Matters arising

Syringomyelia, an hypothesis and proposed method of treatment

Sir: The natural history of syringomyelia seems to march on uninfluenced by surgical procedures. It is suggested that the major determinant of this progression is the ovoid shape imposed upon the cavity by the spinal cord, that it is inherent in the shape of the cavity that it will intermittently extend.

Williams proposed that there was a flap valve mechanism, whereby coughing, sneezing or straining, or even normal pulse waves caused a pressure difference between the inside and outside of the syrinx. The experience of many indicates that opening the syrinx to the subarachnoid space (so that there can be no pressure difference) results in only temporary improvement, even if quite elaborate precautions are taken to make sure no flap valve can reform. It was this experience in two patients of mine with syringomyelia, which prompted the thought that the major determinant of the progress of the syrinx was its shape.

Most cavities in the brain, if they last long enough, become roughly spherical, though influenced by the shape of surrounding structures. In the spinal cord they cannot, but must become ovoid. The reason for the tendency to a spherical shape is that it distributes the tension most evenly around the walls of the cavity. Thus, for a bubble, the most stable shape is spherical.

The pressure difference, \( \Delta P \), across the walls of a cavity at a point is given by

\[
\Delta P = kT \left( \frac{1}{R_1} + \frac{1}{R_2} \right)
\]

where \( T \) is the tension in the walls, \( R_1 \) and \( R_2 \) are the principal radii of curvature at a point, and \( k \) is a constant. If the cavity is ovoid with a relatively narrow pointed end, then as this end the radii of curvature are small, hence their reciprocals are large. If the reciprocals are large then \( \Delta P \), the pressure transmitted across the wall at that point, or \( T \), the tension in the wall may be correspondingly large.

If there is a sharp rise in pressure in the CSF due to coughing, a Valsalva manoeuvre etc., then the pressure differential across the wall at the apex of an ovoid (or even sharper cavity) will be maximal at this point. It may not be balanced by an equal and opposite pressure in the tissue. The tissues of the CNS are semi-solid and it has been clearly shown in the brain that pressure transmission is not instantaneous, but that pressure gradients can persist for an appreciable time.  

Williams replies:

I hope that Graham Martin will forgive me for correcting him on one point. I initially believed that a valvular mechanism was responsible for the commonest kind of syrinx to fill, and thus in a proportion of cases for aiding its progression; but at the moment I believe that the principal cause of progression in the later stages of the disorder is the hydrodynamic effect produced by pressure changes upon fluid which is already in the syrinx. The fluid can surge within the cord as the subarachnoid space becomes compressed by thoraco-abdominal pressure changes. This pressure change is imposed on the dura by engorgement of the epidural veins. The whole mechanism of rapid fluid movement may be called “slosh”. Thus, I find myself in sympathy with parts of Martin’s analysis. I believe that as the fluid sloshes within the syrinx, say in response to a cough, the ends of the cavity must become more circular in response to the pressure rise as he suggests. It may be excessively pessimistic to state that progression of the disease is uninfluenced by surgery. Many cases seem to do very well after cranio-vertebral decompression and quite a few are helped by treatment of associated hydrocephalus with a shunt. Myelotomy, however, is not a reasonable operation unless there is evidence of high pressure within all parts of the syrinx. This state of affairs is exceptional and most syringes are flaccid when at rest. Thus it certainly appears that obliteration of the cavity would be a sensible step in treatment. I sympathise with Martin over the difficulties in being sure that the shunt is working and that the syrinx stays flat.

Several reports have appeared favouring syrinx drainage to an extrathecal site. It seems probable that there will be a vogue for this form of treatment particularly in cases which progress after cranio-vertebral decompression, and it seems likely to be of permanent help in a proportion of such cases. I favour direct drainage into the pleura without the use of a valve.

With regard to strengthening the walls of the syrinx it may be noted that gliosis may be very dense as though strengthening of the wall was the natural response to the fluid. If the cavity can be collapsed by drainage it seems likely that the walls will adhere at least in parts and may become quite strong. Packing the cavity with muscle or inserting irritant fluids to promote adhesion may require more courage than most spinal cord surgeons can summon up. It might be sensible for results of

References

extrathecal shunting to be pooled in a few years time to try to assess whether flattening the syrinx is indeed worthwhile.

References

Ruptured intracranial aneurysms

Sir: There are two points we would like to make with regard to the article "Ruptured intracranial aneurysms: has the incidence of early rebleeding been over-estimated?" J Neurol Neurosurg Psychiatry 1982; 45:774–9. The author attempts to comment on the occurrence of rebleeding and neurological deterioration following an intracranial aneurysmal rupture, having given the majority of his patients tranexamic acid, a drug used to prevent recurrent haemorrhage and a drug suspected of producing ischaemic neurological deterioration. We have recently demonstrated that the pattern of cerebral blood flow (CBF), measured daily over a three week period, differs considerably in patients on tranexamic acid compared to patients on no drug therapy. There is a substantially greater fall in CBF in the patients on tranexamic acid during the second week after haemorrhage. The author, in his study, is clearly examining the effect of tranexamic acid on a group of patients following an aneurysmal rupture and not the natural history of the disease. Thus the comparing of this series with others where tranexamic acid was not used in a similar way is meaningless.

All neurosurgeons are aware of the difficulty of establishing the occurrence of a recurrent haemorrhage in patients already possessing an abnormal CSF and in the limited value of the CT scan in these circumstances. Small haemorrhages causing sudden and transient clinical deterioration, as suggested by the author are extremely difficult to diagnose by these methods. Contrary to the author’s statement it is nigh impossible at the time of surgery, especially as the author apparently does not operate for at least seven days from such a deterioration, to say whether the patient has had a small recurrent haemorrhage.

The paper, we feel, has asked the wrong question, considering the methodology, and not surprisingly produces a very questionable answer.

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Maurice-Williams replies:

Both points raised by Mr Neil-Dwyer and Mr Sharr are dealt with at length in the discussion section of my recent paper. I agree with them that it is possible that the administration of the antifibrolytic agent tranexamic acid may have modified the natural history of the cases studied but I would point out that there is still considerable controversy as to whether or not antifibrolytic agents have any effect on patients who have had a subarachnoid haemorrhage. As pointed out in the article, if the tranexamic acid has had any effect, it has been to replace one problem (rebleeding) with another problem (non-haemorrhagic deterioration), the classically described characteristic time-course of rebleeding having shifted to non-haemorrhagic deterioration while the time-course of rebleeding has become flattened and uniform. This would seem a very surprising effect. Furthermore the total number of episodes of rebleeding and non-haemorrhagic deterioration added together in the first three weeks of the study was very close to the number of "rebleeds" alone that the Co-operative Study would have predicted during this period. The confirmed rebleeds in my study had a very high mortality (90%), but the mortality of the cases of rebleeding and non-haemorrhagic deterioration added together (55%) approximated to the level of mortality of rebleeding reported by earlier papers. All these phenomena would appear to be better explained by the hypothesis that earlier studies had confused episodes of non-haemorrhagic deterioration with rebleeding during the first few weeks after the subarachnoid haemorrhage. It is also possible that confusion between rebleeding and other causes of deterioration may account for the fact that some studies have reported antifibrinolytics to reduce the incidence of rebleeding while other studies have reported no such effect.

With regard to the second point raised by Mr Neil-Dwyer and Mr Sharr, it is indeed possible that some episodes of non-haemorrhagic deterioration were very minor rebleeds, too small in extent to be detected by repeat CT scan or repeat lumbar puncture. However this would explain the totally different time-courses of rebleeding and non-haemorrhagic deterioration, nor the fact that patients who had an episode of non-haemorrhagic deterioration were not more likely to later rebleed than those who had more stable course. In addition, despite the fact Messrs Sharr and Neil-Dwyer say several patients came to surgery within 7–10 days of an episode of non-haemorrhagic deterioration and in no case was there any evidence of a limited local rebleed found. The same was true of the eight patients who died after a period of non-haemorrhagic deterioration and who came to post-mortem examination. I am interested by their statement that tranexamic acid appears to lower the cerebral blood-flow in patients during the second week after haemorrhage. This appears to be at variance with the findings of a recently published study from their unit or on the cerebral blood flow after subarachnoid haemorrhage in older patients which stated that, in this group of patients at any rate, tranexamic acid did not selectively depress the cerebral blood flow. 1

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References

The treatment of acute polyneuropathy by plasma exchange

Sir: The different experiences reported to this Journal in September and August respectively, by the London Hospital