The Presyringomyelic Myelopathic State: A Plausible Hypothesis

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The mechanisms by which nontraumatic, nontumoral syringomyelia develops are not well understood. Although intradural scarring, arachnoiditis, and vascular compromise have been implicated as contributing to syrinx formation, a unified concept to explain this abnormality and the changes in the cord that precede it have not been established. In this issue of the *American Journal of Neuroradiology* (page 7), Fischbein et al affirm an unproved but nonetheless plausible hypothesis that not only furthers the understanding of the underlying pathophysiology of syrinx formation but may also direct treatment of the symptomatic presyrinx state. Specifically, the authors invoke alterations in cerebrospinal fluid (CSF) flow and intraspinal pressure as primary causes of syringomyelia formation. This article, along with Milhorat's accompanying commentary (page 21), deserves close inspection and comment.

Abnormal CSF flow and abnormal intraspinal fluid pulse pressure can result from a number of causes, including the Chiari malformations with low-lying tonsils and prior intradural inflammatory processes. In the former situation there is a downward thrust of the cerebellar tonsils through a narrowed foramen magnum during cardiac systole. In the latter situation there is meningeal thickening and scarring in the subarachnoid space. In either case there is an alteration of the normal fluid pressure relationships in the subarachnoid space (SAS) and the spinal cord. What remains unclear is how these pressure changes, particularly those that may result in large pressure differences at the level of a block, correlate with concurrent anatomic conditions of the central canal that may lead to signal abnormalities in the cord and eventually to syringomyelia. These issues are addressed by Fischbein et al when they describe the consequences of pressure alterations and edema within the spinal cord (the presyrinx state) in the presence of various degrees of central canal patency.

Normal CSF diffusion along perivascular spaces within the spinal cord depends on the maintenance of proper systolic/diastolic pressures in the SAS. When this relationship is disturbed by increased pulsatile pressure, it is possible that fluid directed into the extracellular space of the cord does not become dispersed in a normal fashion. In a process that may develop over time, this pressure gradient can cause an abnormal amount of fluid to enter the central canal, provided that it has remained patent. If the canal is only segmentally patent, a focal syringohydromyelia with cord enlargement may develop; if the canal has totally involuted, the spinal cord may become edematous. In both of these conditions neurologic symptoms can develop, and if the situation persists long enough, those neurologic symptoms may become irreversible. Surgical intervention to prevent irreversible neurologic symptoms would be critical at this juncture.

Why are the symptoms leading up to syringomyelia so often overlooked? CSF flow abnormalities with intraspinal adhesions, canal narrowing, or scarring within the SAS are not uncommon manifestations of syrinx formation. It is possible that a certain degree of flow abnormality is required before significant pressure changes occur or perhaps the cord adjusts to these abnormal fluctuations in pulse pressure. Whatever the explanation for the relatively low prevalence of this abnormality, the proposed pathogenesis has clear implications for treatment. If normal flow and pressure relationships can be reestablished, then cord edema may subside, progression toward a possible syringomyelia may be interrupted, and neurologic symptoms should stabilize or improve. Because intervention is critical at this point, we frequently perform CSF flow studies at our institution when MR findings of intradural abnormalities suggest altered CSF dynamics. Although these MR examinations are most commonly viewed in the midsagittal plane, recent work has allowed us to view sagittal and parasagittal images almost simultaneously, so that lateral and midline flow can be evaluated. Specifically, we look for systolic and diastolic alterations in CSF flow in combination with spinal cord deformity and meningeal thickening. It is clear from Fischbein's article, however, that in certain abnormalities, such as Chiari malformations, we ought to look carefully at cord and tonsillar movement also. With the aid of imaging and the proper clinical circumstances, revealed abnormalities in CSF flow within the subarachnoid space can guide the surgeon. This finding can help to determine if and how normal flow by spine decompression should be accomplished; by dehiscence of subarachnoid scarring, resection of scarred dura, duroplasty with a dural allograft, or a combination of these procedures.

What additional lessons do we take away from Fischbein's study? First, when MR findings reveal structural changes suggesting abnormalities in intraspinal CSF flow, and these changes are seen in combination with a homogeneous, abnormal signal within the spinal cord, cord enlargement, and no abnormal enhancement, the possibility of a presyrinx state should be considered. Second, fluid dynamics (both flow and pressure)