The Role of Intracranial Venous Hypertension in the Pathogenesis of Hydrocephalus *

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The distended scalp veins of a hydrocephalic infant indicate the cephalic venous hypertension which accompanies active hydrocephalus. Although movement of fluid requires relative hydrostatic pressure differences between cerebrospinal fluid (CSF) and intracranial venous structures, in certain instances venous hypertension, rather than being a passive consequence of elevated intracranial pressure, might assume a primary role in the pathogenesis of hydrocephalus. The present study was undertaken to determine the relationship between sagittal sinus venous pressure (SSVP), dural sinus radiographic anatomy and cerebrospinal fluid pressure (CSFP) in a series of patients with infantile hydrocephalus.

Materials and Methods

Thirty hydrocephalic patients, 11 of whom had an associated myelomeningocele, ranging in age from two weeks to 25 months, were studied. Under light sedation [nembutal 1 mg/kg and Vistaril[R] (hydroxyzine palmoate) 1 mg/kg], the superior sagittal sinus and spinal subarachnoid space were cannulated with 20 gauge needles and the pressures were simultaneously recorded (CSFP and SSVP). After obtaining resting pressures, both internal jugular

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veins were compressed separately and pressures were recorded. Normal saline was then infused into the spinal subarachnoid space with a Harvard infusion pump at a rate sufficient to elevate CSFP to approximately 600 mm of water in two to five minutes. The rate and volume of saline infusion were recorded. Following saline subarachnoid infusion, the pressure was allowed to return to pre-infusion levels. To test ventricular-spinal subarchanoid communication, two cc indigo carmine were injected into the lateral ventricle and the color of the fluid obtained from the spinal needle was noted. Superior sagittal sinograms were then performed on 28 out of 30 patients by injecting four cc of 60 per cent Conray over a period of two seconds and taking serial radiographs every two seconds for 12 seconds. As a final procedure, pneumoencephalography or ventriculography (depending on communication) was done.

Results

Resting pressure in the superior sagittal sinus (SSVP) exceeded CSFP in one third of all patients and, in another third, SSVP equalled CSFP. In only the remaining one third did normal pressure relationships exist; i.e., CSFP greater than SSVP.

The rise in SSVP with jugular compression corresponded to radiographic configuration of the sagittal and tranverse sinuses in 27 of 28 cases. Bilateral transverse sinus filling was demonstrated by sinography in 18 patients, although in some the sinuses were slightly asymmetrical. Among the remaining ten sinograms with filling of only one transverse sinus, the right was visualized in seven studies and the left in three studies.

Injection of saline into the spinal subarachnoid space (into the opposite lateral ventricle in two cases) produced a parallel rise in SSVP and CSFP in ten patients. Figure 1 exemplifies all cases with parallel pressure rises. The SSVP (top tracing) and CSFP (ventricular, bottom tracing) begin at 110 and 130 mm of water respectively. Saline was infused into the lumbar subarachnoid space at a rate of 3.1 cc per minute. SSVP and CSFP reach 700 and 720 mm of water while the differential pressure (middle tracing) remains near zero. Nine of the ten patients with parallel pressure changes had an associated myelomeningocele, and only two patients with myelomeningocele demonstrated a non-parallel pressure relationship.

In 20 patients, SSVP remained near baseline values as CSFP rose during saline subarachnoid infusion. The amplitude of venous pulsations did increase in proportion to the rise in CSFP. Figure 2 illustrates the non-parallel pressure change. SSVP and

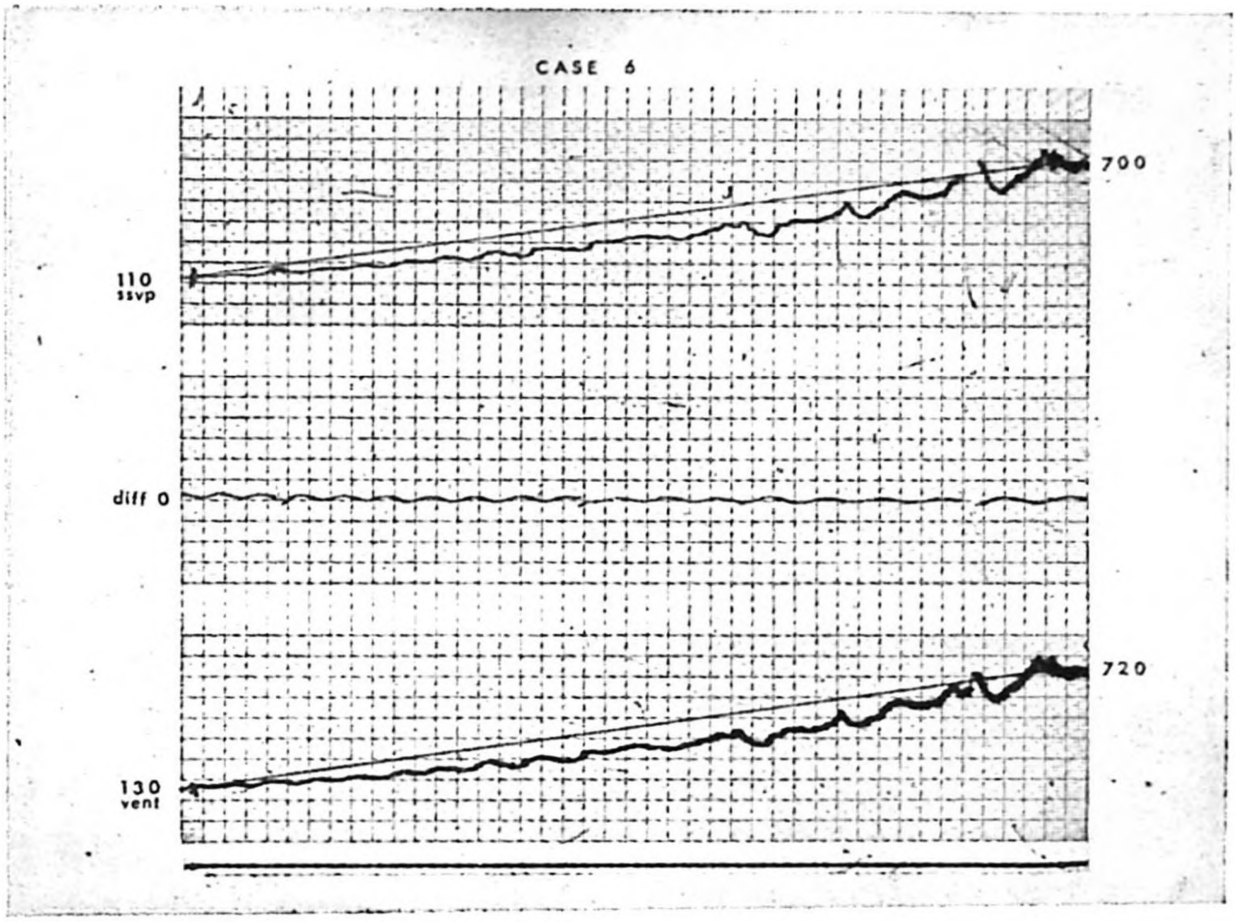


Figure 1. Example of parallel pressure rises.

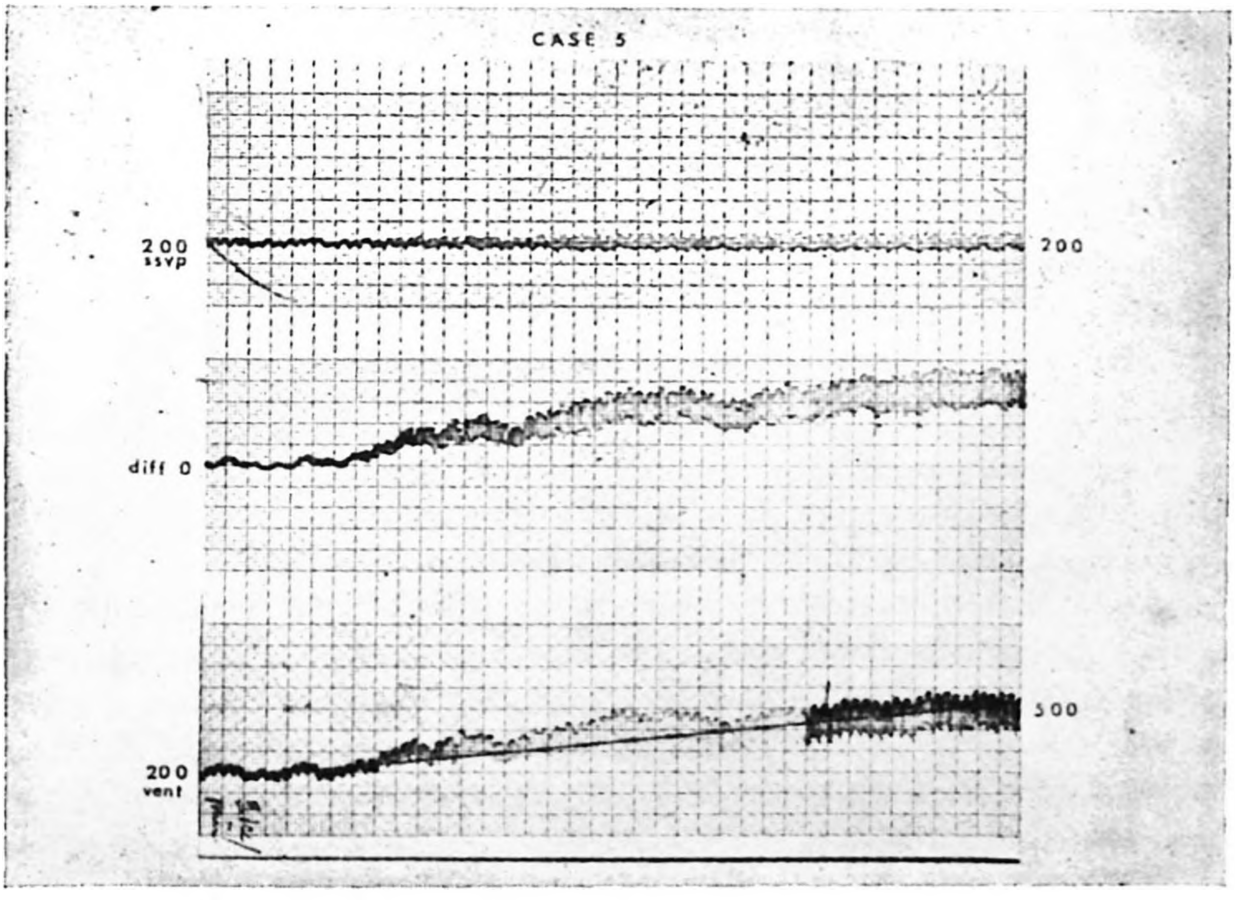
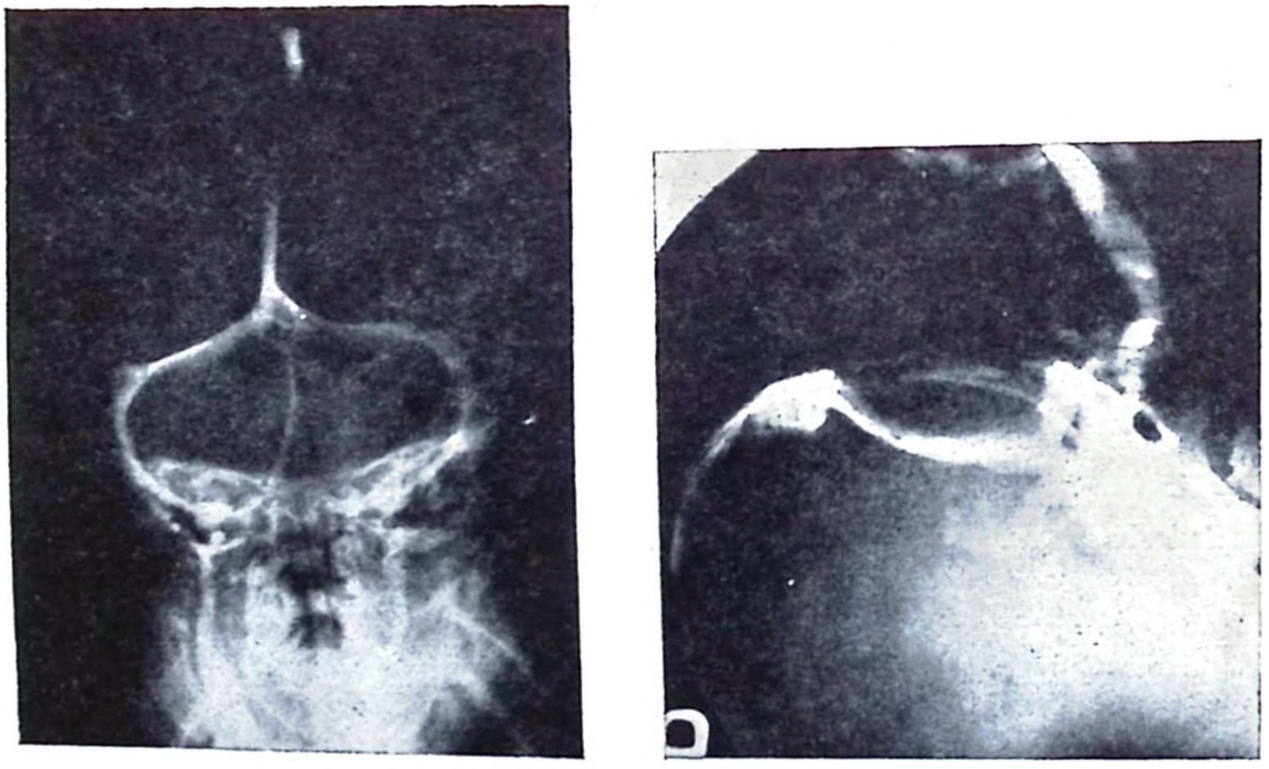


Figure 2. Illustration of non-parallel pressure change.

CSFP begin at 200 mm of water and SSVP remains at baseline value despite elevation of CSFP at 500 mm of water. The pressure differential is reflected in the middle tracing.

Sagittal sinograms fell into two patterns, normal and abnormal. The normal sinus configuration is illustrated in Figures 3 and 4. In the frontal view, the transverse sinuses pass in a horizontal direction from the torcular Herophili and, in the lateral view, the transverse sinus has a slight upward convexity. The second pattern, an abnormal sinus configuration, was observed in the sinograms of all patients with myelomeningoceles. In this pattern (Figures 5 and 6) the torcular Herophili occupies a low position and the transverse sinuses course along the floor of the posterior fossa. In the lateral view the transverse sinus has a downward convexity rather than the normal upward convexity.



Figures 3 and 4. Normal sinus configurations.

Discussion

In 1914 Walter Dandy[1] recognized intracranial venous occlusion as one cause of hydrocephalus. Almost 50 years later Bering[2] produced hydrocephalus in dogs by ligating cephalic veins in the neck. In the same study Bering demonstrated a rise in SSVP as well as CSFP in dogs made hydrocephalic by the injection of kaolin into the cisterna magna.

Shulman and Ransohoff[3] studied 15 hydrocephalic children and found that in 11 cases baseline SSVP was equal to CSFP. Ki-

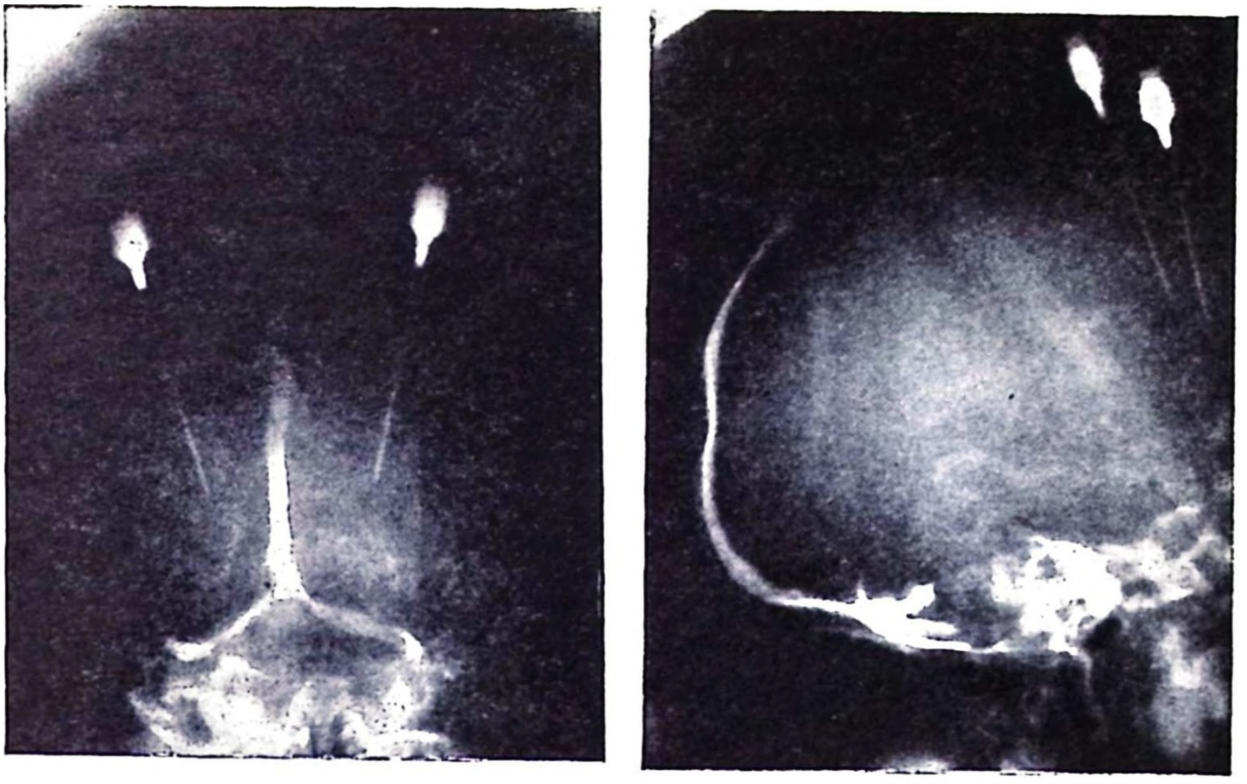


Figure 5 and 6. Abnormal sinus configurations.

nal [1] implicated structural stenosis or atresia of the transverse sinus as the basic defect in certain cases of infantile hydrocephalus, based on sinograms. We interpret these findings as normal.

The parallel rise in the SSVP and CSFP occurred in only two children without lumbar myelomeningocele and in only two children with myelomeningocele did the parallel rise fail to occur with saline infusion. In one of the children without myelomeningocele the superior sagittal sinogram showed the sagittal sinus needle to be in a cortical vein. Shulman and Ransohoff[3] found that the pressure within such veins normally approaches CSFP which would eliminate this case from further consideration.

The parallel pressure rise during saline infusion appears to be independent of resting SSVP, even in the cases with initially elevated SSVP. Our work confirms the clinical and experimental observations of Shulman and Ransohoff[3] that the resting SSVP is frequently elevated in cases of infantile hydrocephalus. They postulated that a collapse of the membranous portion of the sagittal sinus is responsible for the elevation of resting SSVP. Of our 18 patients with initially elevated SSVP, ten showed a parallel rise in SSVP and CSFP during saline infusion. If their postulates had been correct, we would expect to see the parallel rise in all cases with initially elevated SSVP, which did not occur.

Daniel and Strich[5] demonstrated the abnormalities of the posterior fossa occurring with Arnold-Chiari malformation. The attachment of the tentorium may be so low that it often appears to spring from the margin of the foramen magnum, the transverse sinus running near the margin of the foramen magnun. The frequency of the Arnold-Chiari malformation accompanying lumbar myelomeningocele approaches 100 per cent. This abnormality of the transverse sinus was demonstrated in the sinograms of all of our cases with myelomeningoceles. Such a structural deformity of the sinuses might make them liable to kinking or collapse by extrinsic pressure from cerebrospinal fluid or by the ectopic cerebellar tissue accounting for a parallel rise in CSFP and SSVP. We also demonstrated abnormal venous communications between the transverse sinus and extracranial veins in three cases, suggesting that a partial block occurs near the jugular foramen. The development of abnormal venous communication could account for the lack of parallel pressure rise in the one child with myelomeningocele. This child was 19 months old and at this age adequate venous collateral might have developed.

It was impossible to equate pressure rise with rate or volume of saline infusion, head size or cortical mantle thickness.

Sagittal sinus emptying time was prolonged in the children with myelomeningoceles, especially with elevated SSVP. Only later in the study was the procedure standardized to measure venous sinus emptying time.

Recently we have confirmed the dynamic nature of this block by performing sagittal sinography before (Figure 7) and after (Figure 8) elevation of CSFP. With increased intracranial pressure, the abnormal sinus collapses and collateral circulation becomes evident.

Summary

We have shown that sagittal sinus venous pressure is frequently elevated in cases of infantile hydrocephalus. Of more importance, we found that increasing intraventricular pressure is accompanied by a parallel rise in sagittal sinus venous pressure, almost exclusively in children with myelomeningoceles. This rise in pressure is probably related to a dynamic block of the malpositioned transverse sinuses accompanying the Arnold-Chiari malformation. We suspect a structural obstruction of the venous sinus, not related to intracranial

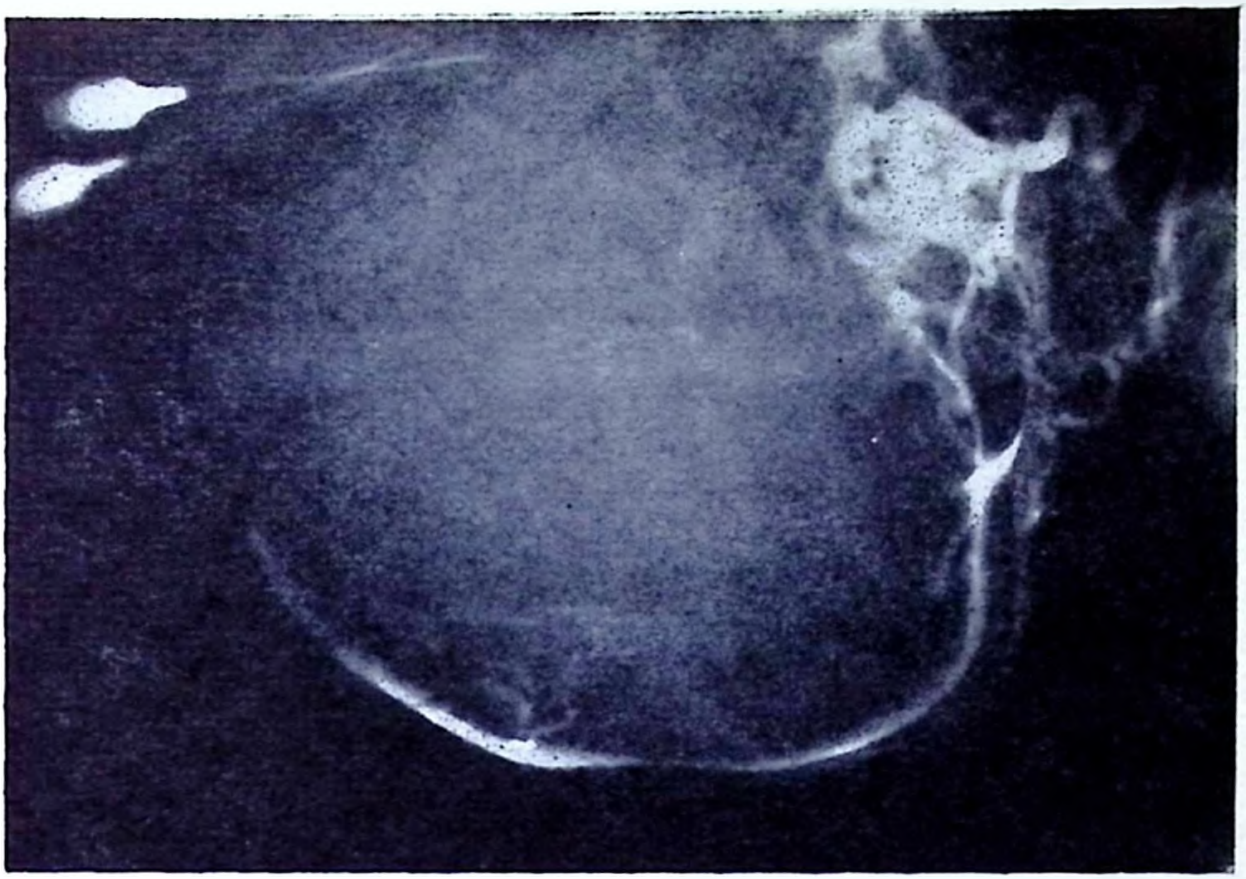


Figure 8. Sagittal sinography after elevation of CSFP.

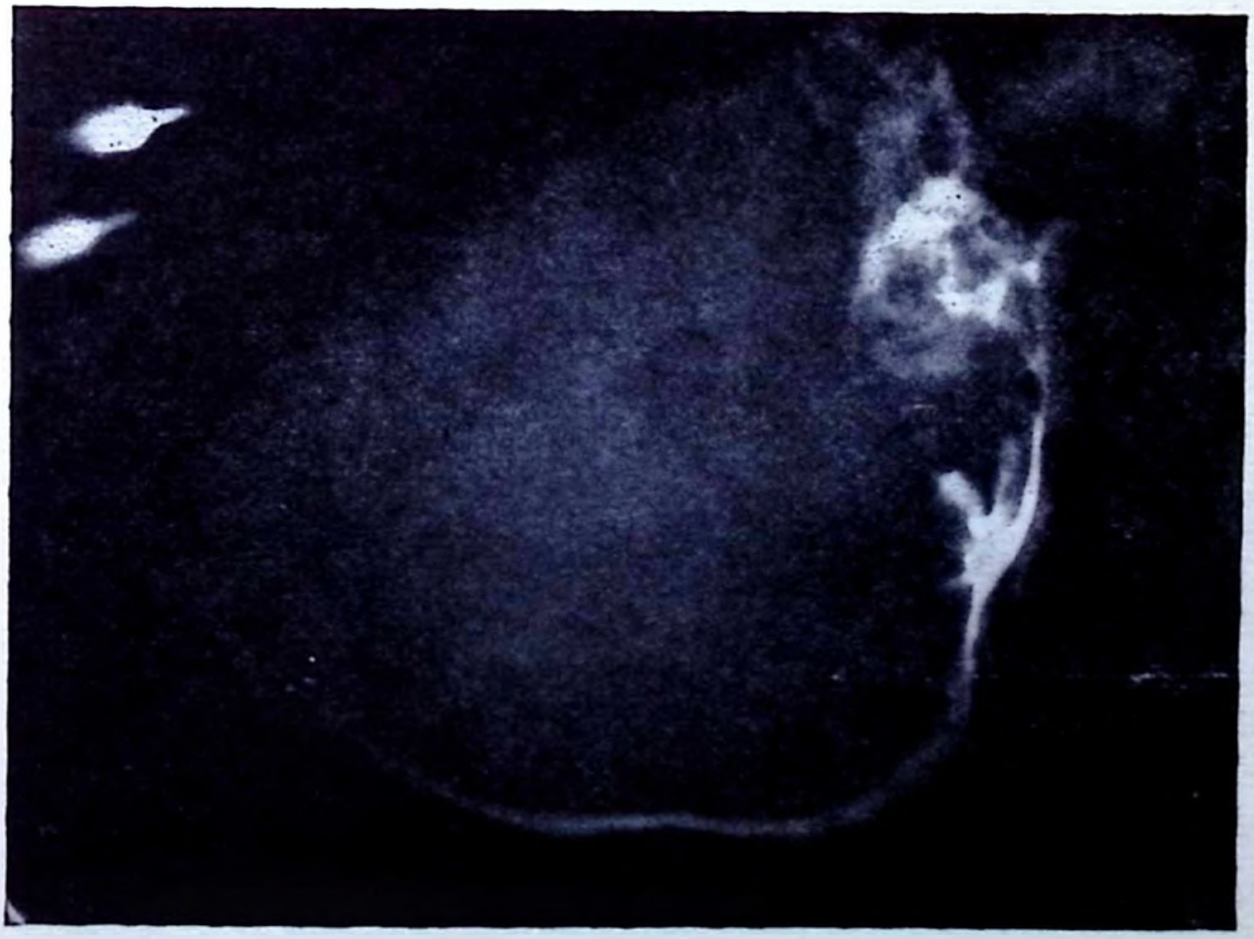


Figure 7. Sagittal sinography before elevation of CSFP.

pressure, in the group of children with elevated baseline SSVP not rising in response to saline infusion. We suggest that venous hypertension may have a primary etiologic role in certain cases of infantile hydrocephalus.

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