Pantopaque Ventriculography in Infants with Myelomeningocele*

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er an 18-month period ending Dec. 31, 1962, 114 newborn infants with myelomeningocele were admitted to the Columbus Children's Hospital, and 91 were judged suitable for surgical treatment. Of these, 70 (76.9 per cent of those repaired) were destined to undergo further operation for significant developing hydrocephalus. The mechanism of this hydrocephalus is little known. Controversy, centuries old, continues. We believe that our studies with Pantopaque clarify some controversial concepts of the circulation of spinal fluid in the posterior fossa.

Infants with myelomeningocele frequently have disorders of the intracranial central nervous system which are not evident upon physical examination or in plain films of the skull. Heretofore, these disorders have been delineated with air ventriculography, but air contrast studies rarely demonstrate adequately the aqueduct of Sylvius or the structures of the posterior fossa and upper cervical area in these tiny infants. Further, the presence or absence of communication of the ventricular system with the subarachnoid space usually cannot be determined. The Blackfan test (observing fluid obtained at lumbar puncture for dye previously placed in the ventricular system) cannot be used to show this communication because lumbar puncture is unsafe in these patients after repair of myelomeningocele.

Morgagni* in the 18th century stated his belief that myelomeningocele results from the pressure of cerebrospinal fluid descending through the central canal of the spinal cord—the basis of the hydromyelic theory of pathogenesis. In 1886 von Recklinghausen* challenged this theory and postulated that the defect results from a primary failure of closure of the neural tube. Weed* in 1917 believed that development of the subarachnoid space in the embryo depends upon fluid of the ventricular system finding its way out of the rhombic roof of the 4th ventricle at the proper time. If this does not occur, or if it occurs too late, the subarachnoid space may fail to develop adequately.

Gardner* pointed out that Weed's work may explain why both obstructive and communicating types of hydrocephalus are seen in patients with myelomeningocele. Indeed, both types of hydrocephalus sometimes occur in the same patient. Further, Gardner felt that this failure of the fluid to issue forth at the proper time from the 4th ventricle can account for the rupture of the neural tube, the hydromyelia, and many of the findings commonly seen in patients with myelomeningocele. Thus, Gardner and Weed were in consonance with the hydromyelic theory of Morgagni.

We felt that positive contrast ventriculography would overcome some of the inadequacies of air ventriculography, and could provide information about pathogenesis. The relative safety of Pantopaque ventriculography is now almost universally accepted.*

Methods and Materials

Fourteen infants with myelomeningocele were subjected to Pantopaque ventriculography after surgical repair of the defect but before shunting procedures for hydrocephalus. All patients were less than 3 months of age except 1, who was 19 months old.

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Pantopaque Studies in Myelomeningocele

Depending on the size of the head, ¾ or 1 cc. of Pantopaque is injected through a No. 18 needle inserted through the coronal suture line into the lateral ventricle, in conjunction with fractional air ventriculography. The head is manipulated so that the dye, if unobstructed, finds its way by gravity into the 3rd ventricle, the aqueduct of Sylvius, the 4th ventricle, and finally through the foramina of the 4th ventricle into the subarachnoid space. This has been done largely without the aid of fluoroscopy, taking serial roentgenograms over whatever span of time is necessary. Only light sedation is used.

Eleven of the procedures were satisfactory technically. Details of the 3 unsatisfactory procedures are described below.

1) Pantopaque ventriculography was performed on a 3-month-old patient who had had several unsatisfactory shunting procedures. Meningitis developed, requiring removal of his shunt. There was immense hydrocephalus, and the dye was lost into the dilated ventricles and pineal recess before it entered the aqueduct, presumably because of the gross distortion of the ventricular system.

2) A 3-week-old infant, who on examination appeared to be free of hydrocephalus, was extremely active during the procedure despite premedication with a dosage that usually produces satisfactory sedation. Because of the infant's vigorous movements during the procedure, the dye was lost into the posterior horns before it could be manipulated into the 3rd ventricle. Since the air study performed concurrently showed a normal ventricular system, a second attempt at Pantopaque ventriculography was not believed warranted.

3) Pantopaque was inadvertently placed in the interhemispheric subdural space in a 5-week-old infant with moderately severe hydrocephalus. The patient appeared to suffer no harmful effects from this injection. A ventriculostriatal shunt was performed with good results. However, at the age of 2 months the infant expired of septicemia arising from urinary-tract infection. At postmortem examination the dura mater stripped easily from the arachnoid. Microscopic studies of the brain failed to demonstrate any evidence of untoward reaction to the Pantopaque.

Results

Fig. 1 shows a Pantopaque ventriculogram of a 19-month-old infant with typical Arnold-Chiari malformation. There is inferior displacement of the stem and buckling of the superior spinal cord. Dye is seen in the spinal subarachnoid space, but not in the intracranial subarachnoid space. This condition persisted even though the infant was placed in the hanging-head position. The cerebellum extends down to the 2nd cervical vertebra. Of particular interest is the puddling of the dye seen in the cervical subarachnoid space.

A photograph of the midsagittal section of a hindbrain with Arnold-Chiari malformation is shown in Fig. 2 for comparison with Fig. 1. The inferior extent of the cerebellar tissue, the S-shaped distortion of the area of medullospinal junction, and the distortion of the 4th ventricle offer strong evidence that the cerebellum is squeezed through the foramen magnum rather than pulled, as was at one time believed.

Fig. 3 shows the Arnold-Chiari malformation in a patient with severe craniolacunia, and also demonstrates the frequently seen

Fig. 1. Infant aged 19 months. Pantopaque ventriculogram showing inferior displacement of 4th ventricle, deformity of the column of dye typical of Arnold-Chiari malformation and puddling of dye with obstruction in cervical subarachnoid space (arrow). Dye did not enter the intracranial subarachnoid space.
thinning or partial agenesis of the corpus callosum. We feel that this combination represents evidence of very early intrauterine hydrocephalus. The dye has reached the intracranial subarachnoid space. There is Pantopaque in a dilated pineal recess.

Fig. 4 demonstrates an aqueduct of remarkably small caliber (relative aqueductal stenosis). The small 4th ventricle is in normal position.

Relative aqueductal stenosis with a deformity of the column of dye suggestive of Arnold-Chiari malformation, but with a 4th ventricle of normal size and position, is seen in Fig. 5. Perhaps this represents a stage in the development of the full-blown malformation. Since we believe that the malformation is a secondary effect of hydrocephalus, it is
postulated that the malformation need not occur only in utero, but also occurs in the early months of extrauterine life of the infant. Fig. 5 also shows puddling of dye and access of dye to the spinal, but only sparsely to the intracranial, subarachnoid space.

Radiographs of a patient who never required shunting are shown in Fig. 6. Of interest are the large amount of dye in the cisterna magna and also the relatively larger and presumably normal aqueduct, as contrasted to those in Figs. 4 and 5, and to those in Figs. 7 and 8 in which the aqueduct appears as a very fine filament of Pantopaque. In Fig. 8 the aqueduct appears to taper gradually to atresia. The large massa intermedia is often seen, as has been reported, and may be related to the general early compression of basal structures.

Gardner has referred to spontaneous relief of the early obstruction at the 4th ventricular foramina by several routes. Rupture can occur through the pineal recess. We believe that Fig. 9 demonstrates a striking enlargement or diverticulum of the pineal recess.

Discussion

Table 1 tabulates some of the findings of the 11 satisfactory ventriculograms. Although some degree of aqueductal stenosis was seen frequently, dye finally entered the

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**Fig. 5.** Infant aged 1 month. Deformity of column of dye suggestive of Arnold-Chiari malformation but with 4th ventricle of normal size and position.

**Fig. 6.** Infant aged 3 months. Relatively larger and presumably normal aqueduct in a patient who never required shunting. Note the large amount of dye in cisterna magna.

**Fig. 7.** Infant aged 1 month. An aqueduct showing zone of relative stenosis.
TABLE 1

Observations in 11 satisfactory studies of Pantopaque ventriculography in infants with myelomeningocele

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<table>
<thead>
<tr>
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<tbody>
<tr>
<td>1</td>
<td>Passage of dye to spinal subarachnoid space</td>
<td>11</td>
</tr>
<tr>
<td>2</td>
<td>Apparent obstruction of flow of dye from spinal to intracranial subarachnoid space</td>
<td>8</td>
</tr>
<tr>
<td>3</td>
<td>Puddling of dye (in cervical subarachnoid space)</td>
<td>3</td>
</tr>
<tr>
<td>4</td>
<td>Fourth ventricle visualized</td>
<td>8</td>
</tr>
<tr>
<td>5</td>
<td>Abnormally small fourth ventricle</td>
<td>6</td>
</tr>
<tr>
<td>6</td>
<td>Low position of fourth ventricle</td>
<td>2</td>
</tr>
<tr>
<td>7</td>
<td>Definite Arnold-Chiari malformation (the same 2 patients as in No. 6)</td>
<td>2</td>
</tr>
<tr>
<td>8</td>
<td>Suggestive of Arnold-Chiari malformation</td>
<td>1</td>
</tr>
<tr>
<td>9</td>
<td>Normal aqueduct</td>
<td>1</td>
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<tr>
<td>10</td>
<td>Attenuated aqueduct</td>
<td>7</td>
</tr>
<tr>
<td>11</td>
<td>Forked aqueduct (included in No. 10)</td>
<td>1</td>
</tr>
<tr>
<td>12</td>
<td>Cranialacunia</td>
<td>6</td>
</tr>
<tr>
<td>13</td>
<td>Enlargement of massa intermedia</td>
<td>4</td>
</tr>
<tr>
<td>14</td>
<td>Thinning or partial agenesis of corpus callosum</td>
<td>8</td>
</tr>
<tr>
<td>15</td>
<td>Pineal diverticuli</td>
<td>3</td>
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spinal subarachnoid space in all cases. The aqueduct appeared stenotic in 7 cases. In 3 cases more than 30 minutes were required for aqueductal passage. Whether the hydrocephalus was the result of the aqueductal stenosis, or poor subarachnoid circulation, or both, is open to question. The fact that dye was seen to enter the intracranial subarachnoid space in only 3 patients, even though the infants were placed in the hanging-head position in all studies, favors subarachnoid obstruction as a causative factor. An apparent inability of fluid to enter the intracranial subarachnoid space is shown. We feel that this sup-

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Fig. 8. Infant aged 1 month. Relative aqueductal stenosis appearing to taper to atresia. However, dye did reach the spinal subarachnoid space some time after 30 min.

Fig. 9. Infant aged 2 weeks. Greatly enlarged pineal recess, or pineal diverticulum.
ports Gardner's hypothesis based on the work of Weed that the inadequate development of or adhesions within the subarachnoid space are of great importance in the continuing development of hydrocephalus. Further support is given by the puddling of dye in the subarachnoid space in 3 patients. It is interesting that 7 of the 8 patients with the apparent defect of subarachnoid circulation required a shunting procedure, while 2 of the 3 patients who showed dye in the intracranial subarachnoid space required no shunt.

There is additional evidence that these children have experienced very early and transient hydrocephalus probably associated with hydromyelia in utero. In children with myelomeningocele the frequent demonstration at birth of cranialacunia, small posterior fossa, partial agenesia of the corpus callosum, and an irregular ventricular configuration is thought to be attributable to the residuals of this phase. We have postulated that postnatal decompensation and further hydrocephalus are related to alteration of the dialyzing membrane of the sac, closure of small spinal-fluid sinuses such as an open central canal, abrupt rise in the neonatal blood pressure, inflammation and infection, and other poorly understood factors of the maternal and infant biological systems.

The 4th ventricle was visualized in 8 patients—a much higher proportion than ordinarily would be seen with air ventriculography alone. The structure was unusually small in 6 patients, presumably because of compression by overlying hydrocephalus as commonly seen at necropsy in patients with myelomeningocele.\textsuperscript{3,10}

A definitely low position of the 4th ventricle was seen in only 2 patients, each of whom had deformities in the column of dye which we thought were diagnostic of the Arnold-Chiari malformation. There was deformity of the column of dye suggestive of this malformation in a 3rd patient who was thought to represent a transitional phase in the postnatal development of the full-blown picture.

Though this is a small series, the proportion of Arnold-Chiari malformations appears to be considerably lower than that reported in the literature from necropsy specimens. We believe that this is further evidence in favor of our postulation that the malformation may develop in early extrauterine life, and that the disparity is attributable to the stage of development at which the disease is observed.

The aqueduct was visualized in 9 patients, but appeared normal in only 2. It was attenuated in 7, and 1 of these demonstrated forking. Unusual angulation of the aqueduct was seen in 2 patients; 1 had a thin aqueduct and the other appeared to have an aqueduct of normal caliber. Additional findings of the studies are listed in Table 1.

**Summary and Conclusions**

A method of positive contrast ventriculography in the infant with myelomeningocele is presented. The findings in 11 technologically satisfactory procedures are discussed. We feel these findings show that positive contrast ventriculography is of value in delineating the intracranial pathology of such infants, and that the risk involved in the procedure is small. The procedure is relatively simple and does not require fluoroscopy. No untoward reactions were attributed to the Pantopaque in any of these patients. One patient, who inadvertently had the dye placed in the interhemispheric subdural space, at necropsy showed no gross or microscopic reaction to the Pantopaque. We believe that combining Pantopaque ventriculography with a less extensive air contrast study not only provides more information but also may reduce the danger to the patient when compared with a more extensive pneumographic study.

Our findings with Pantopaque ventriculography support the concept that Arnold-Chiari malformation may be a secondary defect rather than a primary developmental error, and that it can develop in early infancy as well as in utero.

Inadequate subarachnoid circulation and puddling of dye in apparently partially obstructed subarachnoid spaces strongly sup-
port some aspects of the hydromyelic theory of pathogenesis of myelomeningocele as set forth by Morgagni and as amplified by Weed and Gardner. Hydrocephalus in these children is viewed as a transient phenomenon in utero of early embryonic development which tends to recur in the neonatal period. Some of the factors precipitating the neonatal hydrocephalus are postulated.

References
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