Atresia of the Foramina of Luschka and Magendie: The Dandy-Walker Cyst

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As of 1942, when Taggart and Walker reported three patients with atresia of the foramina of Luschka and Magendie, only six previous cases had appeared in the literature, and only two of these cases had undergone surgical intervention. In the majority of cases published since then, the diagnosis was made either at operation or at postmortem examination.

Treatment consists of cyst resection after posterior fossa exploration, or shunting from the lateral or third ventricle. In 1956, Matson outlined the plain skull film, ventriculographic, and dural sinographic characteristics of prenatal obstruction of the fourth ventricle and emphasized that "intelligent and successful treatment of hydrocephalus depends upon an accurate knowledge of the location and type of obstruction to normal cerebrospinal fluid movement."16

Both the embryologic background and the specific pathological changes associated with this particular clinical entity are controversial. Taggart and Walker maintained that failure in development of the foramina of Luschka and Magendie, in the fourth month of fetal life, produced hydrocephalic enlargement of the fourth ventricle. This, in turn, precluded the development of the inferior vermis and prevented the normal descent of the torcular Herophili and lateral sinuses. Brodal and Hauglie-Hanssen pointed out that the cerebellar anlagen fuse long before the fourth fetal month, and that thus the cerebellar agenesis could not be the result of the later foraminal atresia. They concluded that the entire process was precipitated by a previously existing hydrocephalus of unknown cause. The description of cerebellar embryological development, as outlined by Dow and Moruzzi, supports this concept.

Benda suggested that atresia of the developing fourth ventricular foramina was only a part of the syndrome and not its cause, because in some cases reported, the foramina were found to be patent. He stated that the main pathological process was the "meningomyelocele" sac-like dilatation in place of the posterior medullary velum which results in a cleft cerebellum and hydrocephalus. Gibson, through careful examination of the cystic membranes, concluded that there is no true cerebellar agenesis or aplasia and, therefore, that the whole process may well be secondary to persistent closure of the foramina of Luschka and Magendie. D'Agostino, et al., in a review of their pathological material, came to the same conclusion.

Gardner included the Dandy-Walker cyst among many dysgeneses, all the result of persistent fetal hydrocephalus secondary to impeded flow of cerebrospinal fluid through the attenuated roof of the fourth ventricle. The foraminal atresia mentioned by others, according to him, was therefore merely a more remarkable example of the impermeability of the rhombic roof.

For the purposes of this paper we shall avoid this controversy and consider the basic clinical problem to be the presence of a posterior fossa cyst which is entirely within the confines of the fourth ventricle, and which is caused by complete occlusion of the foramina of Luschka and Magendie. We shall refer to this clinical entity as the Dandy-Walker cyst.

Between the years 1963 and 1968, of 212 hydrocephalic infants diagnosed at Cook County Hospital, eight were found to have...
Dandy-Walker cysts. This group constitutes our clinical series, together with one case of arachnoidal cyst of the cisterna magna which we have added for comparison. We shall describe the salient clinical, neuroradiological, and anatomico-pathological features of these cases, and emphasize their correlation.

**Case Reports**

*Case 1.* A diagnosis of Dandy-Walker cyst was made on this white baby girl at 2½ months of age. Plain skull films revealed that the squamous occipital bone was bulging and that the lambdoidal sutures were maximally split. Ventriculography revealed symmetrical hydrocephalus with a massive fourth ventricle, and free communication between the third and fourth ventricles. The aqueduct was enlarged. A right ventriculo-atrial shunt was performed; the lateral ventricle pressure was 220 mm H₂O.

At 16 months of age she began to develop posterior fossa signs: bilateral nystagmus on lateral gaze, explosive speech, and truncal ataxia. At 18 months she had a broad-based, anteropulsive gait. Revision of the shunt at 19 months did not significantly alter the progression of posterior fossa symptomatology.

At 4 years of age, because of the progressive development of posterior fossa signs despite a well-functioning shunt system, the child underwent another complete neuroradiologic evaluation. These studies demonstrated moderately enlarged lateral ventricles, but the fourth ventricle was now enormous. Air, Conray 60,¹⁹ and Pantopaque* would not pass from the third into the fourth ventricle or vice versa. Since contrast studies performed at birth had revealed free passage of air between the third and fourth ventricles, it was concluded that she had developed a secondary aqueductal occlusion. Therefore, a fourth ventricle peritoneal shunt was inserted; pressure in the fourth ventricle at this time was 150 mm H₂O, and the fluid was xanthochromic. The lateral ventricles contained clear fluid at a pressure of 130 mm H₂O.

*Generic and trade names of contrast media: Conray 60 = Methylglucamine iothalamate 60%; Hypaque 50% = Sodium diatrizoate 50%; and Pantopaque, Myodil = Ethyl iodophenyundicylate.

Subsequently, the child made a dramatic recovery. She became stable on her feet, able to walk without anteropulsion, and steady enough with both hands to feed herself without spilling liquids. Her speech became understandable. Subsequent contrast studies demonstrated that air passed freely from the third ventricle through the patent aqueduct into the encysted fourth ventricle.

*Case 2.* This Negro girl was admitted at the age of 2 months with a history of progressive enlargement of the head. Examination revealed split sutures, bulging fontanels, and a prominent occipital bone. Cerebral angiography (Figs. 1-4) demonstrated changes characteristic of a Dandy-Walker cyst and of agenesis of the corpus callosum. Lateral ventriculography, performed after a lateral ventricle peritoneal shunt, confirmed these diagnoses. The pressure in the lateral ventricle was 210 mm H₂O, and the fluid was clear. No fourth ventricle recording was made.

Nine months later, another shunt revision was performed. The lateral ventricle pressure at this time was 160 mm H₂O. The child has done well during the past 8 months.

*Case 3.* This white girl was born with an occipital encephalocele. Her parents refused surgery until she was 6 months old when a ventriculo-atrial shunt and closure were accomplished without antecedent angiography or pneumoencephalography. Over the following 3 months, her head continued to get larger.

She was admitted to Cook County Hospital when she was 9 months old because of progressive enlargement of her head. She was found to have bilateral optic atrophy, searching nystagmus, the "setting sun" phenomenon, and a markedly enlarged head. The squamous occipital bone was bulging, and the lambdoidal sutures were widely split. Angiographic and pneumographic diagnosis of a Dandy-Walker cyst was made. The lateral ventriculo-atrial shunt was replaced by a ventriculo-peritoneal shunt. The fluid in the lateral ventricles was clear, and the pressure was 270 mm H₂O.

Postoperatively, she developed persistent vomiting in spite of a sunken anterior fontanel. We then removed the lateral ventricle
shunt and performed a fourth ventricle peritoneal shunt, reasoning that it was most logical to drain the Dandy-Walker cyst. The fluid in the fourth ventricle was xanthochromic, and the pressure was 300 mm H₂O. After this procedure the fontanels bulged, and the child developed a hypothalamic syndrome. We began steroid therapy and reinserted the lateral ventriculo-peritoneal shunt. (At this time the child had both lateral and fourth ventricle peritoneal shunts.)

She improved initially, but then developed opisthotonos and gastro-intestinal hemorrhages. An Asambuja procedure (third ventriculogram) demonstrated complete occlusion of the aqueduct (Fig. 5). Posterior fossa exploration revealed occlusion of the foramen of Magendie, a massive Dandy-Walker cyst, and a patent aqueduct that measured approximately 1 cm in diameter. This visual proof of a patent aqueduct in a child with neuroradiologic evidence of occlusion of the
Fig. 3. Case 2. This right brachial angiogram, lateral arterial phase, demonstrating the elevation of the cerebellar hemispheres and the superior displacement of the posterior cerebral arteries and the occipital lobes. It also illustrates the large cystic defect both in the posterior fossa and beneath the occipital lobes, and the characteristic, meandering course of the anterior cerebral artery, which is diagnostic of dysgenesis of the corpus callosum.

Fig. 4. Case 2. The veins over the anteriorly and superiorly displaced cerebellar hemispheres are well-visualized. The thalamic image is displaced anteriorly and superiorly and the internal cerebral vein is lengthened. Notice the cloven vein of Galen, and the posterior-superior course which this vessel follows as it goes to join the straight sinus at the apex of the tentorium. The severe superior displacement of the tentorium results in shortening of its posterior surface and widening of the tentorial incisure. It is through this widened tentorial incisure that extensions of the encysted fourth ventricle protrude to enter the supratentorial compartment.

aqueduct led us to conclude that the obstruction was extrinsic, secondary to enlargement of the fourth ventricle.

The child got steadily worse and died at 14 months of age.

Autopsy examination showed that the meninges were clear and colorless, with no gross evidence of meningitis. Both lateral ventricles, the third ventricle, and the aqueduct of Sylvius (which measured 1 cm in diameter), were enlarged. The fourth ventricle was huge, flattening both the pons and medulla. The choroid plexus of the fourth ventricle was reduced in size and did not extend into the subarachnoid space of the cerebellopontine angle. The two cerebellar hemispheres were separated widely, and between them was a thin membrane covering the fourth ventricle. The inferior third of the cerebellar vermis was dysgenetic, represented only by a thick, membranous, gliotic structure. The superior two-thirds of the cerebellar vermis was displaced superiorly and anteriorly through the tentorial opening.

Case 4. When this Negro boy was born spontaneously after 34 weeks gestation, he weighed 4 lbs 3 ozs. He was noted to have low-set ears, bulging fontanels, micrognathia, and a symmetrical, bulging, squamous occipital bone indicative of a mass within the posterior fossa. He had intermittent episodes
of respiratory arrest, and his feedings were regurgitated through the nose and mouth. On the fourth day of life, a large right-sided abdominal mass was palpated. An intravenous pyelogram demonstrated non-visualization of the right kidney, and barium enema was consistent with a large right-sided mass.

Lumbar puncture revealed cloudy cerebrospinal fluid with 8 lymphocytes per cu mm, under "high pressure with elevated protein levels." Plain skull films, cerebral angiography, and brain scan were diagnostic of a Dandy-Walker cyst. That same night, under local analgesia, a lateral ventricle peritoneal shunt was performed. The fluid was clear and the ventricular pressure was 190 mm H₂O. The following morning the child died immediately after an episode of medullary compression secondary to foraminal impaction.³²

Autopsy revealed a Dandy-Walker cyst with occluded foramina of Luschka and Magendie, a patent aqueduct, and an elevated tentorium with an enlarged tentorial opening. The vermis was compressing the quadrigeminal plate, and a portion of the encysted fourth ventricle was wedged between the occipital lobes. The right-sided abdominal

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Fig. 5. Upper photographs: Third ventriculogram (Asambuja procedure) using gas (left), and Pantopaque (right), show functional occlusion of the aqueduct in a child whose aqueduct actually measured 1.0 cm across (Case 3). Lower photographs: Lateral ventriculography in Case 1 revealed displacement of the culmen of the vermis superiorly through the tentorial opening and into the third ventricle and no passage of gas into the aqueduct. The functional occlusion of the aqueduct is demonstrated in a direct puncture of the Dandy-Walker cyst through the atlanto-occipital membrane, performed in the same child 1 week after the lateral ventriculogram. The gas in the lateral ventricle is residual of the lateral ventricle study, and was so documented with preliminary films.
mass was found to be a huge polycystic kidney.

Case 5. This Negro girl, born at term, had breathing difficulties at birth. Because of a high-pitched respiratory stridor and severe retractions, bilateral vocal cord paralysis was suspected. This was confirmed by laryngoscopy. Following a tracheostomy the child improved dramatically. At 3 weeks of age, the head was noted to be enlarged. There was an elevated external occipital protuberance, bulging squamous occipital bones, bulging fontanels, and split sutures. The lambdoidal sutures were maximally split. Cerebral angiography revealed the presence of a bilateral subdural collection of fluid, agenesis of the corpus callosum, and a Dandy-Walker cyst. The brain scan was suggestive of a cystic mass in the posterior fossa. A serial Conray 60 fourth ventriculogram revealed the presence of a cystic enlargement of the fourth ventricle and no communication between the third and fourth ventricles (Fig. 6).

A shunt between the cystic fourth ventricle and the peritoneal cavity was performed the same night. The fluid was xanthochromic and the pressure 175 mm H₂O. The vital signs improved immediately following the shunt, but feeding problems continued. A gastrostomy was performed. She became steadily worse, with recurrent respiratory infections, and died when 4 months old.

Autopsy examination showed a subdural collection of purulent material over both frontal lobes. The corpus callosum was absent, and the two cerebral hemispheres were held together by frayed strands of the roof of an enlarged third ventricle. The cerebellar vermis appeared well-developed in its superior half but dysgenetic inferiorly. The cerebellar hemispheres were widely separated and displaced superiorly and laterally (Fig. 7). The floor of the fourth ventricle was flattened, and there was maximal dilation of the lateral recesses and stretching of the ninth and tenth cranial nerves. The fibrous tissue bridging across the wall of the encysted fourth ventricle was quite distinct from the dura mater, but it was not possible to identify a cisterna magna. The foramina of Luschka and Magendie were occluded.

The lateral ventricles were dilated and displaced lateralward and inferiorly, separated by the superior expansion of the third ventricle. The aqueduct was patent and mea-
sured 4 mm at its widest point. The fourth ventricle was transformed into a massive cyst (which contained 45 cc of fluid) which occupied almost the entire posterior fossa, displacing the brain stem forward and flattening the pons against the clivus. The superior vermis protruded into the enlarged suprapineal recess. In the brain stem, it was noted that the small volume of the pons made the pontine and medullary nuclei unusually conspicuous. The vocal cord palsy was considered to be the result of the hind brain compression caused by the encysted fourth ventricle.

Case 6. This Negro boy was brought to Cook County Hospital at 2 months of age because of steady increase in head size. At that time, there was a setting sun phenomenon, bilateral atrophy of the optic discs, split sutures (the lambdoidal were most widely split), and an elevated external occipital protuberance. Cerebral angiography revealed findings compatible with a diagnosis of Dandy-Walker cyst and agenesis of the corpus callosum. Lateral and fourth ventricle peritoneal shunts were inserted at the same procedure. The lateral ventricle pressure was 170 mm H2O and the fluid xanthochromic.

Postoperatively the patient did extremely well, and within a few days the “setting-sun” phenomenon disappeared. One week later, Conray 60 lateral ventriculography revealed aqueductal occlusion and the diagnosis of “functional” extrinsic occlusion of the aqueduct secondary to the Dandy-Walker cyst was made. On the 38th hospital day, a Conray 60 ventriculogram of the fourth ventricle confirmed the angiographic diagnosis of cystic enlargement of the fourth ventricle as well as that of “functional” aqueductal occlusion. On the 45th hospital day, the child was discharged in perfectly normal condition. Follow-up at 6 months revealed normal development.

Case 7. This Negro girl was born in Cook County Hospital after a 32-week gestation. At birth she was found to have an enlarged head, bulging anterior and posterior fontanels, and splitting of all sutures. The coronal and sagittal sutures were more widely opened than were the lambdoidal sutures. Following a clinical diagnosis of hydrocephalus, cerebral angiography was performed and a neuroradiologic diagnosis of communicating hydrocephalus and agenesis of the corpus callosum was made.

On the third day of life, a lateral ventricle peritoneal shunt was performed under local anesthesia. The lateral ventricular pressure was 260 mm of H2O, and the cerebrospinal fluid was crystal clear. Following surgery, the child did well initially in that her cry became healthy and her sucking responses quite strong. On the 10th postoperative day, however, the child died during an episode of apparent foraminal impaction.

At autopsy the two cerebral hemispheres were found joined at the cingulate gyrus by a thin membrane (Figs. 8 and 9), and the two lateral ventricles were markedly enlarged. There was no septum pellucidum, and the corpus callosum was represented by a transparent membrane. Transverse section at the level of the cerebral peduncles revealed the aqueductal iter to measure 1.0 × 0.4 cm. The cerebellar hemispheres were widely separated by a cleft which measured 5 cm at its widest point. The vermis of the cerebellum appeared dysgenetic, much smaller than normal, and elevated. Inferiorly, the vermis was represented by a thin membrane which extended from one cerebellar hemisphere to the other. The fourth ventricle was encysted and measured 4 × 2 cm, and the pons was flattened. The foramina of Luschka and Magendie were occluded. The choroid plexus was present but small. The membranous transformation of the inferior vermis, which formed the roof of the fourth ventricle, was frayed, permitting escape of cerebrospinal fluid.

We diagnosed this case as communicating hydrocephalus and were unable to diagnose Dandy-Walker cyst angiographically before autopsy. In his autopsy report, Dr. Orville Bailey stated that “the perforation of the membranous roof of the fourth ventricle may represent the explanation for the failure of the torcular Herophili and lateral and straight sinuses to be displaced superiorly.”

Case 8. This 6-month-old Negro boy was born at Cook County Hospital after a normal full-term pregnancy and delivery. His early progress was unremarkable but during
Fig. 8. Case 7. Photographs and labeled drawing of the brain of a child with a Dandy-Walker cyst, as seen from the superior aspect. In this child there was the associated anomaly of agenesis of the corpus callosum. Upper Left: Anteriorly, the two cerebral hemispheres are held together by bridging arachnoid and at the center by the roof of the third ventricle. The two cerebellar hemispheres are seen separated by a dysgenetic vermis. There is a probe passing from the cystic fourth ventricle through the dilated aqueduct and into the enlarged third ventricle. The roof of the third ventricle ruptured during life. Upper Right: Note severe cystic distention of the lateral recess of the fourth ventricle and thickened pia arachnoid bridging from the superomedial surface of the left cerebellar hemisphere (which is itself displaced superiorly and lateralward) across to the inferomedial surface of the left occipital lobe. This illustrates how portions of the cystic distension of the fourth ventricle may extend into the supratentorial compartment through the enlarged opening in the displaced tentorium.
the 4 months prior to admission, progressive asymmetrical head enlargement was noted. Examination revealed a large head with split sutures, bulging anterior and posterior fontanels, with asymmetrically bulging squamous occipital bones. He had spontaneous nystagmus in lateral and vertical gaze. Cerebral angiography revealed changes characteristic of a Dandy-Walker cyst. This picture was complicated, however, by bilateral supratentorial extensions of the posterior fossa cyst which resulted in elevation of the middle cerebral arteries.

Supra- and infratentorial ventricle-peritoneal shunting procedures were performed simultaneously. Because of technical difficulties, it was not possible to record a reliable pressure in either the lateral or fourth ventricles. However, the pressure was interpreted as high in both, and there were synchronous oscillations in the tubings placed into each of these compartments. The fluid was clear in both the fourth and lateral ventricles.

After this, the child was able to hold up his head, and the nystagmus disappeared. Within 2 weeks he was able to hold his spine erect without weaving. The posterior fossa shunt had to be revised three times over the first 3-month period. These episodes of posterior fossa shunt malfunction were announced by symptomatology reminiscent of the hypothalamic syndrome, in spite of persistently sunken fontanels. At the age of 9 months, the child is now progressing well, is much more alert, and gaining weight. He continues to have unexplained episodes of low-grade fever.

Case 9. This Negro boy was admitted to Cook County Hospital at 3 months of age for evaluation of his enlarged head. The mother stated that she first noticed this and irritability 1 week prior to admission. Clinical examination revealed a large head; bulging anterior fontanel; split coronal, sagittal, and lambdoidal sutures; and a high-pitched cry. There were no posterior fossa signs. Subsequent to cerebral angiography, a diagnosis of communicating hydrocephalus was made. A lateral ventricle peritoneal shunt was performed on the 5th hospital day. The CSF in the lateral ventricle was clear and the pressure recorded only as being high. Postoperatively the child did well.

At 7 months the child was readmitted because of nausea, vomiting, severe nystagmus, progressive lethargy, and a withdrawal from his surroundings. Re-examination of his initial cerebral angiogram resulted in the conclusion that the child had a Dandy-Walker cyst and not simple communicating hydrocephalus. Pneumography performed at this time revealed free communication between the third ventricle and a large cyst in the posterior fossa. It was concluded that this large posterior fossa cyst was a cystic fourth ventricle and, therefore, that it represented a Dandy-Walker cyst. Consequently, lateral and fourth ventricle peritoneal shunts were inserted simultaneously. The lateral ventricle
pressure was not recorded at this time, but the CSF was noted to be crystal clear. The fluid in the posterior fossa cyst was xanthochromic, and the pressure was stated to be high. Postoperatively, the nausea, vomiting, and irritability cleared, the nystagmus diminished markedly, and the child began to relate to his environment.

At 17 months of age, it was necessary to revise the shunt. The child did not improve following this shunt revision, despite the fact that the shunts continued to function well. Conray 60 ventriculography at this juncture revealed marked dilatation of the lateral and third ventricles (Fig. 10), but a normal fourth ventricle. It also showed that there was an enormous cyst of the cisterna magna, which extended into the supratentorial compartment. Air injected into the lumbar subarachnoid space did not enter either the cyst or the ventricular system, and Conray 60 and air injected into the cystic cisterna magna did not go below the foramen magnum. A posterior fossa craniotomy confirmed the presence of a cyst of the cisterna magna. The cyst was resected, and communication between the cisterna magna, the cerebellomedullary cisterns, and the cervical subarachnoid space established. The fourth ventricle peritoneal shunt was removed and the lateral ventricle peritoneal shunt left in place. Postoperatively, the child progressed normally but remained dependent upon the ventriculo-peritoneal shunt.

Clinical Summary of Nine Cases

1. All cases were newborn, except two who were 9 and 6 months old respectively at the time of diagnosis. Five cases showed paralysis of one or more of the lower cranial nerves. Six cases presented with bulging of the squamous occipital bone, and severe splitting of the lambdoidal sutures.

2. All cases but one were diagnosed as Dandy-Walker cysts by cerebral angiography (Figs. 1–4) and then, after the shunting procedure was performed, confirmed with gas or serial Conray 60 ventriculography. The exception was diagnosed clinically as hydrocephalus and neuroradiographically as communicating hydrocephalus.

3. All cases brought to autopsy and all cases that underwent posterior fossa exploration demonstrated complete occlusion of the foramina of Luschka and Magendie (Fig. 8). In seven cases, there was ventriculographic or pneumoencephalographic evidence of occlusion of the foramina of Luschka and Magendie. In four cases there was supratentorial extension of the encysted fourth ventricle.

4. In no case did air enter the fourth ventricle when injected from the lumbar subarachnoid space, nor leave the fourth ventricle to enter the cisterna magna and basal cisterns when injected from above. All cases had encysted fourth ventricles.

5. In all cases, the inferior vermis was dysgenetic, and the cerebellar hemispheres were displaced upward and laterallyward (Figs. 7–9). In all cases but one, the torcular Herophili and lateral sinuses were elevated.

6. All cases had elevated pressure in the lateral ventricles at the time of shunting. In five cases the fourth ventricle pressures were recorded and found to be elevated within the same range as that in the lateral ventricles (Fig. 11).

7. Four cases had xanthochromic fluid in the fourth ventricle and crystal clear fluid in the lateral ventricles. These cases all had a "functional" occlusion of the Sylvian aqueduct.

8. In all cases of "functional" aqueductal occlusion, the aqueduct was enlarged, but neither air, Conray 60, nor Pantopaque passed through it from either the third or fourth ventricles (Fig. 5).

9. All cases had lateral ventricle shunts performed; in five, independent fourth ventricle shunts were also performed. Four cases presented with a clinical picture of hydrocephalus, then demonstrated a picture of posterior fossa tumor after the lateral ventricle shunt was performed.

10. In four cases there was an associated agenesis of the corpus callosum (Figs. 3, 8).

11. The child with the arachnoidal cyst of the cisterna magna demonstrated the
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Fig. 10. Case 9. Gas and Conray 60 ventriculograms in a child with a cyst of the cisterna magna. These studies were performed after lateral ventricle peritoneal and cisterna magna peritoneal shunts had been inserted. **Upper Left:** Air injected into the cystic cisterna magna shows a pattern indistinguishable from that seen in the Dandy-Walker cyst. **Upper Right:** Gas is seen within the supratentorial extension of this cyst. **Lower Left:** After Conray 60 was injected directly into the lateral ventricle, serial films show that both the lateral and third ventricles are dilated, but the fourth ventricle appears normal. **Lower Center:** Conray 60 is seen passing from the fourth ventricle through the vallecula into the massive cyst of the cisterna magna. **Lower Right:** The fourth ventricle is clear and the Conray trapped within the cystic cisterna magna.

angiographic and ventriculographic characteristics of the Dandy-Walker cyst. He also showed clinical signs of hydrocephalus and a posterior fossa mass (Fig. 10).

**Discussion**

Hydrocephalus represents the most obvious clinical observation in the newborn with the Dandy-Walker cyst. There are, however, significant findings in the examination of the skull, cranial nerves, and vital signs which are suggestive or pathognomonic of this congenital anomaly. In addition to this, we have observed that the signs and symptoms of a posterior fossa mass may be precipitated, or made considerably worse, by the performance of an isolated lateral ventricle shunt, without providing for simultaneous direct decompression of the fourth ventricle cyst.

In the newborn infant with atresia of the foramina of Luschka and Magendie who does not have a shunt, the fontanels all bulge equally and the sutures all split. However, the lambdoidal sutures may be disproportionately wider than either the coronal or the sagittal sutures. As a result, the squamous occipital bone bulges and has a “floating” feeling when palpated. The external occipital protuberance is high. Unfortunately, these changes in the skull are not constant.

When only a lateral ventricle shunt has been performed, the anterior fontanel becomes depressed and the sagittal and coronal sutures close. The lambdoidal suture may remain open or split subsequently, and the
squamous portion of the occipital bone may develop a marked concavity of its inner surface.

We have observed such posterior fossa and brain stem signs as nystagmus, paralysis of the fourth through the eleventh cranial nerves, explosive speech, truncal ataxia, and dysmetria in infants after lateral ventricle shunting. One child had bilateral ninth and tenth nerve paralysis at birth. Only in those children who developed a "functional" occlusion of the aqueduct did we observe signs indicative of compression of the quadrigeminal plate and hypothalamic insufficiency. Specifically, two of our eight children with Dandy-Walker cysts had clinical signs of a posterior fossa mass lesion at birth; in four of the remaining six cases these signs became obvious only after isolated lateral ventricle shunting, while in two cases the shunting was followed by signs of hypothalamic insufficiency.

In only one instance did the child do very well with only a lateral ventricle shunt, whereas two died, and one developed incapacitating posterior fossa signs.

The cystic enlargement of the fourth ventricle causes dorsolateral displacement of the cerebellar hemisphere (Figs. 7-9) anterosuperior displacement of the cerebellar vermis (Fig. 9) and ventral displacement of thepons and medulla (Figs. 7 and 9). Cystic distention of the lateral recesses of the fourth ventricle (Fig. 8) causes stretching of the ninth and tenth cranial nerves and flattening of the flocculus. We have observed adequate evidence to justify the conclusion that this enlargement of the fourth ventricle is responsible for the posterior fossa signs that develop. When there is an associated aplasia of the corpus callosum (Fig. 8), the roof of the third ventricle may rupture, thereby allowing the passage of CSF into the subdural space.

In all of our cases the aqueduct was enlarged, the lateral and third ventricles markedly dilated, and the CSF pressure in the lateral ventricles elevated. In addition, in those cases where there was no passage of contrast media through the aqueduct, we found xanthochromic fluid in the encysted fourth ventricle and crystal clear fluid in the lateral ventricle. Therefore, at birth the child with a Dandy-Walker cyst has free communication between the third and fourth ventricles, but he may subsequently develop a "functional" occlusion of the aqueduct. This is secondary to disproportionate increase in size of the fourth ventricle, as compared to the third ventricle, resulting in displacement of either the superior vermis or an extension of the fourth ventricle cyst through the tentorial opening to compress the quadrigeminal plate.

The association of the Dandy-Walker cyst with an agenesis of the corpus callosum has been noted by Van Epps and Whitten, et al. Kirschbaum states that agenesis of the corpus callosum occurs during the third month of gestation secondary to some exogenous factor, and that lipomata and arachnoidal cysts at this site are probably due to changes in the arachnoid of similar etiology. Zettner and Netsky reported lipomas of the corpus callosum in this age group.

It has been stated that if the inner surface of the squamous occipital bone is concave on the lateral skull films, and if the external occipital protuberance is elevated, one may diagnose a large posterior fossa. This radiographic evidence was present in only three of our eight cases.

The angiographic findings in the Dandy-Walker cyst are characteristic, and were present in seven of the eight cases herein reported. They consist of arterial and venous deformity and displacement, and dural sinus displacement.

In the anteroposterior projection, the arterial changes consist of a wide sweep of the posterior cerebral arteries in their course about the enlarged midbrain, and in failure of their internal occipital branches to approach the midline posteriorly (Fig. 8). The former is the result of a midbrain distended by an enlarged aqueduct and the latter of the insinuation of a portion of the encysted ventricle between the posterior cerebri as they pass on to the medial surface of the occipital lobes. In addition to the general characteristics of hydrocephalus, the anteroposterior venous phases reveal a marked elevation of the torcular Herophili and its effluent lateral sinuses which pass symmetrically infero-laterally to the sigmoid sinuses, thereby describing two sides of an isosceles triangle (Fig. 2).

In the lateral projection the posterior cerebral arteries follow a postero-superior course as they run around the brain stem.
and over the superiorly displaced tentorium. The superior, and posterior inferior cerebellar arteries outline the superior displacement of the cerebellar hemispheres and vermis (Fig. 3).

In both the arterial and venous phases (Figs. 3 and 4) a large avascular area indicates the location and extent of the encysted fourth ventricle. In addition to this, the venous phases (Fig. 4) reveal the elevation of the lateral sinuses and of the torcular Herophilus, plus an inversion of the postero-inferior course of the straight sinus as it proceeds to enter the superiorly displaced torcular. The thalamic image, as indicated by the internal cerebral vein, the thalamostriate vein, and the vein of Rosenthal, is displaced superiorly and forward. This results in lengthening of the vein of Galen, which may appear to be cloven.

To illustrate the difficulty of being certain of the roentgenographic diagnosis of Dandy-Walker cyst, we have included the case of a child with a cyst of the cisterna magna (Case 9), whose angiographic and pneumographic studies were indistinguishable from those of a Dandy-Walker cyst (Fig. 10, upper photographs). A serial Conray 60 ventriculogram (Fig. 10, lower photographs) revealed a normal fourth ventricle and cerebellar vermis, but a large encysted cisterna magna which did not communicate with the spinal subarachnoid space. Resection of the cyst resulted in a cure of the child’s posterior fossa syndrome.

Techniumm99 brain scan may reveal a clear area in the posterior fossa and a superior displacement of the densities which correspond to the torcular Herophilus and lateral sinuses. This was noted in two of our eight cases; in the others, we were not able to make a tentative diagnosis of Dandy-Walker cyst using scanning techniques.

In three of our cases, air injected into the lateral ventricles passed immediately from the third ventricle into the cystic fourth ventricle without permitting visualization of the aqueduct. Subsequent gas and Pantopaque ventriculograms, however, performed 2 weeks, 6 months, and 3 years after lateral ventricular shunts had been inserted, revealed complete obstruction to the passage of air from the third ventricle into the aqueduct.

The observation of aqueductal occlusion in one of the children (Case 3) led us to perform a posterior fossa exploration. We found that the foramen of Magendie was occluded, as were the foramina of Luschka. The fourth ventricle was transformed into a massive cyst, flattening the pons and medulla. The aqueduct was patent and measured 1 cm across, so that it was possible to look directly into the third ventricle. The superior displacement of the vermis through the tentorial opening was interpreted as the cause of this “functional” occlusion of the aqueduct.

Regarding this occurrence, Taggart and Walker stated that “there was slight but definite dilatation of the aqueduct of Sylvius. The failure of the fourth ventricle to fill must be due to a mechanical block of the aqueduct, probably the result of compression by the anterior part of the vermis. Such a compression might conceivably occur from sudden relief of pressure above the tentorium, the anterior vermis acting as a valve as the fluid from the fourth ventricle attempted to escape.” This is roentgenographically demonstrated in Figs. 5 and 6 and diagrammatically represented in Fig. 12. In both cases, a fourth ventricle peritoneal shunt was performed on the children. Subse-
sequently, air injected into the lateral ventricle again passed freely from the third into the fourth ventricle and vice versa.

Similar simultaneous lateral and fourth ventricular shunting procedures were performed on an additional five children with resultant disappearance of the posterior fossa clinical signs. These observations led us to conclude that the cystic transformation of the fourth ventricle is not only responsible for many of the posterior fossa signs which develop, but that it is also responsible for the upward displacement of the vermis, which in turn causes a "functional" occlusion of the aqueduct (Fig. 12). This process was either precipitated or aggravated by the performance of an isolated lateral ventricle peritoneal shunt. All but one of the cases were treated this way.

Taggart and Walker, and D'Agostino, et al., warn against the adverse effects of gas ventriculography in children with the Dandy-Walker cyst. One can readily see how alterations in intracranial pressure dynamics, thus produced, would facilitate the valvular action of the dysgenetic vermis precipitating "functional" aqueductal occlusion. It is for this reason that we now shunt our patients on the basis of angiographic evidence and postpone gas ventriculography until the supra and infratentorial compartments are both safely shunted.

Our experience with infantile Dandy-Walker cysts indicates that the presence of adhesive arachnoiditis in the basal cisterns renders these subarachnoid pathways incapable of adequately handling CSF. The anatomical basis for this was described by Russell,21 who predicted that CSF could not be properly circulated through these spaces. Therefore, we conclude that craniotomy, and either opening of the fourth ventricle into the cisterna magna or third ventriculostomy, are of no therapeutic value.

The majority of our cases were Negro (seven of nine), since about 85% of the patients at Cook County Hospital are Negro.

Summary

Eight cases of clinically diagnosed Dandy-Walker cyst in newborn infants have been presented. The salient clinical, neuroradiological, and anatomico-pathological features of these cases have been described and the correlation of the three emphasized. Associated agenesis of the corpus callosum and subdural effusions have been discussed. We have concluded that alterations and displacements of cerebellar structures and the encysted fourth ventricle cause a "functional" occlusion of the aqueduct of Sylvius. The evidence for this has been presented, and particular reference given to its surgical implications. We believe that a combination of fourth and lateral ventricle shunts is the recommended treatment for Dandy-Walker cysts.

Fig. 12. Diagram of the superior displacement of the dysgenetic vermis, caused by continued expansion of the fourth ventricle, which may result in occlusion of the aqueduct of Sylvius. The aqueduct is patent, whereas the displaced vermis has compressed it.
References


