THE ARNOLD-CHIARI MALFORMATION

RADIOLOGICAL EXAMINATION WITH THE "ZIEDSES DES PLANTES" PROCEDURE

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During the last 12 years several publications have appeared on the x-ray diagnosis of the Arnold-Chiari malformation. The ordinary radiological examination of the skull and spinal column in these cases only reveals associated anomalies, such as bony abnormalities of the cranio-vertebral junction, deformation of the cervical spine, and spina bifida. Although these facts in conjunction with the clinical findings may suggest the presence of an Arnold-Chiari deformity, they do not establish the diagnosis. On the other hand several cases have been reported in which the Arnold-Chiari deformity was not associated with other bony abnormalities (Aring, 1938; McConnell and Parker, 1938; Ogryzlo, 1942; Bucy and Lichtenstein, 1945; Epstein, 1948; Swanson and Fincher, 1949; Gardner and Goodall, 1950).

It is apparent that direct radiographic demonstration of the Arnold-Chiari deformity requires special methods. List (1941) was the first to apply myelography as a diagnostic procedure in these cases and he found that the iodized oil was arrested in the high cervical region with a concave filling defect. Adams, Schatzki, and Scoville (1941) gave a more detailed description of the myelographic findings in their case, differentiating it from those found in a high cervical cord tumour. Similar reports were published, for example, by Epstein (1948), by Lewin, Wycis, and Young (1950), and by Malis, Cohen, and Gross (1951). All authors mention that some oil passes to the basal intracranial cisterns.

The Arnold-Chiari deformity can also be demonstrated by lumbar pneumo-encephalography as was shown by Marks and Livingston (1949) and by Swanson and Fincher (1949). The air may be arrested at the second cervical level and even outline the herniated cerebellar tonsils.

Other reports on lumbar pneumo-encephalography in the Arnold-Chiari deformity are less precise on these diagnostic features, but all agree that

little or no air enters the ventricles while the intracranial subarachnoid spaces may be visualized.

Another advantage of myelography and lumbar pneumo-encephalography is that the swelling of the cervical cord caused by a concomitant hydromyelia may be outlined by the air or oil.

As far as we know no mention has been made in the literature of untoward reactions after lumbar puncture. In one of our patients, however (N 7218), a boy aged 17 with basilar impression and Arnold-Chiari deformity, the lumbar puncture was followed by quadriplegia and respiratory failure necessitating immediate operative intervention. Fortunately the patient recovered. This patient had never shown any signs of increased intracranial pressure. When the Arnold-Chiari malformation is associated with signs of increased intracranial pressure in adults, we avoid performing myelography or lumbar encephalography, as we think that lumbar puncture is contraindicated in such cases.

This report is based on ventriculographic studies of 11 patients in whom the presence of the Arnold-Chiari malformation was verified at operation. In all cases the third and fourth ventricles were examined with small quantities of air according to the method described by Ziedses des Plantes (1950).

The Ziedses des Plantes Procedure

A small quantity of air (15–20 ml.) is injected into the right lateral ventricle through the fontanelle or a burr hole. It is important that the air collects in the anterior horn, and if some of the air has escaped to the temporal horn the patient is placed in a sitting position with the head flexed forward; this makes the air travel to the upper part of the body of the lateral ventricle and if the patient subsequently lies down on his back all the air will be collected in the anterior horn. After this the patient is induced to make a slow backward somersault. During the first part of this movement the air travels along the floor of the anterior horn into the third ventricle, where it is collected in its rostral part at the moment that the somersault has progressed to the position of feet upwards, head downwards. While the

somersault is being continued until the patient lies face down on the table, the air travels along the floor of the third ventricle to its posterior part and penetrates through the aqueduct into the fourth ventricle. In this position lateral and postero-anterior *x*-ray pictures are taken. When the fourth ventricle is not obstructed, air will be found in the spinal canal. Should communicating hydrocephalus be demonstrated a second, forward, somersault is carried out, by means of which the air passes to the intracranial subarachnoid spaces.

It is easy to carry out this manoeuvre in little children by seizing the feet by one hand and the head by the other. With adolescents and adults the somersaults are carried out by means of a tilting table.

In order to obtain a maximum contrast the volume of injected air is slightly greater than the amount of ventricular fluid withdrawn and the *x*-ray exposure should be correct and not too heavy. The films are best examined with closely shielded illuminators and in a dimly lighted room (Marks and Livingston, 1949).

This procedure has proved to be very useful for the following three reasons: (1) The intraventricular pressure can be measured at the same time and an impression of the dimensions of the ventricular system is obtained. (2) In some cases the prolongation of the fourth ventricle or the herniation of the cerebellum into the spinal canal can be visualized. (3) It enables us to determine whether there is any communication between the ventricles and the intracranial cisterns and the spinal subarachnoid space respectively. Even if (2) should not materialize, very valuable information is obtained concerning (1) and (3).

Case Material

Five patients were adults. Their clinical pictures consisted of the following chief symptoms:—

Case 1.—Headaches, occipital neuralgia, increased tendon reflexes on the right, paraesthesiae in the right hand.

Case 2.—Occipital headaches radiating to the nape of the neck and increasing with retroflexion of the head. During an attack of headache the head is kept flexed forward. On a few occasions retroflexion of the head was followed by vertigo and a state of collapse. Neurological examination revealed only horizontal nystagmus while looking to the left.

Case 3.—Headaches radiating along the nape of the neck and the back to the arms and legs. Paraesthesiae in hands and feet. Loss of power in right arm and leg. Bilateral papilloedema. Normal tendon reflexes in upper, but increased tendon reflexes on both sides in lower, extremities. Positive Babinski, Oppenheim, and Gordon signs on the right. Bilateral decrease of all types of sensation in the area C 2 to C 8.

Case 4.—Headaches, vertigo, ataxia, and disturbances of gait.

Case 5.—Occipital pains radiating to the left arm and leg, vertigo, disturbances of gait, diminished corneal reflexes, paresis of left trapezius and sternomastoid muscles. Increased tendon reflexes on the right. Babinski, Oppenheim, and Gordon signs on the right. Decrease of all types of sensation in the area C 1 to C 6. Bilateral disturbance of kinaesthesia of hands and feet.

The other six patients were infants, all younger than 1 year old. They were operated upon because of hydrocephalus with rapid enlargement of the head. Other important data, among which are the findings with the Ziedses des Plantes procedure, are summarized in Table I.

The Air Studies of the Malformation

In eight out of 11 patients the malformation could be demonstrated, in one of them with lipiodol, in the others with air according to the Ziedses des Plantes procedure. The most typical pictures were obtained after the first somersault on a lateral film taken with a horizontal beam. Therefore it is essential first to describe briefly the normal picture obtained by the same technique (Fig. 1). The fourth ventricle appears as a triangle. Caudal to its inferior angle it is continuous with the cisterna magna. Normally the inferior opening of the fourth ventricle is situated above the level of the foramen magnum. The cisterna magna lies close to the bone of the posterior fossa, and according to Robertson it usually extends about 3 cm. behind the posterior lip of the foramen magnum. At the level of the foramen magnum the cisterna magna is directly continuous with the spinal subarachnoid space. The fourth ventricle is not always so well visualized, as most of the air may escape from it into the cisterna magna and the spinal subarachnoid space.

There are two characteristic *x*-ray findings in the Arnold-Chiari malformation, namely, (1) visualization of the elongated fourth ventricle, and (2) demonstration of the cerebellar herniation into the spinal canal.

Visualization of the Elongated Fourth Ventricle.—In our first patient (Case 1), examined with the Ziedses des Plantes procedure, the air pictures were not very clear. We repeated the examination with lipiodol injected into a lateral ventricle. This was done just before operation (Fig. 2). The triangular part of the fourth ventricle appeared to be well preserved, but its inferior end was elongated, forming a small canal that projected through the foramen magnum and extended as far as the inferior border of the posterior arch of the atlas. This was confirmed at operation.

In Cases 8, 9, and 10 we succeeded in obtaining good air pictures of the fourth ventricle. The latter was visualized as a small canal that was directly

continuous to the aqueduct of Sylvius and extended downwards as far as the first and the second cervical vertebrae respectively (Figs. 3 and 4). In Case 5 only the elongated inferior part of the fourth ventricle was visualized. It had the appearance of a small canal that projected through the foramen magnum as far as the atlas.

In Cases 1, 5, 8, and 9 no communication with the spinal subarachnoid space was found, so that after the first somersault the air remained within the fourth ventricle.

In Case 10 much air had been injected, so that both ventricle and spinal subarachnoid space were filled with air.

Demonstration of Cerebellar Herniation into Spinal Canal.—This occurred only in cases with a good communication between the fourth ventricle

and the spinal subarachnoid space and in none of them had the air remained within the fourth ventricle. The spinal air column showed typical features that enabled us to make the diagnosis.

In Case 7 the spinal subarachnoid space was only visualized in the area below the posterior arch of the atlas. The upper border of the spinal air column was cut off in an oblique direction, thus delimiting the inferior pole of the malformation (Fig. 5).

In Case 11 the protruding cerebellar tonsils were outlined by air (Fig. 6). A similar picture was obtained in Case 4.

Cerebellar tonsils that are displaced downwards as a result of a space-occupying lesion in the posterior fossa may give a similar shadow to that of the Arnold-Chiari malformation (Fig. 7). However, the differential diagnosis can as a rule be made on the grounds of the clinical picture and the other

TABLE I
DATA ON 11 CASES OF ARNOLD-CHIARI MALFORMATION VERIFIED AT OPERATION

	Sex and Age	Concomitant Anomalies	Intra-cranial Pressure	Ventricles	Arnold-Chiari Malformation Demonstrated by Air	Air Passes to		Results of Suboccipital Decompression
						Spinal Sub-arachnoid Space	Basal Cisterns	
<i>Adults</i>								
Case 1 (D540)	A.v.d.L. Male, 26	Platybasia	Normal	Dilated	By intra-ventricular lipiodol	—	+	Good
Case 2 (D551)	J.H.v.D. Female, 26	—	Normal	Dilated	—	—	—	Died
Case 3 (N6567)	M.P.M.I. Female, 43	Syringoencephalomyelia	Increased	Dilated	—	—	+	Improved
Case 4 (N8429)	H.H. Male, 44	Basilar impression + atlanto-occipital synostosis	Normal	Normal	+	+	+	Good
Case 5 (D809)	J.A.V. Female, 47	Hydromyelia	Normal	Dilated	+	—	+	Good, but only recently operated upon
<i>Infants</i>								
Case 6 (N8805)	J.M. Male, 11 months	Atresia of the aqueduct; diverticulum of left lateral ventricle extending into posterior fossa	Increased	Greatly dilated	—	—	—	Good for a few months; then increasing hydrocephalus (The diverticulum was incised and opened into the subdural space.)
Case 7 (N10377)	W.J.S. Male, 1 month	Myelomeningocele	Very high	Greatly dilated	+	+	—	Died
Case 8 (N10184)	J.B. Male, 3 months	Myelomeningocele	Very high	Greatly dilated	+	—	+	Died
Case 9	J.v.B. Female, 4 months	Myelomeningocele	Very high	Greatly dilated	+	—	—	Increasing hydrocephalus
Case 10 (D739)	J.A.G. Male, 2½ months	Myelomeningocele	Very high	Greatly dilated	+	+	+	Good
Case 11 (D791)	A.R. Male, 6 weeks	Myelomeningocele	Very high	Dilated	+	+	+	Increasing hydrocephalus

FIG. 1.—Picture of the normal obtained by the Ziedses des Plantes technique.

FIG. 2.—Case 1. Lateral and A.P. views of the fourth ventricle filled with lipiodol. The triangular part of the fourth ventricle is preserved, but its inferior end is elongated and extends as far as the lower border of the posterior arch of the atlas.

FIG. 3 (a).—After the first somersault the fourth ventricle appears as a small canal, which is directly continuous to the aqueduct and extends as far as the upper border of the arch of C2. Air has remained in the suprapineal recess.

FIG. 3 (b).—After the second somersault air has entered the pontine and interpeduncular cisterns.

FIG. 3 (c).—Medial section through the brain. The fourth ventricle is reduced to a small canal and the triangular part is poorly developed. The biventral lobule of the cerebellum is herniated into the cisterna pontis. The inferior part of the elongated medulla oblongata has not been adequately removed at the necropsy.

FIG. 4.—Case 10. After the first somersault the fourth ventricle is reduced to a small canal extending as far as C1. The cisterna magna is filled with air and appears to be well developed. Air has remained in the supra-pineal recess and the occipital horns of the lateral ventricles.

FIG. 5.—Case 7: (a) After the first somersault the air has passed from the fourth ventricle into the spinal subarachnoid space. This space, however, is only visualized in the area below the posterior arch of the atlas. The upper part of the spinal air column, which is cut off in an oblique direction, delimits the inferior pole of the malformation. Air in the occipital horns of the lateral ventricles shows that the latter are greatly dilated.

FIG. 5 (b).—After the second somersault more of the air collects in the spinal subarachnoid space. A large volume of air is seen bulging outward between the posterior arch of the atlas and the posterior rim of the foramen magnum. At operation the occipito-atloid ligament appeared to be absent and a very thinned dura over-filled with spinal fluid protruded between atlas and occiput under the neck muscles.

FIG. 6.—Case 11: (a) Taken after the first somersault when the air has passed to the spinal subarachnoid space and outlines the protrusion of the cerebellum into the spinal canal. Air has remained in the occipital horns of the lateral ventricles.

FIG. 6. (b)—After the second somersault the air has passed to the basal cisterns and to the subarachnoid space on the convexity of the frontal lobes. The cisterns are large, the frontal sulci are dilated, and air has accumulated at the tip of the frontal lobe. There has been some reflux of air to the frontal horns of the lateral ventricles.

FIG. 7.—Taken after the first somersault in a case of tumour of the posterior fossa. Air outlines the herniated cerebellar tonsils in the spinal canal.

ventriculographic findings, such as anterior or lateral displacements of the aqueduct of Sylvius in tumours of the posterior fossa.

Unsuccessful Visualization.—In Case 2 the air picture showed a dilated and obstructed ventricle (Fig. 8). Although the inferior part of it was located near the foramen magnum we thought it premature to draw a conclusion from the air studies about the nature of the obstruction, which was the reason why we did not include this case in the series of typical findings.

FIG. 8.—Case 2. Greatly dilated fourth ventricle (arrows). The air does not pass to the cisterns or to the spinal subarachnoid space. At operation the inferior opening of the fourth ventricle was found just below the level of the foramen magnum.

In Case 6 the air was arrested in the third ventricle because of a concomitant stenosis of the aqueduct. In Case 3 insufficient filling was obtained. In Cases 2 and 6 the other clinical and radiological findings were conclusive enough to have the operation performed in the right place.

The association of the Arnold-Chiari deformity with stenosis of the aqueduct as found in Case 6 was described by Lichtenstein. The coincidence of the stenosis of the aqueduct cannot be recognized if only myelography or lumbar encephalography are used as diagnostic procedures. In such cases the operative decompression of the Arnold-Chiari malformation only is ineffective and may even increase symptoms.

Although, on the other hand, ventriculography can only demonstrate the stenosis of the aqueduct and not the co-existing Arnold-Chiari malformation, it has less serious consequences, because ventriculo-

stomy of the third ventricle will relieve the hydrocephalus as well. The only risk is that, should a Torkildsen operation be attempted, it becomes obvious when the posterior fossa is opened that this operation is not feasible owing to the malformation.

The Hydrocephalus

Hydrocephalus was present in all the infants in this group of cases. In four out of the five adult patients the ventricles were dilated, but in only one of them was increased intracranial pressure found.

Most investigators assume that the Arnold-Chiari malformation is the cause of attendant hydrocephalus. It is thought that the malformation may produce hydrocephalus in several ways which are summarized by Ingraham and Scott (1943) as follows:—(1) The foraminae of Luschka and Magendie of the fourth ventricle may be mechanically obstructed by the squeezing of the medulla and cerebellar tails into the narrow cervical canal. Obstructive hydrocephalus results. (2) The subarachnoid space may be obliterated at the level of the foramen magnum by pressure of the herniated hindbrain. As the inferior opening of the fourth ventricle is located below the foramen magnum, there is a free escape of fluid from the fourth ventricle into the spinal subarachnoid space, but there is no communication between the latter and the intracranial subarachnoid space. Communicating hydrocephalus results. (3) The same mechanism is effected by aseptic arachnoiditis of the basal cisterns due to mechanical irritation.

Ogryzlo (1942) suggested that in some cases the relations between the Arnold-Chiari malformation and hydrocephalus might be inverse, hydrocephalus being the cause of the malformation. Gardner and Goodall (1950) reported a case of acquired Arnold-Chiari malformation supposedly due to an acoustic tumour.

The Ziedses des Plantes procedure is an excellent means of examining the communication between the fourth ventricle and the spinal and intracranial subarachnoid spaces. From our case material it appears that the outflow from the third ventricle was obstructed in one case (Case 6). There was no communication between the fourth ventricle and the spinal and intracranial subarachnoid spaces in two cases (Cases 2 and 9).

Communication between the fourth ventricle and the spinal subarachnoid space and not with the intracranial basal cisterns was found in one case only (Case 7). In four cases there was a communication between the fourth ventricle and the basal cisterns, but not between the fourth ventricle and the spinal subarachnoid space (Cases 1, 3, 5, and 8).

In three cases communication was found between the fourth ventricle and both the basal cisterns and the spinal subarachnoid space (Cases 4, 10, and 11).

The last two groups present some difficulty as to the explanation of a co-existing hydrocephalus, because the mechanisms that are active in its production differ from the generally accepted causes of hydrocephalus.

It is interesting to submit the seven cases belonging to these groups to a further analysis. Case 4 offers no difficulties because the intracranial pressure and the size of the ventricular system were found to be normal. In Cases 1 and 5 the ventricles were dilated, but the intracranial pressure was not increased. Possibly there had been a period of hypertensive hydrocephalus, but obviously the intracranial hydrodynamics had returned to normal. With the exclusion of these three cases, four patients remain in whom there was an increase of intracranial pressure. In Cases 8 and 9 air entered the pontine and interpeduncular cisterns only and did not pass more anteriorly or to the convexity of the brain (Fig. 3b). The cause of this blockage is unknown. The operative results were that one patient died, while in the other an increasing hydrocephalus persisted.

Cases 10 and 11 were most remarkable. The examination revealed a perfect communication between the fourth ventricle and the spinal and intracranial subarachnoid spaces (see Fig. 6b). The cause of hydrocephalus in these cases is unknown. We were not astonished that decompression of the malformation did not yield any result in Case 11, but we were much surprised that in Case 10 the intracranial pressure was restored to normal, while the abnormal increase of the size of the head was arrested after this procedure. It is apparent that different mechanisms are involved in the production of hydrocephalus in patients with the Arnold-Chiari malformation, of some of which we are still ignorant.

Our findings demonstrate that not all cases of hydrocephalus with the Arnold-Chiari malformation can be accounted for by an impediment to the cerebrospinal fluid circulation in the area of the malformation. As the causes of hydrocephalus are different in every individual case with Arnold-Chiari malformation, the spinal fluid circulation has to be examined in detail in each of these patients, since this may offer a reasonable basis for treatment. We think that the procedure applied in our cases may be helpful.

Summary

Eleven cases with the Arnold-Chiari malformation were examined by ventriculography with small quantities of air according to the Ziedses des Plantes method. In eight cases the presence of the malformation was demonstrated by the air studies. The intracranial pressure and the size of the ventricular system were estimated at the same time. Another advantage of this procedure is that it enables the investigator to estimate the patency of the communication between the fourth ventricle and the spinal and intracranial subarachnoid pathways. In several cases the presence of hydrocephalus could not be accounted for by the mechanisms generally accepted as producing hydrocephalus in cases with the Arnold-Chiari malformation.

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