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AJNR Am J Neuroradiol 1986, 7 (3) 443-447

<http://www.ajnr.org/content/7/3/443>

This information is current as
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Periventricular Leukomalacia in Combination with Intraventricular Hemorrhage: Sonographic Features and Sequelae

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Periventricular leukomalacia (PVL) is well recognized as a relatively uncommon yet particularly serious complication of prematurity. Although the sonographic features of PVL have been described, its association with intraventricular hemorrhage (IVH) has not been emphasized. Reviewing 26 consecutive cases of PVL in neonates of 34 weeks or less gestational age, significant associated hemorrhage was found in six (23%). Small quantities of blood were also noted in most of the other 20 infants. Of the six infants with both significant IVH and PVL, five required ventricular shunt and all had particularly poor clinical outcomes. In the neonates who required surgical intervention, rapid ventricular enlargement was accompanied by extensive periventricular cyst formation. Eventually, the septations within the cysts and frequently even the ependyma of the superior/posterior lateral ventricles degenerated. Cysts merged imperceptibly with the ventricles giving an appearance that mimicked severe hydrocephalus. This was termed "pseudoventricle formation," as the large intracerebral cerebrospinal fluid spaces are primarily porencephaly and not enlarged ventricles. Response to shunting was minimal by sonography in all five cases and multiple shunt revisions were required in four. Clinical follow-up in children with significant IVH in combination with PVL has shown severe mental retardation and tetraplegia in all cases.

Germinal matrix-related hemorrhage and posthemorrhagic hydrocephalus have been closely associated with the gestationally immature brain for many years [1–3]. Since 1982, however, reports of periventricular leukomalacia (PVL) in the preterm population have become increasingly frequent [4–8]. The sonographic literature thus far has not emphasized the association of PVL with intraventricular hemorrhage (IVH). Most authors have actually described minimal, if any, intraventricular hematoma accompanying PVL [4–8]. This study was undertaken to determine if these two seemingly opposite cerebral insults occur in a significant number of premature infants. The sonographic and clinical follow-up of children with this combination of cerebral pathology was then evaluated to determine its effect.

Materials and Methods

A review of the records of the neonatal intensive care unit between January 1979 and November 1984 revealed that 926 infants less than 34 weeks gestational age had been admitted. All "in-born" neonates received a cranial sonogram within the first 3 days of life. Sonography was performed on all transfer patients at least once during the first week of admission. Follow-up sonograms were obtained at weekly intervals until discharge from the nursery in all neonates with intracranial abnormalities other than those of obvious congenital origin. Because of the potential insensitivity of early sonograms in diagnosing PVL, pre-discharge sonograms became routine during the latter half of the study. Scans were obtained using commercially available real-time units. Before 1982, sonography was performed exclusively with a 5-MHz-transducer system. Since then, a combination of both 5 and 7.5 MHz scanning has been used in almost every examination.

The diagnosis of PVL was based upon early demonstration of abnormally increased periventricular echogenicity and/or later development of typical regions of cystic periventricular

Received May 20, 1985; accepted after revision August 6, 1985.

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AJNR 7:443–447, May/June 1986

0195–6108/86/0703–0443

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TABLE 1: Clinical Summary of Infants with Periventricular Leukomalacia (PVL) and Intraventricular Hemorrhage (IVH)

Case No. (Weeks Gestation)	Birth Weight (g)	Day of Sonographic Manifestation		Shunt/Revisions	Present Age (in months)
		Of PVL	Of IVH		
1 (31)	1300	3	3	+/-2	Decreased 22
2 (34)	1940	3	3	+/-1 (two shunts required for loculated ventricles)	
3 (26)	700	6	2	+/-8	18
4 (29)	1210	1	1	+/-3	27
5 (28)	1200	1	3	+/-3	23
6 (26)	1100	1	3	No shunt	16

Note.—The clinical outcome was quadriplegia and severe developmental delay in cases 2–6; case 1 died.

degeneration [4–8]. The differentiation between significant and minor IVH proceeded along the lines outlined by Shankaran et al. [9] and Fleischer et al. [10]. Reiterated, significant IVH completely filled and expanded the lateral ventricles. Minor, or insignificant, IVH was diagnosed on the basis of abnormal echogenicity within the ventricles but in a quantity insufficient to cause ventricular expansion. Ventricular enlargement by cerebrospinal fluid (CSF) after the initial hemorrhage had no bearing on the classification of hemorrhage.

Results

Among the 926 premature neonates, films were available for review in 26 who were diagnosed as having PVL and who met the criteria set forth under Materials and Methods. Various facets of the sonographic abnormalities encountered in these children have been reported previously [8, 11]. Six (23%) of these children had large IVHs in addition to PVL. Small quantities of IVH were identified in most of the other 20 subjects. The pertinent clinical data and follow-up information on the six children with significant IVH are summarized in Table 1. Early scans showed no particular chronologic relation between the appearance of PVL and IVH. In three neonates they were identified simultaneously; in two, PVL preceded the hemorrhagic event (Fig. 1); and in one, IVH was identified first. All six neonates experienced rapid enlargement of the ventricles. In two cases, periventricular cysts were identified as early as 8 days of age (Fig. 2). Five of the six neonates eventually underwent insertion of a ventricular shunt because of progressive ventricular enlargement, increasing head circumference, and development of a bulging fontanelle. The ventricular size and head circumference stabilized without intervention in only one patient (case 6). This child, however, had moderately enlarged ventricles, extensive parenchymal cystic degeneration, and signs of cerebral atrophy (Fig. 3). It is of note that in two children (cases 2 and 3), abnormal parenchymal echogenicity and early cyst formation about the ventricular trigones remained relatively subtle until 5 and 3 weeks, respectively, when massive periventricular cystic degeneration occurred (Fig. 4).

In four of the five children requiring shunts, the ependyma

separating the periventricular cysts from the ventricles degenerated. The ependyma was frequently totally absent over the superior aspects of the lateral ventricular bodies, trigones, and occipital horns. Only one of the shunted neonates had clear preservation of the ependyma (Fig. 5). In one child, fenestrations in the ependyma were already visible at 14 days. As the infants became older, the septations typical of the cystic phase of PVL also disappeared. The net result of this degeneration of the septae and ependyma was the formation of large ventricular outpouchings. In some cases, these areas of periventricular degeneration were so extensive that the demarcation between actual ventricle and the attendant porencephaly became impossible (Fig. 6). The most severely affected areas were usually the posterior lateral ventricles; the anterior tip of the frontal horn was usually preserved. In some cases these cystic areas extended so far superiorly that they were not even visible on parasagittal sector scans. We have termed this conglomeration of true ventricle and periventricular cysts a "pseudoventricle" because frequently its appearance closely mimics a hydrocephalic ventricle. Review of previous sonograms, however, shows it to actually represent massive porencephaly. Long-term follow-up in the five patients who underwent shunting revealed minimal response to the procedure by sonography. Four of these five children required multiple shunt revisions, and the large "pseudoventricles" persisted.

Discussion

Germinal matrix-related hemorrhage and PVL are two forms of intracranial pathology closely associated with prematurity. In the former, friable vessels in a rudimentary fetal structure rupture. Although the hematoma may be self-contained, extension into the adjacent ventricular system or even the cerebral parenchyma is common [2]. Conversely, PVL is an ischemic event; infarction in the delicate watershed zones that exist between the ventriculopedal and ventriculofugal arteries in the gestationally immature brain is believed to be the underlying lesion [12]. Armstrong and Norman [13] described cerebral bleeding in association with PVL in 25% of their subjects but ascribed it to hemorrhage into a preexisting periventricular infarction. Therefore, IVH could arise on this basis. Pape and Wigglesworth [2], on the other hand, described the coexistence of PVL and germinal matrix hemorrhage, raising the possibility of two independent lesions in the same patient. Certainly the dynamics of cerebral blood flow in the preterm neonate are such that lesions secondary to both hyper- and hypoperfusion could be postulated.

The causes for such severe sequelae in patients with significant IVH and PVL are most likely multifactorial. One could postulate the occurrence of marked ventricular enlargement in these patients on the basis of hemorrhage alone since the severity of posthemorrhagic hydrocephalus is directly proportional to the size of the original IVH [14, 15]. The increased CSF pressure that accompanies posthemorrhagic hydrocephalus could also contribute to the enlargement of the periventricular cysts [16]. Similarly, increases in CSF

Fig. 1.—Case 5. Coronal sonograms. **A**, Day 1. Abnormal echogenicity typical of PVL about both lateral ventricles (*arrowheads*). **B**, 2 days later. Large left frontoparietal intraventricular/intraparenchymal hemorrhage (*arrows*). Hematoma (H) in contralateral ventricle and persistence of abnormal periventricular echogenicity (*arrowheads*). Parasagittal sonograms further demonstrated large amounts of blood in trigones and occipital horns of lateral ventricles (cf. Fig. 2B).

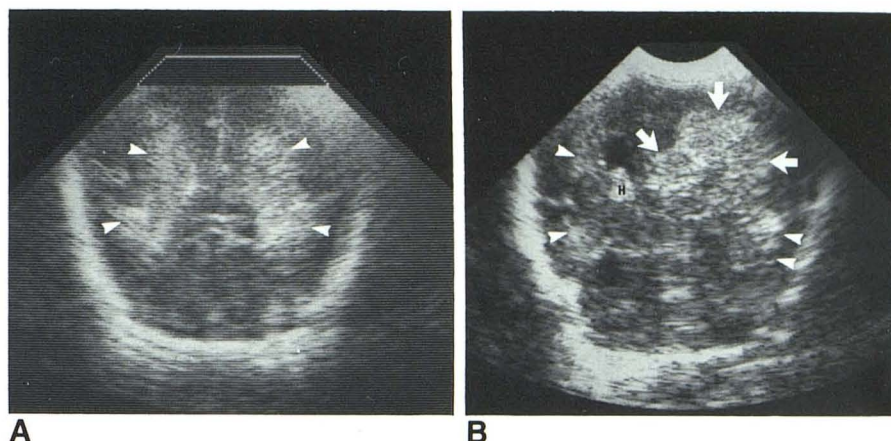


Fig. 2.—Semi-axial (**A**) and left parasagittal (**B**) sonograms at 8 days of age in cases 1 and 5, respectively. Extensive periventricular cystic degeneration (*arrows*) has already taken place. Lateral ventricles have rapidly enlarged; large intraventricular hematomas (H) persist.

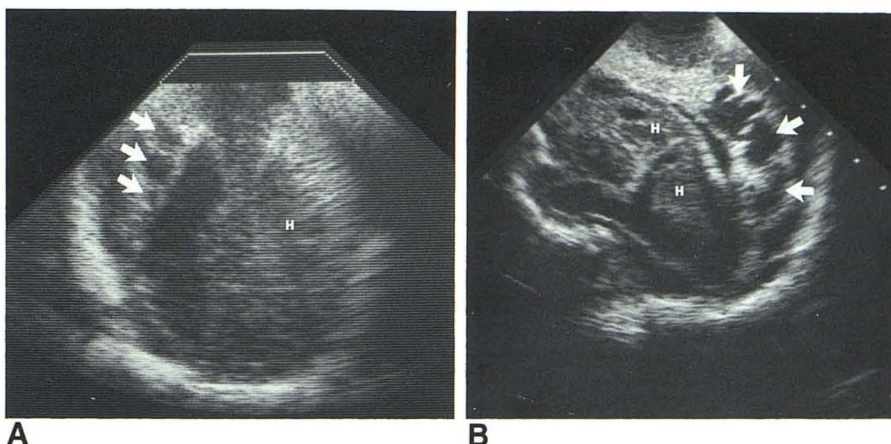
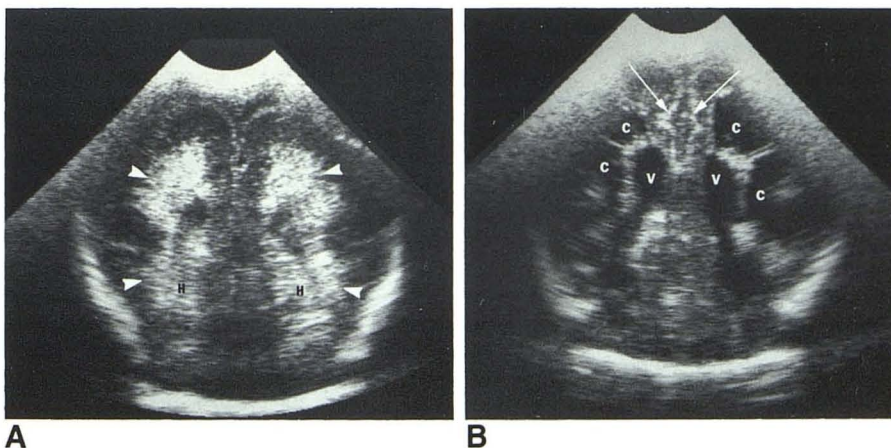


Fig. 3.—Case 6. Semi-axial sonograms. **A**, 3 days. Bilateral echogenic PVL (*arrowheads*) and large intraventricular hematomas (H). Parasagittal sonograms showed the hematomas to be in the atria of the lateral ventricles. **B**, 6 weeks. Moderate ventriculomegaly (V), extensive periventricular cyst formation (C), and widening of interhemispheric fissure (*arrows*). Latter finding is typical of cerebral atrophy.



pressure with early posthemorrhagic hydrocephalus may accentuate the severity of the infarction; in this case the two forms of pathology may actually be synergistic. Finally, one might postulate that these children are merely unfortunate enough to have had both severe hyper- and hypoperfusion.

Regardless of the underlying causes, five of six neonates with PVL and significant IVH required ventricular diversion. Despite functioning shunts, the ventricles never returned to normal size, most likely on the basis of extensive pseudoventricle formation. Although abnormal intracranial pressure may be

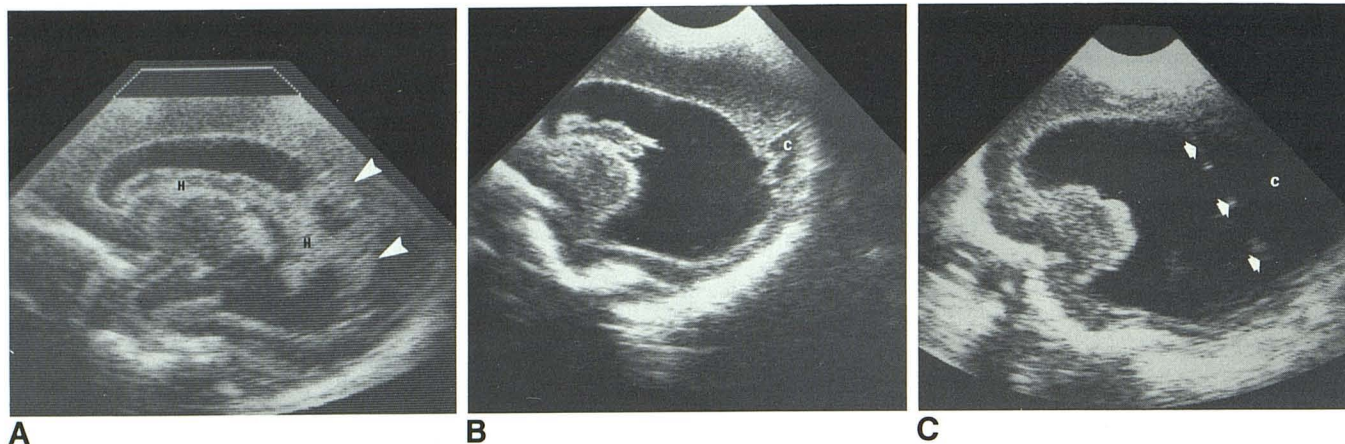


Fig. 4.—Case 4. Parasagittal sonograms. **A**, 14 days. Moderate ventricular enlargement, intraventricular hematoma (H), and abnormal periventricular echogenicity (arrowheads) above ventricular trigones. **B**, 5 weeks. Marked ventric-

ular enlargement; periventricular cystic degeneration (C) has taken place but remains relatively subtle. **C**, 2 weeks later. Large periventricular cysts (C) bilaterally. Ependyma (arrows).

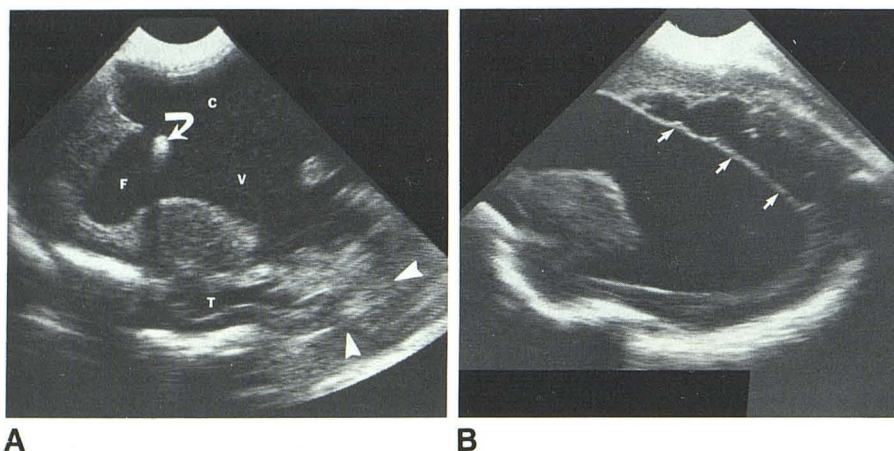


Fig. 5.—Parasagittal sonograms reveal extensive periventricular cyst formation at 6 weeks of age. **A**, Case 1. Apparent open communication between ventricle (V) and cyst (C). Note persistence of intraventricular hematoma (arrowheads) and presence of shunt device (arrow). Only frontal (F) and temporal (T) horns remain distinct. Among the five infants requiring shunt, only case 4 (**B**) showed clear preservation of ependyma (arrows).

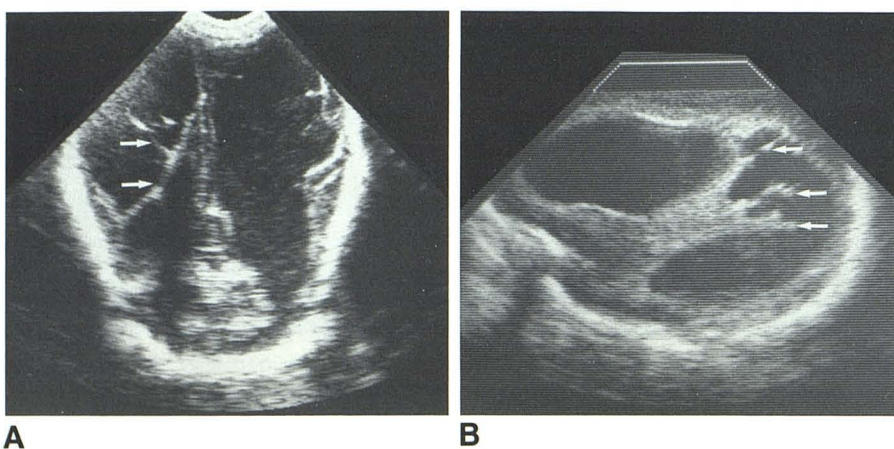


Fig. 6.—Case 5 at 4 months of age. Semiaxial (**A**) and parasagittal (**B**) sonograms show pseudo-ventricle formation; demarcation between hydrocephalic ventricle and porencephaly is lost. Note persistence of sparse internal septations (arrows); most have degenerated. Patient had functioning shunt at this time.

relieved and the ventricular system reduced in size, massive porencephaly will remain.

Pseudoventricles may be particularly difficult if not impossible to differentiate from true hydrocephalus. Pseudoventri-

cles are frequently bilateral and relatively symmetric. Eventually even the internal septations typical of cystic PVL disintegrate. In many cases, the ependyma also degenerates leaving extensive areas of porencephaly in apparent open communi-

cation with the lateral ventricles. The situation is compounded by the sector scanner; as shown in Figures 4 and 5, the superior extent of the cysts may not even be visible. Assessment of the true lateral ventricular size is, however, important preoperatively as porencephaly responds poorly to shunting [17]. Our series shows that although extensive pseudoventricle formation may be present about the bodies and occipital horns, the frontal horns are generally spared. This is best demonstrated on parasagittal sections and seems the only adequate method of assessing true ventricular size.

On the basis of this study, it seems that one may tailor scanning routines in patients with PVL according to the size of the attendant IVH. Cases of PVL with small IVHs require fewer follow-up sonograms; the ventricles do not expand significantly nor do these children seem to require intervention. Children with significant IVH and PVL, on the other hand, require frequent and often long-term follow-up with cranial sonography. Children with significant IVH and PVL have also been particularly difficult to manage neurosurgically because of the frequent need for shunt revisions. Finally, the clinical outcome in children with significant IVH and PVL appears worse than that in children having PVL without significant IVH. Certainly any child with PVL has a poor prognosis. Our follow-up in children with PVL and large IVHs, however, has shown particularly poor results, with spastic tetraplegia and severe developmental delays in all six infants.

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