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The "Keyhole": A Sign of Herniation of a Trapped Fourth Ventricle and Other Posterior Fossa Cysts

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When a cystic structure in the posterior fossa increases in size, the accompanying increase in pressure may cause it to herniate upward through the tentorial hiatus. In our experience this happens most commonly with a dilated trapped fourth ventricle secondary to infection or intraventricular hemorrhage. However, herniation of an arachnoid cyst or a Dandy-Walker malformation through the tentorium may also occur. When herniation occurs, the cystic structure assumes a "keyhole" configuration, indicating that it is trapped and that surgical intervention is necessary. Five cases are presented that illustrate this point, including two patients with dilatation of the fourth ventricle secondary to hemorrhage, two patients with Dandy-Walker malformation, and one patient with an arachnoid cyst.

With the advent of modern imaging techniques, cystic structures of the posterior fossa have been identified with increasing frequency. In our experience the most common cystic structure of the posterior fossa is a dilated fourth ventricle. This can be congenital (a Dandy-Walker cyst) or acquired secondary to hemorrhage or infection. In a patient with communicating (external) hydrocephalus, the fourth ventricle can dilate along with the other ventricles. Occasionally, after shunting of the lateral ventricles for hydrocephalus, the fourth ventricle will become selectively dilated, which is referred to as a "trapped" or "isolated" fourth ventricle [1]. This is thought to be due to either kinking of the aqueduct or an inflammatory reaction to the shunt tube [1]. The diagnosis of a trapped fourth ventricle by sonography and CT has previously been made by inference from persistent dilatation of the fourth ventricle after successful shunting of the lateral ventricles [2, 3]. However, Collada et al. [4] have shown that persistent dilatation of the fourth ventricle is not sufficient evidence for the diagnosis of a "trapped" fourth ventricle, and CT ventriculography has been used to make a definitive diagnosis.

We observed four children with trapped fourth ventricles confirmed by CT ventriculography in whom the trapped fourth ventricle had assumed a distinctive "keyhole" configuration on sonographic or CT examination. In addition, a posterior fossa cyst in another child was noted to have a similar "keyhole" configuration. The "keyhole" configuration is indicative of herniation of the dilated trapped fourth ventricle or cyst through the tentorial hiatus; further study, such as contrast ventriculography, may not be necessary in the preoperative work-up of these patients.

Case Reports

Case 1

A 27-day-old boy was admitted to the hospital for placement of a ventriculoperitoneal shunt for hydrocephalus secondary to intraventricular hemorrhage. He was born at 27 weeks gestation by emergency cesarean section for abruptio placentae; his birth weight was 936 g. The sonographic examination done on admission showed dilatation of all ventricles including

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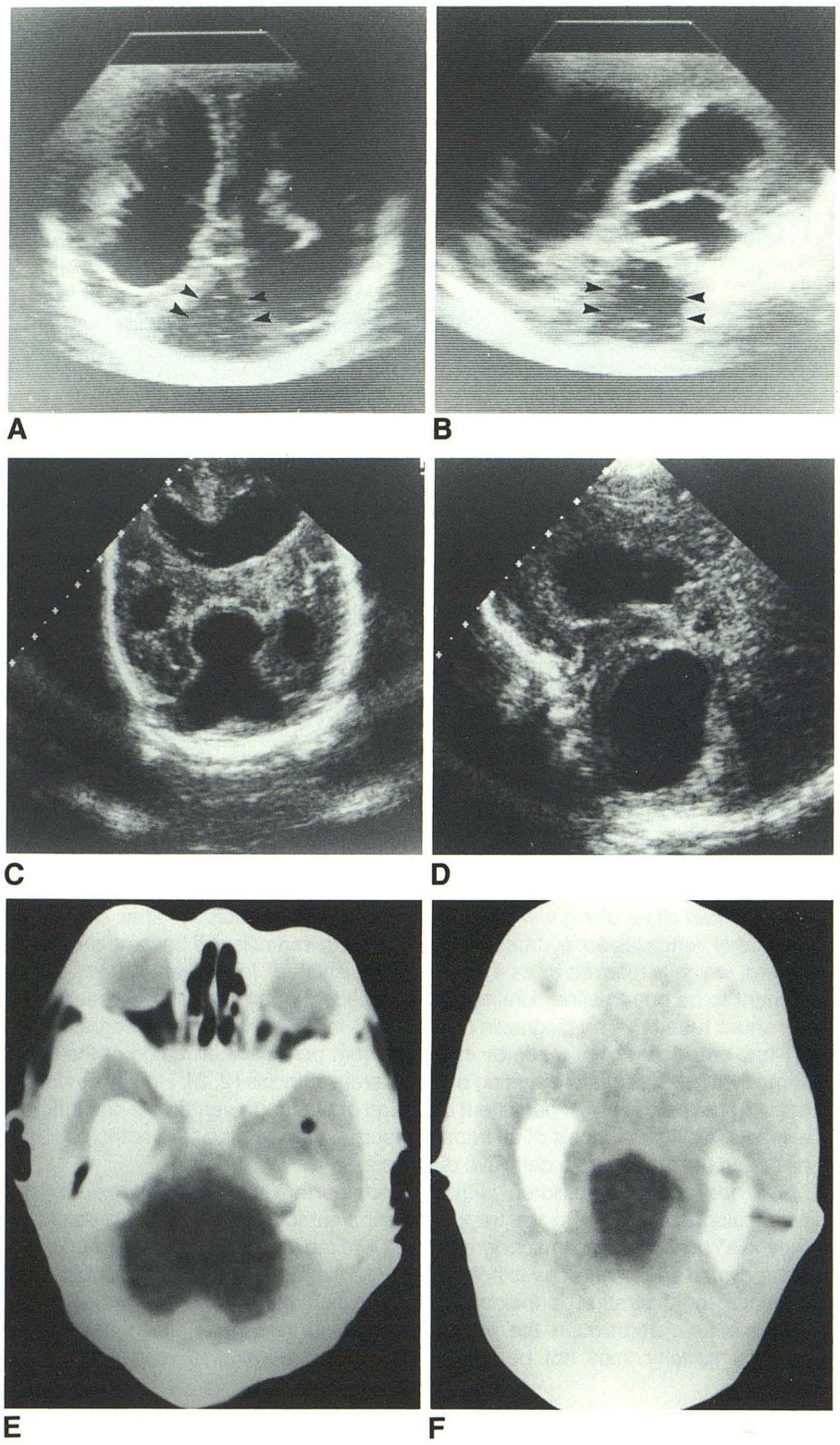

Fig. 1.—Case 1. Sonographic examination at age 27 days shows dilatation of all ventricles including fourth ventricle (*arrows*) in coronal (*A*) and sagittal (*B*) planes. Sonographic examination at 4 months old, 3 months after initial shunt, shows "keyhole" fourth ventricle in coronal plane (*C*). Herniation of fourth ventricle is not easily identified on sagittal view (*D*). CT contrast ventriculography shows that fourth ventricle does not communicate with other ventricles—i.e., infratentorial portion (*E*) and supratentorial portion (*F*) of fourth ventricle.

the fourth ventricle (Figs. 1A and 1B). After placement of the ventriculoperitoneal shunt, the patient developed Klebsiella ventriculitis, which was difficult to eradicate, and several shunt revisions were performed. At age 4 months the patient lost the ability to swallow. Sonographic examination at that time demonstrated adequate decompression of his lateral and third ventricles, but the fourth ventricle had increased in size and had assumed a "keyhole" configuration (Figs. 1C and 1D). A CT ventriculogram was performed and showed isolation of the fourth ventricle (Figs. 1E and 1F).

Case 2

A 43-day-old boy was admitted to the hospital for control of hydrocephalus secondary to grade IV intraventricular hemorrhage.

Sonographic examination on admission showed hydrocephalus with dilatation of all ventricles. A ventriculoperitoneal shunt catheter was placed at that time and the hydrocephalus was adequately decompressed. The patient was reevaluated at 8 months of age because of the death of his twin sister to sudden infant death syndrome. The patient was neurologically impaired but did not have symptoms specifically referable to the posterior fossa. At that time the sonographic examination showed the fourth ventricle to have a "keyhole" configuration. CT ventriculography was then performed and confirmed the presence of a trapped fourth ventricle.

Case 3

An infant girl was admitted to the hospital shortly after birth because of overt macrocephaly. She was the product of a full-term pregnancy with labor complicated by a breech position necessitating a cesarean section. The breech position was thought to be secondary to the hydrocephalus. The initial CT examination showed massive hydrocephalus with a dilated fourth ventricle characteristic of a Dandy-Walker cyst. A ventriculoperitoneal shunt was placed when she was 3 days old. The child required several shunt revisions during her first 3 years of life, but CT examinations during that period did not demonstrate a change in size of the fourth ventricle.

At 3 years, 3 months of age the patient again presented with signs of increased intracranial pressure, and the CT examination done at that time showed that the fourth ventricle had assumed a "keyhole" configuration. CT ventriculography was performed and demonstrated that the fourth ventricle was trapped.

Case 4

A 3-week-old infant was referred to the hospital with evidence of increased intracranial pressure manifested by increasing head circumference and diastasis of the cranial sutures. The patient had been the full-term product of an uncomplicated pregnancy, labor, and delivery. Sonographic and CT examinations at that time demonstrated hydrocephalus with dilatation of the fourth ventricle characteristic of Dandy-Walker malformation. A posterior fossa-peritoneal shunt was placed initially but was revised to a lateral ventriculoperitoneal shunt at 9 months of age.

At 2 years, 7 months of age the patient presented with lethargy, nausea, vomiting, headache, and opisthotonic posturing. A CT scan demonstrated decompression of the lateral ventricles and an increase in the size of the fourth ventricle, which had a "keyhole" configuration. CT ventriculography confirmed that the fourth ventricle had become trapped (Fig. 2).

Case 5

A 1-day-old boy was admitted to the hospital for management of an occipital meningocele. He was born at 36 weeks gestation and was delivered by cesarean section because of his large head. Sonographic and CT examinations in the first week of life showed dilatation of the lateral and third ventricles, with a normal-sized fourth ventricle and a large posterior fossa cyst. The meningocele was repaired at 12 days of age and a ventriculoperitoneal shunt was placed at 18 days of age. At 6 weeks of age, sonography showed that the posterior fossa cyst had increased in size and had assumed a "keyhole" configuration (Fig. 3A). The fourth ventricle was compressed by the large cyst (Fig. 3B). CT ventriculography showed that the cyst did not communicate with the ventricular system (Figs. 3C and 3D).

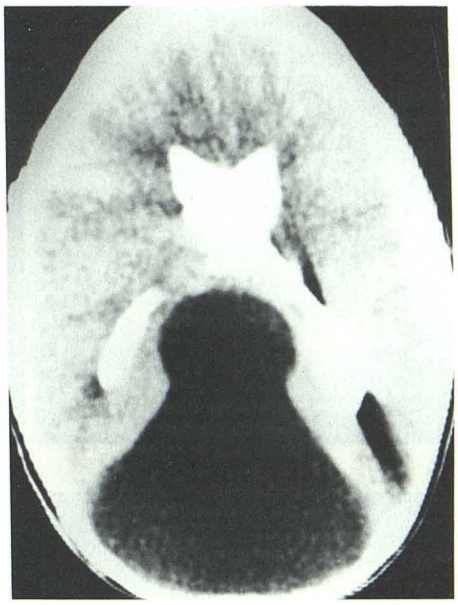

Fig. 2.—Case 4. CT ventriculography shows "keyhole" configuration of herniated Dandy-Walker cyst and that trapped cyst does not communicate with lateral ventricles.

Discussion

Dilatation of the fourth ventricle may be congenital or acquired. When it is congenital it is referred to as a Dandy-Walker malformation and there may be obstruction of the foramina of Luschka and Magendie [5]. Congenital dilatation of the fourth ventricle is associated with dysplasia of the cerebellum and enlargement of the posterior fossa [6]. Dilatation of the fourth ventricle may be acquired as a complication of hemorrhage or infection, and various mechanisms have been postulated, including organizing ependymitis, basilar arachnoiditis, or clot [7, 8]. Dilatation of all four ventricles is most commonly caused by obstruction of CSF flow at the arachnoid granulations [9]; however, the fourth ventricle may also become dilated if the foramina draining it become obstructed. When the fourth ventricle becomes isolated from the other ventricles it may continue to enlarge even after the lateral ventricles have been shunted. This entity has been called the "trapped" fourth ventricle [10], and its appearance on CT scans and by CT ventriculography has been well described [3, 4, 8]. Dilatation of the fourth ventricle may be recognized by sonographic examination as well [2]. Bejar et al. [2] stated that persistent dilatation of the fourth ventricle observed by sonographic examination after the lateral ventricles have been shunted is indicative of a trapped fourth ventricle and no further studies need be done. Scotti et al [3] made the same observation using CT studies. Collada et al. [4], however, reported two cases in which ventriculography was needed to distinguish dilatation of a communicating fourth ventricle from a trapped fourth ventricle. In addition, Brooks and El Gammal [11] reported a case in which ventriculography was necessary to differentiate a midline posterior fossa ependymal cyst from the fourth ventricle.

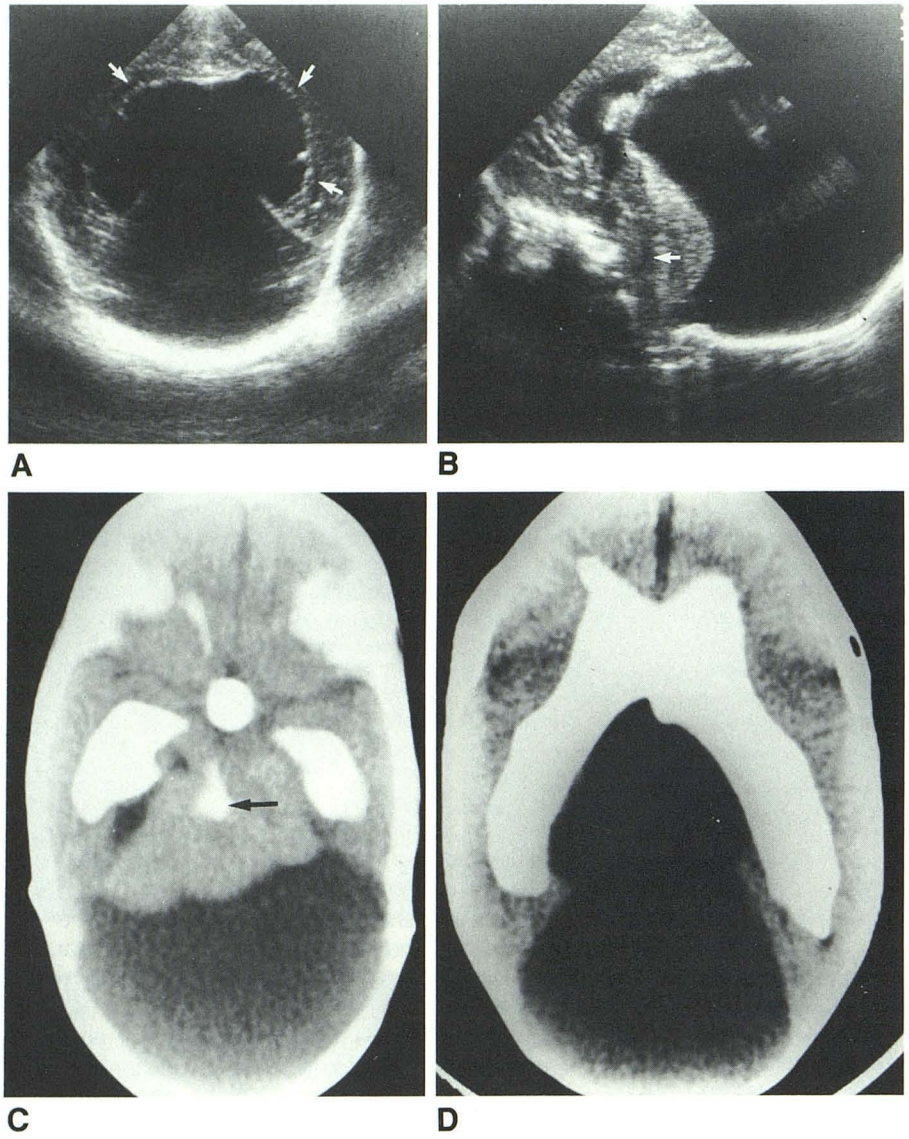

Fig. 3.—Case 5.
***A*, Coronal sonographic image shows large posterior fossa cyst compressing lateral ventricles (*arrows*).**
***B*, Sagittal sonographic image shows anterior displacement and compression of cerebellum and fourth ventricle (*arrow*) by cyst.**
***C* and *D*, CT ventriculogram shows fourth ventricle (*arrow*) filling with contrast and a lack of communication between cyst and ventricular system.**

In our experience, dilatation of the fourth ventricle alone is not sufficient to indicate that the ventricle is trapped. However, when the ventricle herniates and assumes a "keyhole" configuration on sonographic or CT examination then it is trapped and no further studies need be done. The triangular portion of the "keyhole" is made up of the infratentorial portion of the ventricle bordered by the tentorium, and the rounded portion is the supratentorial herniated portion of the ventricle. This herniation is a known potential complication of Dandy-Walker cysts and therefore Raimondi et al. [5] recommended that infra- and supratentorial shunts be placed simultaneously.

Other cystic entities in the posterior fossa may herniate through the tentorial hiatus and assume a "keyhole" configuration. As a rule, however, such cysts can be distinguished from a trapped fourth ventricle because the fourth ventricle can be demonstrated by sonographic or CT examination to be separate from the cystic mass.

We first recognized the "keyhole" configuration on sonographic examination. In older patients in whom the fontanelle has closed, the "keyhole" configuration of the fourth ventricle may be demonstrated by CT scanning if the plane of scanning is tangential to the tentorium.

In summary, when the pressure in a dilated fourth ventricle or other posterior fossa cyst is increased, these cystic structures may herniate through the tentorial hiatus. In so doing they assume a "keyhole" configuration that can be identified on sonographic or CT examination. The "keyhole" configuration is indirect evidence that the fourth ventricle or posterior fossa cyst is "trapped" and does not communicate with the ventricular system. If this configuration is present then there is no need for more invasive methods of study, such as water soluble contrast ventriculography.

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