Syringomyelia: A Look at Surgical Therapy

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Syringomyelia and syringobulbia are characterized by an accumulation of fluid in a cyst within the substance of the spinal cord or medulla or both. These cysts arise from various pathological processes and have been treated by surgical incision and drainage and by irradiation. Different materials have been inserted between the syrinx and the subarachnoid space to provide permanent drainage.

In 1953, while attending a meeting of the Society of Neurological Surgeons in New Orleans, one of us heard Dr. Dean Echols explain his technique of using a tantalum wire to maintain a fistula between a syrinx of the spinal cord and the spinal subarachnoid space and saw his patients who had had this operative procedure. The results were impressive, and we have employed this technique at our clinic since that time.

In an effort to determine whether results have improved since the introduction of this technique, we have reviewed the records of patients with syringomyelia or syringobulbia and syringomyelia but without associated neoplasm who were operated on at the Mayo Clinic between 1948 and 1962.

Evaluation

Forty patients underwent 44 operative procedures for syringomyelia or syringomyelia and syringobulbia. These patients do not represent all those with syringomyelia who were seen at the Mayo Clinic, as only surgically verified cases were studied. We have analyzed the subjective complaints and objective neurological findings both before and after operation and throughout the follow-up period. In addition we have reviewed diagnostic studies, operative findings, and surgical methods employed. All patients considered as having adequate follow-up studies had neurological re-examinations at least 1 year after operation; the longest follow-up was 10 years. The majority of our patients had complete follow-up for 2 years or more.

Thirty-five patients with syringomyelia did not have associated anomalies and were without evidence of other pathologic processes related to the central nervous system. Three of these, however, had minimal platybasia, but this did not contribute to the patients’ symptoms. The remaining 5 patients had syringomyelia or syringobulbia and syringomyelia associated with Arnold-Chiari malformations. These 5 cases will be discussed separately.

Symptoms

The common complaint in syringomyelia is one of an asymmetrical diminution of sensation plus muscular weakness at and below the level of the cyst. Because of the preference of the syringomyelic process for the cervical and cervicothoracic region, diminution of sensation in the hands and forearms is a common early finding. Nineteen of our 40 patients said that radicular pain, corresponding to the level of the syrinx, had been an early symptom. The pain, as described, was indistinguishable from radicular pain produced by other space-occupying intraspinal masses. The symptom-complex most typical of syringomyelia includes bilateral diminution of temperature perception and asymmetrical hypalgia. Proprioception and vibratory sensation are generally reduced to a moderate degree, whereas diminution of touch and pressure may be minimal. Characteristically, the motor disturbances in syringomyelia are those of weakness at or below the upper level of disturbed sensation. The weakness at the upper level of sensory disturbance is generally associated with atrophy and hyporeflexia indicative of denervation, whereas in the lower extremities there are hyperreflexia, pathologic reflexes, and spasticity indicative of corticospinal-tract involvement. The major motor disturbances distally are generally opposite the side of major sensory disturbance.

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Sex and Age

In syringomyelia without complications, males tend to be affected more frequently than females (23 males to 12 females). Of the 5 patients with associated Arnold-Chiari malformation, 3 were males and 2 were females. Twenty-one of the 40 patients were between 20 and 40 years of age. The ages of patients at the time of surgery ranged from 12 to 60 years. A few patients did not have onset of symptoms prior to the age of 40 years. One of the 5 patients with syringomyelia-syringobulbia and associated Arnold-Chiari malformation had a Klippel-Feil anomaly with fusion of the C-1 and C-2 vertebrae as well. Of the other 35 patients, 30 patients had cervical syrinxes, 4 patients had thoracic syrinxes, and 1 patient had a cervical and thoracic syrinx.

Diagnosis

In all 35 patients with uncomplicated syringomyelia, the history and neurological examination suggested the diagnosis. In this group, 27 lumbar punctures were performed. Jugular compression done at the time of the lumbar puncture revealed no evidence of subarachnoid-space block in 19 cases. There was evidence suggestive of partial block in 5 and of complete block in 3 cases. Seventeen contrast-medium (Pantopaque) myelograms and 1 air myelogram were done in this group of patients. Of these 12 demonstrated widening of the spinal cord at the level of the syrinx, and 2 showed no abnormal changes. Three of the contrast-medium myelograms and the 1 air myelogram revealed complete block at the level of the spinal-cord cyst. In the majority of patients with syringomyelia or syringobulbia and syringomyelia, the level of the lesion can be determined by neurological methods, and thus contrast myelography is not necessary.

The value for cerebrospinal-fluid protein was elevated above the normal of 45 mg./100 ml. in 18 of the 27 patients in whom it was studied. Four patients had values of 750 mg. or greater; 2 of these had thoracic cysts and 2 had cervical syrinxes. Values for protein content in fluid removed from syringomyelic cavities ranged from 10 to 180 mg. /100 ml. in 10 recorded specimens. The 1 specimen of 180 mg. was taken from a patient who had a cerebrospinal-fluid protein value of 900 mg. In the remaining 9 patients, the protein values of the fluid from the cysts was at least 20 mg. below the values of the cerebrospinal-fluid protein. The range of values of syrinx-fluid protein in these 9 patients was 10 to 40 mg./100 ml.

Surgical Methods

The surgical procedures were performed by several neurological surgeons, providing various approaches for comparison. The procedures included the following: laminectomy alone, laminectomy with simple evacuation of the syrinx, laminectomy with syringostomy for drainage by the use of various materials to maintain cyst and subarachnoid communication (rubber drain, polyethylene tubing, cotton wicks, silk sutures, and tantalum-wire sutures), and suboccipital craniectomies with cervical laminectomies alone and with both simple drainage of cysts and syringostomies with permanent-drain materials. In all but 15 of the 44 operative procedures, some form of permanent drainage of the syrinx was attempted. Two of these operations were reoperations in which the spinal cyst was found to be collapsed. The initial procedure in 1 patient had been simple drainage of a thoracic cyst; and the second patient had had a tantalum-wire syringostomy for a cervical syrinx previously.

Results of Surgical Treatment

Results of surgical therapy were classified as excellent, good, or poor. Results were considered excellent when detailed examination revealed definite objective improvement in neurological status and when subjective improvement was noted by the patients themselves. Results were considered good when the neurological deficit did not increase or decrease and no subjective increase in deficit was noted by the patients themselves. However, most patients of this second group believed that they had definitely been improved by operation. Results were classified as poor if the neurological deficit increased after surgery even though the patients themselves may have considered their condition improved.

Of the 35 patients with syringomyelia alone, 5 were lost to follow-up. The neurological status of these 5 patients 7 to 10 days after operation was essentially unchanged