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. Presumably the spinal cord is similar.

In contrast, pressure is transmitted instantaneously through a fluid such as CSF, whereas there may be a delay in transmitting it through tissues. Also, from the formula above the pressure difference across the walls is likely to be greater at the narrowest end of the syrinx, when there is a rise in CSF pressure.

Hence, if the ovoid shape of the syrinx by itself can cause the cavity to enlarge, then obliteration of the cavity would be the proper treatment. These thoughts led me to shunt the syrinx into the peritoneum in two patients, in order to close the cavity. For reasons which are diverse, different and irrelevant to this argument, it has not been possible to determine success or failure—in particular, one problem is that it is hard to know if the shunt is working. This difficulty suggests that there is a practical objection to shunting as a treatment. Even if one can determine whether or not the shunt is working, the significance of the "blocked" shunt would be unknown. A shunt that had completely done its duty and collapsed the cavity would feel the same as a blocked one which was causing the cavity to re-inflate.

Consequently, if the cause of the unpredictable progression of these cavities is their shape, and the instantaneous transmission of pressure through the fluid CSF, with slower transmission through the semisolid tissues—then perhaps the best treatment would be to pack the cavity with muscle to exclude CSF and strengthen the walls by fibrosis.

In short, if a cavity develops in the cord, whether due to developmental failure of the canal to close, or due to trauma producing a haematomyelia, or from a cavity in a tumour, then its shape gives it an inherent tendency to enlarge, limited only by the strength of the tissue at its ends. Treatment should be obliteration of the cavity and strengthening of the walls.

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References


Williams replies:

I hope that Graham Martin will forgive me for correcting him on one point. I initially believed that a valvar mechanism was responsible for causing 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 subarachnoidal 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 syrinxes 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. Pacing 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