# Technologist Case Report

## Graphic Demonstration of a Dandy-Walker Cyst by the Use of Radionuclide Scan With $^{99m}$Tc-GHT

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***A 4-month-old male infant with a diagnosis of progressive hydrocephalus since birth received a radionuclide brain scan using $^{99m}$Tc-GHT. The study showed a very large posterior fossa with displacement of the transverse sinus. The diagnosis of a Dandy-Walker syndrome was substantiated by cerebral angiograms, pneumoencephalogram, and by axial tomography. Subsequent surgery proved difficult, with the ventriculoperitoneal shunt being placed and later removed. The shunt was then replaced and the infant's condition and prognosis improved after the last surgical procedure.***

The Dandy-Walker syndrome is a cerebella anomaly in which there is a cystic balloon-like dilatation of the fourth ventricle, which can be associated with hydrocephalic dilatation of the lateral and third ventricles. The foramens of Magendie and Luschka may be occluded or individually patent. The anterior part of the vermis is displaced upward but is otherwise normal. The posterior vermis is underdeveloped and it is continuous caudally with a membrane consisting internally of epidermis and externally of connective tissue. The large posterior fossa contents prevent the tentorium in making its normal descent during fetal life. This lack of tentorial descent produces a characteristic high position of the lateral sinuses *(1)*.

The Dandy-Walker syndrome may be detected by brain scanning. The brain scan demonstrates the presence of large intracranial cysts since they distort the normal anatomy and exhibit decreased isotopic activity *(2)*. The torcular Herophili and lateral sinuses are not visualized on skull roentgenograms before the age of 1 year and are seen in only approximately 50% of patients between the ages of 1 and 2 years. Brain scanning, with its lack of morbidity, constitutes an easy method for visualizing these structures.

The neuroradiologic findings associated with the presence of a Dandy-Walker cyst relate to the cystic dilation of the fourth ventricle, which accompanies atresia of the foramen of Magendie and, on occasion, atresia of the foramen of Luschka *(3)*.

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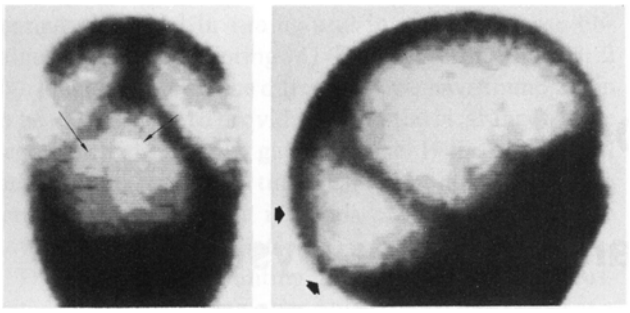
### Case Report

A 4-month-old male infant was admitted with a progressive enlargement of the posterior fossa, swelling over the right eye, accompanied with a loss of vision in that eye, and loss of use of right arm. He also suffered from the floppy-infant syndrome. The child was the product of a normal full-term pregnancy and a 24-year-Gravida III, Para II with 6 h of spontaneous labor. He was a breech presentation, but elective Cesarean section was done because of a previous Cesarean. In the postpartus period the child had some mild respiratory difficulties and was in an incubator the first day of life. He was initially described as having a poor suck. Growth and developmental parameters revealed the child only able to head follow a hand movement at 3 months of age. He held his head up at 1 month of age but was unable to roll over. The mother reports that as the occipital area increased, the child's growth patterns became retarded. At 4 months of age the infant was admitted to this facility for evaluation.

Physical examination revealed an extremely large head well beyond the 97% range, with a circumference of 46.7 cm *(4)*. General physical examination was unremarkable. No bruits were heard over the head but transillumination revealed marked increased illumination frontally, and especially on the right and the posterior fossa. The infant had marked head lag and followed rather poorly.

Two hours following the injection of 5 mCi of $^{99m}$Tc-GHT, six views of the brain were taken including a vertex and hyperflexion view of the posterior fossa, The brain scan revealed a large posterior fossa with superior displacement of the torcular herophili (Fig. 1). The brain scan was read out as grossly abnormal, and further correlation was needed to confirm the diagnosis of the Dandy-Walker syndrome versus a posterior fossa arachnoid cyst.



(A) (B)

**FIG. 1.** Posterior and R lat brain scan taken 2 h after $^{99m}$Tc-GHT injection. Arrows indicate large posterior fossa.

A subsequent EEG and echoencephalogram were read out as normal. A bilateral internal carotid and right vertebral angiogram were done. The positions of the cerebellar arteries, vein of Galen, straight sinus, and lateral sinuses are keys to correct angiographic diagnosis. Especially important in identifying the Dandy-Walker syndrome are the appearances of the vein of Galen and the position of the dural sinuses.

The normal vein of Galen is 1.5 to 2 cm long *(5)*. Stenosis of the vein of Galen may be elongated but it does not ascend greatly because of the straight sinus, which is a fixed structure. The straight sinus demarcates the junction line of the Falx cerebri and the tentorium cerebelli, while the lateral sinus outlines the lateral attachment of the tentorium to the skull. Thus, in the Dandy-Walker syndrome the failure of the tentorium to descend in fetal life determines the observed changes in the vein of Galen, straight sinus, and lateral sinus. The angiogram showed the anterior cerebral arteries as well as the sylvian angle to be displaced upward and forward (Fig. 2). The posterior temporal and midtemporal branches of the middle cerebral artery are also displaced upward and laterally to a similar degree as on the opposite side. There are double superior cerebella arteries bilaterally which are displaced upward. The distal branches of the superior cerebellar arteries do not appear to approximate the midline as they normally should. Both posterior cerebral arteries, filled from the right vertebral injection, appear to be displaced upward and also spread laterally in their distal course. There is a large extra-axial nonvascular mass displacing the entire cerebellum forward and upward. The cerebellum is markedly compressed, as is the brain stem. The large posterior fossa cysts with associated compression of the cerebellum, upward displacement of the brain stem, upward herniation of the temporal lobes, and absence of the corpus callosum are most compatible with the diagnosis of a Dandy-Walker formation.

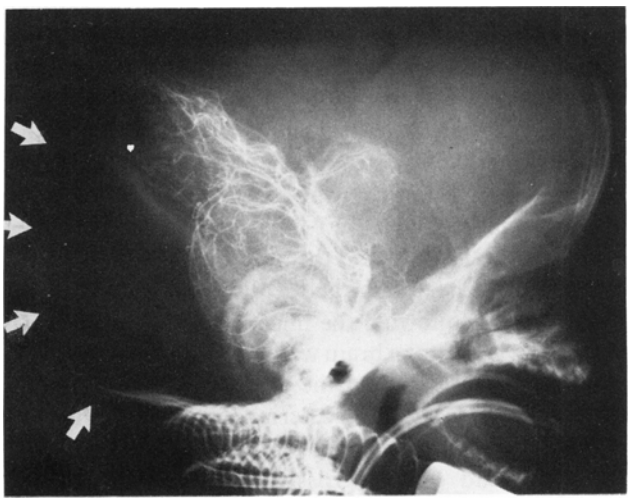


**FIG. 2.** Right lateral view of vertebral angiogram showing forward displacement of vessels due to enlarged posterior fossa. Arrows indicate enlarged posterior fossa owing to Dandy-Walker cyst.

After the completion of the cerebral angiogram a pneumoencephalogram was ordered. Air studies permit the differentiation between the Dandy-Walker syndrome and posterior fossa cysts. The key to these studies is the position and size of the fourth ventricle. In the instance of the Dandy-Walker syndrome, the fourth ventricle is markedly dilated and fills the enlarged posterior fossa *(6)*. The fourth ventricle may herniate inferiorly through the foramen magnum and/or superiorly throughout the tentorial notch. The pneumoencephalogram showed air flowing into the large posterior fossa cyst. The air always seemed to percolate into the cystic space, but always from the anterior aspect of the posterior fossa. The roof of the third ventricle was higher, with a large third ventricular chamber, characteristic of agenesis of the corpus callosum. The size of the lateral ventricle on the frontal horn area suggested mild lateral ventricular enlargement. The cerebellum was small, as outlined by air in the brow up or modified Waters' presentation filming. The aqueduct of Sylvius is exposed. There is partial foramen of Luschka obstruction with large cystic posterior fossa (Fig. 3).

To complete the diagnosis of Dandy-Walker syndrome, computerized axial tomograph was done. Contrast material was not used. Scan section thickness was 13 mm. These images (Fig. 4) reveal the skull to be abnormal with enlargement of the posterior fossa. The posterior fossa is occupied almost entirely by cerebrospinal fluid. The cerebellar hemispheres are extremely small. There is gross enlargement of the third ventricle and portions of both lateral ventricles. The intrahemispheric fissure is grossly enlarged anteriorly and in the sagittal region. The Sylvian fissures are also abnormally widened. The medulla appears relatively small for the size of the foramen magnum, and considerable fluid density is noted in this area.

After completion of all other examinations, the diagnosis made after the brain scan was proven correct. This is the first Dandy-Walker cyst syndrome to be so completely evaluated at this institution. The radionuclide scan proved a very useful tool in the diagnosis of a Dandy-Walker cyst.

The infant was subsequently scheduled for surgery for

**FIG. 3.** Pneumoencephalogram shows enlarged third ventricle (arrows), displaced upward due to union with corpus calossum.

the placement of a V-P shunt. Due to the infant's poor physical condition the shunt was removed several days after initial surgery. As the infant's condition became more stable another shunt was placed. After the second surgery the infant's condition was good and the prognosis was good. This infant will be followed as closely as possible by this department for additional evaluation.

## References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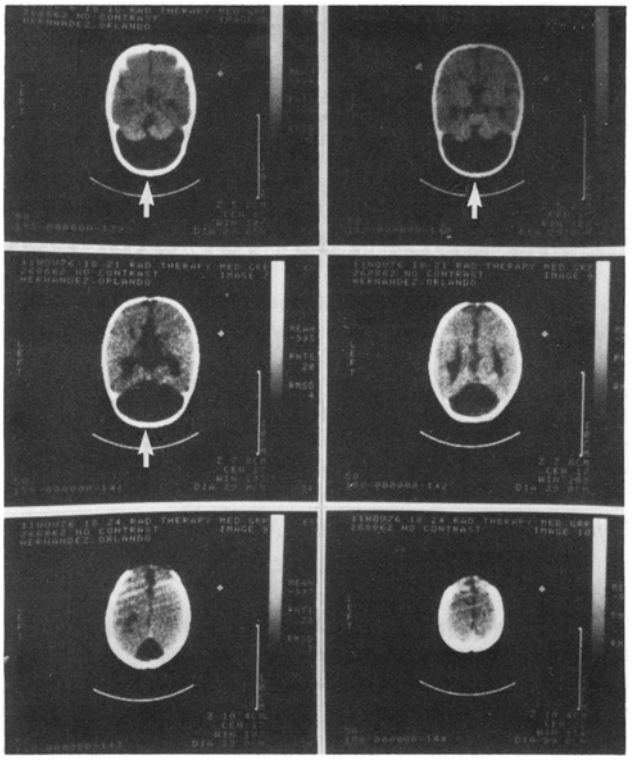

**FIG. 4.** CT scan with arrows pointing to area of Dandy-Walker cyst.


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