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## ***Caffey Award: Cranial Sonographic Findings in Meningomyelocele***

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# Caffey Award

## Cranial Sonographic Findings in Meningomyelocele

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Children with meningomyeloceles have problems involving several organ systems and often require repeated radiologic examinations. They have multiple cranial defects, particularly the Arnold-Chiari II malformation and associated hydrocephalus. Cranial sonographic findings in 29 patients included hydrocephalus with pointing of the frontal horns, asymmetry of the lateral ventricles, and relative enlargement of the occipital horns compared with the frontal horns. Partial absence of the septum pellucidum, abnormalities of the third ventricle including a prominent massa intermedia, prominent anterior commissure, and prominent suprapineal recess were also seen. A V-shaped tentorium cerebelli was demonstrated on axial scan as well as low position of the tent on coronal scan. The posterior fossa pseudomass was demonstrated. A prominent interhemispheric fissure was also seen in some patients. Sonography is an excellent method for following ventricular size and shunt function since repeated examinations can readily be performed.

Children with meningomyeloceles are a special group with problems involving several organ systems and repeated radiologic examinations are often required. They have numerous cranial defects, particularly the Arnold-Chiari (type II) malformation [1-3], and associated hydrocephalus, which can be evaluated with newer imaging techniques. Several recent reports have demonstrated the cranial computed tomography (CT) findings in these patients [4-7]. Sonography of the head is now a recognized diagnostic imaging method that offers a noninvasive, safe, inexpensive way of studying these meningomyelocele patients without the danger of ionizing radiation, while providing useful neuroanatomic information. We report our experience with B-mode gray scale sonography in the meningomyelocele patient.

### Materials and Methods

We retrospectively reviewed 65 sonograms obtained on 29 meningomyelocele patients. The Cincinnati Center for Developmental Disorders of the University of Cincinnati is a referral center for patients with this abnormality and infants are referred for multidisciplinary treatment within a few hours after birth. The spinal defect is repaired, if possible, within 24 hr after birth. The initial cranial sonogram is usually obtained within 2-4 days after birth. The sonograms are obtained with a commercially available contact B-mode gray scale ultrasonoscope or a high-resolution mechanical sector, real-time ultrasonoscope and recorded on video tape and x-ray film. The examinations are usually performed in the radiology department in the ultrasound room, which is equipped so that critical life support measures can be maintained. Bedside examinations are occasionally performed on severely ill infants using the real-time equipment; 3.5 and 5.0 MHz medium-focused transducers are used. Sedation is usually unnecessary, although chloral hydrate in a dose of 50 mg/kg is used on older infants. Technically adequate head examinations are performed only in children under the age of about 2 years who have open sutures and fontanelles. Serial scans of the entire head are obtained at 5 mm intervals in axial, coronal, posterior fossa, and sagittal projections. Our technique was described previously [8].

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## Results

Table 1 lists abnormalities we found associated with meningocele. The findings represent the frequency with which we detected the abnormality, not necessarily its frequency of occurrence in this group of patients.

### Lateral Ventricular Size

Although the spinal defect had been repaired, most patients were not shunted at the time of the initial examination and the size of the lateral ventricles varied considerably. In nine patients there was no or mild ventricular dilatation (fig. 1). There was moderate to marked dilatation in 15 patients and severe dilatation in five patients (fig. 2). In many patients, the ventricular size was shown to gradually increase on serial examinations until the patient was shunted. Examinations were not routinely performed after shunting; however, in several patients the ventricles decreased markedly in size after shunting. The lateral ventricles were mild to moderately asymmetrical in 17 of the 29 patients (fig. 2).

TABLE 1: Summary of Findings in Meningocele

| Finding                                                 | No.<br>(n = 29) |
|---------------------------------------------------------|-----------------|
| Hydrocephalus:                                          |                 |
| Not present                                             | 1               |
| Mild                                                    | 8               |
| Moderate to marked                                      | 15              |
| Marked to severe                                        | 5               |
| Anterior pointing of the frontal horns on axial scans   | 23              |
| Inferior pointing of the frontal horns on coronal scans | 25              |
| Occipital horns more dilated than frontal horns         | 19              |
| Asymmetry of the lateral ventricles                     | 17              |
| Cava septi pellucidi present                            | 4               |
| Partial absence of the septum pellucidum                | 16              |
| Dilated third ventricle                                 | 21              |
| Prominent massa intermedia                              | 5               |
| Prominent anterior commissure of the third ventricle    | 2               |
| Prominent suprapineal recess                            | 3               |
| Low position of the tent on coronal view                | 23              |
| V-shaped tent on axial view                             | 8               |
| Tentorial-cerebellar pseudomass                         | 9               |
| Prominent interhemispheric fissure                      | 12              |
| Herniation of third ventricle into parasella cistern    | 1               |

The occipital horns and atria were dilated more than the frontal and temporal horns in 19 of the patients (figs. 3 and 7). This has been noted in congenital hydrocephalus from other etiologies as well. The cerebral mantle was thinnest over the occipital horns (fig. 3).

### Configuration of the Lateral Ventricles

On axial view, a characteristic anterior pointing and medial concavity of the frontal horns has been described on CT examination [4]. This was seen in 23 of our patients (fig. 4). Inferior pointing of the frontal horns resulting from prominence of the caudate nuclei, which bulge medially into the lateral walls of the lateral ventricles has been described on ventriculography [2] and was seen in 25 of our patients (fig. 5).

### Evaluation of the Septum Pellucidum

The septum pellucidum was completely or partially absent in 16 of the patients (figs. 4, 7, and 11). A cavum septi pellucidi, or visible separation between the leaves of the septum pellucidum, was identified in four of the patients (fig. 6). This was the same incidence as seen in other patients with hydrocephalus and may not be related to the Arnold-Chiari II malformation.

### Third Ventricle Abnormalities

The third ventricle was dilated in all but seven of the patients with hydrocephalus. It was noted to be only mildly dilated in most patients, although it may become large in some cases (fig. 7). The massa intermedia has been described as being unusually enlarged in 82%–90% of Arnold-Chiari II brains [1]. The massa intermedia was not demonstrated in many of our patients, however, a prominent massa intermedia was identified in five (figs. 7 and 8) and appeared to lie unusually close to the foramen of Monro.

A prominent anterior commissure has been described by ventriculography and was identified in two patients (fig. 8A). Herniation of the third ventricle into the parasellar cistern (fig. 8B) was seen in one patient and an enlarged suprapineal

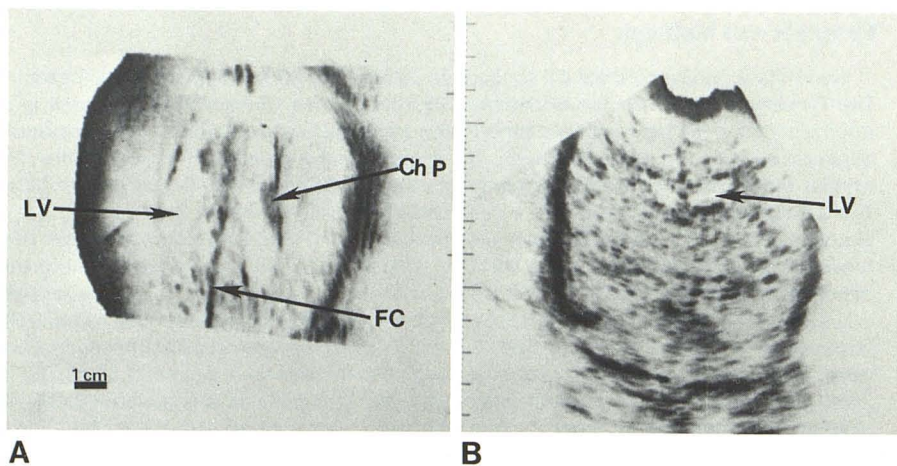
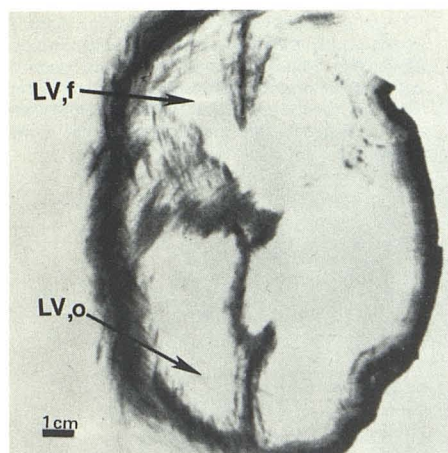
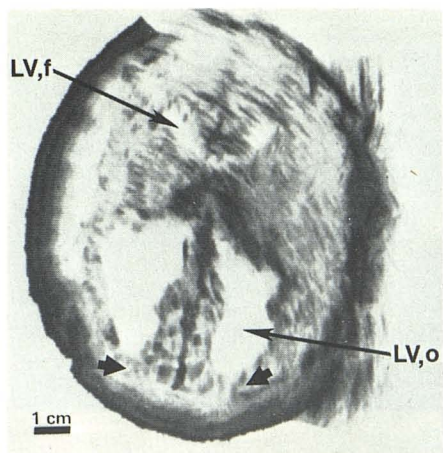


Fig. 1.—Mild hydrocephalus. Axial (A) and coronal (B) scans. Mild dilatation of lateral ventricles (LV). FC = falx cerebri, ChP = choroid plexus.

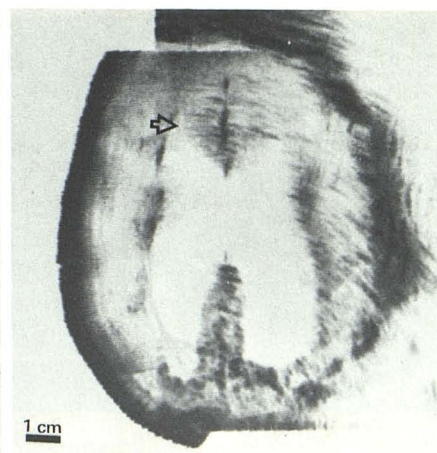




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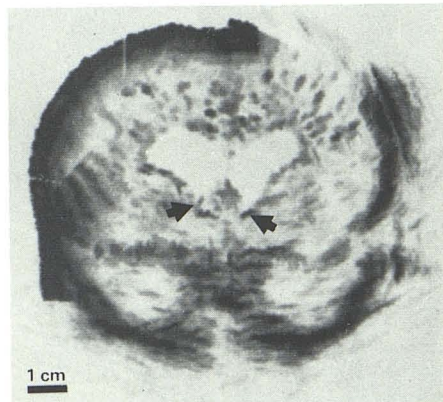
Fig. 2.—Asymmetric, marked hydrocephalus. Low axial scan. Asymmetric dilatation of lateral ventricles. LV,f = frontal horn, LV,o = occipital horn.

Fig. 3.—Lateral ventricles. Axial scan. More dilatation of occipital horns (LV,o) than frontal horns (LV,f) and thinnest cerebral mantle over occipital

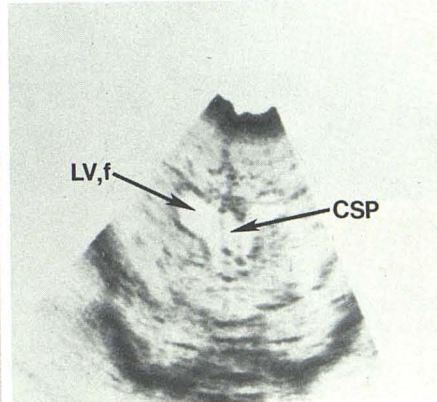
horns (arrows).

Fig. 4.—Lateral ventricles. Axial scan. Characteristic anterior pointing and medial concavity of frontal horns (arrow).

Fig. 5.—Lateral ventricles. Coronal scan through frontal horns of lateral ventricles. Inferior pointing of frontal horns (arrows).

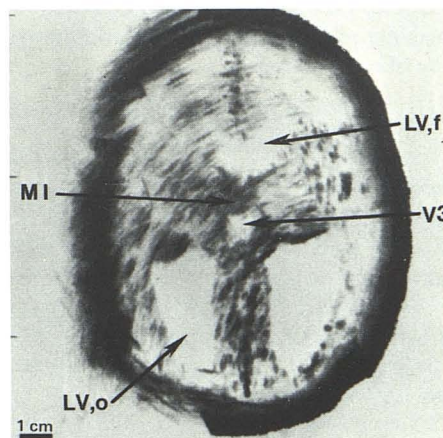


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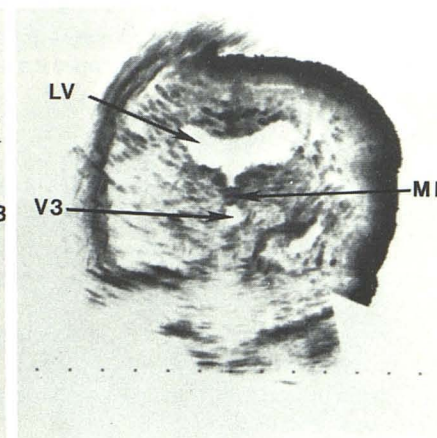


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Fig. 6.—Cavum septi pellucidi. Coronal scan. Cavum septi pellucidi (CSP) between dilated frontal horns of lateral ventricles (LV,f).



A



B

Fig. 7.—Absent septum pellucidum. Dilated third ventricle with prominent massa intermedia. Axial (A) and coronal (B) scans. Absent septum pellucidum and moderate dilatation of lateral and third ventricles (V3). Occipital horns (LV,o) more dilated than frontal horns (LV,f). MI = massa intermedia.



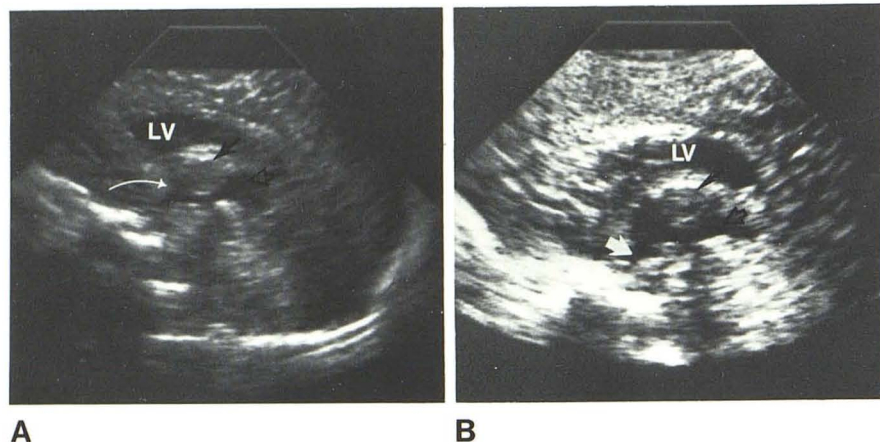


Fig. 8.—Third ventricle abnormalities. Sagittal scans on two different patients. Enlarged massa intermedia (black arrows), prominent anterior commissure (curved arrow), herniation of third ventricle into the parasellar cistern (straight white arrow), and enlarged suprapineal recess (open arrows).

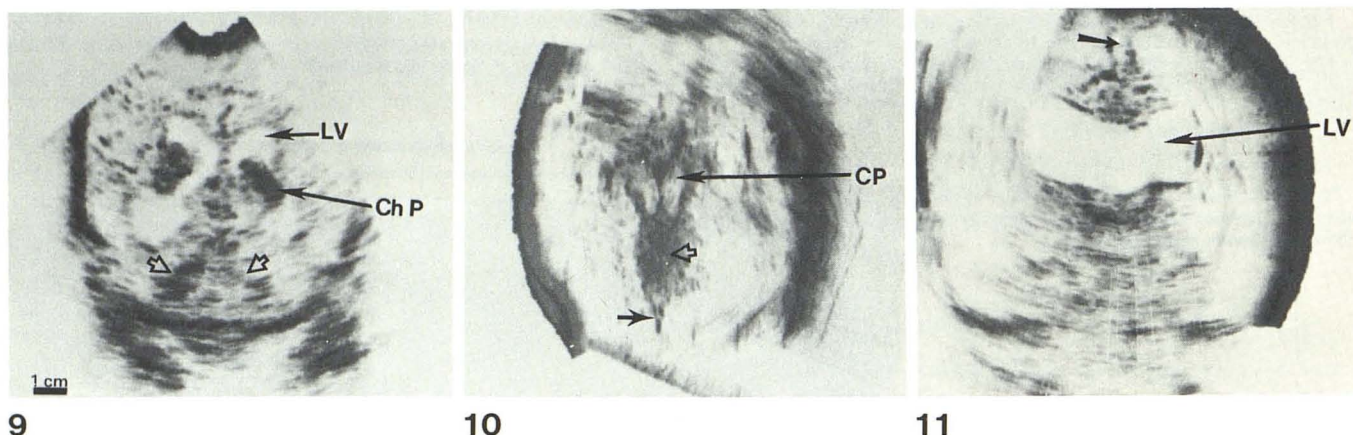


Fig. 9.—Low position of tentorium cerebelli. Coronal scan. Relatively low position of tentorium cerebelli (arrows) and small posterior fossa. Dilated lateral ventricles (LV) with choroid plexuses (ChP).

Fig. 10.—Tentorial-cerebellar pseudomass. Low axial scan. Heart-shaped tentorial-cerebellar pseudomass (open arrow) and V-shaped tentorium cere-

belli posteriorly (solid arrow). CP = cerebral peduncles.

Fig. 11.—Prominent interhemispheric fissure. Coronal scan. Prominent interhemispheric fissure (arrow) and moderate dilatation of lateral ventricles (LV) with absence of septum pellucidum.

neal recess was noted in three patients (fig. 8A and 8B). Most of these abnormalities of the third ventricle were identified only on sagittal views, and this view was not available on all of the patients. Beaking of the tectum and herniation of the third ventricle into the quadrigeminal cistern has been described but was not demonstrated on any of our patients.

#### Tentorial-Cerebellar Pseudomass

The posterior fossa abnormalities in the Arnold-Chiari II malformation include downward displacement of the cerebellum, medulla, and fourth ventricle, with a relatively small posterior fossa. In 23 of our patients, a relatively low position of the tentorium cerebelli was seen on coronal scans, with a small posterior fossa (fig. 9). The fourth ventricle was not identified in any of our patients, probably because of its low position in the posterior fossa and because it is not dilated in this malformation.

The tentorial-cerebellar pseudomass recently described on CT examinations [4, 6] was seen in nine of our patients (fig. 10). On axial scans the leaves of the tentorium come

together to form a sharp angle posteriorly, producing a V-shaped configuration, rather than the normal U shape. The tentorium is hypoplastic, with a wide incisura through which the cerebellum bulges upward, producing the heart-shaped tentorial-cerebellar pseudomass.

#### Interhemispheric Fissure

The interhemispheric fissure was identified as being prominent in 12 of the patients and was associated with hydrocephalus (fig. 11). It has been postulated that communicating hydrocephalus with distal block at the foramen magnum may result in simultaneous dilatation of both the ventricles and the interhemispheric fissure [7]. It was noted in several cases that the interhemispheric fissure enlarged when the ventricles diminished after ventricular shunting.

#### Ventricular Shunt Function

Occasional patients returned for study after ventricular shunting with symptoms of shunt malfunction. Figure 12 [8]



Fig. 12.—Ventricular shunt function. Axial (A) and coronal (B) scans at age 2 days. Moderate to severe dilatation of lateral (LV) and third (V3) ventricles. SP = septum pellucidum, LV,b = lateral ventricle, body.

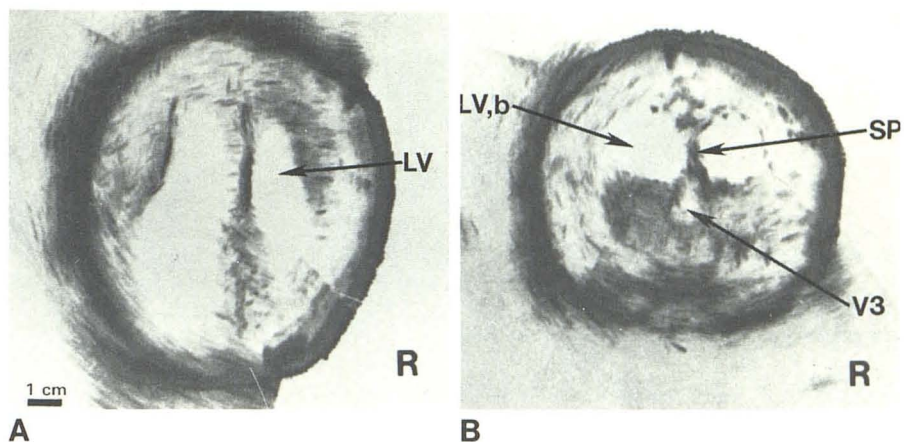


Fig. 12.—C and D, Follow-up 9 days later after temporary ventricular shunt. Lateral and third ventricles diminished, although still mildly enlarged.

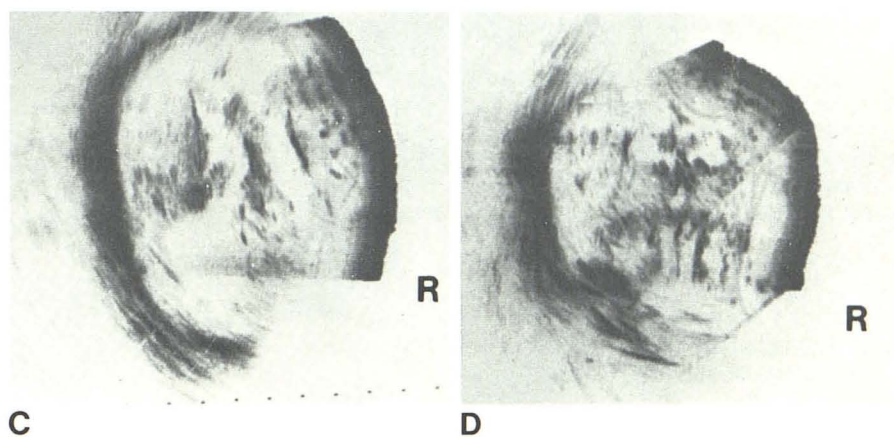


Fig. 12.—E and F, 2 weeks follow-up for increasing head size. Increased ventricular size indicates shunt (S) malfunction. Malfunctioning shunt replaced.

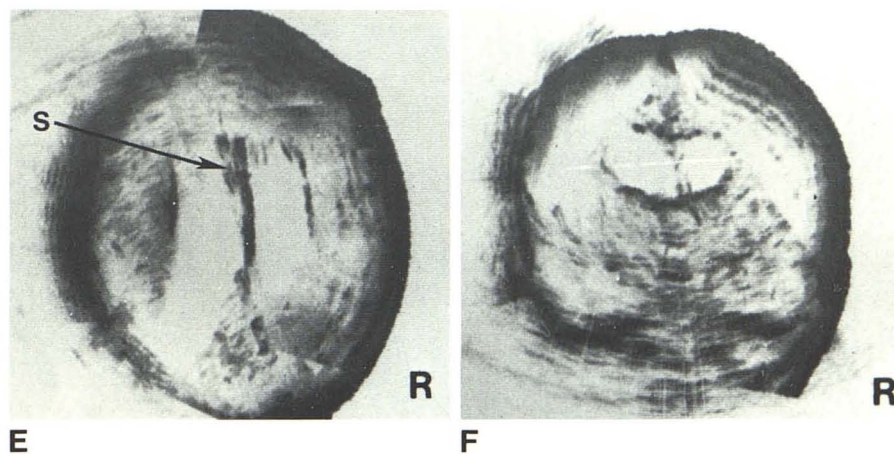
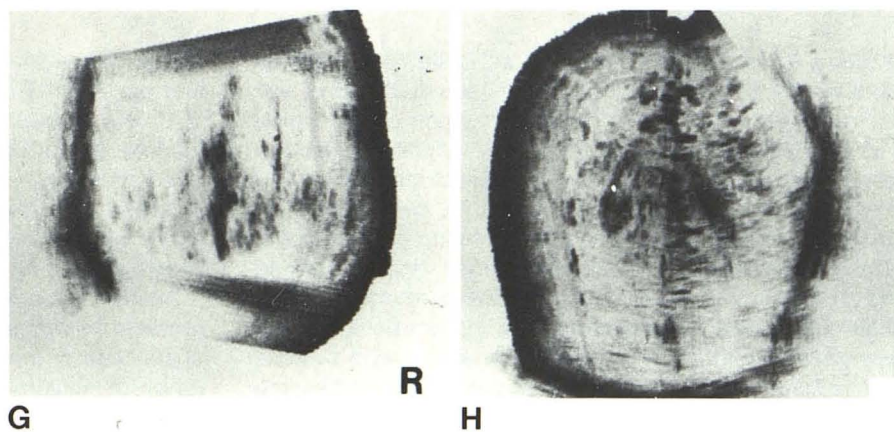


Fig. 12.—G and H, Postoperative examination 1 month later. Ventricles collapsed and slitlike. (Reprinted from [8].)





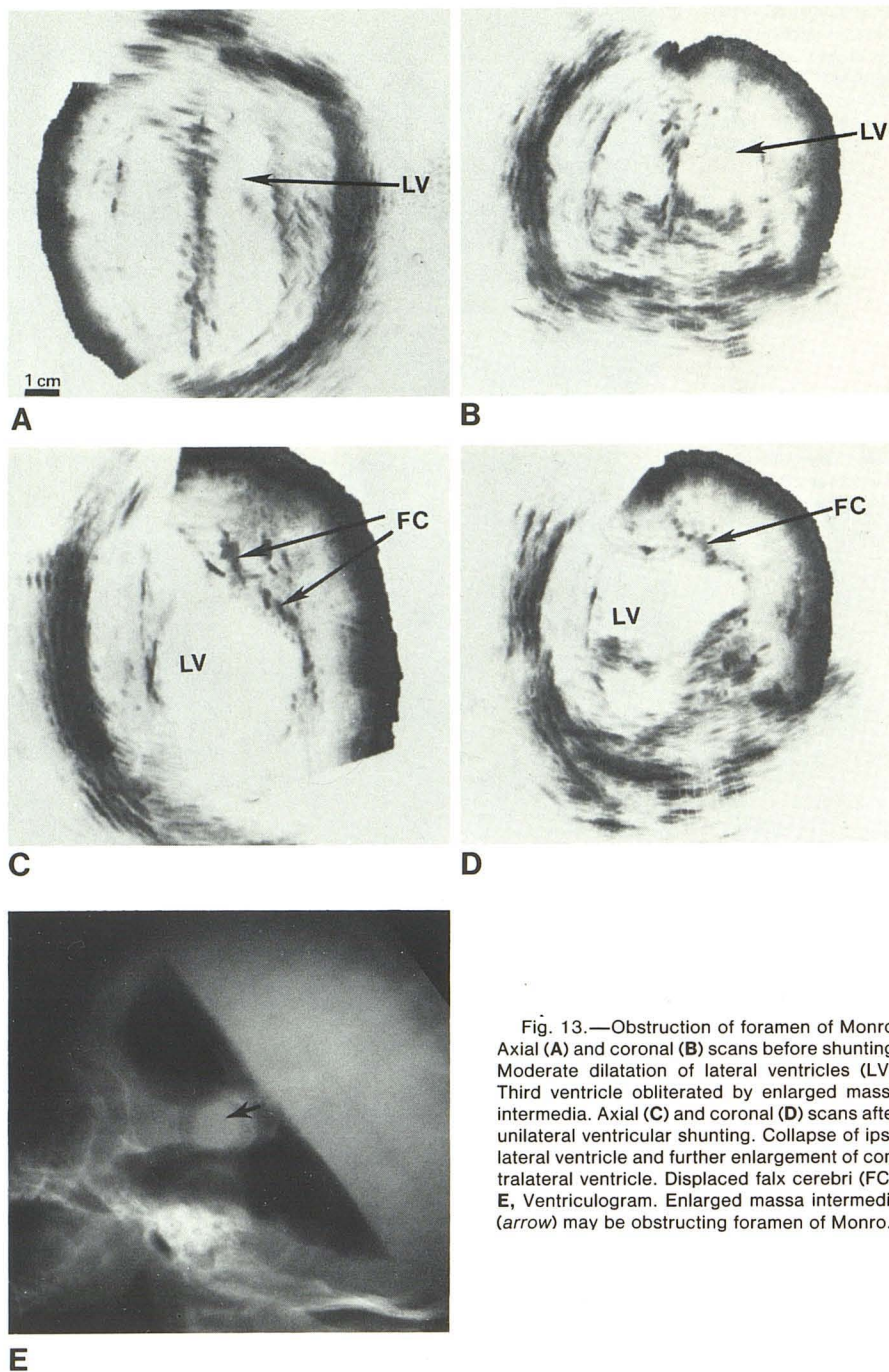


Fig. 13.—Obstruction of foramen of Monro. Axial (A) and coronal (B) scans before shunting. Moderate dilatation of lateral ventricles (LV). Third ventricle obliterated by enlarged massa intermedia. Axial (C) and coronal (D) scans after unilateral ventricular shunting. Collapse of ipsilateral ventricle and further enlargement of contralateral ventricle. Displaced falx cerebri (FC). E, Ventriculogram. Enlarged massa intermedia (arrow) may be obstructing foramen of Monro.

shows a 37 week gestation infant with mild head enlargement whose first sonographic examination at 2 days of age demonstrated moderate to severe dilatation of the lateral and third ventricles. After a temporary ventricular shunt, a follow-up examination 9 days later demonstrated diminished lateral and third ventricles, although they were still mildly enlarged. The patient returned for follow-up examination 2 weeks later because of increasing head size suggesting shunt malfunction. Increasing ventricular size since the last examination was demonstrated. The malfunctioning shunt was replaced and a final postoperative examination 1 month later showed that the ventricles had collapsed and become

slitlike. When the ventricles collapse, the skull diminishes and the sutures close and even tend to overlap, causing technical problems in obtaining good quality scans. Zimmerman et al. [4] and Naidich et al. [5-7] reported that even after shunting, the ventricles tend to remain moderately enlarged and on serial studies their size seems to correlate with the variations in shunt function rather than the underlying disease process.

Another patient with a ventricular shunt developed progressive enlargement of the contralateral ventricle indicating obstruction of the foramen of Monro (fig. 13). Ventriculography demonstrated only an enlarged massa intermedia,

which may have been obstructing the foramen of Monro sufficiently to require bilateral ventricular shunts.

### Discussion

Meningomyelocele patients with Arnold-Chiari II malformation have been reported to show a variety of abnormalities on cranial CT and ventriculographic examinations [2-7]. We found that many of these features can also be demonstrated by cranial sonographic examination. The primary indication for performing cranial sonography in our patients was to determine ventricular size, and this was successfully determined on all examinations. Therefore, cranial CT and ventriculography were unnecessary in most patients and performed only rarely in unusual or complicated situations such as shunt malfunction and unilateral ventricular enlargement (fig. 13) to evaluate the patency of the third ventricle and foramina of Monro.

Sonography has the advantage of being a rapid, safe, and less expensive method for studying these patients with no radiation exposure with repeated examinations. This is particularly important for following ventricular size and shunt function. The need for deep sedation and anesthesia is also avoided. Sonography has one disadvantage in that only patients with open sutures and fontanelles can be evaluated by this method.

Since the advent of cranial sonography at our institution, ventriculography and CT are rarely performed on infants with this malformation.

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