The Chiari Malformation in Adults

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SUMMARY: *The clinical features of the Chiari Malformation in seven adult patients are presented. It is suggested that the clinical syndromes associated with this malformation, in adults, can be classified as (a) compression of structures at the level of the foramen magnum (with or without radiologically demonstrable associated bony anomaly at the cranio-vertebral junction) (b) increased intracranial pressure or obstructive hydrocephalus and (c) intramedullary cervical cord syndrome. The usefulness of tomography, and demonstration of the vertebro-basilar circulation in the neuro-radiologic investigation of these patients is emphasized. The surgical procedures performed in the management of these patients are outlined.*

RÉSUMÉ: *Les manifestations cliniques de la malformation de chiari chez sept adultes sont presentées en details. Les syndromes cliniques peuvent être classifiés de la façon suivante (a) compression des structures nerveuses au niveau du trou occipital (avec ou sans anomalies osseuses de la charniére cranio-vertébrale) (b) hypertension intracrânienne ou hydrocéphalie obstructive. (c) syndrome cervical intramedullaire. L'importance de la tomographie osseuse et l'angiographie vertébrale est soulignée. Les lésions operatoires et la conduite therapeutique sont analysées.*

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To most clinicians, the eponym, the Arnold Chiari Malformation, suggests an infant with a lumbar myelomeningocele, hydrocephalus, and associated morphologic abnormalities of the brainstem, cerebellum and perhaps other regions of the cerebro-spinal axis.

Several case reports over the past three decades have drawn attention to the occurrence of the Chiari Malformation in adults, usually without associated spina bifida or overt hydrocephalus (Aring, 1938; McConnell & Parker, 1938; Ogryzlo, 1942; Ray, 1942; Bucy & Lichtenstein, 1945; Hurteau, 1950; Spillane et al, 1957; Teng & Papatheodorou, 1965; Appleby et al, 1968; Carmel & Markesbery, 1969; Millar, 1974). In this paper we emphasize the clinical syndromes associated with the Chiari Malformation in seven adult patients, the radiologic investigations and the findings at surgery.

CASE REPORTS

Case: 1

M.R.:, a 32-year-old housewife, had been aware of a mild unsteadiness of gait for about four years. Two years prior to entry she had been told by her family physician of an abnormality of her eye movements (nystagmus). She had also noted clumsiness of her hands for the same period of time, and this had become more marked in the two months preceding admission. This clumsiness interfered with her domestic activities such as sewing and knitting. She had been aware of occasional episodes of sub-occipital headaches, which radiated to the frontal regions. Abnormalities on physical examination were limited to the central nervous system where there was nystagmus on horizontal and vertical gaze, ataxia and spasticity chiefly in the lower limbs, and a slight diminution of position sense in the fingers of both hands. X-rays of the skull and spine were normal. A positive contrast myelogram showed a normal flow of the contrast material in the spinal subarachnoid space. At the level of the odontoid process, the contrast was slightly restricted on the left side; (Fig. 1) the upper cervical nerve root sleeves followed a normal course. Pneumoencephalogram showed prolapse of cerebellar tonsils which were seen in a tongue-like outline to the level of the posterior arch of C_2. Air was also seen to demarcate the anterior inferior aspects of the prolapsed tonsils; no filling of the ventricular system was obtained. Surgery confirmed prolapsed cerebellar tonsils, which were markedly gliosed. At follow-up, over the past four years, there has been no progression of the neurologic deficit. She is able to manage all her domestic duties without functional

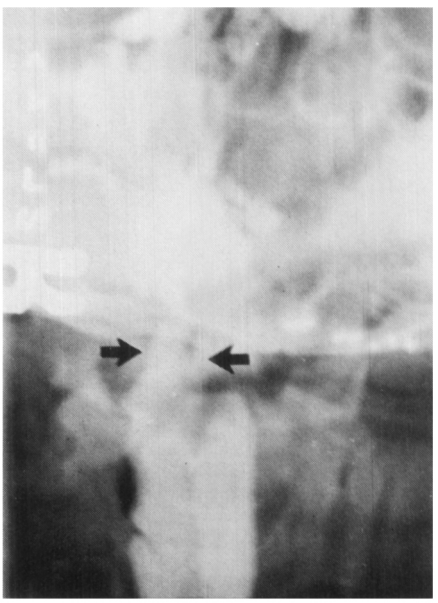

Figure 1—Myelogram: Constriction of contrast material at the odontoid level.

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deficit. The nystagmus persists on lateral gaze.

Case: 2

W.C.:, a 34-year-old man had complained of low back discomfort for about ten years, which he attributed to carrying heavy loads on his back. He has been aware of "cramp" and a feeling of tightness in the lower limb muscles for about five years, particularly with activity. He consumed large quantities of alcohol, and the unsteady gait which had been observed for several years was attributed by his colleagues (and by him) to excessive alcohol intake. On examination, he was of below average intelligence, with a short neck, low hairline, and restriction of neck movements in all directions (Fig. 2a). There was a marked spastic tetraparesis; his gait was also ataxic. X-ray of the skull and spine showed the following: there was platybasia; the odontoid and the anterior arch of C_1 could not be identified as separate structures; the anterior rim of the foramen magnum was fused with subjacent bone structures; the posterior arch of C_1 lay within the foramen; the lateral aspects of the foramen seemed fused with the lateral aspect of the arches of C_1 and C_2. Myelogram (Fig. 2b) showed maximal constriction of the contrast column on the left side at the level of the bony deformity. The upper cervical nerve root sleeves were directed cranially. Pneumoencephalogram showed filling of the fourth ventricle, aqueduct, and posterior third ventricle. The fourth ventricle was of normal size and appeared normal in position; the lateral ventricles were not outlined.

The bony abnormalities were confirmed at surgery; in addition the dura was thickened, with new vessel formation; there were dense arachnoid adhesions, and marked cerebellar tonsilar prolapse (Fig. 2c). At follow-up, there had been no progress in the neurologic deficit. Though his gait remained spastic and mildly ataxic, these features were less marked.

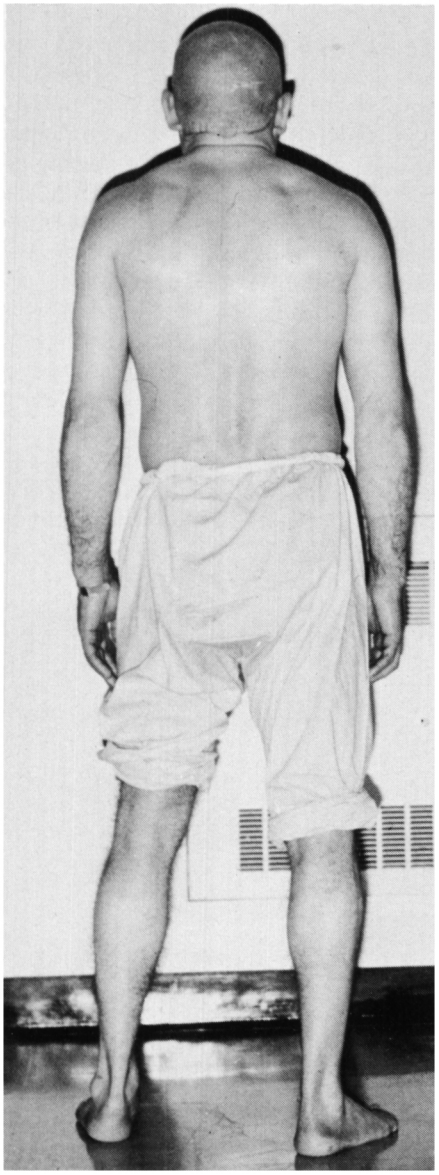

Figure 2 (a)—Clinical photograph, dorsal view. Note short neck, wide based stance.

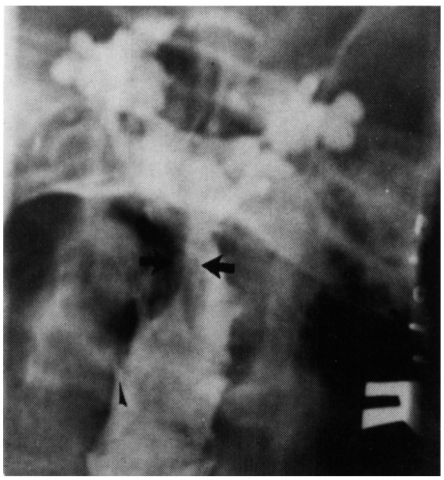

Figure 2 (b)—Myelogram—constriction of contrast indicated by two upper arrows. Small arrow on left points to cranially directed nerve root sleeve.

Figure 2 (c)—Operative photograph — note herniated tonsils, arachnoid adhesions and neovascularisation.

Case: 3

A.M.:, This 53-year-old man had been investigated on several occasions at different hospitals for episodic vomiting which had recurred over twenty years. The vomiting had been attributed to peptic ulceration, although an ulcer could not be demonstrated on several appropriate, radiologic examinations. Low back pain had been noted for about ten years, and an unsteady gait for about eight years. A year prior to entry he was treated at a mental hospital with a psychiatric diagnosis of a depressive illness. On examination he was of below average intelligence. He had a short neck and restricted neck movements in all directions. There was nystagmus on horizontal and vertical gaze and a marked spastic tetraparesis. Similar bony anomalies at the craniovertebral junction were evident in this patient as in patient number two. A myelogram showed marked restriction of the contrast column in the A.P. views, maximum at C_1-C_2 levels. The upper cranial nerve root sleeves were directed cranially; there was a soft tissue density suggesting tonsillar prolapse which was

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noted on supine, prone and oblique views. At pneumoencephalography, the fourth ventricle could not be filled with air. Air passed from the cisterna magna over the superior aspect of the cerebellum and into the quadrigeminal cisterns. There was evidence of cerebellar atrophy. At surgery, the bone at the foramen magnum was markedly thickened, particularly in the midline where it caused severe extradural compression. At follow-up, the degree of spastic tetraparesis was less marked than pre-operatively. The nystagmus persisted, chiefly on horizontal gaze.

Case: 4

C.S.:, Six months prior to admission, this 61-year-old man developed episodic vertigo and vomiting, while undergoing treatment for pulmonary tuberculosis. These symptoms worsened and in addition his speech became slurred. Five weeks prior to entry, there were episodic, severe, occipital headaches, and there were three episodes of momentary loss of consciousness associated with bouts of vomiting. On examination, he had a short neck with a low hairline. His speech was dysarthric and of the cerebellar type. There was bilateral papilledema, horizontal nystagmus, ataxia of trunk and gait and incoordination which was more marked in the left arm and leg than on the right. X-ray of the skull suggested increased intracranial pressure. C_1 was occipitalised and associated with synostosis of the atlanto-occipital joint on the right (Fig. 3). The posterior arch of C_1 was open, the foramen magnum appeared large. Bilateral brachial angiograms and a ventriculogram showed evidence of a right sided cerebellar lesion. At surgery, the left half of the atlas was absent. Both cerebellar tonsils were swollen, herniated to the level of C_2, and markedly gliosed. In addition to the congenital bony anomalies and the prolapsed tonsils, a metastatic tumor was removed from the medial aspect of the (R) cerebellar hemisphere. This patient died six months following the surgical procedure, presumably due to widespread metastatic disease. An autopsy was not performed.

Case: 5

L.G.:, this 34-year-old housewife was initially seen eight years previously by a neurologist and a diagnosis of myasthenia gravis was made. The patient was treated with anticholinesterase drugs. Despite this medication, constant fatigue persisted, together with episodic weakness and paraesthesiae of the right leg. Three years prior to entry, she had sought medical attention with the additional complaints of occipitonuchal pain and weakness in the upper limbs. A myelogram was performed and interpreted as showing a herniated cervical disc which was treated surgically. She presented to us with increasingly severe occipital pain, blurring of vision, and unsteadiness of limbs and gait. On examination there was bilateral papilledema, nystagmus on horizontal and vertical gaze, mild weakness of the left upper limb, diminution of sensation over the right trunk arm and leg, and hyperreflexia in both lower limbs. X-rays of skull and spine were normal. A brachial angiogram (Fig. 4a) showed the vertebro-basilar complex to be displaced forwards. The tonsillo-hemispheric division of the posterior inferior cerebellar artery was herniated through the foramen magnum. The choroid point was displaced forwards and slightly upwards. The ventriculogram confirmed the angiographic findings of hydrocephalus of the lateral and third ventricles. The aqueduct was not outlined, air was seen in the cervical spinal canal outlining prolapsed cerebellar tonsils. The myelogram done three years previously was reviewed and showed diffuse expansion of the cerivcal cord from C_2 down to the lower cerivcal levels, and upward inclination of the

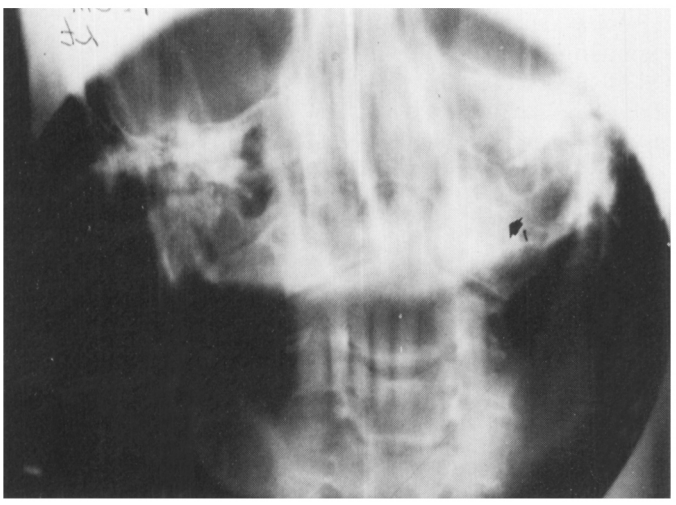

Figure 3—Tomogram of craniovertebral junction C_1 is occipitalised and there is synostosis of the (r) atlanto-occipital joint.

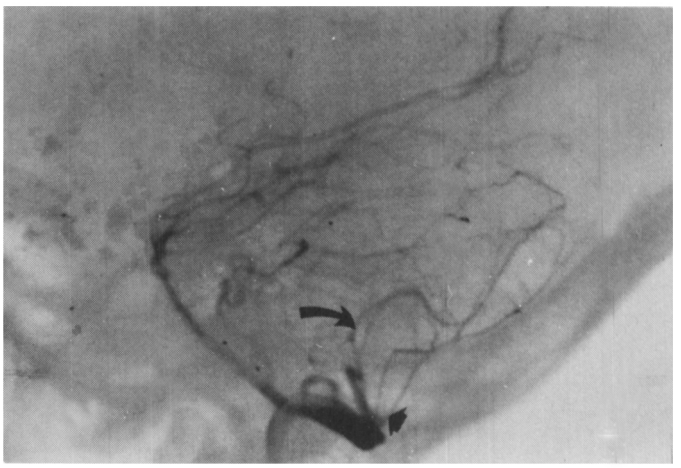

Figure 4 (a)—Brachial angiogram — Basilar artery is pushed against clivus; tonsillo-hemispheric branch of post inf. cerebellar artery indicated by lower arrow. Choroid point indicated by upper arrow.

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nerve root sleeves was evident to the level of C_3. Surgery (Fig. 4b) confirmed prolapse of the cerebellar tonsils which were markedly gliosed and bound down with dense arachnoid adhesions. This patient has shown remarkable improvement at follow-up examinations. The symptoms and signs of increased intracranial pressure have cleared; the sensory deficit in the right upper limb has cleared, and the weakness and hyperreflexia in the lower limbs have improved.

Case: 6

T.C.:, This 19-year-old technical college student was admitted to the gynecology service for investigation of amenorrhoea. She had noted recurrent headaches and vomiting for about a year. Despite this she had continued at her studies, and had also gained about 20 lbs. in weight. She had been aware of an asymmetry of her tongue, noted incidentally while brushing her teeth. On examination she was short, mildly obese, and there was a mild paresis of the right hypoglossal innervated muscles, with deviation of the tongue to the right. X-rays suggested chronic increased intracranial pressure. Pneumoencephalogram (Fig. 5a) failed to outline any part of the ventricular system. The pontomedullary cistern was narrow, the circummesencephalic communicating cistern seemed splayed and the quadrigeminal cisterns were dilated. There was tonsillar prolapse to the level of C_1. Ventriculo-atrial shunting was followed at a later date by suboccipital craniectomy. The suboccipital squama was thin, the external surface of the dura was thickened and reddish. Numerous adhesions were present between the inner surface of the dura and the arachnoid at the level of the foramen magnum. The cerebellar tonsils were enlarged, gliosed and prolapsed to the level of C_2. The foramen of Magendie was occluded by thick membranous adhesions (Fig. 5b). Revision of the ventriculo-atrial shunt was necessary at follow-up, as signs suggesting intracranial pressure recurred consequent on malfunction of the shunt system. She completed her studies, and is presently employed as a laboratory technologist. The right hypoglossal paresis has improved; she remains amenorrheic.

Case: 7

J.G.C.:, This 24-year-old mechanic had noted episodic pallor of the fingers of both hands when working in a cold environment, for the preceding four years. The following year, at an annual physical examination, a curvature of the spine was detected. Two years prior to entry he had noted weakness of hand grip bilaterally, and decreased perception of temperature in both hands. The weakness in the upper limbs and impairment of sensation had been progressive, and on a couple of occasions he had burnt his fingers without being aware of the injury. Abnormalities on examination were as follows: There was marked nystagmus on horizontal and vertical gaze. The trapezius, supra and infraspinati muscles were wasted and weak, as were the muscles of the right arm and forearm. A claw hand deformity was evident on the right. Pain and temperature sense were markedly impaired distal to the elbows bilaterally, while touch was relatively preserved. The tendon reflexes were absent in the arms and abnormally exaggerated in the legs. There was a slight thoracic scoliosis. X-rays of the skull showed no abnormality. There was scoliosis of the thoracic and lumbar spine. There was a slight expansion of the spinal canal at the level of C_5 and C_6. A myelogram suggested narrow channels on both sides of the odontoid, slightly more marked on the right. On supine positioning there was a free flow of contrast material through the foramen magnum. The myelogram did not suggest an enlarged cervical spinal cord. An air myelogram showed herniation of the cerebellar tonsils. Surgery (Fig. 6) confirmed displacement of cerebellar tonsils to C_2 with the caudally displaced cerebellar tissue appearing large, white and markedly gliosed. Two cc's of clear fluid was aspirated from the widened cervical spinal cord, and a drainage tube inserted through the myelotomy connecting with the spinal subarachnoid space. At follow-up this man had been able to return to work; there had been no progression regarding the sensory deficit or muscle weakness in his limbs. Horizontal nystagmus persisted, but was not marked.

DISCUSSION

In our series, Case 1 is an example of the Chiari Malformation presenting in adult life. The clinical syndrome is explicable on the basis of compression of neural structures at the cranio-vertebral junction. The absence of radiologically demonstrable bony anomalies at the craniovertebral junction has been documented in several previous reports of similar cases (Aring, 1938; Ogryzlo, 1942; Busy, 1945; Gardner and Goodall, 1950; Teng and Papatheodorou, 1965; Appleby et al, 1968; Banerji and Millar, 1974).

Case 2 illustrates the Chiari Malformation in association with congenital bony abnormalities at the craniovertebral junction. These bony abnormalities can be suspected, when a short neck, low hairline and relative restriction of neck movements are present. When these clinical signs co-exist with radiologically demonstrated craniovertebral junction anomalies, an associated Chiari Malformation is highly likely.

Case 3 illustrates the co-existence of the clinical and radiological features suggesting a lesion at the craniovertebral junction. In addition, the marked thickening of bone at the level of the foramen magnum (noted at surgery) added a further element to the compression of neural structures already compromised by the presence of the Chiari Malformation.

In Case 4, the bony abnormalities at the foramen magnum, demonstrated radiologically and confirmed at surgery, together with the prolapse of the cerebellar tonsils to the level of the second cervical spine and marked gliosis of these herniated cerebellar structures, confirm the existence of the Chiari Malformation. The additional problems of bronchogenic malignancy, pulmo-

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nary tuberculosis, and a cerebellar metastatic lesion may have been precipitating factors leading to the emergence of the clinical syndrome.

Cases 5 and 7 are examples of a cervical syringo-myelic syndrome in association with the Chiari Malformation. Gardner (1957) has postulated that developmental atresia or occlusion of the outlet foramina of the fourth ventricle causes diversion of the CSF pulse waves from the sub-arachnoid space into the central canal of the spinal cord. The hydrodynamic effect of these pulse waves then leads to the development of syringomyelia. This view has been supported Appleby et al (1968), and Hankinson (1970). Arguments against this hypothesis have been advanced by Ball and Dayan (1972) and "vascular factors" have been implicated in the pathogenesis of syringomyelia (Banerji and Millar,

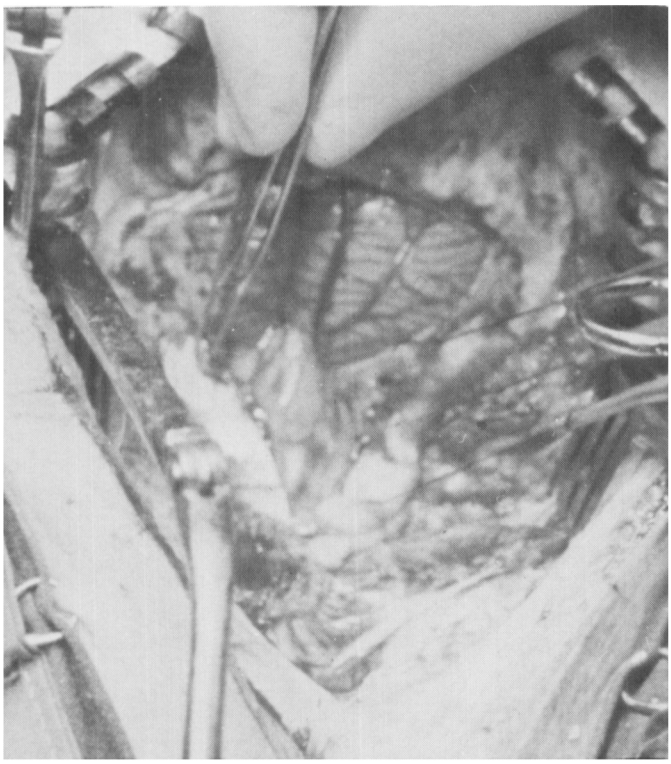

Figure 4 (b)—Operative photograph — note herniated, gliosed, pale looking tonsils.

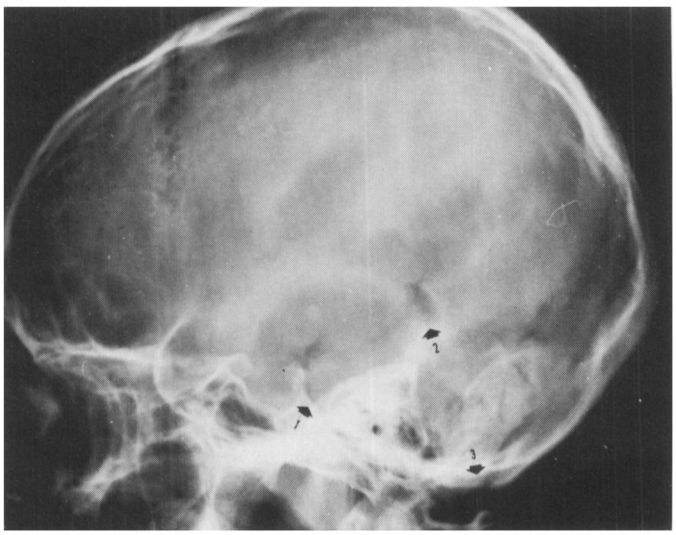

Figure 5 (a)—Lumbar pneumoencephalogram. Arrow 1 points to the pontine cistern; 2, quadrigeminal cistern, 3, air outlining herniated tonsils.

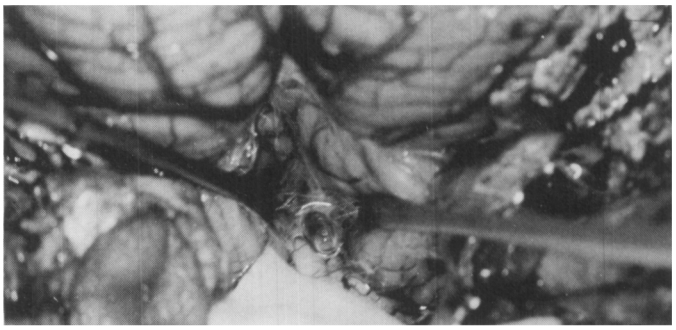

Figure 5 (b)—Operative photograph. Note the membrane overlying the foramen of Magendie demonstrated after separation of tonsils.

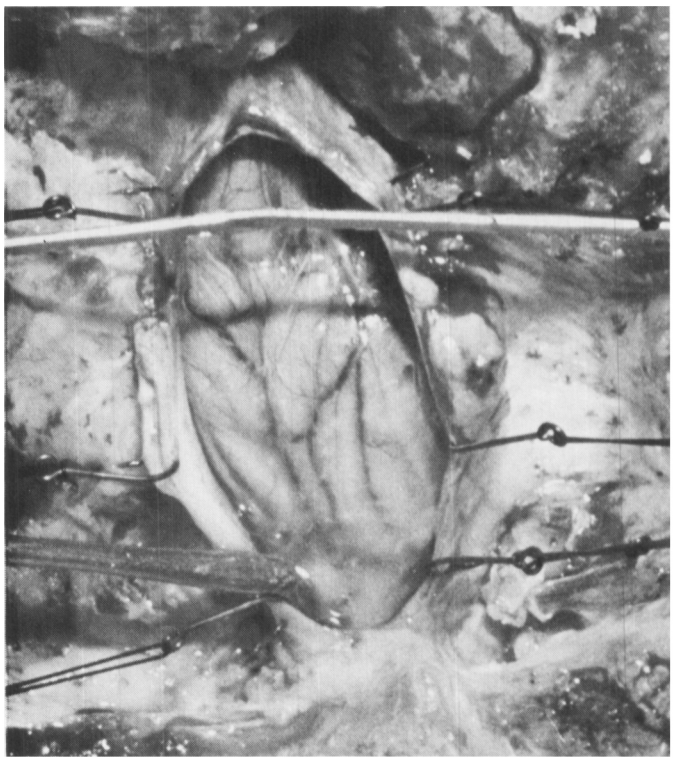

Figure 6—Operative photograph showing gliosed, markedly herniated tonsils. The horizontally placed instrument indicates level of foramen magnum.

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1974). Our experience would lend further support to Gardner's theory.

Marked internal hydrocephalus may be the most prominent feature of the morphologic changes in the central nervous system consequent on the Chiari Malformation, and Case 6 illustrates such a presentation. The amenorrhoea and obesity in this patient could represent hypothalmic dysfunction consequent on the marked internal hydrocephalus (including third ventricle); the asymmetric hypoglossal nerve involvement suggests a further lesion in the brainstem, or in the extramedullary course of the nerve. Banerji and Millar (1974) record dementia as the main manifestation of an adult with the Chiari Malformation, the dementia is ascribed by them as due to "normal pressure" hydrocephalus.

Thus, from our experience the clinical syndromes in adults associated with the Chiari Malformation can be classified as follows:

(a) Compression of structures at the level of the foramen magnum.
  (1) Without associated bony abnormality at the craniovertebral junction.
  (2) With associated bony abnormality at the craniovertebral junction.
  (3) With associated bony abnormality, and an independent pathologic process.
(b) Increased intracranial pressure or obstructive hydrocephalus.
(c) Intramedullary cervical cord syndrome.

In any one patient the clinical features may represent one or more of the above syndromes.

Tomography is a valuable, and often essential, technique in the radiologic studies of these patients. It is invaluable to elucidate the often complex bony abnormalities at the craniovertebral junction. When used in conjunction with air contrast examination to outline the fourth ventricle, cervical subarachnoid space and posterior fossa subarachnoid cisterns it is most helpful. Appleby et al (1968) comment on the usefulness of the supine position in myelographic examination of these patients, particularly to demonstrate the herniation of the cerebellar tonsils. We have found air encephalography and demonstration of the posterior inferior cerebellar artery and its tonsillohemispheric branches by angiography as helpful as myelography in outlining the cerebellar tonsils. The surgical procedures performed in our series of patients have included the following:

(a) Posterior decompression of the osseous structures at the craniovertebral junction.
(b) Further decompression of the structures at the craniovertebral junction by incising the thickened atlanto-occipital membrane and dura.
(c) Inspection of the degree of herniation of the cerebellar tonsils, and the degree of 'arachnoiditis'.
(d) Since attempts to 'break' the dense arachnoid adhesions seem to be associated with changes in respiratory and cardiac rhythm, and probable insult to the many fine vessels in the same area, it has been our policy not to attempt to 'unroof' the foramen of Magendie or to attempt to plug the orifice at the apex.
(e) Instead, a ventriculo-atrial shunt has been performed to relieve the hydrocephalus. This has been done as a preliminary step, prior to the posterior decompression at the craniovertebral junction in those cases where hydrocephalus is particularly prominent (in the radiologic studies).
(f) When the intramedullary cervical cord lesion is the predominant clinical feature, aspiration of the syrinx, myelotomy, and establishment of a 'shunt' between the syrinx and the spinal subarachnoid space has been achieved.

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