LP Shunting in Cases of Chiari Malformation

To the Editor: I enjoyed the carefully worded article from Park and coworkers (Park TS, Cail WS, Broaddus WC, et al: Lumboperitoneal shunt combined with myelotomy for treatment of syringohydromyelia. J Neurosurg 70:721–727, May, 1989). Parts of it were provoking. The authors have identified the biggest weakness of the Gardner hypothesis, which is that a communication from the fourth ventricle to the syrinx cavity is just not present in the majority of cases, and therefore is not found on radiological investigation or at necropsy. There is of course much experimental as well as embryological evidence to support the idea that a functional communication of some sort probably existed at some time and this does not require repetition.

Absence of a communication, when it seems only common sense that such much have existed, is also found lower in the spine. It is increasingly clear that, in cases of posttraumatic syringomyelia associated with spinal paraplegia, there is quite frequently a syrinx present away from the site of blockage with no sizeable communication between the cord cavity and the site of the paraplegia. At the fracture level there is sometimes a separate cyst and sometimes an arachnoid pouch with no radiologically or surgically demonstrable communication to the syrinx. These are very testing cases and obviously must lead us to question some of the ideas put forward by Gardner and championed by others, including myself.

There is a serious objection, however, to the idea that fluid is forced into the spaces of Virchow-Robin or comparable clefts and chinks in the spinal cord, thus producing a distending cavity. It is impossible to imagine on mechanical grounds how, when a tube (be it never so porous) is suspended in a fluid the pressure of which is then intermittently raised, the tube becomes distended and dilates a cavity within it. It seems more likely that the cavity develops from a mechanism such as fluid being pumped into a patent central canal and that maintenance of the cavity is due to the production of undulating movements in the wall of the syrinx causing excavation at both ends. Violent movement of fluid inside the cavity may be called “slosh.” The walls of the cord contain no tight junctions and clearly must allow easy transfer of such fluids as water-soluble contrast medium and cerebrospinal fluid (CSF) itself. The physics of replenishment of the syrinx cavity by transudation across the walls of the cord appear to be complex, but may involve transient pressure differences across the walls associated with slosh which could provide active filling.

The opinion expressed in the article that my pressure recordings were wrong and that lumbar pressures are either equal to or higher than pressures in the head in cases of syringomyelia with myelomeningocele is disturbing. It is almost certain that low pressure in the spine causes the hindbrain hernia in the first place. The authors’ view is certainly incorrect for the majority of newborn infants with spina bifida aperta, as was pointed out in 1932 by Grätjik.1 The majority of these babies when seen in the early hours after birth have a lumbar sac which is at atmospheric pressure while the head is full. If the sac is removed, the pressure in the lumbar subarachnoid space is deprived of its vent to atmosphere and the pressure swings become very great, as shown in the article by Park, et al. In the majority of babies, there is both established dissociation and also valvular effect while the hydrocephalus is building up in the time between closure of the back and ventricular shunting. This means that the spinal spaces act as a pump and keep forcing CSF up past the hindbrain hernia into the head. It seems that as the valvular action persists the hindbrain is more tightly impacted and then, as the outlets of the fourth ventricle are more severely compressed, hydrocephalus becomes very marked and ventricular shunting is usually performed.

When the ventricles are shunted, usually the whole situation changes and the baseline pressures in the head and the spine become equal. I have not seen any child with properly controlled hydrocephalus in whom I could show significant valvular effect. For ethical reasons I have not pursued a large number of cases in this way. I certainly agree that, when a child is tested during crying after the spine is closed, the envelope of the traces is much higher in the lumbar sac because the peaks of this pressure fluctuation are higher than the peaks of pressure produced in the head. Thus, during a period of prolonged crying the mean intracranial pressure may be less than the mean lumbar spinal pressure. The baseline pressure, however, tends to rise in the head because of the valvular mechanism posed by the morphology of the hindbrain hernia. I was able to observe this in most of my patients who were developing hydrocephalus. When the hydrocephalus had been successfully treated by shunting, the pressure was equal with the exception of the envelope changes already mentioned.

I do not find in their article a description of whether Park, et al., recorded these pressures in the patients who had been treated by shunting, or whether the patients were in a stage of active hydrocephalus. It seems from the pressure changes that the lumbar sacs had been closed off; otherwise, the intraspinal pressure
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Pulses would be dampened to almost flat. In a technical comment, I would suggest that they refer their pressures to atmospheric pressures not from different points as they presently do, but from the same point. The hydrostatic differences are thus eliminated. The use of differential manometry makes the recordings much easier to understand and allows frequent checking of baselines during the recordings. I suggest putting the lumbar trace on top and the intracranial trace beneath. If the spinal pressure is subtracted from the head pressure and the difference is displayed, then an upward pressure difference implies a force likely to produce upward movement across the foramen magnum; conversely, a downward displacement implies a downward force.

I am prepared to share all of their doubts about filling mechanisms, but I am not prepared to doubt my belief in the prime importance of “suck” across the foramen magnum or craniospinal pressure dissociation in all diseases associated with hindbrain herniation. It seems dangerous to suggest that subarachnoid lumboperitoneal (LP) or lumbopleural shunting is a preferred method of treatment for any aspect of this symptom complex. The authors could do no better than to reread the article by Welch, et al., where the disastrous effects of LP shunting are reported. Clearly the important thing is whether the Chiari malformation becomes impacted. In over half of my adult patients with hindbrain-related syringomyelia, the malformation could be made to impact at least briefly during strenuous testing at the time the patients presented for treatment. If it becomes impacted so that there is a pressure difference between the head and the spine and then the lumbar sac is shunted, there will be increased “suck” and the Chiari malformation will be pulled tightly into the hole, which will distort the syrinx and securely fix the herniated tissues. The impaction of the hindbrain may lead to lower cranial nerve palsies with swallowing and voice difficulties, tinnitus or deafness, long-tract compression, oscillopsia, severe vertigo, ataxia, abasia, respiratory irregularities, and sometimes death. This disastrous result occurs not infrequently; the problem is that it is not reported in the literature. Surgeons who produce these disasters usually keep quiet about them. It is difficult to build a big series of such cases, but my own experience and that of others indicates that thecoperitoneal shunting in the presence of active hindbrain herniation is a quick way into severe trouble. This is not to say that this procedure does not help syringomyelia in certain cases. Park, et al., have reported four such patients with good results, and this is obviously interesting. It cannot be denied that, as they say, LP shunting combined with myelotomy can effect shrinkage of the syrinx. Myelotomy is probably not necessary. I believe that the explanation is well expounded in another of my articles of some interest to those who consider the theoretical aspects of pulsation in the neuraxis. The explanation offered there is that if the overall CSF pressure is markedly lowered then the veins throughout the neuraxis become distended. This can occur with little change in baseline pressure. If the veins are thus predistended as it were, the injection of blood to the intravenous compartment by coughing and straining is markedly less, and thus the amplitude of “venous” pulsation is greatly reduced; additionally, the total volume of CSF is lessened. The energy transmission through the CSF pathways thus being favorably influenced, it is not surprising that syringomyelia and indeed the herniation itself may be markedly lessened.

If a surgeon desires to treat syringomyelia by moving his patient to the left along the pressure-volume curve, then please let him do it by ventricular to extrathecal shunting and not by lowering the pressure below the herniation of the hindbrain. Even after decompression, suck may persist.

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References

RESPONSE: I am pleased to respond to the comments of Mr. Williams, who has made significant contributions to our knowledge of syringomyelia over the years. He concurs that a major shortcoming of Gardner’s hydrodynamic theory is the lack of conclusive evidence that a communication between the fourth ventricle and syrinx exists in patients with syringomyelia. In fact, Dr. Gardner proposed his theory on the basis of operative findings and he never personally demonstrated the communication either radiographically or pathologically. To advance his theory, he usually cited sporadic case reports of others which showed the communication. Mr. Williams himself has examined the question of the communication, and could delineate radiographically the communication in only seven of 171 patients with presumed “communicating syringomyelia.” The results led to a conclusion: “a sizeable communication is rare at the time when patients seek treatment.” Previous pathological investigations and all recent magnetic resonance (MR) studies, including our own, are in complete agreement with the conclusion. The absence of
the communication at the time of clinical presentation explains why the obex-plugging procedure achieved no better results than did posterior fossa decompression alone.  

When considering the pathogenesis of syringomyelia, it would be appropriate to distinguish between two separate processes, namely: the initial formation and the subsequent maintenance and expansion of the syrinx. In the case of “noncommunicating syringomyelia,” such as posttraumatic syringomyelia, clearly the communication is not required for formation and expansion of the syrinx cavity. Since the ventricular fluid is unlikely to be a source of syringeal fluid in such cases, there must be alternative sources of the syringeal fluid. In the case of “communicating syringomyelia,” especially those associated with myelomeningocele, I agree with Mr. Williams that sometimes the communication may exist and participate in initial formation and maintenance of syrinx cavity. On the other hand, it is also possible that the communication may never exist or, if it exists in early stages, it may close later. Therefore, even in “communicating syringomyelia,” we need to resolve a question of how syrinx cavities are maintained and enlarge in the absence of the communication.

According to Mr. Williams, the “suck” mechanism underlies the initial formation and subsequent expansion of a “communicating syrinx;” the “slosh” mechanism is responsible for expansion of the syrinx. Since MR imaging has clearly demonstrated pulsatile movement of cerebrospinal fluid (CSF) and a flow-void sign in syrinx cavities, the concept of “slosh” appears to be a correct one. In contrast, the validity of the “suck” phenomenon is still debatable. Mr. Williams suggested that CSF is “sucked” from the subarachnoid space of the posterior fossa into the syrinx via the fourth ventricle as a result of a pressure gradient between the posterior fossa and the spinal canal. Some of the clinical features of syringomyelia that contradict the “suck” mechanism are as follows: the outlet of the fourth ventricle is often completely obliterated in older children and adults whose syringomyelia accompanies the Chiari II malformation. Metrizamide-computerized tomography examinations cannot delineate the communication even in cases where the fourth ventricle is filled with contrast medium. As shown in our report, the pressure gradient across the foramen magnum may not be sufficiently great to force CSF into the spinal canal. Finally, a functioning communication is not demonstrable in the majority of patients at the time of clinical presentation.

How certain substances move into the spinal cord is still unclear and Mr. Williams’ objection to considering spinal CSF a source of syringeal fluid is legitimate. However, many studies have unequivocally demonstrated that a variety of substances move into the spinal cord or syringeal cavity. For example, in a recent study using horseradish peroxidase and lanthanum, the extracellular space was shown to provide a pathway for the substances to move into the normal spinal cord and syringeal cavity. At this time, it is thus difficult to rule out the possibility that CSF can also traverse the extracellular space of the spinal cord and reach the syrinx cavity.

With regard to pressure recordings, we did not state either implicitly or explicitly that the pressure recordings of Mr. Williams were “wrong.” Our point was that the data from pressure recordings in our patients do not reconcile with his notion of the valvular effect related to a pressure gradient across the foramen magnum. Our patients were myelodysplastic children with Chiari II malformation. The only reported data on the pressure gradient in Chiari II malformation that can be interpreted with some accuracy have been provided by Mr. Williams. The study included 10 neonates or infants and the pressure differential was examined in 23 recordings. In 13 of the 23 recordings, intracranial pressure was greater than intraspinal pressure; in the other recordings intracranial pressure was equal to or lower than intraspinal pressure. Thus, the pressure gradient even in his patients was not a universal phenomenon. Since submission of our paper, we have examined more patients with Chiari II malformation and ventricular shunts. No definite pressure gradient as suggested by Mr. Williams was observed in our patients.

Our patients who had the pressure recording had undergone ventricular shunting prior to the pressure recording. I agree with all of Mr. Williams’ comments about the effect of shunting on pressure dynamics and advantages of a pressure differential recorder. He suggested that the transducer be placed at different positions to eliminate a factor of hydrostatic pressure. We took that technical aspect into account but, nevertheless, our conclusion regarding the pressure gradient remained unchanged.

The possible impaction of the brain stem following lumboperitoneal (LP) shunting in patients with Chiari malformation is indeed a concern. Being aware of that particular complication, in our patients with Chiari malformation shunting was employed as the last resort. Since posterior fossa decompression alone can reduce the syrinx cavity and LP shunting carries some risk, we have suggested that the decompressive procedures may have to precede the LP shunting. Fortunately, none of our patients has thus far developed this complication. At this time it is unknown whether LP shunting will prove useful in the management of syringomyelia, but the demonstrated reduction of syrinx cavities coupled with clinical improvement following LP shunting deserves further examination. Myelotomy may be necessary to eliminate a pressure gradient across the wall of the syrinx, thereby achieving rapid reduction of the syrinx.

Mr. Williams has stated “The necessity of establishing whether or not there is usually a functioning communication must be clear for the theory of syringomyelia. The propagation of convenient untruths because they allow a ‘rational’ explanation is a besetting difficulty in medical science and is responsible for sustaining
TO THE EDITOR: I read with interest the recent article by Dr. Park and his colleagues (Park TS, Cail WS, Broadus WC, et al.: Lumboperitoneal shunt combined with myelotomy for treatment of syringohydromyelia. J Neurosurg 70:721–727, May, 1989). The authors’ series included 11 patients with Chiari II and I malformation, of whom six were treated with lumboperitoneal (LP) shunting. In the discussion, they suggested a potential occurrence of caudal displacement of the brain stem and cerebellum aggravated by the LP shunting. I would like to comment on this complication.

In my experience of 207 patients with LP shunting (in preparation), two hydrocephalic patients developed postoperative respiratory abnormalities and disturbance of consciousness. Both patients recovered rapidly after ventricular drainage and infusion of saline solution into the spinal subarachnoid space. This clinical process suggested the occurrence of tonsillar herniation. Retrospective evaluation of their computed tomography scans was highly suggestive of Chiari malformation. Based on this experience and on theoretical considerations, we consider LP shunting to be contraindicated in patients with Chiari malformation. Fortunately, the few patients with Chiari malformation described by Park, et al., did not suffer adverse effects from LP shunting; however, further experience might be complicated by symptomatic tonsillar herniation.

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REFERENCES


RESPONSE: We appreciate Dr. Aoki’s comments. Welch, et al., originally brought to our attention the possible caudal displacement of the brain stem following lumboperitoneal (LP) shunting. Their report led us to suggest in our paper that, in cases where LP shunting is considered for syringohydromyelia associated with Chiari malformation, it may be necessary to decompress the foramen magnum before shunting. The six patients in our series who had insertion of LP shunts had undergone decompressive procedures prior to the shunting procedure.

Clinical observations of neurological deterioration following LP shunting, such as those described by Dr. Aoki, will likely help to determine whether LP shunting can be carried out safely in patients whose syringohydromyelia is associated with Chiari malformation. One may argue that the rapid reversal of the symptoms in his patients could have resulted from malfunctioning of the LP shunts rather than drainage of cerebrospinal fluid through the shunt.

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REFERENCE


Hypothermia and Circulatory Arrest in Aneurysm Surgery

TO THE EDITOR: I was fascinated with two recent articles describing hypothermia and circulatory arrest in aneurysm surgery (Chyatte D, Elefteriades J, Kim B: Profound hypothermia and circulatory arrest for aneurysm surgery. Case report. J Neurosurg 70:489–491, March, 1989; Spetzler RF, Hadley MN, Rigamonti D, et al: Aneurysms of the basilar artery treated with circulatory arrest, hypothermia, and barbiturate cerebral protection. J Neurosurg 68:868–879, June, 1988). These articles would certainly seem to herald a rebirth, indeed a renewed interest in the utilization of this technology for the surgery of difficult or nonoperable cerebral aneurysms. In addition, the authors provide us with a brief but important historical overview of the development of these techniques, as well as the experimental work that demonstrated the permissible limits of circulatory deprivation to brain at reduced cerebral temperatures. Our congratulations to the authors of both of these seminal papers.

In examining the historical development of the methodologies for intravascular cooling and the experimental studies that demonstrated the salutary effects of low temperature in protecting the brain under conditions of circulatory arrest, we would like to bring to the