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Prominent choroid plexus in meningomyelocele: sonographic findings.

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Prominent Choroid Plexus in Meningomyelocele:
Sonographic Findings

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Of 18 infants with meningomyelocele who were examined by cranial sonography, 17 were found to have a prominent choroid plexus, especially in the region of the glomus. In almost half of the cases this prominence assumed the configuration of a stalk arising from the glomus and terminating in a clublike ending. This prominence can be easily confused with intraventricular hemorrhage. The literature dealing with meningomyelocele and the Arnold-Chiari malformation makes no mention of this finding, but literature related to choroid plexus pathology describes similar findings in hydrocephalic infants, some of whom had documented meningomyelocele. This unusual choroidal configuration seen on sonography might be part of the spectrum of brain malformations seen in infants with meningomyelocele.

Cranial sonographic findings in patients with meningomyelocele and Arnold-Chiari malformation have been described, and include such features as pointed frontal horns, a large massa intermedia, an absent septum pellucidum, a relatively small posterior fossa, and hydrocephalus [1]. In reviewing the cranial sonograms of 18 infants and neonates with meningomyelocele, we noted a previously undescribed radiologic finding—a prominent, and often lobulated, choroid plexus. This prominence is most pronounced in the region of the glomus and it may extend for some distance posteriorly, or even outward to the walls of the bodies of the lateral ventricles. This configuration assumes particular importance to the sonographer because it easily can be taken to represent intraventricular hemorrhage. The sonographic features of the choroid plexus in children with spinal dysraphism form the basis of this report. The possible significance of this finding and its differentiation from intraventricular hemorrhage are discussed.

Materials and Methods

Eighteen infants with meningomyelocele were studied by cranial sonography using commercially available real-time sector scanners. Examinations were performed with both 5.0 and 7.5 MHz transducers. Two of the 18 were born prematurely, one at 32 weeks and the other at 33 weeks gestational age. The other 16 were considered term infants. Altogether, 51 sonographic studies were performed, all using the anterior fontanelle as an acoustic window. Fifteen of the infants had their first sonographic study within 4 days after birth (most on the first day), and the other three had their initial scans at 7 days, 3 weeks, and 3 months. Follow-up sonograms were obtained in 13 patients, with the greatest interval between the first and last study being 7½ months. A CT scan of the head was also obtained in half the patients, all within the first 2 weeks of life. Ten of the 18 underwent insertion of a ventriculoperitoneal shunt tube. This procedure was done in seven cases within the first 12 days of life and in the others at 19 days, 28 days, and 10 months.

Results

In 17 (94%) of the 18 infants the choroid plexus echo complex was believed to
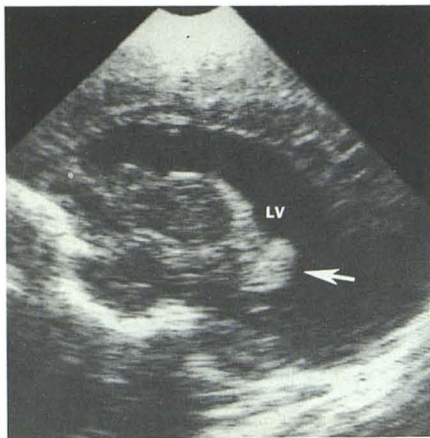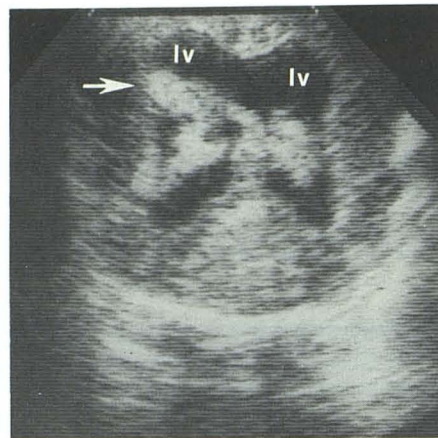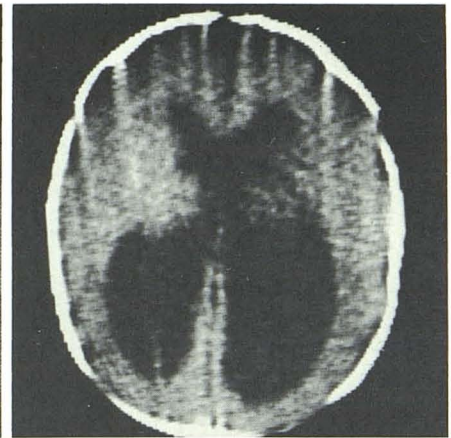
Fig. 1.—Parasagittal sonogram through anterior fontanelle in infant with meningomyelocele. Prominent and somewhat lobulated choroid plexus (*arrow*) extending posteriorly from region of glomus. Lateral ventricle (LV) is enlarged.

Fig. 2.—Prominent choroid plexus. **A**, Coronal sonogram on first day of life. Choroid plexus is prominent bilaterally, more on one side (*arrow*), and extends laterally into bodies of dilated lateral ventricles (lv). **B**, CT scan 10 days later revealed no evidence of hemorrhage, and inspection of CSF showed no xanthochromia.
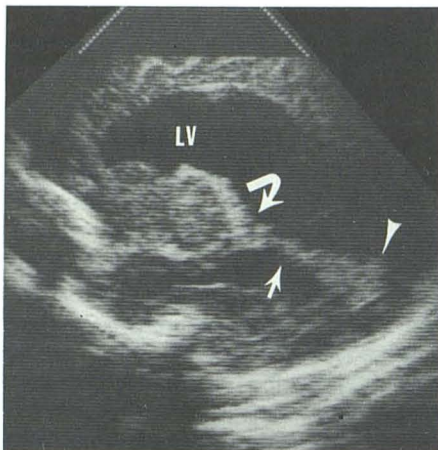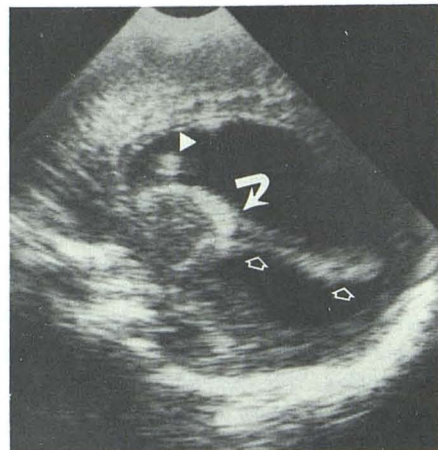

Fig. 3.—Parasagittal sonogram. Stalk (*straight arrow*) arises from glomus (*curved arrow*) and terminates in wider, clublike ending (*arrowhead*). LV = lateral ventricle.

Fig. 4.—Bilateral stalk-and-nodule configuration of choroid plexus (*open arrows*) extending from glomus (*curved arrows*). **A**, Right lateral ventricle. Tip of shunt tube anteriorly (*arrowhead*). **B**, Left lateral ventricle.

be more prominent than normal. The choroid plexus appeared to be large, unusually irregular, and somewhat lobulated (figs. 1 and 6). The prominence was usually most marked in the region of the glomus and best demonstrated on sagittal sonograms. On coronal sonograms, the more anterior parts of the choroid plexus often appeared large and extended laterally toward the wall of the lateral ventricle (fig. 2). In eight of the infants in whom the choroid plexus was enlarged, a peculiar configuration could be seen arising from the glomus on some or all of the sonograms localized to the area of the trigone. This configuration consisted of a long stalk that seemed to arise from the glomus, extended mostly poste-

riorly, and usually terminated in a small, clublike nodule (fig. 3). This "stalk-and-nodule" configuration could be seen either bilaterally (fig. 4) or unilaterally. Where it was unilateral, it was usually associated with a prominent contralateral choroid plexus. In almost every case in which we found a prominent choroid plexus, the prominence was bilateral, although not necessarily of equal size.

The sonographic appearance of the choroid plexus described above is similar to what may be seen in intraventricular hemorrhage. The irregular and lobulated choroid plexus may appear identical to the sonographic manifestation of intraventricular hemorrhage, and the stalk-and-nodule configuration
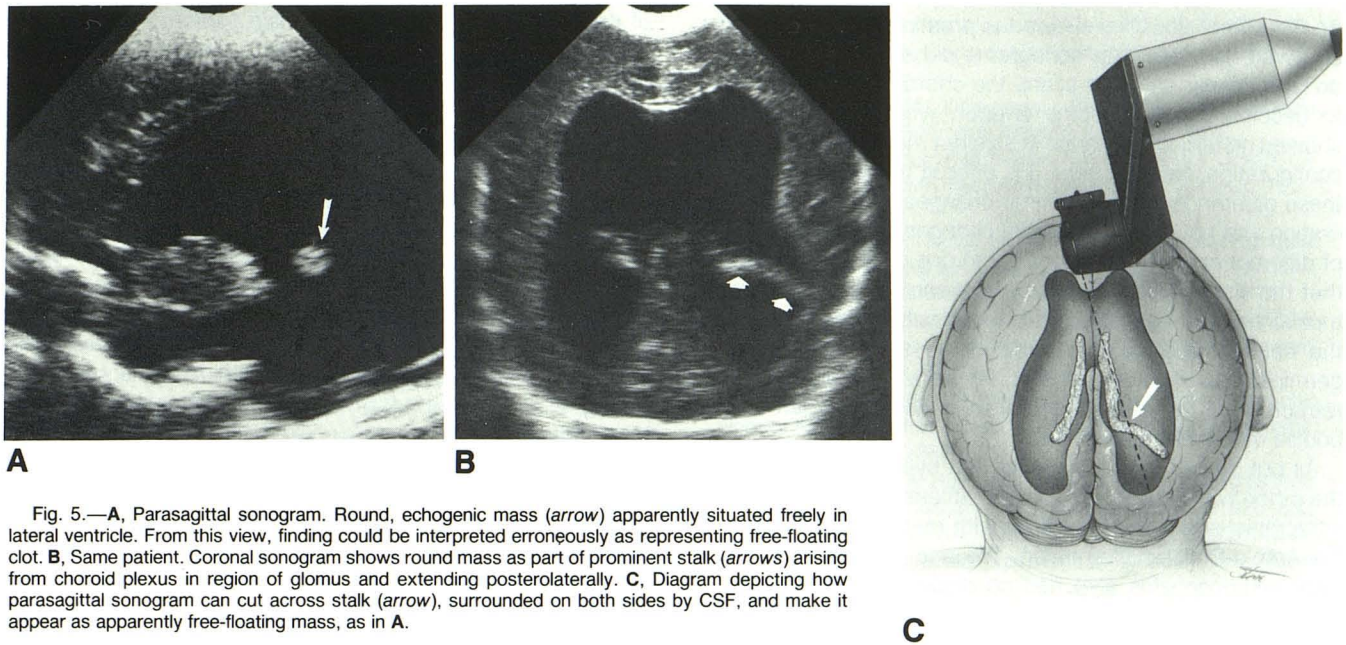
A   **B**   **C**

Fig. 5.—**A**, Parasagittal sonogram. Round, echogenic mass (*arrow*) apparently situated freely in lateral ventricle. From this view, finding could be interpreted erroneously as representing free-floating clot. **B**, Same patient. Coronal sonogram shows round mass as part of prominent stalk (*arrows*) arising from choroid plexus in region of glomus and extending posterolaterally. **C**, Diagram depicting how parasagittal sonogram can cut across stalk (*arrow*), surrounded on both sides by CSF, and make it appear as apparently free-floating mass, as in **A**.
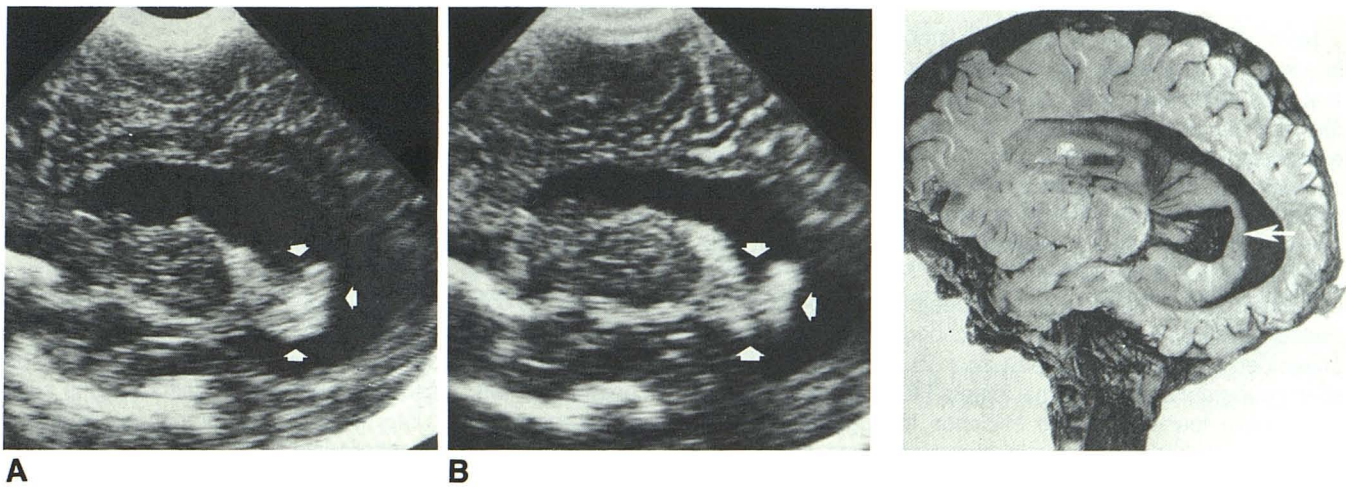

A   **B**

Fig. 6.—Parasagittal sonograms. Prominent, lobular choroid plexus in region of glomus has similar size and shape on sonograms 2 months apart. **A**, Prominent choroid plexus in region of glomus (*arrows*), situated in dilated lateral ventricle. **B**, 2 months later. Similar size and configuration of choroid plexus (*arrows*). Interval decrease in ventricular size.

Fig. 7.—Gross specimen in Arnold-Chiari malformation. Choroid plexus (*arrow*) in region of glomus has appearance similar to what may be seen sonographically. (Reprinted from [20].)

could be construed as representing a clot attached to the glomus by a thick fibrin strand. One may even be misled, occasionally, to believe that the ventricle contains a free-floating clot (fig. 5). In an effort to determine whether these unusual choroidal configurations were inherent or secondary to adherent intraventricular hematoma, we reviewed all available operative reports to determine the appearance of the cerebrospinal fluid (CSF) in these children when shunt was performed. Only in two cases was the CSF believed to be xanthochromic, and examination revealed crenated red blood cells. In addition, no evidence of bleeding was detected on CT. The choroid plexus was never imaged on the CT scans

since no intravenous contrast material had been administered. Altogether, 12 of the 17 infants either had a CT scan, CSF inspection, or both.

In none of the 13 infants who had more than one scan did we see the development of an anechoic center with an echogenic rim in the prominent choroid plexus. This change has been described with maturation of intraventricular clots [2–6]. Moreover, six of those who had follow-up scans showed no evidence of appreciable change in the size and appearance of the choroid plexus (fig. 6), two of them over the course of more than 7 months. Such changes would have been expected in cases of intraventricular hemorrhage [3, 5–

7]. The other six who showed a prominent choroid plexus and who had follow-up sonograms did show some change on later scans; on some scans the choroid plexus appeared to become larger, in some smaller, while still in others it showed no apparent change in size but merely had a different configuration. Nevertheless, it is difficult to say how much of these differences represent true changes in size and configuration and not merely apparent changes caused by the use of different scanning angles. Only in one case did we believe that there was strong evidence of a component of intraventricular hemorrhage; this child had a subependymal cyst at the head of the caudate nucleus, possibly from previous germinal matrix hemorrhage [8, 9], as well as unmistakable retraction of the large choroid echo complex seen in the bodies of the lateral ventricles.

All but one of the 18 patients had hydrocephalus, and of the other 17, 15 appeared to have at least some of the other intracranial stigmata associated with meningomyelocele and the Arnold-Chiari malformation. The one patient who did not have hydrocephalus also had no discernible abnormality in the appearance of the brain and was the only patient with a normal appearing choroid plexus.

Discussion

Normally, the choroid plexus of the lateral ventricle in infants is seen sonographically as an echogenic, smooth structure that runs posteriorly from the foramen of Monro, widening to form the glomus at the atrium, hugging the thalamus, and extending into the temporal horns [10]. Occasionally, the choroid plexus in the region of the glomus can be normally somewhat prominent [6, 10, 11] and has been described sonographically as being exceedingly large in one normal infant [6]. Such excessive prominence, however, is unusual in normal infants. A pedunculated appearance to presumably normal choroid plexi has been described in adults in earlier pneumoencephalographic literature [12], but has not been seen sonographically in normal infants [10].

We have found very little evidence to suggest that the excessive prominence seen in our cases was caused by intraventricular hemorrhage. Even in the patients in whom crenated red blood cells were seen in the CSF, one could postulate that these old red cells entered the CSF from previous meningomyelocele repair. The possible etiology of the prominent choroid plexus in meningomyelocele patients, therefore, remains unclear.

The literature dealing with the Arnold-Chiari malformation gives little aid in elucidating the nature of this finding. When describing the features of the Arnold-Chiari malformation or the intracranial findings of patients with meningomyelocele, neither radiologic [1, 13–19] nor pathologic [20–28] reports offer specific descriptions of the appearance of the choroid plexus in the lateral ventricles. In reviewing autopsies of 26 cases of spina bifida, Cameron [22] noticed a thickened choroid plexus in the region of the *fourth* ventricle, with frequent hemosiderin deposits and connective tissue proliferation in its caudal aspect. These changes were attributed to venous obstruction, presumably caused by compression

of the brainstem by the Arnold-Chiari malformation, accompanied by congestion, ischemia, and hemorrhage. Similarly, Feigin [24], in reviewing three cases of Arnold-Chiari malformation, noticed in one that the choroid plexus of the fourth ventricle was "considerably hypertrophied and hyperplastic." This finding was labeled a "malformation." However, neither author mentioned any abnormality in the appearance of the choroid plexus in the lateral ventricles.

Pathologic literature dealing with the choroid plexus itself, and not primarily with meningomyelocele or the Arnold-Chiari malformation, contains two reports that may describe findings similar to ours. In 1940, Liber and Lisa [29] described three infants with meningomyelocele, all with hydrocephalus, as having a "stalk" arising from the glomus of each choroid on both sides and ending in a "clublike" terminal enlargement. A similar finding was described in a fourth infant with unilateral hydrocephalus but without meningomyelocele: The choroidal abnormality was on the hydrocephalic side. Histologically, these excrescences were called "stromal nodular hyperplasia" and were assumed to represent neoplastic processes, precursors perhaps to intraventricular meningiomas [29]. Liber and Lisa ascribed the ventriculomegaly to these tumorlike processes. In contrast, Netsky and Shuangshoti [30] thought that these same findings represented a congenital malformation of the choroid plexus related to meningomyelocele, rather than "stromal nodular hyperplasia."

In 1935 Dandy [31] described "small primary tumors" in the lateral ventricles found incidentally at autopsies. Among his seven cases were four hydrocephalic infants who had "almond-sized" firm tumors attached to, or dangling "like earrings" from, the glomus of the choroid plexus. In two cases the findings were bilateral. Three of the four were interpreted histologically as "fibromas" and one as an "adenoma." Dandy hypothesized that these entities represented benign tumors arising in the connective tissue of the choroid plexus, perhaps forerunners of symptom-producing tumors later in life. He further hypothesized that, at least in infants, they might be congenital "rests" associated with other malformations such as hydrocephalus. Unfortunately, Dandy did not mention whether these hydrocephalic infants had associated meningomyelocele nor did he comment on the gross appearance of the brain. Dandy found a large amount of iron pigment in all of these "tumors" and speculated that what he saw was in fact an organized clot. He discounted this as being unlikely, both for lack of apparent cause and for reason of the marked symmetry of the findings.

Lastly, a specimen showing the choroid plexus in the lateral ventricle of an infant with Arnold-Chiari malformation was shown by Peach [20]. The choroid plexus appears morphologically larger than normal and is similar to some of our sonographic images (fig. 7).

It is perplexing not to find more extensive descriptions in the pathologic literature of a feature that we found sonographically to be almost universal in infants with meningomyelocele. It is tempting to propose, especially in the cases with a definite stalk-and-nodule configuration, that this finding represents a congenital malformation, perhaps even a reflection of the findings reported by Liber and Lisa [29] or by Dandy [31]

over 40 years ago. Perhaps the large choroid plexus represents early embryonal arrest, inasmuch as the choroid plexus is enlarged and lobular during certain stages of embryonal life [24, 25, 30]. Other factors, such as edema or an inflammatory response, should not be dismissed. One might also consider the possibility that these findings are in some way related to the hydrocephalus itself.

It is still possible that, at least in some infants, the prominent choroid plexus was in fact caused by hemorrhage, as suggested, for example, by the iron pigment found by Dandy [31]. Although not universally accepted [32], some maintain that intraventricular hemorrhage in term infants originates in [33, 34] and is confined to the choroid plexus, and therefore is of little clinical significance [34]. Perhaps neonates with Arnold-Chiari have a predilection for intrauterine or perinatal choroid plexus hemorrhage. This supposition, however, is questionable, in view of the lack of associated findings.

Further work is needed to corroborate the findings related to this communication and to clarify their nature. The unusual appearance of the choroid plexus in these patients is of great significance to the sonographer, because it may be almost identical in appearance to intraventricular hemorrhage. Knowledge of the appearance of the choroid plexus in children with spinal dysraphism should assist in differentiating between this apparent congenital malformation and intraventricular hemorrhage.

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