ARNOLD-CHIARI DEFORMITY WITHOUT
BONY ANOMALIES

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SINCE the early and original contributions of Arnold and Chiari on the
malformation that now bears their names, numerous articles have ap-
peared pertaining to the mechanism, diagnosis and treatment of this
anomaly. The occurrence of this deformity has usually been found in infants
to be associated with extreme degrees of spina bifida and cranium bifidum.
When encountered in adults this pathologic entity has generally been seen
in conjunction with some type of bony anomaly of the cranio-vertebral
junction, such as basilar impression (platybasia) or the Klippel-Feil deform-
ity.

A survey of the literature, however, reveals several cases in which this
malformation has been proven, either at operation or autopsy, to exist with-
out spina bifida or coexisting bony anomalies of the spine. The case reported
here illustrates the fact that this anomaly may exist in the absence of either
bony deformities or spina bifida, and furthermore that it may occur in the
very young, our patient being the youngest of those recorded. It is because
of this fact and in view of the unusual progress and diagnostic difficulties
presented by this case that we set forth our experiences in dealing with this
problem of the Arnold-Chiari malformation without demonstrable bony
anomaly.

CASE REPORT

History. A 5-year-old white female was seen in consultation on April 28, 1947
She was the first born of the family. Her birth and developmental history were
entirely normal, and she had had none of the usual childhood diseases. In November
1946, the mother first became aware of the fact that the child was less active than
usual, that she appeared listless and preferred not to run and play as she had for-
merly. She offered no specific complaints other than that her head ached and that
exercise caused pain in her neck. By December 1946 such general lassitude and
weakness of the lower extremities had developed that her schooling was discontinued.
It was then that the mother noted a change in the child’s gait, described as a tend-
dency to drag her feet when walking. Following tonsillectomy in February 1947 there
was noted for the first time a weakness in the upper extremities similar to that seen
in the lower. In the latter part of March, 1947 the child had to be confined to bed
because of profound weakness of all extremities. She was unable to elevate her arms
above the shoulder level. She could not hold her head erect nor was she able to hold
her body erect upon attempting to stand upright. Two weeks before the initial
examination the child had, on two occasions, regurgitated liquids through her nose.
At about this same time there developed huskiness of her voice. This illness had not
been attended by fever; she had not complained of any specific pain and had not
experienced any cranial nerve disturbances other than the dysphagia.

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**Neurological Examination.** The appearance of this child was similar to that of an adult in an advanced stage of myasthenia gravis. She was carried into the examining room by her father, and when placed in the erect position she immediately sagged against the support of his arms, the knees flexed and were adducted, the arms hung limply by her side, and the chin rested against the chest. When she attempted to look at the examiner she did so by elevating her eyes and lids but not her head. She breathed through her mouth with her tongue partially protruded, and there was a salivary drooling. Her breathing was rapid and labored and was performed entirely by the intercostal muscles. When held in an upright position she was able to offer some support of herself on her toes and heels. The handgrips and finger movements were likewise weak. Individual muscle tests revealed that the most marked handicaps were apparently confined to the group supplied by the upper and middle cervical cord. There were no fibrillary twitchings noted and the only demonstrable muscle atrophy was confined to the intrinsic muscles of the left hand. This produced a modified ulnar type of deformity. There was no involuntary mucal rigidity, but she complained of pain when her head was moved.

Examination of the vertebral column revealed no palpable deformities of the cervical spine, and there were no findings to suggest a spina bifida at a lower level. The cranial nerve study was not noteworthy aside from obvious palate and tongue weakness which was not attended by atrophy or fibrillary twitching of the tongue. The sensory examination was entirely normal for all modalities. There were no abnormal cerebellar objective evidences. Reflex studies elicited brisk triceps responses bilaterally, but the biceps reflexes and the abdominal reflexes were absent. There was a marked exaggeration of the knee and ankle reflexes. Transient ankle clonus was present and the Babinski reflex was bilaterally positive.

**Course in Hospital.** The child was immediately hospitalized with a presumptive diagnosis of an infiltrating tumor of the upper cervical spinal cord and medulla oblongata. Roentgenograms of the spine revealed no evidence of a congenital anomaly. There were no suggestions of a widening of the interpedicular measurements, laminal erosions or traumatic damages; nor did X-rays of the skull depict abnormalities. A lumbar puncture was attempted and although the operator was certain the needle was within the canal, clear fluid was obtained only by resorting to abdominal compression, and the CSF pressure was recorded at 30 mm. of water. Fluid sufficient for a protein determination was finally removed and 1 cc. of lipiodol was then introduced. The protein was reported as 63 mg. per cent, the cell count 0. An attempt was made to fluoroscope the movement of the lipiodol but it was found to be immobile, apparently having been introduced into the subdural space. A 2nd attempt was made to determine the CSF dynamics, with similar results except that on the 2nd occasion jugular compression did increase the flow of the spinal fluid measurably.

At this point a neurological consultant confirmed the previous findings and corroborated the probable diagnosis of glioma of the brain stem and cervical cord. A trial of roentgen therapy was agreed upon, inasmuch as there had been a distinct improvement clinically. Even before changes secondary to the X-ray therapy could be expected, the child became more active, was able to engage in minor activities on the ward and no longer exhibited the positive Babinski reflexes. At the completion of the course of roentgen ray therapy she was ambulant with increasing power, no longer exhibited difficulty in swallowing or breathing, and could elevate her arms above her head. She was discharged from the hospital to be observed at frequent intervals.