normal or less than normal. They then become progressively decompensated within a few weeks after birth; in many cases removal of a meningocele and closure of the back defect seems to hasten the onset of the hydrocephalus. The typical case with severe myelomeningocele therefore has a biphasic course, the second episode being brought on at birth or immediately afterwards.

In cases in which the hydrocephalus compensates early the patient may survive. Such patients may have mild forms of spina bifida and be prone to syringomyelia or posterior fossa presentation of chronic hindbrain hernia in later life.

Aetiology

Most hernias seem to be due to failure of the orifice to hold the contents. Femoral, inguinal, and incisional hernias are examples in which the contents of the hernia are initially normal, the extruding force is the increase in pressure which accompanies the normal events of life, and the essential cause is weakness of the orifice. The situation at the foramen magnum might be thought to be quite different because only in Chiari Type II is the foramen magnum commonly enlarged and that is most probably a response to the tissue jammed in it during development of the skull.

Perhaps because of the use of the term 'Arnold–Chiari malformation' the idea has become widespread that the Chiari Type I deformity is also a 'malformation' and that it reflects an intrauterine pathological process. Uusually, however, a deformity is either Type I or Type II and it seems that the milder deformity is later in its onset and occurs after the cerebellum is fully formed.

The commonest cause of herniation is an abnormality in the head pushing the brain out, such as a space-occupying lesion of the type shown in Figures 1 and 2; such cases are most commonly acute. In patients with no space-occupying lesion and a chronic hernia hydrocephalus was implicated by Chiari. Studies of patients with syringomyelia, however, have shown that only 29% have even mild hydrocephalus *(3)*, while in those with posterior fossa presentation the proportion with hydrocephalus is similar. It seems probable that the hindbrain hernia causes the hydrocephalus rather than the other way round.

With Chiari Type I herniation it is commonly not possible to determine the cause. It seems likely that birth injury is much the commonest cause and comparison with a control series supports this contention *(4)*. Compression of the bones of the vault may cause downward displacement of the brain and, particularly if associated with tentorial tears, may cause herniation of the hindbrain *(5)*. Local compression of the bones of the base by the expulsive force of the uterus or obstetric forceps may cause fracture or greenstick deformity of the occipital bone. Local haemorrhage around the cisterna magna, brain swelling due to anoxia, and transient hydrocephalus may all play a part in starting off the hernia.

The Chiari Type II malformation of infancy is an intrauterine abnormality, but the aetiology is as obscure as that of the spina bifida. The simplest explanation would seem to be that the mesodermal abnormalities of spina bifida are primary and that when the ectodermal tube fails to close the pressure in the spine remains low compared with that in the head. A similar explanation was proposed by Cameron *(6)*. Certainly the brain tissues seem almost to have flowed down into the spine (Fig. 4). It is not necessary to postulate a primary high pressure in the head; the embryo is a high-pressure pulsating system and a breach in its coverings and resultant low pressure may produce widespread abnormality.

Mechanism of advance in hindbrain herniation

Both the adult and the infantile herniation therefore show a stable period sometimes followed by exaggeration of the hernia symptoms and even death.

Why does the hindbrain recommence herniation or even continue to herniate through a normal orifice when the brain is free of hydrocephalus or space-occupying lesions? It might be thought that the brain does not move in and

out of the foramen magnum and could not be exposed to such pressures as those in the abdomen for instance. A fundamental principle of neurosurgery is that the total contents of the intact skull must be constant at all times because of the incompressibility of the brain and its associated fluids. When a person coughs, for example, CSF moves sharply upwards through the foramen magnum and then rebounds downwards to return to normal. This is because the epidural veins around the spinal theca become distended when the thoracoabdominal pressure is raised. During Valsalva's manoeuvre, for example, the CSF pressure is equal in the head and the spine, but the veins in the spinal canal are distended, the CSF is partly displaced into the head, and veins within the head are compressed. The jugular and facial veins are distended not only because of damming back of blood from the thoracoabdominal cavity but also because the diploic veins draining the intracranial system to the extracranial tissues are fully open.

Recording intracranial and intraspinal pressures simultaneously in patients with hindbrain hernia may show a pressure difference between the head and the spine which may be permanent or transitory. This is called craniospinal pressure dissociation *(7)*. Observations may be made easily on babies with spina bifida. A meningocele may be slack and subject only to atmospheric pressure at the time when the fontanelle is tense. If a meningocele is squeezed the fluid from it may be displaced into the head and then be greatly delayed in its return to the spine *(8)*. The measurement of such pressure changes by electrical methods allows classification of Chiari Type II malformations into four groups. In the normal group the pressures are equal in the head and the spine. In the second, with mild impaction of the hernia, the pressures are equal at rest, but delayed equalisation indicates a valvular effect. In the next worse group the baseline (resting) pressures are unequal, the pressure in the head being higher, but valvular action is still detectable. In the most severely impacted group the pressures are entirely dissociated so that pressure events in the spine and the head are independent. Once the recordings become abnormal there is a tendency for the situation to grow worse and for the patients to move into the entirely dissociated group, in which there is progression of the hydrocephalus *(9)*.

In adults craniospinal pressure dissociation may be recorded in the sitting position with a ventricular and lumbar puncture needle and differential manometry performed by subtracting the ventricular pressure from that in the lumbar sac. Figure 5 shows an abnormal result in a patient with severe basilar impression, headache, ataxia, and oscillopsia. The pressure difference after a single cough took about 25 s to disappear.

In most cases of chronic hindbrain herniation in adults a valvular action can be detected. It is almost always of the 'baseline equality with valvular effect' type; the baselines have only been separated in 3 recordings out of 168 and no case of total dissociation has been seen in adults. Another example is given in Figures 6 and 7, taken from a patient who presented with a sudden onset of diplopia caused by a syringobulbia. There were long-standing changes of syringomyelia and radiographic signs of hindbrain hernia with arachnoiditis due to birth injury.

Thus it can be seen that the valvular action of the chronic hindbrain hernia has a high explanatory value for several associated clinical features. Firstly, the maintenance and progression of the morphological deformity of the hindbrain hernia itself: the moulding provided by the pressure difference between the spine and the head causes downward moulding of the lower cerebellum and makes it fit more closely

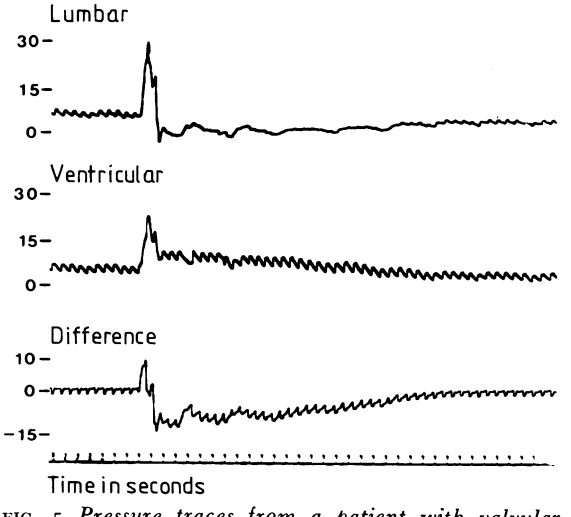

FIG. 5 *Pressure traces from a patient with valvular effect from a hindbrain hernia showing the effect of a single cough. When the differential trace is displaced downwards—that is, below zero—there is a downward force acting across the foramen magnum. Reading the trace from left to right, for 7 s after the trace begins only cardiac pulsation is seen, then a cough produces a pressure pulse up to 30 mm Hg (4 kPa) in the lumbar sac which does not go so high in the head. There is a downward force present thereafter for some 25 s.*

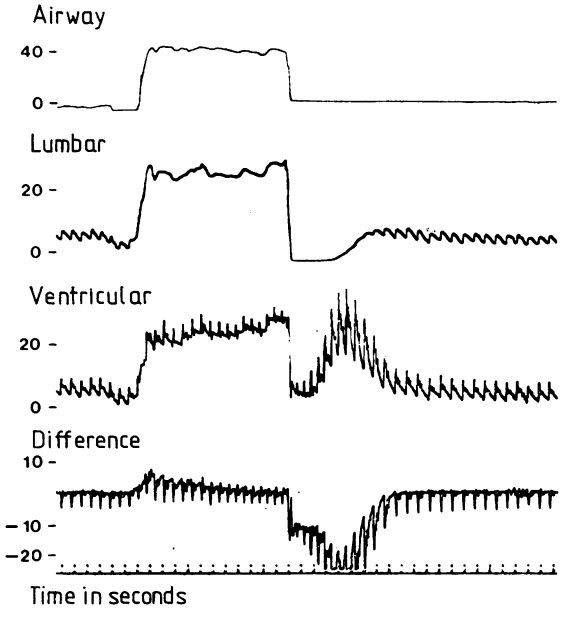

FIG. 6 *Effects of blowing for 12 s to a height of 40 mm Hg (5.3 kPa). The top trace shows the expiratory effort and provides a guide to the abdominothoracic pressure. After the blow is over a post-Valsalva rebound occurs which is confined to the head. The downward force across the foramen magnum is in excess of 25 mm Hg (3.3 kPa). This is severe dissociation.*

into the foramen in both adults and babies with spina bifida.

Secondly, the downward traction provides an explanation for the progressive cranial nerve deformities and the pain which accompanies the maximum pressure dissociation in patients with cough headache.

Thirdly, the filling of syringomyelia may be explained on the basis of a communication between the fourth ventricle and the inside of the spine. Such a communication is commonly present as the central canal which, although of small size, can be expected to transmit increments of fluid under the influence of pressures such as those shown in Figures 5 and 6 thoughout a lifetime. Because the transient pressure increase above the hernia is equally dispersed through all compartments of the head there is no tendency to hydrocephalus.

Finally, the relapse of compensated hydrocephalus after birth is almost certainly related to the onset of respiration, crying, sucking, and so on producing an increase in the ebb and flow through the foramen magnum. If the valvular action causes the hernia to become forced down into the foramen and upper spine this constricts and compresses the exit of the fourth ventricle, thus causing exacerbation of the hydrocephalus. When the meningocele is closed by surgical repair the pressure swings in the spine in response to crying are higher because the damping effect of the meningocele has been removed *(9,10)*, causing impaction of the hernia to be speeded up.

Operative treatment

Although treatment of a hernia by making the rim of the orifice larger might seem contrary to surgical principles, this is the basis of craniovertebral decompression. Part of the occipital bone is removed from the back of the foramen as also is the arch of the atlas. The exposed dura is removed and the impacted tissues decompressed. The wound is closed leaving the dura widely open. If a free flow of CSF is re-established, then the pressure difference is abolished (Fig. 7) and the intermittent force moulding the hindbrain is removed. Relief of downward dislocation may alleviate traction on cranial nerves or pain-sensitive structures; similarly the foramen of Magendie may be decompressed and pressure may be taken off the cord if the tonsils have been compressing it directly.

The hindbrain is suspended in CSF of almost

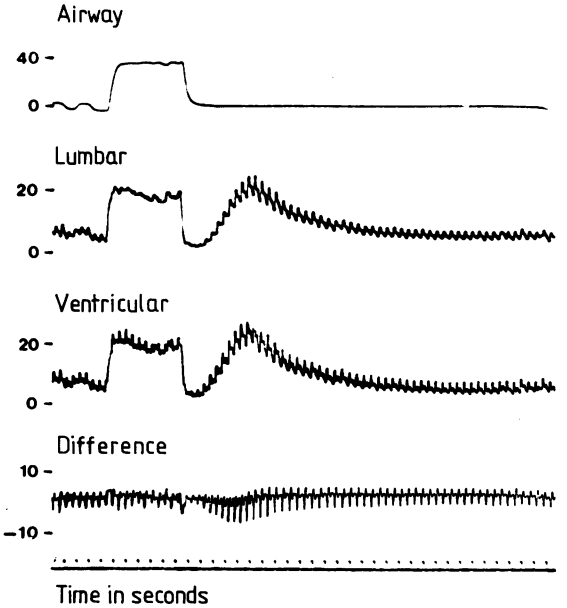

FIG. 7 *Same case as in Fig. 6 after operative decompression of the hindbrain hernia. Note that the post-Valsalva rebound is shared equally between the head and the spine and that the differential trace shows little deviation from zero compared with Fig. 6. The trace is indistinguishable from normal and was accompanied by an excellent clinical result.*

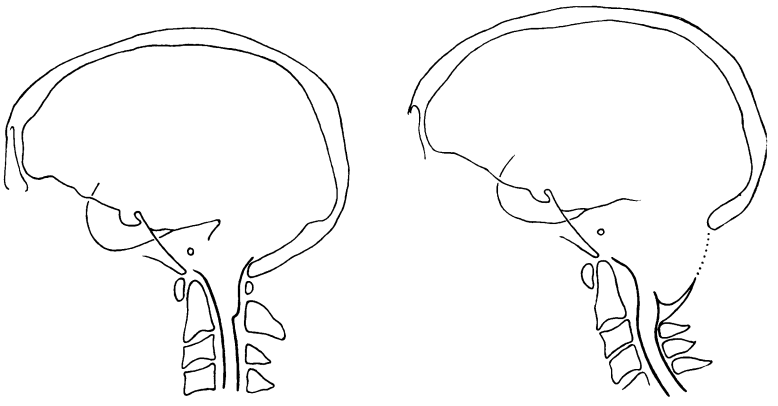

FIG. 8 *Drawings made from lateral radiographs of a patient before and after a generous decompression of the tonsils. The cerebellum has slumped badly into the decompression and carried the brainstem down with it producing postoperative headache.*

the same density, the cerebellum is held on to the brainstem by the cerebellar peduncles, and if the decompression is modest the remaining parts of the occipital squamous bone support the lateral parts of the cerebellum. If the decompression is too large or if craniospinal pressure dissociation is still occurring, then the hindbrain may 'slump' into the decompression, producing a worse hernia than the original lesion. An example is given in Figure 8 of a case described elsewhere *(11)*. Because of the possibility that free egress of CSF from the head to the spine may not be re-established it is tempting to open the foramen of Magendie widely and hold it open. This may be done by making a slit in the tonsils and sucking out the contents, suturing the pia together so that the tonsils are plicated, and taking the sutures upwards and outwards and fastening them to the dura. Alternative schemes include the insertion of a tube to hold the foramen of Magendie open *(12)*. A series of soft pliable funnels have been made from woven silicone rubber *(13)*. The funnel is cut into petal-like flanges and is put in upside down with the hollow stem holding open the foramen of Magendie. The petals are bent back and sutured to the dural edges so as to support the lower edge of the cerebellum. This prosthesis has not been successful, the postoperative course usually being stormy. One patient died from an associated tumour and necropsy showed that the funnel had caused inflammatory reaction and fibrosis with dense blockage of both the tube and the foramen of Magendie.

Although it is possible to evacuate the bulk of the tonsils and suture them back without causing any neurological deficit, there is sometimes a difficult postoperative period, particularly if the brainstem has been manipulated. The results after decompressing the craniovertebral junction seem as good as after more radical interventions and this is at present the preferred policy.

Results

ADULTS

Operative treatment of hindbrain hernia in adults has been encouraging. My personal series of 53 cases of chronic hindbrain hernia in adults is broadly in line with the experience of others *(14–20)*. The most impressive results have been with posterior fossa presentations such as cough headache, but 42% of patients with syringomyelia have shown unequivocal improvement after operation and in others the progression of the disease has seemed to be arrested or delayed. Clinical improvement may be pleasing; all symptoms may be abolished without perfect correction of craniospinal pressure dissociation.

The complications of decompression of adult Chiari deformity for syringomyelia have been commoner in patients who had hydrocephalus, syringobulbia, or arachnoiditis around the herniated tissues *(11,14,19)*. In such patients posterior fossa operations may be avoided. A ventriculoatrial shunt for hydrocephalus may help not only posterior fossa types of presentation but also syringomyelia *(11,20)*.

BABIES

The hernia seems likely to be the cause of the exacerbation of the hydrocephalus in babies and although attempts to control it by hindbrain decompression have been made, failure has been almost universal. The rewards of allowing the normal pathways to open would be considerable. The arachnoid villi make a more satisfactory absorptive mechanism than any mechanical valve. If the head could develop normally for a few months the normal pathways might be remoulded to limit dependence on any foreign device.

A small series of cases has been treated in conjunction with monitoring of craniospinal

pressure dissociation using hindbrain decompression and insertion of a valved shunt, developed by Eric Turner, posing no resistance and allowing fluid to move from the head to the cervical subarachnoid space. This, it was hoped, would prevent the development of craniospinal pressure dissociation and reverse the trend to hindbrain impaction. Although some of the results were initially encouraging, the immediate morbidity and complication rate was higher than for ventriculoatrial shunting and the method has been abandoned.

Future developments

The treatment of chronic hindbrain hernia at present is frequently beset by the difficulty that when the diagnosis is made in the adult the damage continues to progress inexorably despite the apparently successful correction of the hindbrain hernia. This is particularly true of syringomyelia. Emphasis therefore should be upon prevention and earlier diagnosis of hindbrain herniation.

If, as seems probable, difficult birth is the commonest aetiological factor, then it does not give any cause for complacency that obstetrics is advancing and the perinatal mortality rate is falling. The perinatal mortality rate in England and Wales has not improved as well as in other developed countries *(5)*. Perhaps more important is the consideration that as neonatal intensive care units become more successful, so does the number of children at risk who survive increase. The most spectacular increase in survival has been seen in infants with birth weights of 1500 g or less and it is this group which is particularly prone to neurological sequelae such as spastic diplegia, hydrocephalus, epilepsy, and mental retardation. Thus it seems unlikely that the occurrence of a group at risk can be lessened, short of euthanasia for low-birth-weight infants. The hindbrain hernia is usually silent for many years and it is perhaps advisable to use modern techniques to screen babies who have been at risk.

Lateral skull radiographs of newborn infants may show occipital bone fracture. It may be possible to decide whether basilar invagination can occur as part of moulding during birth.

Computer-assisted tomographic scanning gives more information : intraventricular, paraventricular, and subarachnoid haemorrhage may be seen *(5)*. Transient hydrocephalus is likely to be common in infants after difficult birth with hindbrain hernia. It is possible that in the majority of cases of hindbrain hernia which become chronic a hydrocephalus is present in the early stages which may later become compensated.

If babies after difficult labour develop hydrocephalus or a cisternal haematoma it would seem sensible to image the tonsils by Amipaque. If the tonsils are herniated, then pressure recordings from the ventricles and the spine simultaneously are easy to carry out in the first year, using the anterior fontanelle.

If craniospinal pressure dissociation can be demonstrated in conjunction with chronic hindbrain hernia, then surgical correction may be advisable in childhood. Even if surgery is thought inadvisable it would seem sensible to follow up such children carefully as outpatients.

There is a need for research into chronic hindbrain hernia. Although there is a clear association with difficult birth a proportion of hernias have no known aetiology. It may be that the desire to explain deformity in terms of pressure is unduly simplistic and that teratogenic factors are at work in the hindbrains of many persons who present with mild forms of the Chiari deformities in adult life. In many cases, however, the pressure disturbances seem to be important and indicate the opportunity for minimising a deal of suffering, provided that the lesions are promptly recognised.

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