Significance of positive Queckenstedt test in patients with syringomyelia associated with Arnold-Chiari malformations

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Ten patients with syringomyelia associated with Arnold-Chiari Type I malformations were evaluated. In each patient, a manometric Queckenstedt test was performed with the neck in various positions. No patient showed evidence of a block to the flow of cerebrospinal fluid (CSF) with the neck in the extended position; however, all showed a complete CSF block with the neck in a flexed position. Posterior fossa decompression with a C1-2 laminectomy was performed in nine cases, after which Queckenstedt test demonstrated free CSF communication in all nine with the neck in extension, in a neutral position, and in flexion. Postoperative magnetic resonance imaging showed shrinkage of the syrinx in the patients who underwent surgery. It is suggested that obstruction of the CSF pathway at the foramen magnum produced by neck movement is of importance in the formation and progression of a syrinx.

KEY WORDS • Arnold-Chiari malformation • Queckenstedt test • syringomyelia

SINCE Gardner7 first proposed his hydrodynamic theory as a cause of syringomyelia associated with Arnold-Chiari malformation, the dynamics of cerebrospinal fluid (CSF) circulation have attracted considerable attention. However, controversy continues as to the initiating cause of syringomyelia, whether it be ventricular pulse waves3 or transient pressure dissociation above and below the foramen magnum.1,2,13 Williams14 has also proposed a “sloshing” mechanism for progression of a syrinx. This confusing situation is partly due to the fact that, in patients with a communicating syringomyelia associated with Arnold-Chiari malformation, a CSF block has rarely been demonstrated on Queckenstedt testing.2,3,8 The following studies were undertaken to determine if a block at the level of the foramen magnum could be demonstrated in relation to the position of a patient’s neck.

Clinical Material and Methods

Ten patients with syringomyelia associated Arnold-Chiari malformations confirmed by magnetic resonance (MR) imaging were included in this study (Table 1). All were women, ranging in age from 32 to 75 years. Computerized tomography (CT) scans revealed normal ventricular size. No clinical signs of raised intracranial pressure were detected. Patients with syringomyelia associated with ventricular dilatation were excluded from this study.

Queckenstedt Test

A graduated electric manometric Queckenstedt test was carried out in a manner described elsewhere.9 Each patient was asked to lie on her side. Lumbar CSF pressure was recorded through an electric pressure sensor connected to a No. 20 or 21 needle. The patient’s jugular vein was compressed by inflating an ordinary blood pressure cuff wrapped about the neck. The cuff was pumped up as rapidly as possible to a pressure of 20, 40, or 60 mm Hg, which was held for 10 seconds then released. Inflation of the cuff was repeated with the neck in the neutral, extended, and flexed positions.

Operative Procedures

Posterior fossa decompression with a C1-2 laminectomy was performed in nine of the 10 cases. The operation was performed with the patient prone, keeping her neck in a neutral position. Duroplasty was carried out with a patch graft for the purpose of widening the foramen magnum. In five patients, the funnel-shaped opening of the central canal was plugged with a piece of muscle. In one 75-year-old patient in poor general condition (Case 7), a syringosubarachnoid...
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TABLE 1

Neurological symptoms and signs in 10 patients with syringomyelia associated with Arnold-Chiari malformation*

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<th>Case No.</th>
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* Abbreviations: + = sign or symptom present; - = sign or symptom absent.

TABLE 2

Queckenstadt test in 10 patients with syringomyelia associated with Arnold-Chiari malformation*

<table>
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<tr>
<th>Test</th>
<th>neck position</th>
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<td>CSF protein level (mg/dl)</td>
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<td>32</td>
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</table>

* Abbreviations: CSF = cerebrospinal fluid; - = no block; ± = partial block; + = major block; ++ = complete block.

shunt with silicone tubing was placed. The radiological, operative, and Queckenstedt test results were analyzed.

Results

Neurological and Radiological Findings

The patient data are summarized in Table 1. Magnetic resonance imaging revealed ectopia of the cerebellar tonsils in all cases. No associated abnormalities were demonstrated such as spina bifida, hypoplasia of the cerebellar vermis, or low position of the fourth ventricle, or of the medulla oblongata. Except for syringomyelia, there was no neck pathology such as spondylosis, herniated discs, or neoplastic lesions.

Cerebrospinal Fluid Flow Dynamics

The results of the manometric Queckenstedt test are summarized in Table 2. No block to CSF flow was demonstrated in any patient with the neck in the extended position and, with the neck in the neutral position, a major CSF block was observed in only one case. In contrast, testing with the neck in the flexed position demonstrated severe to complete CSF block in all cases. Analysis of the lumbar CSF showed that protein concentrations were normal to slightly elevated, with a mean value of 45.3 ± 27.0 mg/dl (range 17 to 112 mg/dl). The CSF block with the neck in the flexed position resolved after posterior fossa decompression (Fig. 1).

Operative Results

In all patients the obex was located approximately 1.5 cm below the foramen magnum, concealed by herniated cerebellar tonsils. No abnormal membrane, severe thickening, or adhesion of the arachnoid membrane at the foramen magnum was observed. There was no postoperative mortality or morbidity. Shrinkage of the syrinx was confirmed in all patients by postoperative MR imaging.

Discussion

Cerebrospinal Fluid Flow Dynamics

Williams13 drew attention to CSF dynamics in relation to syringomyelia with a theory that syrinx formation is caused by obstruction of CSF flow at the foramen magnum rather than at the outlet of the fourth ventricle as previously proposed by Gardner.1 On measuring the ventricular and the spinal CSF pressure simultaneously,14,15 he found that in some, but not all, cases a transient pressure dissociation between the intracranial
Significance of positive Queckenstedt test

CSF and that of the spinal canal exists for a certain period of time after a Valsalva maneuver. He concluded that such transient pressure dissociation is produced by the cerebellar tonsils plugging the foramen magnum.

With regard to the block found on the positional Queckenstedt test in the present investigation, it is of interest that, in patients with neck lesions such as cervical spondylosis, CSF block is exaggerated with the neck in the extended position. In our series, no abnormalities in the neck other than the syrinx were present. Although, with high cervical lesions, a Queckenstedt test has little value when it shows no block, the positional block observed in the present cases should be considered to be located at the level of the foramen magnum. Thus, when the neck is flexed, the cervical spinal cord is elongated and at the same time the medulla, the pons, and even the cerebellar tonsils move downward and exert pressure on the foramen magnum resulting in obstruction of the CSF pathway. Such obstruction cannot be limited strictly to the sharp edge of the foramen magnum, but must also spread to some extent down to where the herniated cerebellar tonsils pack the spinal canal (Fig. 2). With maximum neck flexion, the central canal (which communicates between the syrinx and the subarachnoid space) would therefore most likely be obstructed by compression.

Pathogenesis of Syrinx Formation

Williams concluded that a “sucking” phenomenon occurs for a certain period of time after a Valsalva maneuver, because the CSF pressure in the spinal canal is lower than the intracranial pressure, and that this is the main cause of syrinx formation. According to his idea of pressure dissociation, the opening of the central canal must be located above the foramen magnum. In the present series, however, a low obex position was confirmed at the time of operation in all cases with posterior fossa decompression. Such a caudal dislocation of the obex was also observed by Gardner in 28 of his 74 patients with syringomyelia. The roof of the fourth ventricle is easily visualized by CT or MR imaging, but the exact position of the obex is rather difficult to ascertain with these techniques. None of the patients presented here was predicted preoperatively to have a low obex position, the location being confirmed only when the herniated cerebellar tonsils were retracted during surgery.

In addition to pressure dissociation between the cran-
nial cavity and the spinal canal, both a low position of the obex and packing by the herniated cerebellar tonsils of the funnel-shaped opening of the central canal are candidates to play important roles in the formation of communicating syringomyelia. The “milking” phenomenon caused by intracranial arterial pulsation-associated downward movement of the cerebellar tonsils as proposed by du Boulay, et al., also requires consideration; such a “milking” of the central canal might be produced by neck movement, squeezing the obex and opening the central canal. If such a “milking” phenomenon is produced by neck flexion, pressure dissociation produced by jugular compression might not be needed to explain syrinx formation.

**Syrinx Progression**

Patients with syringomyelia associated with hindbrain hernia often complain of exaggerated radiating pain associated with sneezing, coughing, or simply flexion of the neck. An increment of venous pressure does not appear necessary and these effects might just be due to neck flexing, which also occurs automatically to a certain extent with sneezes and coughs. In contrast to the neutral or extended positions, when the neck is in the flexed position the lower cervical spinal cord is stretched resulting in an increment of intramedullary pressure. In addition to this stretching, the syrinx loses its communication to the subarachnoid space and the CSF pressure inside the syrinx might increase sufficiently to destroy the syrinx wall. Williams proposed a “slosh” mechanism as a factor underlying syrinx growth, based on the theory that the syrinx communicates with the subarachnoid space. He suggested a syringoperitoneal shunt as the treatment of choice for communicating syringomyelia, doubting the efficacy of syringosubarachnoid shunting. Ellertsson and Greitz observed that pressure inside the syrinx was higher than that in the subarachnoid space at the same level. For ethical reasons, the existence of such a high-pressure phase inside the syrinx clearly cannot be proved, but the shrinkage of the syrinx observed after placement of a simple syringosubarachnoid shunt does offer strong support for this theory.

**Theory of Surgical Treatment**

Theoretically, in patients without hydrocephalus, operative decompression of the foramen magnum should be the treatment of choice. Although a considerably high risk of mortality and morbidity associated with direct surgery on the foramen magnum has been reported, no patient in this series deteriorated postoperatively. We would like to stress that the most important precaution for avoiding operative complications is to maintain the patient’s neck in the neutral position during surgery. Plugging the opening of the central canal (Gardner’s operation) is probably unnecessary.

**References**


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