Hydrodynamic Studies in Syringomyelia

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Although intramedullary cavitation of the spinal cord was recognized earlier, the term "syringomyelia" was first used by Oliver d'Angers in 1837 for cavities within the cord, regardless of etiology. Using the term in this manner, Schlesinger in 1902 noted the occurrence of intramedullary cavities in relation to trauma, inflammation, vascular disorders, tumors, malformations of the nervous system, pachymeningitis, and leptomeningitis, while in other cases he could identify no associated pathology. Holmes, Collier, and Schneider have studied the role of trauma in causing central cord cavitation. Tauber and Langworthy reported cavities associated with arterial insufficiency. McLaurin, et al., have experimentally produced cavitation secondary to adhesive arachnoiditis. Netsky has suggested arterial pathology as a basic cause of idiopathic syringomyelia, although, in general, the thickening and hyalinization of the blood vessels in the walls of syringes have been regarded as a secondary reaction, possibly due to hydrostatic pressure within the cyst.

There is a high incidence of syringomyelia in association with intraspinal neoplasms. Kernohan, et al., and Poser reported an incidence of 53.5% and 31% respectively. This association has been ascribed to necrosis within the tumor, obstruction of vital arterial and venous channels by the tumor, or transudation of fluid from such neoplasms as hemangioblastomas. Poser in his recent exhaustive review concluded that both the syrinx and tumor result from abnormal glial and mesodermal elements included in the cord as a result of faulty closure of the dorsal raphe of the neuraxis during embryonic development.

Syringomyelia is often present in cases of dysraphic and dysplastic phenomena within the central nervous system, such as meningocele, duplication of the spinal cord, and the Arnold-Chiari malformation, and in the body as a whole in cases of skeletal abnormality, asymmetry of the two sides of the body, genitourinary abnormality, and intestinal duplication. Accordingly, syringomyelia is often regarded as part of the syndrome of "status dysraphicus" of Bremer.

Mackay and Favilli considered abnormal glial proliferation followed by degeneration and cavitation to be the primary pathogenesis of spinal cord cavitation. Hassin suggested that the process was primarily one of degeneration, defective glial undergoing atrophy. Taylor, et al., concluded that disintegration of congenitally unstable glia at the union of the embryonic basal and alar plates of the primitive neuraxis explained the distribution of the cavities in the cord and brain stem.

Recognizing the high incidence of other malformations in association with syringomyelia, many investigators have questioned whether spinal cord cavitation is a part of the dysplastic process or whether it is due, partially or completely, to cerebrospinal fluid hydrodynamic changes imposed by the other malformations.

Taylor, et al., quoted Grund (1908) as being the first to consider cerebrospinal fluid pressure as an important factor in the formation of syringomyelia. Taylor, et al., (1922) suggested that pressure might be a contributing factor. They described the clinical course and pathological findings of two patients. One had a progressive course and evidence of hydrocephalus at postmortem examination. The second patient, 4½ years after the onset of his symptoms, subsequently had no further deterioration during 24 years of further observation. The patient committed suicide by hanging. Postmortem examination demonstrated a rupture of the syrinx into the subarachnoid space, presumably at
the time of the arrest of symptoms. The authors felt that the specimen showed evidences of previous hydrocephalus and previous increased pressure which were also presumably relieved at the time of the spontaneous rupture of the syrinx.

It has been established that, in 71% to 80% of normal subjects, the central canal of the spinal cord closes and becomes a vestigial structure represented by a core or nests of ependymal cells. Cameron\(^5\) regarded the majority of the spinal cord cavities occurring in dysraphic states as due to the delayed closure or persistent opening of the lower neuraxis (meningomyelocele). He believed that, during embryogeny, the cerebrospinal fluid ran down the persisting central canal and out the defect, thus dilating the canal (hydromyelia).

Syringomyelia has frequently been described in association with hydrocephalus due to blockage of the outlets of the fourth ventricle, with or without the Arnold-Chiari malformation.\(^1\) It has been postulated that the majority of the syrinxes found in this location are due to blockage of the outlets of the fourth ventricle, and that some cases are due to an underlying Arnold-Chiari malformation.\(^6\) In other reports, the cause of hydrocephalus has not been apparent.\(^3\)\(^6\)\(^4\)

Gardner\(^1\)\(^5\)\(^-\)\(^19\) considered the Arnold-Chiari malformation, diverticula and cysts of the outlets of the fourth ventricle, and the Dandy-Walker syndrome to be varying expressions of embryonal atresia of the fourth ventricle. He reported that such malformations were found in each of his 74 cases of syringomyelia in which the posterior fossa was explored.\(^19\) Some cases had hydrocephalus due to blockage of all outlets of the fourth ventricles, but others had normal ventricular volume and were felt to have only the midline foramen of Magendie embarrassed.\(^17\) Gardner hypothesized that blockage of this foramen prevented free egress into the subarachnoid space for the fluid pulse waves initiated by the pulsating choroid plexus. Thus, the pulsation was diverted down the central canal of the spinal cord, leading to cavitation of the cord. This produced hydromyelia if the cavity was contained by ependyma, or syringomyelia if it broke through the ependymal lining and dissected into surrounding nerve tissue. Gardner stated that this entity accounted for the majority of cavities within the spinal cord. He recommended the surgical treatment of opening the foramina of the fourth ventricle and blocking the connection between the fourth ventricle and the spinal cord cavity.

There is no unanimity of thought as to either the etiology or the best therapy for spontaneous cavitation within the spinal cord. Radiation therapy is used by those who regard the basic pathology to be glial proliferation.\(^13\)\(^\)\(^2\)\(^7\)\(^4\)\(^10\) Others drain the cyst directly, presumably regarding the syrinx to be isolated and under tension.\(^12\)\(^\)\(^27\)\(^29\)\(^36\)\(^50\)\(^53\) Both methods have met with some success.

In cases where the cavity is connected with the ventricles, Gardner’s method has merit. The purpose of our study was to find criteria by which to diagnose cases of this type preoperatively.

Surgical therapy to correct the proposed hydrodynamic problem had been undertaken in over half the cases. We have used the term “syringomyelia” to indicate a proven or presumed cavity in the spinal cord regardless of etiology, with the exception of an acute central cord syndrome secondary to trauma, which we have excluded. Proven cervical intramedullary tumors will be discussed separately.

**Material**

Between 1958 and 1962, 17 cases with a spontaneous progressive central syndrome of the cervical cord were seen on the neurosurgical service at the University of California at Los Angeles. During the years 1963 to 1966, five additional cases were seen at the Medical College of Virginia.

Of this total of 22 patients, two were initially diagnosed as having intramedullary cervical cord tumors which subsequently were proven to be Grade I astrocytomas, one cystic and one probably solid. Of the remaining 20 patients, it was possible to study 12 in the manner listed below (Table 1). Six of these underwent posterior fossa exploration and cervical laminectomy of C-1 through C-4 and were found to have cysts of the cervical cord in connection with the fourth ventricle. A seventh patient had had a cervical laminectomy 10 years previously; at that time, clear fluid was aspirated from a cyst in the cord but the posterior fossa was not visualized. The remaining five had similar radiological findings and appear to represent similar pathology. The
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TABLE 1
Summary of 12 cases of syringomyelia

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age, Sex</th>
<th>Duration Before Investigation</th>
<th>Operation</th>
<th>Follow-Up</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>15 M</td>
<td>progressive—3 years</td>
<td>none</td>
<td>9 yrs</td>
<td>progression</td>
</tr>
<tr>
<td>2*</td>
<td>20 M</td>
<td>progressive—7 years</td>
<td>posterior fossa cranietomy; cervical laminectomy</td>
<td>9 yrs</td>
<td>improvement</td>
</tr>
<tr>
<td>3</td>
<td>62 F</td>
<td>progressive—15 years</td>
<td>none</td>
<td>9 yrs</td>
<td>progression</td>
</tr>
<tr>
<td>4</td>
<td>14 M</td>
<td>progressive—7 years</td>
<td>none</td>
<td>9 yrs</td>
<td>progression</td>
</tr>
<tr>
<td>5*</td>
<td>34 F</td>
<td>progressive—7 years</td>
<td>posterior fossa cranietomy; cervical laminectomy</td>
<td>8 yrs</td>
<td>improvement</td>
</tr>
<tr>
<td>6*</td>
<td>28 M</td>
<td>progressive—16 years</td>
<td>posterior fossa cranietomy; cervical laminectomy</td>
<td>2 yrs†</td>
<td>improved</td>
</tr>
<tr>
<td>7*</td>
<td>19 M</td>
<td>progressive—10 years</td>
<td>posterior fossa cranietomy; cervical laminectomy</td>
<td>6 yrs</td>
<td>improved</td>
</tr>
<tr>
<td>8*</td>
<td>11 M</td>
<td>progressive—3 years</td>
<td>posterior fossa cranietomy; cervical laminectomy</td>
<td>6 yrs</td>
<td>improved</td>
</tr>
<tr>
<td>9</td>
<td>45 M</td>
<td>progressive—20 years</td>
<td>(laminectomy 10 yrs before low-pressure ventriculojugular shunt)</td>
<td>10 yrs</td>
<td>progression</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>3 mos</td>
<td></td>
<td>improved?</td>
</tr>
<tr>
<td>10</td>
<td>38 M</td>
<td>progressive—3 years</td>
<td>none</td>
<td>1½ yrs</td>
<td>progression</td>
</tr>
<tr>
<td>11</td>
<td>32 F</td>
<td>progressive—2 years</td>
<td>none</td>
<td>3 mos</td>
<td>no change</td>
</tr>
<tr>
<td>12*</td>
<td>30 M</td>
<td>progressive—11 years</td>
<td>posterior fossa cranietomy; cervical laminectomy</td>
<td>1 yr</td>
<td>improved</td>
</tr>
</tbody>
</table>

* Syrinx proven at surgery to be connected to the fourth ventricle.
† Patient’s symptoms progressed for 4 years after contrast studies. Then, he underwent surgery and has continued to improve to present, 2 years.

The former group will be termed “proven” (meaning that the cyst has been surgically demonstrated to be in connection with the fourth ventricle) and the latter group will be termed “suspected” (implying that a similar connection exists).

Suspected and Proven Cavities. 1. Sex and Age: The twelve cases of syringomyelia include nine men and three women. Their ages range from 11 to 62 (proven, 11 to 34; suspected, 15 to 62).

2. Symptoms: Symptoms had progressed from 2½ to 20 years (proven 2½ to 7 years; suspected 3 to 20 years). The first symptom was a vague difficulty in using the arms, soon followed by loss of pain sensation, trophic changes, motor weakness, and atrophy in the arms. Within 1 to 2 years severe neck and shoulder pain, often accompanied by torticollis, was an invariable and persistent symptom. Within months of the onset of this pain, symptoms of long tract involvement of the legs became evident. Sphincter disturbances accompanied pronounced bilateral long tract impairment.

3. Findings: Each of the cases had classical findings of a disassociated sensory loss to pain and temperature over the shoulders and arms. All had involvement of the descending tract of the fifth cranial nerve and all had varying degrees of long tract motor and sensory involvement. Case 9 had pronounced brain stem malfunction which in addition to bilateral impairment of the descending tract of the fifth cranial nerve included unilateral involvement of the sixth and seventh cranial nerves and bilateral involvement of the ninth, tenth, and elev-
enth cranial nerves, nystagmus, and intermittent difficulties with respiration.

**Proven Intramedullary Cervical Cord Tumors.** Two patients with proven intramedullary tumors of the cervical cord were also seen. Both were women, aged 28 and 30. The course of their disease was much more rapid (6 and 9 month); pain was not a feature, trophic changes were not prominent, and both had marked quadriplegia at the time of investigation.

**Method**

Each of the 12 cases of syringomyelia with proven or suspected fourth ventricle connection, plus both cases of proven intramedullary cervical cord tumors, had routine skull and cervical spine films with the lateral view of the cervical spine obtained at a target film distance of 72 inches.

Each patient had a lumbar puncture. The pressure, cells, and protein were determined. A blood pressure cuff was placed around the subject's neck and inflated to a pressure of 60 mm of mercury. A rise in pressure of at least 100 mm of CSF within 10 seconds and a fall to baseline levels within 10 seconds of relieving the pressure was considered evidence of no block. A complete block was defined as no rise in pressure.

Ten of the 12 patients had positive contrast myelography by the lumbar route with 9 to 16 cc of Pantopaque. In three of the cases, Pantopaque was directed into the fourth ventricle in an attempt to fill the syrinx. The fourth ventricle filled in two cases, and the syrinx was partially demonstrated in one case.

Each patient underwent air myelography by the lumbar route using the method of Young and Scott. The patients were placed in the sitting position and air was exchanged with cerebrospinal fluid in 5 cc increments via the lumbar subarachnoid space. By extreme flexion of the head, a large portion of the air was maintained in the cisterna magna and cervical canal. Attempts to introduce or exchange 10 to 20 cc of air and outline the cervical spinal cord were unsuccessful. The rapid exchange of 60 to 85 cc of air followed by stereolateral films of the cervical spine and sagittal laminograms through the area were successful in all cases.

In three of the cases, an additional increment of air was exchanged. In all cases, there was filling of the ventricular system, and pneumoencephalography was performed.

Six patients had bilateral posterior fossa exploration and laminectomy of C-1 through C-4. Four of these cases were explored by the author, each in the sitting position. A burl hole was placed in the right occipital region and 1 cc of indigo carmine injected into the right lateral ventricle. Appearance of the dye in the syrinx was noted. In one of the six cases, the cerebellar tonsils were resected. In the other five cases, the abnormal tonsillar mass was dissected free of the brain stem, and the fourth ventricular foramina were opened widely. A piece of siliconized rubber tubing, approximately 2 mm in diameter and 5 cm in length, was then placed with one end in the fourth ventricle and the other in the cervical subarachnoid space. The tubing was held with fine silk sutures attached to the thickened arachnoid of the cervical cord and the cerebellar tonsils. In each case, a malleable silver probe could be passed from the fourth ventricle into the syrinx. A piece of muscle was used to obliterate this opening.

One patient, not suitable for posterior fossa exploration, underwent a ventriculojugular shunt with a low-pressure valve.

**Results**

**Cervical Spine X-rays.** Figure 1 indicates the sagittal diameter of the cervical canal in 11 of the 12 cases of syringomyelia. Case 9 had a previous laminectomy. The shaded area represents the values recorded by Wolf, et al., for the normal sagittal diameter of the cervical canal in 200 subjects equally proportioned as to sex, at a target film distance of 72 inches. Included for comparison are the two cases of intramedullary cervical cord tumor encountered during this period of study. In the cases of proven and suspected syringomyelia, there was a marked irregularity of the measurements of the spinal canal, usually not paralleling the normal values. The lower segments were large, none below average values.

This enlargement is even more pronounced if Cases 3, 8, and 11 are deleted. Cases 3 and 8 were subjects of less than average stature, and Case 11 was a patient with mild symptoms and a short history. Case 3, a 62-year-
old woman with a 15-year history of syringomyelia, was 5 ft 4 inches tall and weighed 72 pounds. Case 8, an 11-year-old boy with a 3-year history, was 4 ft 9 inches tall and weighed 65 pounds. Case 11, a 32-year-old woman of average height and weight, had minimal signs of syringomyelia consisting of a disassociated sensory loss involving the left face, left shoulder, and arm with mild lower motor neuron weakness in the left forearm and hand, and a left Babinski’s sign. She also had the shortest history, 2 years.

Bulbous enlargement of the sagittal diameter of the lower cervical segments was seen in Cases 6, 7, and 12, all patients whose symptoms began before the age of 19 and had been present for at least 10 years at the time of investigation. These cases also had scalloping of the posterior margins of the lower cervical vertebrae. Chronicity of the disease when it begins at an early age is associated with erosion and/or expansion of the lower cervical segments. When symptoms begin at a more advanced age, expansion does not occur. In Case 3, a woman whose symptoms began at age 47, despite a 15-year history of progressive and severe symptomatology there was no bulbous enlargement and no scalloping of the posterior margins of her vertebrae. All instances of enlargement of the lower cervical canal were found in men.

In the majority of the cases, regardless of age or chronicity, the upper cervical segments were usually not a great deal larger than the lower segments, sometimes no larger. This was most striking in Cases 8 and 11 in which values for the sagittal diameter were the same throughout the cervical canal.

The sagittal diameter and configuration of the spinal canal of the cases with spinal cord tumor were within normal limits.
Lumbar puncture pressure ranged from 80 to 220 mm of CSF; it was below 140 mm of CSF in one patient and above 150 mm of CSF in one patient. Cerebrospinal fluid protein ranged from 20 to 54, and there was no increase in cells. Bilateral jugular compression, as described above, revealed no block in any cases of syringomyelia. A complete block was present in both cases of intramedullary tumor.

Positive contrast myelography showed a definitely enlarged cord (using the criteria of Taveras and Wood) in nine of the 10 cases in which it was performed. A normal size was seen in Case 11, the patient with the shortest history (2 years) and minimal signs.

Air myelography in the sitting position in each case revealed a narrow cord in sagittal diameter, varying in width from 2.5 to 6 mm at its narrowest point (film target distance of 40 inches). Case 11, the case with the normal Pantopaque myelogram, also had a narrow cervical cord visualized by air myelography. The diameter of the cord was 5 mm. In three cases, the upper portion of the cervical cord was narrow, and the lower portion remained enlarged. Subsequent exchange of additional air and cerebrospinal fluid caused the lower portion of the cord also to become narrow.

Illustrative Myelograms. Figure 2 is the positive contrast and air myelogram of Case 1, an 15-year-old boy with a 3-year progressive history consistent with an intramedullary cavity of the cervical cord. Positive contrast myelography with 9 cc of Pantopaque revealed an enlarged cervical cord. Subsequent air myelography in the sitting position (65 cc of air were exchanged with 55 cc of fluid in 5 cc increments) showed an ill-defined abnormality of the posterior fossa and an irregular narrow cord measuring 6 mm in diameter at its narrowest portion.

Figure 3 is a positive and air contrast myelogram of Case 3, a 62-year-old woman with a 15-year history consistent with syringomyelia; 85 cc of air were exchanged with 75 cc of cerebrospinal fluid in 5 cc increments. The positive contrast myelogram revealed an enlarged cervical cord, and the air myelogram revealed an extremely narrow spinal cord measuring 2.5 mm in diameter. The fourth ventricle was normal in position, but there was abnormality of the cerebellar tonsils.

Figure 4 is the air contrast myelogram of Case 8, an 11-year-old boy with a 3-year history of a central cord lesion; 60 cc of air were exchanged with 50 cc of fluid in 5 cc
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The cord shadow measures 4.5 mm in diameter. A previous oil myelogram revealed an enlarged cervical cord, and subsequent posterior fossa exploration and cervical laminectomy of C-1 through C-4 showed a syrinx in connection with the fourth ventricle.

Figure 5 is the air myelogram of Case 6, a 28-year-old man with a 16-year history of a progressive intrinsic lesion of the cervical spinal cord, which proved to be a syrinx in connection with the fourth ventricle. Positive contrast myelography revealed a bulbous cord. Air myelography with the exchange of 60 cc of air with 50 cc of cerebrospinal fluid revealed the upper portion of the cord to be narrow, but the lower portion, broad. A wait of 30 minutes did not change the size of the cord shadow. Exchange of an additional 40 cc of air with 40 cc of fluid, however, caused further narrowing of the lower cervical cord.

FIG. 3. Positive (top) and air contrast (bottom) myelograms of Case 3. The portion of the cord is shaded in the diagram.

Fig. 4. Air contrast myelogram of Case 8.

Fig. 5. Air myelogram of Case 6.

TABLE 2

<table>
<thead>
<tr>
<th>Vertebral Level</th>
<th>Cord Diameter (mm) after Air Exchange</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>60 cc</td>
</tr>
<tr>
<td>C-1</td>
<td>4.5</td>
</tr>
<tr>
<td>C-3</td>
<td>7</td>
</tr>
<tr>
<td>C-5</td>
<td>11</td>
</tr>
<tr>
<td>C-6</td>
<td>18</td>
</tr>
</tbody>
</table>
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FIG. 6. Air contrast myelogram, a proven Grade 1 cystic astrocytoma of the spinal cord, Case 1.

FIG. 5. Positive contrast myelogram of Case 6 after the exchange of 60 cc of air (above) and an additional 40 cc of air (below).

shadow. Table 2 lists the diameter of the cervical cord shadow at multiple levels of the cervical canal, after the first and second exchange.

Both cases of proven intrinsic cervical cord tumors seen during the same period of study underwent air myelography in an identical manner.

Figure 6 is the air myelogram of Case A, a 30-year-old woman with a proven cystic Grade 1 astrocytoma of the cervical cord containing an isolated cyst of high protein content. Exchange of 50 cc of air with 40 cc of fluid, followed by the exchange of an additional 40 cc of air with 40 cc of fluid, revealed the cord to be bulbous. Findings in the case of solid astrocytoma were similar.

Surgical Therapy. Six of the patients underwent bilateral craniectomy of the posterior fossa and cervical laminectomy of C1-C4. In each case, a malformation of the tonsils and roof of the fourth ventricle and a syrinx of the cervical cord in connection with the fourth ventricle were found. Four cases were explored (by the author) in the sitting position. A burr hole was placed in the right occipital region and 1 cc of indigo carmine was injected into the right lateral ventricle. In each case, the dye could be seen to discolor the fluid within the thin-walled syrinx in a matter of minutes. In the same four cases, the cord remained bulbous until the cisterna magna was opened and cerebrospinal fluid allowed to drain. Within 30 to 60 seconds of draining the cisterna magna, the spinal cord collapsed and became ribbon-like, held laterally by the dentate ligaments.

Table 3 lists the protein values of the fluid of the syrinx in each of the three cases in which it was obtained. The protein values of the ventricular and lumbar subarachnoid fluid are also recorded. The protein values in the case of cystic astrocytoma are also listed for comparison.

Table 1 lists the 12 cases in this study. Those undergoing posterior fossa exploration, cervical laminectomy, opening of the foramina of the posterior fossa with placement of a tubular shunt and blockage of the
TABLE 3

Protein content

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>CSF Fluid Content (mg%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Ventricle</td>
</tr>
<tr>
<td>Syringomyelia (proven)</td>
<td></td>
</tr>
<tr>
<td>Case 5</td>
<td>10</td>
</tr>
<tr>
<td>Case 8</td>
<td>6</td>
</tr>
<tr>
<td>Case 12</td>
<td>12</td>
</tr>
<tr>
<td>Cystic Astrocytoma (proven)</td>
<td>—</td>
</tr>
</tbody>
</table>

inlets to the syrinx, as previously described, are noted (Cases 2, 5, 6, 7, 8, and 12). Surgery was performed from 1 to 9 years ago. Information regarding the progress of these patients was recently reviewed. After initial recuperation from surgery, all patients showed progressive improvement from their preoperative status for at least 2 years, or are still improving. In no case has there been subsequent regression. In all but one case, the neurological improvement has represented a functional improvement in the patient’s daily living. The exception is Case 5 who had marked weakness of both arms and spastic paralysis of both legs with incontinence at the time of surgery. She remains incontinent and confined to bed and wheelchair. She no longer has neck pain and has had some improvement in the strength of her right arm and both legs and has lost the marked spasticity of her legs. Of the remaining five patients, three were moderately incapacitated and two mildly incapacitated preoperatively. None has more than minimal incapacity at this time; one is going to school, and the other four are employed. Case 6 refused surgery for 4 years during which time he showed significant progression. He has shown marked improvement in the past 2 years since surgery.

Case 9, a 45-year-old man with a 21-year history of syringomyelia and progressive involvement of the bulb for 3 years prior to investigation, was thought to be too poor a risk for posterior fossa exploration. A previous laminectomy had revealed that the cyst contained clear fluid resembling cerebrospinal fluid. Positive contrast and air contrast myelography showed a bulbous cord on the former study and a narrow cord on the latter. Pneumoencephalography revealed mild dilatation of the ventricles. The cerebrospinal fluid pressure was 220 mm of CSF. Under general anesthesia, a routine ventriculojugular shunt was performed using a low pressure Holter valve, the opening pressure measured at 20 mm of water. The patient’s postoperative lumbar puncture pressure at the time of surgery and again on the seventh postoperative day was 80 mm of cerebrospinal fluid, and there was attenuation of the spinal fluid pulse wave. Now, 3 months after surgery, there has been some improvement in his condition. He has less neck pain and torticollis, less spasticity, and more strength in the right hand, and he no longer uses his accessory muscles of respiration at bedrest. Preoperatively, he could count to 4 in a single breath. He is now able to count to 12. Pulmonary function of blood gas studies remain markedly impaired.

Four of the five patients who have not had surgery have shown significant progression of their disease. Case 11, early in her course, has shown no change to date, under 3 months of observation. Surgery is planned if progression occurs.

Discussion

Values for the sagittal diameter of the cervical canal have been reported by Boijsen and Wolf, et al. The latter group evaluated the sagittal diameter of the normal cervical canal of 200 subjects, approximately equally apportioned as to sex with a standard target film distance of 72 inches. These values are produced in the shaded area of Figure 1. Widening of the sagittal diameter of the cervical canal in syringomyelia has been reported by Boijsen. He described three
cases, each of which had a diameter that was either at the upper limits of normal or enlarged. Wells, et al.,31 studied 32 cases of syringomyelia and found that the majority of those patients whose symptoms began before the age of 30 had enlarged sagittal diameters of the cervical canal, whereas this phenomenon was rare in those cases whose symptoms began later.

Only two of the cases in our series had the onset of symptoms after the age of 30. Cases 3 and 10. In Case 3, a 62-year-old woman, despite a 15-year history the canal size was well within normal limits, although rather large for the patient’s general body physique. Case 10, a 38-year-old man, had symptoms that began at age 35, and there is a suggestion of slight early expansion of the lower segments. All other cases in our series had symptoms that began between the ages of 7 and 30. In this group there is an approximate correlation between the degree of expansion of the lower sagittal canal, as compared with the upper cervical segments, and two factors: an early age of onset, and a long history of symptoms. Pronounced enlargement and erosion of the lower cervical canal was found in cases whose symptoms began before the age of 19 and were at least 10 years in duration at the time of study. There was no correlation between the degree of canal expansion and the severity of symptoms or signs, including neck pain, degree of sensory loss, or degree of lower motor neuron weakness in the arms.

Both cases of intrinsic tumor had no enlargement of the sagittal diameter of the cervical canal, probably reflecting the age of the patients and the short history. Intradural cervical tumors in childhood, even with a relatively short history, are reported to produce frequent enlargement of the cervical canal.41

Definition of the pedicles in the anteroposterior views of the cervical spine was not clear in most cases, and was particularly difficult in those cases with torticollis. This has been the experience of others.1,34 The cervical interpedicular distance, therefore, could not be accurately measured.

The normal sagittal diameter of the cervical cord in air myelography has been evaluated in 20 subjects by Klefenberg and Saltzman.30 They used the method of Lingren,32 introducing air by the cisternal route with the patient in the lateral Trendelenberg position. Plain films and laminograms taken at a target film distance of 40 inches revealed direct mensuration of the cervical cord to be between 8 and 12 mm. We have measured the presumably normal cervical cord at the time of pneumoencephalography on 10 subjects in the sitting position at a target film distance of 40 inches, and have found values to vary between 9 and 12 mm. Klefenberg and Saltzman30 have evaluated the size of the cervical canal in 28 cases of syringomyelia using air myelography in the lateral Trendelenberg position and injecting air via the cisternal route. They found the cord to be characteristically irregular, usually wider than normal in diameter; occasionally it was narrower than normal.

Greenwald, et al., (1958)33 were the first to record that during pneumoencephalography in the sitting position the shadow of the spinal cord is narrow in syringomyelia. Gardner, who was one of the authors of that paper, in 196519 again referred to the same phenomenon and suggested it indicated a syrinx in connection with the fourth ventricle. He proposed that fluid leaks through the thin wall of the syrinx, which leads to collapse of the cavity and the narrow image on x-ray. If this were to occur it would probably happen regardless of whether the syrinx was in connection with the fourth ventricle. Indeed, a cavity so connected would be likely to refill from ventricular fluid.

In 1966 Heinz33 again described this phenomenon and attributed it to fluid seeking its own level in the cyst, but again felt it related to a connection of the cyst to the fourth ventricle, “hydromyelia.” In 1961 we9 suggested that this collapse was due to active aspiration of the cyst during air exchange. Robertson42 has shown that during pneumoencephalography air enters the ventricles at the expense of the subarachnoid fluid over the hemispheres, and the lateral ventricles are actually inflated during air study. In none of our air myelograms was the air and fluid completely trapped by the head posture. There was gradual loss of air into the ventricular system. We suggest that, in each of the cases
Hydrodynamic Studies in Syringomyelia

studied, there was a cyst in connection with the ventricles via the fourth ventricle and associated with a partial block of the outlets of the fourth ventricle. This has been confirmed in six cases by surgical exploration.

It seems likely that, during the exchange of air and fluid in the sitting position, fluid is withdrawn from over the hemispheres and the ventricles expand. The ventricular expansion allows air to enter the ventricular system. Due to the partial blockage of the outlets of the fourth ventricle, however, and the acute flexion of the head, air is not readily available and fluid is also aspirated from the syrinx, causing it to collapse (Fig. 7). The drainage of fluid from over the hemispheres is more effective with the patient in the sitting posture. With the subject upright, the effect of gravity on the intracranial spinal fluid and blood creates negative pressure in the cranial cavity and ventricles. This assists the effect of ventricular expansion in draining the cyst. Air fluid exchange in the Trendelenberg position would presumably neither remove fluid from the surface of the cerebral hemispheres nor produce a negative pressure in the lateral ventricles and, therefore, would not internally decompress the cyst.

FIG. 7. Proposed mechanism of collapse of the cervical syrinx during air myelography. A. Diagram of a subject undergoing air (circles) and fluid (arrows) exchanged via the lumbar route. Fluid appears at the lumbar puncture needle at the expense of fluid over the cerebral hemispheres. This causes expansion of the cerebral hemispheres, dilatation of, and negative pressure in the lateral ventricles. Air, having risen to the cisterna magna, enters the ventricular system with difficulty due to the partial obstruction of the outlets of the fourth ventricle and the flexed head position. B. Accordingly, fluid is aspirated from the syrinx causing it to collapse, particularly in its upper portion.
Thus, the air studies of Klefenberg and Saltzman\textsuperscript{9} did not lead to collapse of the cavities. If the mechanism of collapse were one of seepage of fluid through the cyst wall, as suggested by Gardner,\textsuperscript{19} the horizontal cisternal air myelogram of Klefenberg and Saltzman\textsuperscript{10} would have also led to collapse of the syrinx, regardless of its connection with the fourth ventricle.

The three cases reported in which serial exchanges of air caused progressive collapse of first the upper and then the lower portion of the cord while the passage of time, without exchange, had no effect can be explained by the additional exchange of air and fluid leading to further ventricular expansion and cyst aspiration. The first exchange only partially evacuated the cavity, and the lower portion of the cavity remained expanded, with the fluid seeking its own level.

Opening the cisterna magna at the time of surgery had a similar effect on the syrinx. If the integrity of the subarachnoid space was maintained, the cervical cyst was under tension. Opening the cisterna magna and allowing cerebrospinal fluid to drain (presumably from over the hemispheres, particularly as the fourth ventricular outlets are partially blocked) caused the syrinx to gradually collapse over 30 to 60 seconds.

The six cases of proven connecting cavities represent almost one-third of the cases seen by the author in an 8-year period (6 of 20 cases). If the validity of the collapsing cord sign for fourth ventricular connection is accepted, then this syndrome accounts for more than half of the cases seen during this period (12 of 20). As the pathology of the remaining eight cases remains undetermined in seven,* and as only two cases of proven intrinsic cervical cord neoplasms were found during the same period, we suggest that cysts connected with the fourth ventricle are a common cause of progressive cervical central cord disease.

Six patients have had surgical exploration with blockage of the inlet of the syrinx and opening of the outlets of the fourth ventricle. All have shown progressive improvement over their preoperative levels and maintained this improvement to the present time.

We believe that, if positive contrast myelography shows a normal or bulbous cord and air myelography shows a narrow cord in sagittal diameter, a cyst connected with the fourth ventricle is likely. In one case, as reported, a cord that appeared normal by Pantopaque myelography was found to be narrow on air myelography. We believe, therefore, that the latter procedure is a more sensitive test for the presence of a syrinx.

All of the cases had evidence of a posterior fossa malformation similar to that described by Chiari. None of these cases had the most severe type of malformation of Chiari, frequently referred to as the Arnold-Chiari malformation. Six of the 12 air studies, including three of the cases explored, revealed clear-cut evidence of a posterior fossa malformation. We have found that the collapsing cord sign, therefore, not only gives a functional test for continuity between the cervical syrinx and the fourth ventricle, but that it may also be present when pneumoencephalography fails to identify malformations of the posterior fossa.

In one case in which severe bulbar signs prevented safe posterior fossa exploration, a low pressure ventriculo-jugular shunt was performed. Lumbar puncture pressure and pulse wave were found to be lowered by this procedure. The patient has shown some improvement during 3 months of postoperative observation.

In cases in which air study has revealed a cyst connected with the fourth ventricle, the surgical approaches described above seem more rational than direct surgical opening of the cyst or radiation therapy.

**Summary and Conclusions**

A cystic dilatation of the cervical cord is commonly connected with the fourth ventricle and associated with partial or complete closure of the outlets of the fourth ventricle. In view of the frequency with which syringomyelia has been reported in association with hydrocephalus or posterior fossa malformations, it is likely that this

* * The eighth case did undergo subsequent exploration and was found to have a syrinx in connection with the fourth ventricle. The patient is excluded from the study as he did not have preoperative positive and air contrast myelography. He has had dramatic improvement in his neurological deficit since surgery.
connection has frequently been overlooked. Symptoms of such a cavity can appear in childhood or during adult life. They usually progress over a matter of years, and severe neck and shoulder pain becomes a common complaint. Usually there is no evidence of a spinal fluid manometric block, and the pressure and protein are normal to slightly elevated.

In cases starting at an early age and of long duration, cervical spine films often show dilatation of the lower portion of the cervical canal with erosion and scalloping of the posterior margins of the lower cervical vertebrae. Pantopaque myelography in the prone position usually shows a large cervical cord. Air myelography in the sitting position revealed collapse of this enlarged cord. We have suggested a hydrodynamic cause for this collapse supported by surgical observations.

We recommend Gardner's procedure of opening the outlets of the fourth ventricle and blocking the opening between the fourth ventricle and the syrinx. In advanced cases, the use of a low-pressure ventriculo-jugular shunt as a permanent or temporizing procedure is worthy of consideration.

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References


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