Angiographic differentiation between Dandy-Walker cyst and arachnoid cyst of the posterior fossa in newborn infants and children

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Elevation of the tentorium and its dural sinuses, originally considered a diagnostic sign of Dandy-Walker cyst, may also occur in arachnoid cysts of the posterior fossa. Differentiation between these two lesions may be achieved angiographically by the evaluation of the posterior inferior cerebellar artery and its vermian branch, and of the inferior vermian vein. All these vessels are displaced forward and upward by an arachnoid cyst, while in the Dandy-Walker cyst the posterior inferior cerebellar artery is miniature and the vermian branch and the inferior vermian vein are absent.

KEY WORDS Dandy-Walker cyst arachnoid cyst retrograde brachial angiography posterior inferior cerebellar artery vermian vein hydrocephalus

In many clinics the neuroradiological investigation of infants with suspected hydrocephalus is confined to a demonstration of the fluid pathways by lumbar pneumoencephalogram and air or Pantopaque ventriculography. In our institute, since March, 1967, right retrograde brachial angiography has always been performed as a preliminary or exclusive investigation; other diagnostic studies may follow, depending on angiographic findings. Up to February, 1972, 100 hydrocephalic infants less than 1 year old were investigated under this policy; only 40 required more than one diagnostic study. The various final diagnoses in this series, largely derived from angiographic findings, are shown in Table 1.

This paper emphasizes the findings in five patients whose basic problem was a posterior fossa cyst. Most of these cases were originally considered as Dandy-Walker cysts; this diagnosis was based on the angiographic evidence of the high position of the torcular Herophyli and transverse sinuses,6,12,17 the straight sinus and elongated vein of Galen,10 the upward and forward deviation of the posterior cerebral arteries, and the huge avascular area within the posterior fossa. One of these cases was erroneously reported as a Dandy-Walker cyst in an earlier paper.14 Further experience including operative verification of angiography and especially postoperative angiography following a shunt procedure revealed that all of these angiographic findings may occur in both Dandy-Walker and arachnoid cysts, a feature recently confirmed by others.5,26 Both cysts represent developmental anomalies present during fetal life before the tentorium and its dural sinuses have reached their final posi-
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tion in the third fetal month. However, review of our material showed that these lesions may be differentiated by other angiographic findings, and to emphasize this differentiation we are including an example of inflammatory 4th ventricle occlusion (Case 6).

Case Reports

Case 1
This 20-day-old boy was admitted because of steady increase in head size and vomiting. The cranial circumference was 35 cm; the fontanels were bulging. Left retrograde brachial angiography revealed supratentorial hydrocephalus and a huge avascular space in the posterior fossa (Fig. 1); sinography showed high position of the torcular and transverse sinuses. A diagnosis of Dandy-Walker cyst was made and a shunt between the right lateral ventricle and the peritoneal cavity performed. The protein content of the ventricular fluid was 200 mg%. Postoperatively, vomiting became worse in spite of a sunken anterior fontanel. The posterior fossa cyst was punctured; its fluid had a protein content of 1000 mg%.

Exploration of the posterior fossa through a small craniectomy revealed the findings of a Dandy-Walker cyst; the aqueduct was occluded when tested by a thin rubber catheter. The occlusion was finally bypassed and a silicone catheter left in place. The patient did not tolerate the procedure well and died a few hours later. No autopsy was performed.

Case 2
This baby girl had been delivered surgically because of macrocephaly. When she was seen at 4 days the cranial circumference was 48.5 cm; sutures and fontanels were bulging. There was nystagmus in every direction of gaze. Right retrograde brachial angiography was diagnostic of Dandy-Walker cyst (Fig. 2). A shunt was placed between the lateral ventricle and the right atrium. On discharge, at 20 days of age, the cranial circumference had decreased to 40 cm. The patient did well, and at the age of 2 years psychomotor development was quite adequate and the shunt appeared to be functioning.

Case 3
This baby girl was 10 days old when first admitted. Normal delivery had followed full pregnancy but the mother, previously operated on for a thyroid tumor, had received x-

<table>
<thead>
<tr>
<th>General Diagnosis</th>
<th>Specific Pathology</th>
<th>No. of Cases</th>
</tr>
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<tbody>
<tr>
<td>supratentorial mass (20)</td>
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<tr>
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<td>porencephaly</td>
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<tr>
<td></td>
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</tr>
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<td></td>
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<td>Arnold-Chiari malformation</td>
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<tr>
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<td>foramen of Monro (occlusion)</td>
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</tr>
<tr>
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<td>4th ventricle (occlusion)</td>
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</tr>
</tbody>
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TABLE 1
Diagnoses in 100 hydrocephalic infants 1967-72
FIG. 1. Case 1. Arteriogram. Left: Anteroposterior view showing evidence of supratentorial hydrocephalus. There is good filling of the vertebrobasilar system but none of the posterior inferior cerebellar artery. Right: Lateral view showing upward deviation of the posterior cerebral arteries and of the vermian branch of the anterior superior cerebellar artery. Again, no posterior inferior cerebellar artery is visualized.

Ray therapy during pregnancy. Head size was large at birth; when seen, the cranial circumference was 45 cm and there was evidence of dolicocephaly. Right retrograde brachial angiography revealed a huge avascular area involving both infra- and supratentorial compartments (Fig. 3). Combined air and opaque ventriculography showed the entire ventricular system pushed forward by a cyst occupying two thirds of the intracranial space. The 4th ventricle was small and pushed against the clivus (Fig. 4 left), a finding which was not appreciated at the time. Sinography revealed a high position of the torcular and transverse sinuses (Fig. 4 right). A diagnosis of Dandy-Walker cyst was made and a shunt placed connecting the cyst with the right atrium. The patient did well, and 1 year later psychomotor development was adequate for her age. The fontanels were closed; the shunt was functioning. The angiographic study was repeated and offered evidence of decreased size of the cystic area (Fig. 5). The child was readmitted at the age of 2 yrs 8 mos because of repeated vomiting. There was evidence of inadequate functioning of the shunt. Chest films revealed the tip of the cardiac catheter at the level of T-3. At this time all diagnostic studies were reviewed, and the diagnosis of arachnoid cyst became obvious. Following shunt revision the child recovered and now, at 4 years, is asymptomatic.

Case 4

This baby girl was born at the end of the 36th week of gestation; her mother had had...
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German measles during early pregnancy. Steady increase of head size had been noted since birth. When admitted at the age of 2 months the cranial circumference was 41 cm and there was evidence of dolicocephaly. The anterior fontanel was $5 \times 5$ cm and was not under pressure. Transillumination was positive in the posterior part of the head; the EEG showed widespread flattened rhythms. Right retrograde brachial angiography revealed findings compatible with supratentorial hydrocephalus secondary to an arachnoid cyst of the posterior fossa (Fig. 6 upper left and right). A shunt procedure was performed, draining the cyst. The patient did well and when 15 months old was readmitted for complete evaluation. Neurological examination and cranial circumference were normal, the shunt was functioning, and the EEG was normal. The angiographic findings at this time revealed that hydrocephalus was no longer present and the cystic area had disappeared (Fig. 6 lower left and right).

Case 5

This 7-month-old boy was admitted for evaluation of an enlarged head, noted since the third month of life. He had been deliv-
Case 3

The cyst communicates with the 4th ventricle and pushes the entire normal ventricular system anteriorly. Sinography shows enormous elevation of the torcular Herophili and transverse sinuses. The sagittal diameter of the cyst may be appreciated by observing the limits of air and contrast media (arrow).

Case 6

This 3-month-old boy had meningitis immediately following birth. Recovery occurred within 3 weeks, but 15 days before admission the head began to rapidly increase. The fontanels were bulging. Right retrograde brachial angiography suggested obstructive tetraventricular hydrocephalus and normal position of the tentorium (Fig. 8). Pantopaque ventriculography confirmed this diagnosis, revealing a markedly enlarged 4th ventricle with occlusion of its foramina. Ventricular fluid had contained no cells and a protein content of 50 mg%. A shunt was performed between the lateral ventricle and the peritoneal cavity. No problems have occurred during the following 7 months.

Discussion

The Dandy-Walker cyst is a developmental anomaly in which a balloon-like dilatation of the 4th ventricle is associated with cerebellar anomalies. Although it is a congenital lesion, neuropathologists disagree on the primary mechanism in its development.\textsuperscript{1,2,9,11,24}

The arachnoid cyst is a developmental anomaly involving a huge dilatation of the normal space between the two layers of the posterior medullary velum. Arachnoid cysts should not be confused with so-called "cerebellar cystic arachnoiditis" which is a huge dilatation of the subarachnoid space resulting from inflammatory adhesions of the arachnoid membrane.

Both arachnoid and Dandy-Walker cysts may become symptomatic in infancy as well as in later years; obviously, the milder cases are encountered late in life while the severe cases come to medical attention early.

The pathological differentiation between these lesions is based upon two points: first is the location of the cyst with respect to the 4th ventricle. Dandy-Walker cysts are intraaxial, entirely confined within the 4th ventricle; arachnoid cysts are extra-axial, dorsal to the 4th ventricle. The second point is the de-
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formity of the posterior vermis which is typi-
cal of a Dandy-Walker cyst, but unusual
with arachnoid cysts. Other factors such as
patency or occlusion of the foramina of the
4th ventricle may be misleading, since in
Dandy-Walker cysts one or all of the normal
foramina may be patent, and arachnoid
cysts may or may not communicate with the
subarachnoid space.

The relation of the cyst to the 4th ventri-
cle may be assessed by examinations visual-
izing the fluid pathways; but identification of
the 4th ventricle by pneumoencephalography
or ventriculography may fail due to the un-
derlying pathology. For instance, occlusion
of the foramina of the 4th ventricle or for-
ward displacement of the vallecula by an
arachnoid cyst may preclude filling of the
4th ventricle during an air study by the lum-
bar route; or the cyst may push the upper
ermis forward and upward producing aque-
ductal occlusion and preventing passage of
contrast medium from the third ventricle.

Sometimes these difficulties may be sur-

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**Fig. 5. Case 3. Postoperative angiographic study.** *Upper Left:* Anteroposterior arteriogram showing the reduced distance between the P-2 segments of both posterior cerebral arteries and their occipital branches. *Upper Right:* Lateral arteriogram showing the supratentorial vascular pattern to be grossly normal. However, the upper part of the basilar artery is flattened, as well as segments P-1 and P-2 of both cerebral arteries, indicating posteroinferior migration of the brain stem through the tentorial notch. The posterior inferior cerebellar artery is now seen running parallel to the occipital bone and very near to it, indicating posterior mi-
gration of the vermis. *Lower Right:* Lateral phlebogram showing the new position of the thalamus, the location of dural
sinuses remaining unchanged. The infraten-
torial space is markedly avascular.
mounted by injection of the contrast medium directly into the cyst. However, all of these examinations risk interference with fluid pathways and harmful alterations in intracranial pressure dynamics.

When a radionuclide injection of the ventricle fills the cyst without visualization of the subarachnoid space, the diagnosis of a Dandy-Walker cyst is supported. If the cyst fills from a radionuclide subarachnoid injection, an arachnoid cyst is more likely. Since recent evidence indicates that in some cases of congenital obstructive hydrocephalus the subarachnoid space is normally formed but mechanically compressed and that a temporary shunt may allow re-expansion of this functioning space, the studies just described may help candidates for definitive surgery after a temporary shunt.

Angiography can demonstrate the cystic

Fig. 6. Case 4. Preoperative and postoperative angiographic studies. Upper Left: Preoperative lateral arteriogram indicating supratentorial hydrocephalus and upward deviation of the posterior cerebral arteries. The posterior inferior cerebellar artery is normal size and its vermian branch appears displaced forward and upward, indicating similar displacement of the posterior vermis. Upper Right: Preoperative lateral phlebogram showing the high position of the torcular Herophili and transverse sinuses, and forward displacement of the thalamic image and the posterior vermis; the latter is outlined by the inferior vermian vein and a large avascular area. Lower Left: Postoperative lateral arteriogram showing great changes in the vascular pattern. There is no supratentorial hydrocephalus. The upper part of the basilar artery is deviated backward and the P-1, 2, 3 segments of the posterior cerebral arteries are flattened. All findings indicate posteriorinferior migration of the brain stem through the tentorial notch. The posterior inferior cerebellar artery now follows a course parallel and near to the occipital bone. The vermian branch of the artery indicates the new position of the posterior vermis, well above the inion. Lower Right: Postoperative lateral phlebogram, showing a posterior shift of the thalamus and of the cerebellar vermis, well outlined by the inferior vermian vein (arrows).
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Fig. 7. Case 5. Upper Left: Anteroposterior arteriogram indicating supratentorial hydrocephalus and a midline position of the vermian branch of posterior inferior cerebellar artery. Upper Right: Lateral arteriogram showing a parallel course between the middle and posterior cerebral arteries, indicating an expanding lesion within the posterior fossa. The posterior inferior cerebellar artery is pushed forward and upward; there is evidence of a wide avascular area within the posterior fossa. Lower Right: Lateral phlebogram. The infratentorial cyst is quite evident, as well as the forward and upward deviation of the inferior vermis vein. The straight sinus is elevated and the torcular Herophili is above the inion, indicating the congenital origin of the lesion.

avascular area, its location with respect to the 4th ventricle, and the deformity of the posterior vermis; this requires consideration and visualization of the course and size of the posterior inferior cerebellar artery and its vermian branch. In infancy, this is normally the largest among the cerebellar arteries and is well-visualized on anteroposterior, lateral, and submentovertical views. Moreover, its situation is strategic for recognition of infratentorial masses. In the lateral view the choroid arc points to the roof of the 4th ventricle (while the midline position of its vermian branch occupies a midline position and supplies the posterior vermis).

In each of our three cases of arachnoid cyst an otherwise normal posterior inferior cerebellar artery had been carried forward and upward by the displacement of the 4th ventricle and cerebellar hemispheres. In the case of acquired Dandy-Walker cyst this artery was displaced downward and the choroid arc or apex of the cranial loop was displaced backward, indicating an enlarged 4th ventricle. In our two cases of Dandy-Walker cyst this vessel was miniature, deviated
Fig. 8. Case 6. Upper Left: Anteroposterior arteriogram indicating supratentorial hydrocephalus; note the well-formed posterior-inferior cerebellar artery (arrows) whose choroid arc is in the midline. Upper Right: Lateral arteriogram showing evidence of both supratentorial and infratentorial hydrocephalus. The latter is outlined by the parallel course of the middle and posterior cerebral arteries. Moreover, the posterior-inferior cerebellar artery (faintly visualized) is displaced backward and downward, indicating an enlarged 4th ventricle. A 4th ventricle tumor is ruled out by the normal course of the anterior superior cerebellar arteries. Lower Right: Lateral phlebogram showing a normal relationship of the thalamus to the straight sinus. The superior vermian vein outlines the upper limit of the vermis and cerebellar hemispheres; the avascular area outlined within the cerebellar blush offers evidence of the size of the 4th ventricle. A markedly enlarged 4th ventricle was documented by opaque ventriculography.

downward, and had no vermian branch, all reflecting the enlarged 4th ventricle and absent posterior vermis.

These basic differential angiographic criteria prove valid when applied to pertinent cases reported by others in which surgery, pneumoencephalography, or ventriculography are available for review.\textsuperscript{3,26} Moreover, the operative photograph of the case of arachnoid cyst reported in 1961 by Cassinari and Migliore,\textsuperscript{3} which had no angiography but whose air study resembles that of our Case 3, shows a well-developed vermis with a cyst dorsal to the 4th ventricle, and thus adds indirect evidence of the diagnostic accuracy of the above criteria. On the other hand, it is clear that some angiographic diagnoses reported as Dandy-Walker cysts, but with inadequate verification, are obvious instances of arachnoid cyst since the posterior inferior cerebellar artery is clearly shown displaced forward and upward.\textsuperscript{5,23}

Another angiographic finding of diagnostic significance concerns the inferior vermian vein, a vessel which is seen on 85% of normal pediatric angiographic studies.\textsuperscript{16} This vein was well visualized as displaced forward and upward in our cases of arachnoid cyst, but was absent in the cases of Dandy-Walker cyst. Moreover, this vein is absent in other well-documented cases of Dandy-Walker cyst\textsuperscript{5,10,26} and fully visualized but anterosu-
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...displaced in cases of arachnoid cyst and in the cases we have quoted as instances of erroneous diagnosis of Dandy-Walker cyst.

Other angiographic findings, like the course of the anterior superior cerebellar arteries and the size and position of the cerebellar blush, have been implicated in the differentiation under discussion. Our experience suggests that forward and upward displacement of the anterior superior cerebellar arteries as well as displacement and decreased size of the cerebellar blush are not characteristic of the Dandy-Walker cyst alone. Both kinds of cysts are able to produce anterosuperior displacement of the cerebellar hemispheres and upper vermis and reduction of the size of the cerebellar blush. The latter may not only depend on impaired injection within the field of the posterior inferior cerebellar artery, but on other factors such as the size of the cyst and the degree of the infratentorial pressure.

Postoperative roentgenological evaluation of the pathology under discussion has received little attention and postoperative angiography none at all. In our two cases of arachnoid cyst in which postoperative angiography was performed, there was evidence that the hydrocephalus had disappeared. The vertebrobasilar system showed anatomical changes following the relief of pressure from the cyst. Backward displacement of the upper part of the basilar artery, backward deviation of segments P-1, P-2, P-3 of the posterior cerebral arteries, and backward deviation of the precentral cerebellar vein are findings indicating posteroinferior migration of the brain stem through the tentorial notch. The course of the posterior inferior cerebellar artery and of the inferior vermian vein revealed the new position of the posterior vermis, well above the inion. The postoperative cerebellar blush appeared flattened, with a large sagittal diameter.

References

3. Cassinari V, Migliore A: (On median cystic formations of the posterior cranial fossa (observations apropos of a case)). Minerva Neurochir 5:71-76, 1961 (It)
14. La Torre E, Occhipinti E: Angiographic findings in some malformations of the brain: report of seven cases. Europ Neurol 4:210-225, 1970
20. Moss ML, Noback CR, Robertson GG:


22. Pia HW: (Pseudocysts of the cisterna cerebel-lomedullaris. A congenital dilation of the cisterna and its relation to cerebellar dysraphias.) Deutsch Z Nervenheilk 184:1–22, 1962 (Ger)


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