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DEPARTMENT OF PATHOLOGY
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A lad of 5 was admitted to University Hospital because of progressive weakness of his legs and right arm.

The prenatal course of his mother was uneventful. Delivery was by difficult forceps. Birth weight was 5 pounds 10 ounces. Development appeared normal until age 4 when his mother noted that he began to walk stiffly, abduct his right leg and to throw his foot outward while walking. The child complained of stiff neck. He was unsteady on his feet and fell down frequently.

Past history revealed admission to another hospital for tonsillectomy; however, this was not performed because a displaced cervical vertebra was discovered. He had frequent bouts of vomiting since birth. However, x-ray examination of the GI tract was normal. He had pneumonia & tracheotomy at age 2. The child wore a cervical cast and collar for awhile and the same year underwent a cervical laminectomy with fusion. At age 4 he began to show the progressive stiffness of his legs and weakness of his right arm. For about 5 to 6 weeks prior to the last admission, he had been unable to walk and for 3 weeks prior to admission he complained of headache and vertigo.

Physical examination showed a chronically ill, irritable, alert white male of stated age. He lay on his right side. The following neurological abnormalities were noted: Decreased tone in the right arm; the right wrist held extended with elbow flexed; very limited use of right arm; increase in muscular tone in both legs which were held in extension. There was inability to dorsiflex in right foot. Deep tendon reflexes were absent in the right arm; hyperactive in both legs. The plantar response was extensor. There was no nystagmus or gross ataxia. The gait was markedly spastic, and the child was unable to walk without support.

Cervical myelogram with Pantopaque was performed 4 days after admission and revealed complete obstruction of the subarachnoid space at the level of the 10th thoracic vertebra. Lumbar puncture revealed slightly xanthochromic fluid with an abnormal Queckenstedt sign. Pantopaque was next injected into the cisterna magna where it was retained.

2 weeks later a laminectomy from 3rd cervical through first thoracic vertebrae was performed. There was an intrinsic swelling in the cervical spinal cord which was very hard and firm. The wound was closed without biopsy.

2 weeks later laminectomy was performed from the 8th through the 10th thoracic vertebrae. After the dura was opened, a large, soft, swollen spinal cord was noted. A needle was introduced into the cord and 20 cc. of xanthochromic fluid was withdrawn. Incision was next made over the dorsal portion of the cord in the midline and a cystic space was exposed A tiny fragment of the cyst wall was removed. A pathologic diagnosis of syringomyelia was made. Two silk setons were left in the cystic cavity and were embedded in the perispinal muscles.

Following operation, spasticity remained. His right arm now became completely paralyzed and flaccid. He was discharged on post-operative day 23 but was readmitted 1.5 months later because of difficulty in breathing. The interval since discharge had been characterized by progressive paralysis, so that now he was quadriplegic and in respiratory distress, with recurrent bouts of cyanosis, apnea and loss of consciousness. Spastic paralysis continued in the lower extremities with ankle clonus and extensor plantar response. The upper extremities were flaccid. The patient died quietly a few days later.