Occluded Fourth Ventricle After Multiple Shunt Revisions for Hydrocephalus

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ABSTRACT. Trapped occluded fourth ventricle has been considered a rare occurrence. Intraventricular hemorrhage followed by repeated shunt revisions may increase the risk (8/47 cases). Because premature infants with intraventricular hemorrhage and shunted hydrocephalus often have preexisting neurologic abnormalities, dilation may produce clinically undetected further neurologic damage. Shunting improved function in both currently treated as well as 13 of 14 previously treated patients. In light of this observation, the importance of recognition is stressed. *Pediatrics* 1989;83:981–985; *occluded (trapped) ventricle, intraventricular hemorrhage complication, ventriculoperitoneal shunt complication.*

Periventricular-intraventricular hemorrhage occurs in approximately 35% to 45% of premature infants of less than 32 weeks' gestation.[1,2] Hydrocephalus following intraventricular hemorrhage will occur in about half and will become progressive in approximately 25%.[3] The placement of a ventriculoperitoneal shunt is often the mode of treatment. Complications of this procedure include mechanical obstruction, infection, intraabdominal CSF pseudocyst, inguinal hernia from CSF ascites, and slit ventricle syndrome.[4–7] In addition, a blockage of the aqueduct of Sylvius plus the foramina of Luschka and Magendie can isolate the fourth ventricle producing progressive enlargement, a rare complication of ventricular shunting.[8,9] Occluded fourth ventricle (commonly known as "trapped" ventricle) was reported in six of 950 children who received shunts for hydrocephalus because of multiple causes studied by Palmieu et al.[10] We postulated a greater frequency of this complication after shunted posthemorrhagic hydrocephalus.

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METHODS

All CT scans performed on pediatric patients between 1984 and 1987 and reported by the radiologist as showing hydrocephalus were identified. If a ventriculoperitoneal shunt was present, the neonatal record was reviewed. Cases in which hydrocephalus resulted from infection, malformation, and other known causes were excluded. Only children with prior intraventricular hemorrhage greater than grade I and proven by ultrasound or CT scan were included. The fourth ventricle was considered occluded if there was radiologic evidence of upward herniation under the tentorium.

RESULTS

A total of 47 children were identified postintraventricular hemorrhage shunt. Their records were collected from a large neonatal referral center at which 544 premature infants weighing less than 1,500 g had been admitted in the last 4 years. Of these, 163 had intraventricular hemorrhages, of which 101 were grades III to IV. Evidence of an occluded fourth ventricle occurred in eight (17%) and resulted in massive dilation in four (Table). Fourth ventricular enlargement developed only after shunting. Symptoms potentially related to the occluded fourth ventricle occurred in the four children (patients 1 to 4) with massive enlargement. A progressive spastic quadriparesis was present in three children, and signs of increased intracranial pressure with lethargy and vomiting occurred in two children.

Patient 1 was observed to progress from a left hemiparesis to quadriparesis. Despite worsening,

TABLE. Occluded Fourth Ventricle

Case No.	Gestational Age (wk)	Grade of Intraventricular Hemorrhage*	Chronologic Age, Sex	No. of Shunt Revisions	Age at Onset of Symptoms (mo)	Clinical Appearance	Treatment	Present Function
1	30	IV	3 yr, F	7	22	Lethargy, vomiting hemiparesis progressing to quadriparesis	None—vomiting resolved after 3 d	Nonverbal, spastic quadriparesis
2	25	III	6 yr, M	5	24	Spastic quadriparesis, progressive	None	Single words, spastic quadriparesis
3	31	IV	1 yr, F	7	11	Lethargy, bilateral 6th nerve palsy, spastic quadriparesis, mute	Shunted 4th ventricle	Resolved 6th nerve palsy, single words, resolving quadriparesis
4	24	IV	6 mo, F	2	6	Spastic quadriparesis, progressive	Shunted 4th ventricle	Mild improvement in motor function
5	31	IV	2 yr, F	3	None			Normal
6	30	III	11 yr, M	3	None			Educable, mental retardation, no cerebral palsy
7	27	IV	7 yr, M	8	None			Nonverbal, spastic quadriparesis
8	28	IV	6 mo, F	2	None			Spastic quadriparesis

* Determined by computed tomography or ultrasound.

the fourth ventricle was not shunted. Lethargy and vomiting spontaneously resolved within three days, but the loss of motor function was persistent (Fig 1). Patient 2, who also did not receive shunting, had a severe neurologic deficit with a progressive spastic quadriparesis prior to the diagnosis of an occluded fourth ventricle (Fig 2). Two children underwent a fourth ventricular shunt (patients 3 and 4). One became more alert and less quadriparetic, and regained language development within three days (Fig 3). The other improved gradually in motor function following resolution of postoperative status myoclonus (Fig 4).

DISCUSSION

Symptoms and signs of an occluded fourth ventricle may include headache, lethargy, vomiting, ataxia, spastic quadriparesis, cranial nerve palsies, and head tilt.[8,11,12] Infection, inflammation, hemorrhage, posterior fossa brain malformation, or tumor may be responsible for the obstruction. Following intraventricular hemorrhage, inflammation of the basilar cisterns and fourth ventricular outflow (foramina of Luschka and Magendie) may occur. This is due to a reaction to the blood products with resulting scarring and subsequent constriction of the subarachnoid space (obliterative arachnoiditis).[3] The placement of a ventriculoperitoneal shunt allows drainage of the fourth ventricle through the aqueduct. However, secondary aqueductal closure may occur because of chronic inflammation due to bleeding accompanying repeated shunt revisions.[7]

Intraventricular instillation of contrast was performed in three of the eight patients to document aqueductal obstruction with isolation of the fourth ventricle. Support for ventricular isolation in the remaining five patients is provided by the disproportionate enlargement of the fourth compared with lateral ventricles plus the keyhole sign (Fig 5). This sign is due to upward herniation of the most superior part of the fourth ventricle above the ten-

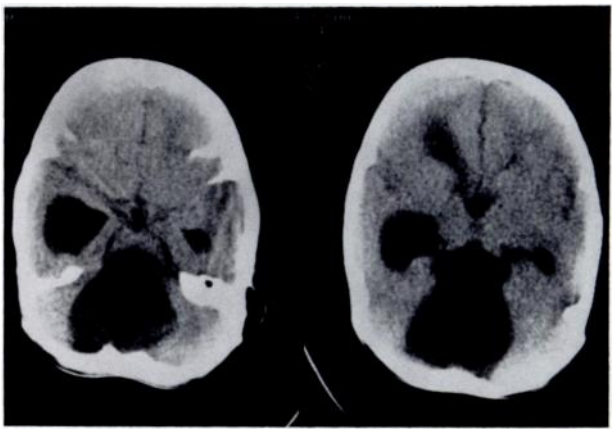

Fig 2. Right temporal horn and fourth ventricle are massively enlarged with supratentorial herniation of rostral fourth ventricle (patient 2).

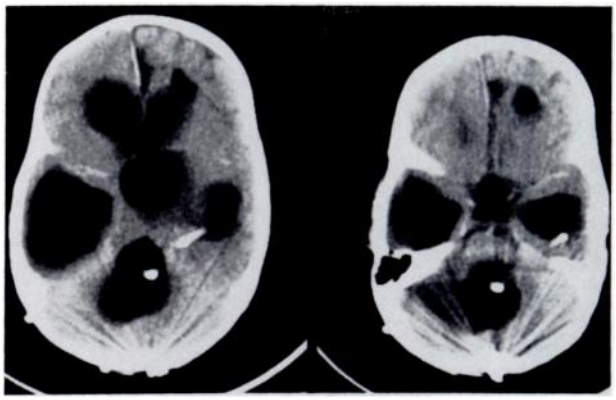

Fig 3. Following shunting of fourth ventricle, speech returned and sixth nerve palsy, lethargy, and quadriparesis improved. In this patient, fourth ventricle dilation is not disproportionate to lateral ventricles (patient 3).

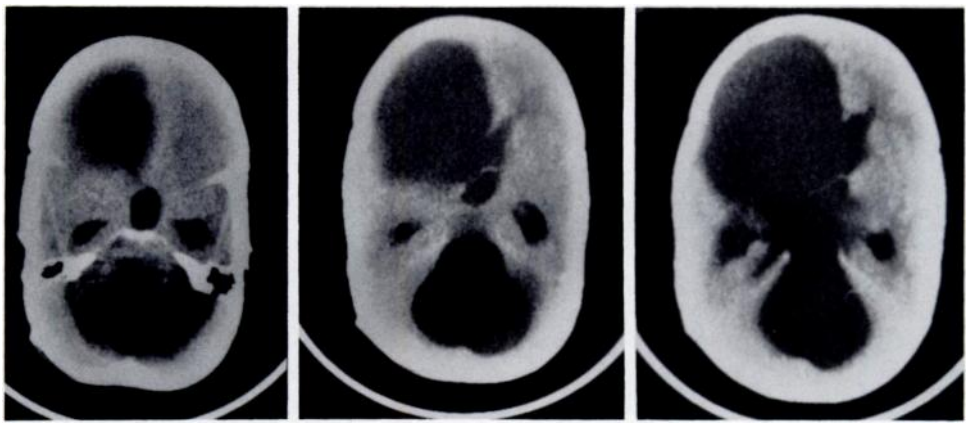

Fig 1. Note massive dilation of fourth ventricle with moderate enlargement of temporal horns and large right porencephalic cyst. Two right scans show supratentorial herniation of rostral fourth ventricle (patient 1).

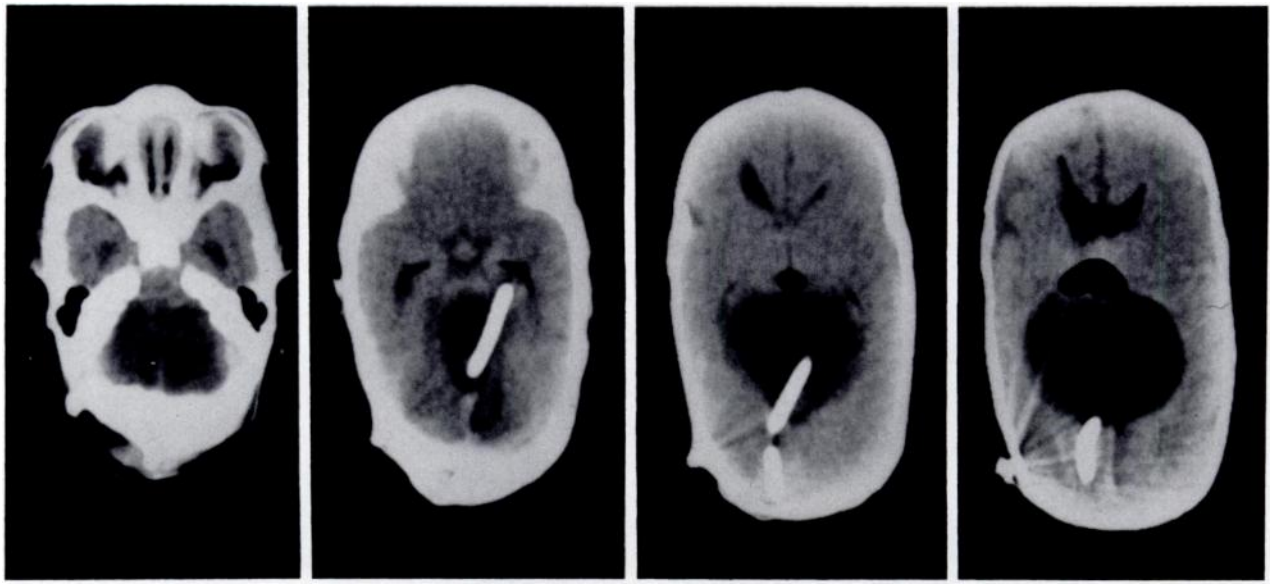

Fig 4. Only mild decrease in size of fourth ventricle occurred after shunting; however, quadriparesis improved (patient 4).

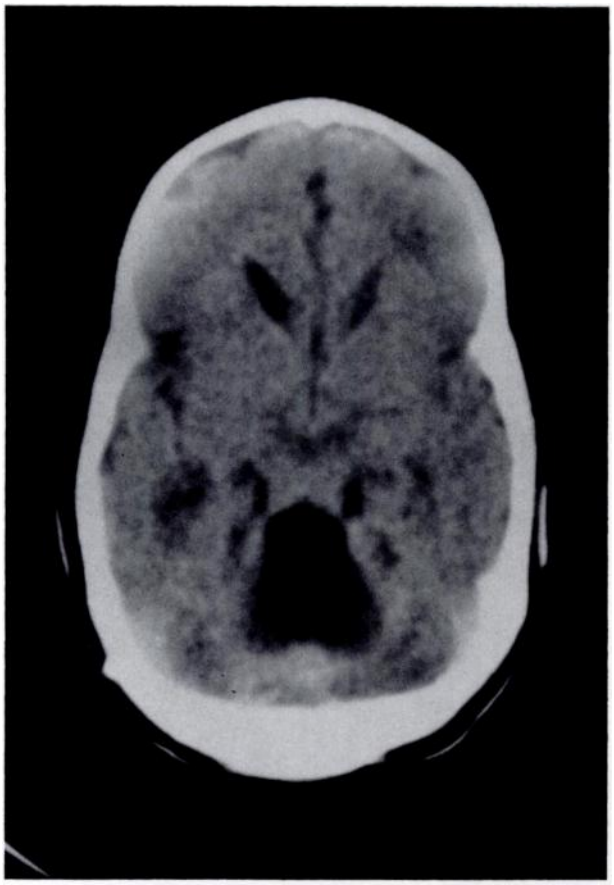

Fig 5. Lateral ventricles are normal in size. Fourth ventricle is enlarged to moderate degree with keyhole sign in this asymptomatic 2-year-old child (patient 5).

torium. Contrast documentation of obstruction is not necessary when this sign is present.[13]

Occluded fourth ventricle is a common (17%) complication among children with intraventricular hemorrhage who have received ventriculoperitoneal shunting. All of our patients received more than one shunt revision, previously described as associated with occluded fourth ventricle.[14] The percentage we described is probably an underestimation, because, although all patients who received shunts received scans, the patients were not all scanned after each revision. Also, the retrospective design allowed clinically asymptomatic cases to go undetected.

The reason dilation of the fourth ventricle did not continue to progress in the six untreated patients is unknown. Perhaps the choroid plexus in the fourth ventricle stopped secreting CSF because of progressive pressure or the aqueduct or perhaps the foramina reopened.

Recognition of this complication is difficult in premature infants with shunted hydrocephalus. Most of these infants have preexisting neurologic problems, some of which may be severe. The clinician ordinarily does not thoroughly document neurologic function at each follow-up visit because the deficits are considered static. The shunt pumps well, the fontanelle remains flat, but the child may insiduously worsen as the fourth ventricle dilates. Just as progressive lateral ventricular enlargement can precede signs of increased intracranial pressure or macrocephaly by days to weeks,[15] so might fourth ventricular enlargement be silent. Experimental studies of "silent" lateral ventricular dilation have shown signs of brain injury with axonal stretching and gliosis, impaired blood flow, and changes in evoked potentials.[16–18] Progressive fourth ventricular dilation is likely to do the same and postshunt CT or magnetic resonance imaging are necessary

to rule it out. Perhaps brainstem auditory-evoked responses could serve as a baseline and be repeated in instances of fourth ventricular enlargement to indicate progression.

Neurologic improvement occurred following a fourth ventricular shunt in 13 of 14 previously reported cases[6,8–10,19] and in both children who received shunting described in this article. Although cessation of progressive paresis and improvement in neurologic function may have been due to resolution of a supratentorial process or age-related development, most children have shown acute improvement in signs of increased intracranial pressure and neurologic function following the placement of a shunt.

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