Hypothesis on the pathophysiology of syringomyelia based on simulation of cerebrospinal fluid dynamics

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Objective: Despite many hypotheses, the pathophysiology of syringomyelia is still not well understood. In this report, the authors propose a hypothesis based on analysis of cerebrospinal fluid dynamics in the spine.

Methods: An electric circuit model of the CSF dynamics of the spine was constructed based on a technique of computational fluid mechanics. With this model, the authors calculated how a pulsatile CSF wave coming from the cranial side is propagated along the spinal cord.

Results: Reducing the temporary fluid storage capacity of the cisterna magna dramatically increased the pressure wave propagated along the central canal. The peak of this pressure wave resided in the mid-portion of the spinal cord.

Conclusions: The following hypotheses are proposed. The cisterna magna functions as a shock absorber against the pulsatile CSF waves coming from the cranial side. The loss of shock absorbing capacity of the cisterna magna and subsequent increase of central canal wall pressure leads to syrinx formation in patients with Chiari I malformation.

Methods

Figure 1 shows our electric circuit model superimposed on the actual anatomical structures. The CSF pathways are broken up into multiple nodal points starting from the fourth ventricle and prepontine cistern, followed by obex and cisterna magna, and then by nine nodal points and finally the lumbar theca. (The number of nodal points is arbitrary.) We represented the compliance (or the temporary fluid storage capacity) of the cisterna magna with a capacitor named Ccist. In the spine, two pathways of CSF, namely the central canal and the extramedullary subarachnoid space, are represented as series of multiple resistors. To represent the compliance of the spinal cord and the dural sac, we inserted two arrays of capacitors: one between these two CSF pathways, and the other between the subarachnoid CSF pathway and the ground. Thus, the capacitors between the central canal and the subarachnoid space (C1 to C9 in fig 1) represent the elasticity (compliance) of both the central canal wall and the spinal cord. Similarly, the capacitors between the subarachnoid space and the ground (D1 to D9 in fig 1) represent the elasticity of both the dural sac and the root sleeves. Table 1 shows the values of the parameters that we used for the calculations in this study.

We could solve this electrical diagram with the standard analysis technique of a linear system. After setting the initial voltage accumulated in C1 to C9 to zero, and that in Ccist, D1 to D9 and Ccist, to 100, we applied a sudden increase of voltage on the cranial side of this model, simulating the CSF pressure obtained from the CSF pressure transducers taken as unknown variables. These equations were solved on a personal computer using a software package: Mathematica version 4.0 (Wolfram Research, Champaign, IL, USA).
We then examined the effect of changing the temporary fluid storage capacity (or compliance) of the cisterna magna. Because reduced compliance at the cisterna magna is equivalent to reduced capacitance of $C_{\text{cist}}$ (fig 1) in our model, we calculated the response of our circuit with three different values: the original value, one tenth of the original value, and one hundredth of the original value.

To simulate the effect of syringo-subarachnoid shunting, we inserted a small resistor valued 0.2 bypassing the capacitor $C_4$ in figure 1 in addition to the reduced value of $C_{\text{cist}}$ to one hundredth of the original value.

RESULTS

The time course of the pressure wave propagated along the central canal in response to a sudden increase of voltage on the two cranial leads is shown in figure 2. In biological terms, the voltage accumulated in these capacitors corresponds to the pressure difference between inside and outside the spinal cord. The time course of the accumulated voltage at each nodal point, a positive value of the voltage indicating higher pressure inside. In the following figures, the horizontal coordinate shows the nodal points (1 to 9 in fig 1) in our model, the left side being the cranial side and the right side being the caudal side. The time course of the accumulated voltage at each nodal point is shown in the figure starting from the time of application of the pulsatile wave at time zero. Figure 2A, B, and C show the responses of the system with three different values of capacitance at the cisterna magna ($C_{\text{cist}}$ in fig 1). As the capacitance of $C_{\text{cist}}$ was decreased, there was a dramatic increase of the central canal wall pressure. The peak of this pressure increase was at the mid-portion of the spinal cord.

The result of the simulation of syringo-subarachnoid shunting is shown in figure 3. In this case, we set the value of $C_{\text{cist}}$ at one hundredth of the original value. In this setting, the pressure response is increased as in figure 2C. We placed an electrical shunt at the nodal point 4 (fig 1) between the central canal and the subarachnoid space. As shown in figure 3, this shunting effectively reduced the central canal wall pressure, although it was less effective on the cranial side.

DISCUSSION

Our results showed that reduced compliance at the craniovertebral junction increased the pressure wave propagated through the spinal cord. This phenomenon is analogous to the Windkessel model of the cardiovascular system. In the Windkessel model, the aorta functions as an elastic tube with temporary fluid storage capacity. This temporary fluid storage capacity is understood with an analogy to a Windkessel—a reservoir of a water pump of a fire engine (fig 4). When the aorta becomes stiff and loses this temporary fluid storage capacity, the arterial pulse wave becomes steep resulting in systolic hypertension. Likewise, our model provides a hypothesis explaining why the constriction at the craniovertebral junction causes the formation of syrinx in the spinal cord. When the temporary fluid storage capacity of the cisterna magna is reduced as in Chiari type I malformation, the pressure wave propagated through the central canal is increased. In other words, we can think that the cisterna magna normally functions as a shock absorber, which absorbs the pulse pressure of the CSF coming from the cranial side. If this shock absorbing capacity is lost, because of the overcrowding of the posterior fossa in Chiari 1 malformation, the CSF pressure wave propagated along the central canal is markedly increased; it will then lead to leakage of CSF into the parenchyma, which precedes the formation of syrinx. Thus, we may consider that our hypothesis is a modified version of Gardner’s water-hammer theory. The difference is that our theory can explain why extradural decompression at the craniovertebral junction without intradural procedure can reduce the size of the syrinx in the spinal cord.

Our hypothesis is better in some regards than the other hypotheses presupposing the role of the Virchow-Robin space. Firstly, it can explain the phenomenon of syringomyelia with no tonsillar herniation, and why foramen magnum decompression is effective in reducing the size of the syrinx. Tubbs et al13 showed that these patients had moderately small posterior fossae. Our hypothesis can explain this phenomenon well, because we can assume that the temporary fluid storage capacity at the cisterna magna is reduced in such patients because of the mild overcrowding of the posterior fossa. On the other hand, other hypotheses have difficulty explaining this phenomenon, because they require either a one way valve mechanism or a piston-like movement of the cerebellar tonsils, which are less likely to occur in those patients with no Chiari malformation. Secondly, our model is compatible with the fact that syringo-subarachnoid shunting is effective in reducing the size of the syrinx. Although the syringo-subarachnoid shunting is known to have poor long term effects, our model suggests that it might be effective in reducing the size of the syrinx.
results mainly because of obstruction of the tube, it is usually
effective in short-term. If we assume that the CSF enters
from the subarachnoid space to the syrinx through the
Virchow-Robin space, we cannot easily explain this phenom-
enon; a larger conduit made by a syringo-subarachnoid shunt
will, simply interpreted, rather aggravate the syrinx.

The weak point of our hypothesis might be that it assumed
the patency of the central canal, or an existence of some other
channel connecting the fourth ventricle and the syrinx. Only
in about 14% of the patients with syringomyelia associated
with Chiari type I malformation, was the connection between
the syrinx and the fourth ventricle demonstrated on MRI. Also,
cadaver studies showed that the central canal was oblit-
erated in a large proportion of the human population as the
age increased.

We can make several arguments concerning
this point. Firstly, to say the least, our theory can explain the
pathophysiology in those 14% of patients who have communi-
cating syringomyelia associated with Chiari type I malforma-
tion. Secondly, MRI may not be able to demonstrate the con-
nection between the syrinx and the fourth ventricle. There is
some evidence that the current resolution of MRI cannot
detect the normal sized central canal.

In that case, we may
have larger proportion of patients with communicating syrin-
gomyelia that can be explained by our theory. Actually, Milho-
rat et al found in their pathological study that, in 42% of syrin-
gomyelia associated with Chiari I malformation, the syrinx
was rostrally communicating with the patent central canal.
Thirdly, our model showed that if there was stenosis of the
central canal at some point, the pressure wave was markedly
increased immediately rostral to that stenotic point (data not
shown). If we postulate that this increased pressure wave
ruptures the ependyma of the central canal, and drives the
formation of syrinx inside the parenchyma, our theory does
not necessarily contradict the necropsy finding of progressive
obliteration of the central canal in human. Fourthly, even if
the central canal is obliterated in a large percentage of the eld-
erly human population, it does not necessarily mean that the

Figure 2 Three dimensional plots showing the wall pressure of the
central canal [represented as the voltage of the capacitors Cₐ to C₉]
after application of a step input on the cranial leads at time 0. The
voltage at each nodal point 1 to 9 in figure 1 is plotted. The value of
Ccw is set at the original value in A, one tenth of the original value in
B, and one hundredth of the original value in C respectively.

Figure 3 Simulation of syringo-subarachnoid shunt. The same plot
as figure 2C except for an insertion of a small resistor shunting the
capacitor C₄ in figure 1 simulating the syringo-subarachnoid shunt.
The increased wall pressure response seen in figure 2C is much
reduced.

Figure 4 Cisterna magna functions like a Windkessel of a fire
engine’s water pump. If it becomes “stiff” and loses its temporary
fluid storage capacity as in Chiari I malformation, the pressure wave
propagated along the central canal is increased, leading to syrinx
formation.

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same thing happens in the patients with Chiari type I malformation. In those patients, our results suggest that the comparatively small volume of the posterior fossa causes increased pressure waves along the central canal; this increased pressure wave may prevent the normal process of occlusion of the central canal. Fifthly, if we closely observe our model, we find that the patency of the central canal is not absolutely required. Our model only assumed some fluid channel inside the spinal cord, and this fluid channel may well be the Virchow-Robin space.

Our theory may also be criticised because we used rather arbitrary values on the resistors and capacitors of our circuit. However, we were only interested in the qualitative behaviour of our circuit, not the quantitative determination of the amplitude and the time course of the pressure waves. The qualitative behaviour of our circuit was basically unchanged with different sets of parameters that we tested. Our study is theoretical in nature; we believe that a theory that can elegantly explain the actual phenomena can be justified as far as its assumptions do not absolutely contradict the experimental findings.

Our theory is important in the clinical perspective. Even though we have a standard surgical approach to patients with syringomyelia associated with Chiari malformation, it is not satisfactory if we do not clearly understand why it works. Better understanding of the pathophysiology will certainly improve the results of our surgery. In addition to syringomyelia associated with Chiari type I malformation, we have other types of syringomyelia such as that associated with adhesive arachnoiditis. Our theory was also effective in explaining the pathophysiology of syringomyelia associated with adhesive arachnoiditis, thus providing a unifying theory on those two types of syringomyelia. We believe that our theory can serve as a working hypothesis for further clinical and experimental works.

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