

ARNOLD-CHIARI DEFORMITY IN AN ADULT
WITHOUT OBVIOUS CAUSE

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The term "Arnold-Chiari Deformity" is generally used to designate a
specific modification in the configuration and location of the hind-
brain. With this deformity this portion of the nervous system is
narrowed laterally and elongated in its axial diameter so that the lowermost
portions of the medulla oblongata and the cerebellum extend through the
foramen magnum into the vertebral canal. This deformity is frequently asso-
ciated with a relative stenosis of the aqueduct of Sylvius and with abnormal
stretching of the lowermost cranial and the upper cervical spinal nerve roots
(Lichtenstein).

It is found very frequently in infants suffering from extreme degrees of
spina bifida (Arnold,3 Chiari,4 Lichtenstein,7 Schwalbe and Gredig,10 and
Jacob5). Its occurrence in adults has generally been in conjunction with bony
anomalies of the craniovertebral junction, such as basilar impression, or
platybasia, and fusion abnormalities of the cervical spine (Klippel-Feil’s
syndrome). Examples of such combinations have been reported by List8
and by Gustafson and Oldberg.5 The occurrence of an Arnold-Chiari de-
formity of sufficient magnitude to produce very marked neurological symp-
toms with no evidence of a spina bifida or of any bony anomaly in the cer-
vical region is most unusual.

REPORT OF CASE

Case #151-279. The patient A.B.J., an American negress with part Indian blood, aged
40 years, was admitted to the outpatient division of the Department of Neurology and
Neurological Surgery on Oct. 26, 1942. She stated that she had been perfectly well until
August 1942, when, after a day of unusually hard work, her legs gave way and she slumped
to the floor. In a few minutes she arose and seemed perfectly well. The following morning she
experienced weakness in both lower extremities and was forced to use a cane for support.
Two days afterwards she experienced weakness in the entire right upper extremity and had
particular difficulty in moving her fingers. This was followed by tingling paraesthesias in the
right upper and both lower extremities as well as by sharp knife-like pains which were
most severe in the calf of the right leg and in the region of the right elbow. The paraesthesias
persisted and the pain occurred intermittently. She developed a subjective feeling of numb-
ness in the affected parts.

In her past history nothing of significance was noted. She had suffered from otitis media
on the right side since childhood and had had a single attack of acute arthritis of the left
ankle in August 1932. In 1923 some pelvic operation had been performed at the Cook County
Hospital. She had given birth to six children and had had one spontaneous miscarriage. Her
family history revealed nothing of significance.

Neurological examination on Oct. 26, 1942 showed deviation of the tongue to the right,
decreased motor power in the right upper and in both lower extremities, stereoanesthesia

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in the right hand, patchy hypesthesia and hypalgesia in the right upper extremity, and loss of vibration sense at the ankles. On Oct. 30, 1942 it was noted that she walked slowly and laboriously and that Hoffmann’s sign could be elicited on the right side. On Jan. 25, 1943 atrophy and fibrillar twitchings of the right side of the tongue were noted for the first time. The deviation of the tongue to the right was marked and hypalgesia was found on the right side of the body with the exception of the face. At this time a vascular lesion at the level of the hypoglossal nucleus was considered as a probable diagnosis. A subsequent examination on Mar. 1, 1943 revealed, in addition to the aforementioned findings, bilateral foot drop with absent Achilles’ reflexes. On July 19, 1943 nystagmus was noted and on Oct. 4, 1943 her gait was very feeble and marked pitting edema of both feet and ankles had developed. Because of the atypical clinical picture the patient was admitted to the Illinois Neuro-psychiatric Institute, the Neurological and Neurosurgical Division, on Nov. 15, 1943 for further studies.

A summary of the positive findings in the detailed neurological examination performed on Nov. 15, 1943 follows.

Cranial nerves. III, IV, and VI: Sustained rotary counterclockwise nystagmus in all directions of gaze but most marked on looking to the right. XI: Weakness of elevation of the right shoulder. XII: Deviation of the tongue to the right with marked atrophy of the right side and very obvious fibrillar twitchings. Fine fibrillar twitchings in the left half of the tongue.

Motor. Marked weakness of the right upper and lower extremities.

Sensation. Hypesthesia and hypalgesia of the right side of the body, excluding the face, of patchy distribution but most marked in the leg, hand and shoulder. Hypesthesia and hypalgesia of the left foot. Loss of vibration sense in both ankles. Stereoanesthesia in the right hand. The coordination test of placing the finger to the nose revealed marked ataxia with decomposition of movement on the right side.

Reflexes. 

<table>
<thead>
<tr>
<th>Superficial</th>
<th>Abdominals absent</th>
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</thead>
<tbody>
<tr>
<td>Deep</td>
<td>Patellar Rt. +++  Left +</td>
</tr>
<tr>
<td></td>
<td>Achilles Rt. 0    Left 0</td>
</tr>
<tr>
<td></td>
<td>Biceps Rt. +++    Left +</td>
</tr>
<tr>
<td></td>
<td>Hoffmann Rt. +++  Left +</td>
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<tr>
<td></td>
<td>Babinski Rt. 0    Left 0</td>
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</tbody>
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Spinal puncture performed on Nov. 17, 1948 revealed a complete subarachnoid block, the initial fluid pressure being 40 mm. of fluid. The cerebrospinal fluid was clear, colorless, and contained only one cell per c. mm. The Pandy and Wassermann reactions were negative and the total protein was 40 mg. per cent.

Because of the spinal block, diagnoses of syringobulbia and of intramedullary tumor at the level of the foramen magnum were proposed as well as the diagnosis of a neurinoma arising from the right hypoglossal nerve.

Roentgenograms of the skull, the foramen magnum and the cervical spine failed to reveal significant alterations.

A suboccipital exploration was performed on Nov. 26, 1943 under local anesthesia. Upon removal of the central portion of the occipital bone overlying the cerebellum and including the posterior part of the foramen magnum, the dura mater was found to be taut and bulging. The posterior arch of the first cervical vertebra was then removed. Upon incision of the dura mater a typical Arnold-Chiari deformity was encountered. The medulla oblongata and the tonsils of the cerebellum were displaced into the upper portion of the cervical spinal canal (Fig. 1). The right hypoglossal and spinal accessory nerves were found to be elongated and taut. The fourth ventricle and the space beneath the cerebellum on either side of the bulb was carefully explored. No tumor was found. The lower part of the fourth ventricle lay below the level of the foramen magnum. Blunt cannulae were inserted into the substance of the cerebellum without encountering any unusual resistance or any cystic cavity. Except for the downward displacement the cerebellum and the brain stem appeared normal.