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Chiari III Malformation: Imaging Features

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Purpose: To analyze and discuss the MR and CT features of Chiari type III malformations. **Patients and Method:** MR and CT studies in nine neonates born at term with Chiari type III malformations were retrospectively reviewed. **Results:** High cervical/low occipital encephaloceles were present in all cases. Hypoplasia of the low and midline aspects of the parietal bones was seen in four patients. The encephaloceles contained varying amounts of brain (cerebellum and occipital lobes, six cases; cerebellum only, three cases), ventricles (fourth, six cases; lateral, three cases), cisterns, and in one case, the medulla and pons. Associated anomalies included: petrous and clivus scalloping (five cases/nine cases), cerebellar hemisphere overgrowth (two cases/nine cases), cerebellar tonsillar herniation (three cases/seven cases), deformed midbrain (nine cases), hydrocephalus (two cases/nine cases), dysgenesis of the corpus callosum (six cases/nine cases), posterior cervical vertebral agenesis (three cases/eight cases), and spinal cord syrinxes (two cases/seven cases). In four patients who underwent surgical resection and closure, aberrant deep draining veins and ectopic venous sinuses within the encephaloceles were found. Pathology examination of the encephalocele (four cases/nine cases) showed multiple anomalies (necrosis, gliosis, heterotopias, meningeal fibrosis) that were not demonstrable by either MR or CT. The marked disorganization of the tissues contained within the cephalocele may account for the lack of MR sensitivity to these abnormalities. **Conclusion:** Preoperative determination of the position of the medulla and pons is essential and is easily accomplished by MR. To avoid surgical complications, the high incidence of venous anomalies should be kept in mind.

Index terms: Brain, hernia; Chiari malformations

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Encephaloceles occur in approximately one out of every 4000 to 5000 newborns (1). Those located in the occipital region are the most common type in the western world. In 1891, Chiari described the pathologic features of what today is known as the type I and type II Chiari malformations (2). This paper also contains a single case in which cervical spina bifida combined with multiple cerebellar and brain stem anomalies was present. This spectrum of abnormalities has been called a Chiari type III malformation. Recently, the definition of Chiari type III malformations has been expanded to include those patients with herniation of the hindbrain into a low occipital (below the inion) and/or high cervical encephalocele in combination with pathologic and imaging features of Chiari II malformations (3). For the purpose of this paper, we analyzed the magnetic resonance imaging (MR) and computed tomography (CT) studied obtained in nine patients who harbored a combination of abnormalities conforming to both the classic and the more recent definitions of Chiari type III malformations.

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Materials and Methods

MR scans of nine patients with Chiari III malformations were reviewed retrospectively (Table 1). All patients were neonates born at term (three males and six females). Five patients had prenatal ultrasounds. The patients underwent preoperative imaging with MR (n = 7) (1.5 T systems), CT (n = 2), and plain films of the skull (n = 9). Sagittal, coronal, and axial (5 mm) spin-echo T1-weighted images were obtained (500–750/20–30/2 (TR/TE/excitations)) in

all cases. Axial spin-echo proton density and T2-weighted (2000–2400/30–80/1) were also obtained. Two patients underwent preoperative evaluation with only noncontrast axial (5 mm) CT sections. One CT study included images of upper five cervical vertebrae. The upper cervical spine was also included in the saggital and axial MR T1-weighted images in all cases. In one case, the cervical spine was not evaluated.

The MR and CT studies were then assessed for the contents, location and signal abnormalities of the encephaloceles, presence of lacunar skull, scalloping of the posterior aspect of the petrous pyramids and the clivus, overgrowth of the cerebellar hemisphere, towering of the cerebellum through a wide tentorial incisura, deformities of the midbrain (specifically beaking of the tectum and/or fusion of the colliculi), hydrocephalus, morphology of the corpus callosum, cerebellar tonsillar herniation, and abnormalities in the cervical spine and cervical spinal cord. The MR and CT findings were then compared to the surgical findings in four cases.

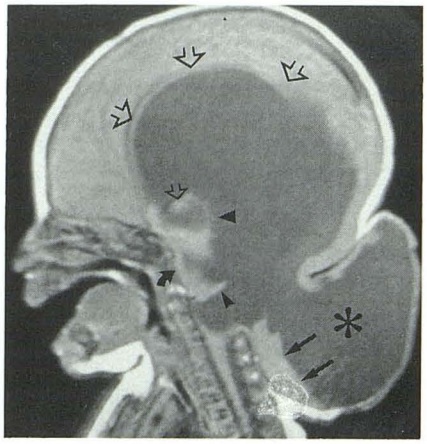

Fig. 1. Case 7: Sagittal spin echo 750/20 slightly off-center image shows a large low occipital/high cervical encephalocele. The CSF density (*) inside the cephalocele is believed to be a markedly dilated fourth ventricle, with the roof of the fourth ventricle displaced superiorly (*small arrowhead*). The third ventricle (*large arrowhead*) and massa intermedia (*small open arrow*) are mildly prominent. The corpus callosum (*large open arrows*) is thin and the splenium is absent. Residual cerebellar tissue (*small arrows*) are present inside the encephalocele. Note that there is absence of the posterior elements of C1, C2, and C3. Scalloping (*curved arrow*) of the clivus is present. There is marked dilatation of the lateral ventricles.

Results

In five instances, prenatal sonography revealed encephaloceles, and all of these patients were born by cesarean section. Apgar scores were available in six cases, and all but two were abnormally low (the median score was 3 at 1 minute and 5 at 5 minutes). At birth, microcephaly (head circumference below the fifth percentile for age) was present in five newborns. Four patients were considered normocephalic, two of these patients had hydrocephalus. Surgical resection of the aberrant tissues contained within the encephaloceles was performed in four cases. At surgery, these four patients had abberantly located venous sinuses and deep cerebellar veins and one patient

TABLE 1: Imaging features of Chiari III malformation

Case No.	Imaging Study	Encephalocele Contents[a]	Encephalocele Location[b]	Lacunar Skull[c]	Bone Scalloping[d]	Cerebellar Overgrowth	Towering Cerebellum	Midbrain	Hydrocephalus	Corpus Callosum[e]	Tonsillar Herniation[f]	Cervical Spine[g]	Cervical Cord[h]
1	MR	B, V, CSF	HC, O	–	–	–	–	Deformed	–	PA	+	–	Syrinx
2	MR	B, V	HC, O	+	–	–	–	Deformed	–	CA	–	+	–
3	MR	B, V, CSF, M	HC, O, P	–	–	–	–	Deformed	–	CA	–	–	–
4	CT	B, V	O, P	–	P, C	Medial and anterior	–	Deformed	–	CA	NE	–	NE
5	CT	B, V	O, P	–	–	–	–	Deformed	–	PA	NE	NE	NE
6	MR	B, V	HC, O	+	P, C	–	–	Deformed	–	CA	+	+	Syrinx *
7	MR	B, V	HC, O, P	–	P, C	–	–	Deformed	+	THIN	–	+	–
8	MR	B, V, CSF	O	+	P, C	Medial and anterior	–	Deformed	+	NL	+	+	–
9	MR	B, CSF	O	–	P, C	–	–	Beaked	–	NL	–	–	–

[a] B, brain; V, ventricles; CSF, cerebrospinal fluid containing spaces different than ventricles; M, medulla.
[b] HC, high cervical; O, occipital; P, parietal.
[c] Evaluated with plain films.
[d] P, petrous bones; C, clivus.
[e] PA, partial agenesis; CA, complete agenesis; THIN, corpus callosum is thinned but present; NL, normal.
[f] NE, not evaluated.
[g] Refers to agenesis of the posterior elements of C1 through C3.
[h] *, extending into the pons.

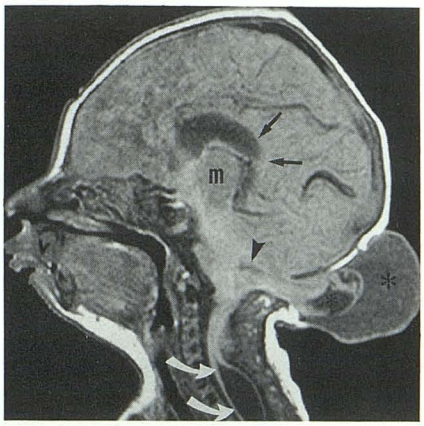

Fig. 2. Case 1: Midline sagittal spin echo 700/20 image demonstrates a low occipital encephalocele containing cerebellar tissues. The cystic portions (*) within the herniated cerebellum are of uncertain etiology. The posterior aspect of the corpus callosum (*arrows*) is not clear and is probably dysgenetic. The third ventricle is not seen, but the massa intermedia (*m*) is very prominent. The tectum is deformed and not readily identified. The fourth ventricle (*arrowhead*) is deformed and displaced posteriorly. A syrinx (*curved arrows*) is present in the mid to lower cervical spinal cord.

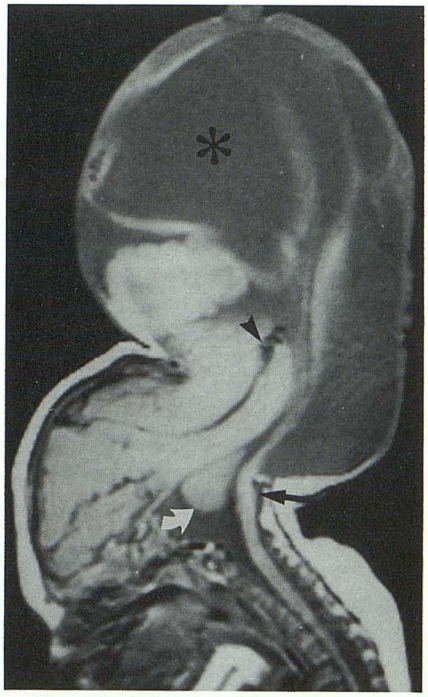

Fig. 3. Case 3: Midline saggital spin echo 800/20 image in a patient with a large occipital and parietal encephalocele. The encephalocele contains a large amount of brain tissues and dilated ventricles (*). The intracranial contents are dysmorphic and the corpus callosum is not identified. Note the presence of a venous structure (*arrowhead*) inside the encephalocele. The medulla (*arrow*) and pons (*curved arrow*) herniate into the encephalocele. Recognition of these structures is critical for the surgical planning.

had aplasia of the posterior falx cerebri. These vascular structures were preserved in all cases. All patients required duroplasties and skin grafts. Two patients died in the immediate postoperative period; one to diffuse edema probably due to thrombosis of the superior saggital sinus and one of aspiration. One patient who had surgery is alive, but severely retarded. The remaining five patients were lost to follow-up. In four cases, pathologic examination of the tissue removed from the encephalocele showed cerebral and cerebellar tissues with areas of pressure necrosis, ventricles with normal choroid plexuses, gray matter heterotopias, small focal areas of gliosis, meningeal inflammation and fibrosis, and reactive astrocytosis.

Imaging studies showed that all encephaloceles involved the occipital bones; five extended into the high cervical (C1–C3) spine (Fig. 1). In four cases, the low and midline parietal bones were also involved (Table 1). All encephaloceles contained brain (cerebellum, nine cases; occipital, five cases; and parietal lobes, one case). Portions of the ventricular system (lateral ventricles, three cases; fourth ventricle, six cases) extended into six encephaloceles. Three encephaloceles contained no ventricles (Fig. 2). In one case, the brainstem and medulla herniated into the encephalocele (Fig. 3). Three encephaloceles contained large cerebrospinal fluid (CSF) spaces unrelated to the ventricular system and, although these are of uncertain etiology, we believe they could represent distorted and enlarged basilar cisterns. Lacunar skull was present on plain films in three patients. The abnormal venous structures identified during surgery in four patients were only seen by MR in two cases (Figs. 3 and 4A). Angiograms were not performed on any patients. The multiple pathologic findings (necrosis, grey matter, heterotopias, gliosis, reactive astrocytosis, and meningeal inflammation and fibrosis) found within the encephaloceles could not be individually identified preoperatively by MR on either T1- or T2-weighted images, possibly due to the marked disorganization of these tissues. Specifically, the encephaloceles did not show abnormal areas of increased signal intensity (corresponding to gliosis) on the T2-weighted images. Five patients had scalloping of the posterior petrous pyramids and the clivus (Figs. 4A and 4B).

Overgrowth of the cerebellum, defined as the extension of the cerebellar hemispheres medial and anterior to the brainstem and pons, was detected in two instances (Table 1 and Fig. 4A).

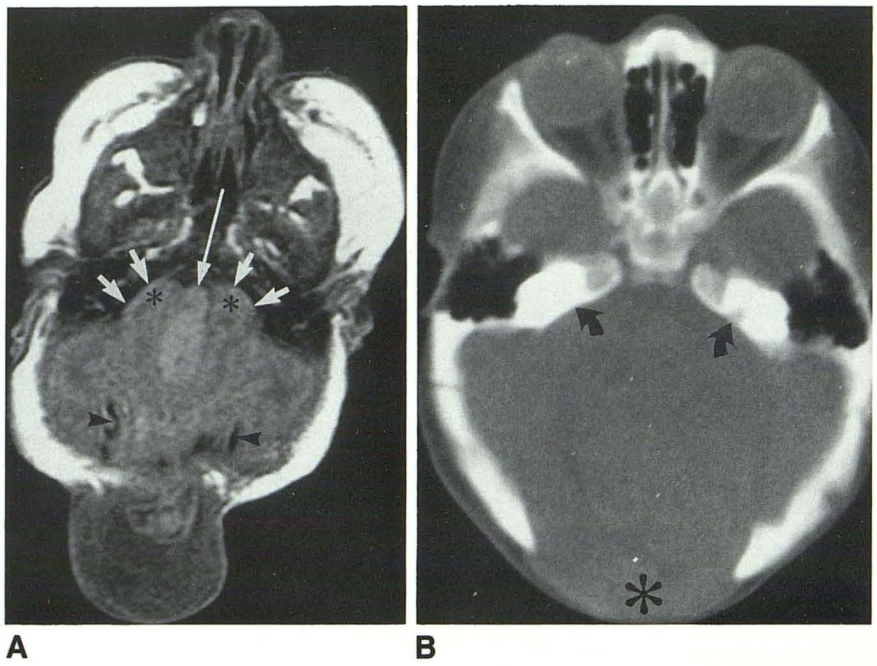

Fig. 4. *A*, Case 8: Axial spin echo 700/20 image at the level of the mid posterior fossa shows scalloping (*short arrows*) of the posterior aspect of the petrous bones. The cerebellar hemispheres (*) show medial and anterior overgrowth with respect to the brainstem (*long arrow*). Also seen are aberrant paired venous structures (*arrowheads*) bordering the skull defect. Note the posterior encephalocele.

B, Case 4: axial CT scan (bone windows) shows scalloping (*arrows*) of both petrous bones. Also note the encephalocele (*).

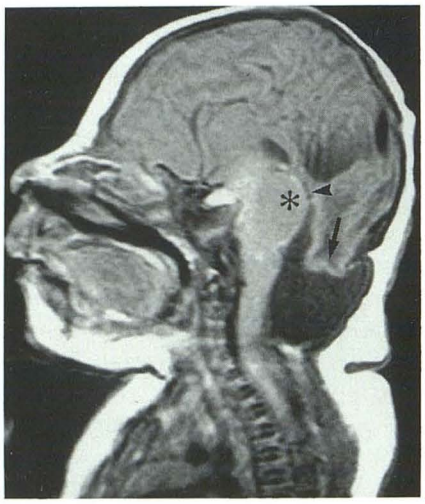

Fig. 5. Case 2: Sagittal spin echo 750/20 image in a microcephalic patient with a high cervical/low occipital encephalocele. There is herniation of the occipital lobes (*arrow*) into the defect. The cerebellum is not seen and the tectum is not clearly identified. The midbrain (*) is deformed and bulbous and is displacing the aqueduct (*arrowhead*) posteriorly. The splenium of the corpus callosum is absent. Agenesis of the posterior elements of C1 through C3 is present.

Towering of cerebellum (supratentorial extension of cerebellar tissues through a widened tentorial incisura) was not documented in any case. The cerebellum was not identified in one patient (Fig. 5). The midbrain was considered abnormal if it was beak-like in configuration (n = 1), or if the colliculi could not be identified individually (n = 8) (Fig. 5). A deformed midbrain was seen in eight cases (Fig. 5).

Hydrocephalus (dilation of the intracranial ventricular system) was seen in two cases (Fig. 1). Colpocephaly (localized enlargement of the atria and occipital horns of the lateral ventricles) was seen in one case. Abnormalities of the corpus callosum included: partial agenesis (always involving the splenium), two cases; complete agenesis, four cases; and extreme thinning probably secondary to marked hydrocephalus, one case (Table 1) (Figs. 1, 2, and 5). A normal corpus callosum was present in two cases. Extension of the cerebellar tonsils downward below the foramen magnum was present in three cases (Fig. 2). In one case of cerebellar aplasia, the tonsils were not identified. The position of the tonsil could not be evaluated in two patients who underwent only preoperative CT. In three patients, the position of the cerebellar tonsils was considered normal. Absence of the posterior elements of the first three cervical vertebrae was identified in three cases (Figs. 1 and 5). Spinal cord fluid filled cavities were seen in two cases; in one of these, the syrinx extended into the pons (Figs. 2 and 6). Two patients died due to surgical complications in the immediate postoperative period.

Discussion

The CT and MR features of Chiari type I and II malformations are well known (4–9). Chiari type

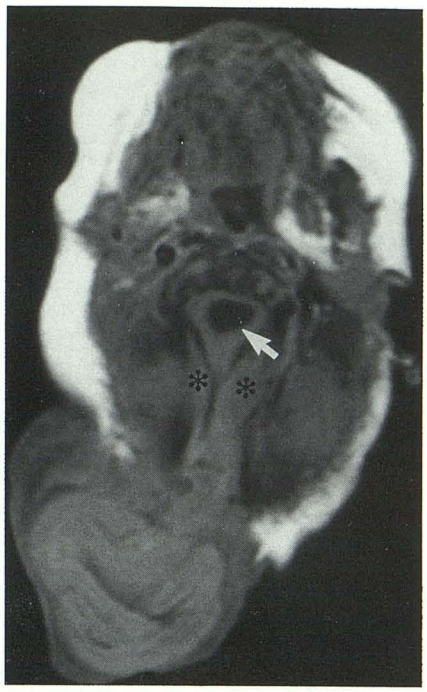

Fig. 6. Case 6: Axial spin echo 800/20 image through the low occipital regions clearly shows the encephalocele. Notice that the tissue within the encephalocele are markedly distorted and it is not possible to recognize any normal structures. A syrinx (*arrow*) is present in the medulla. In this patient, the syrinx extended into the pons. Note the cerebellar tonsils (*) displaced inferiorly.

III malformation is rare and only scattered cases (included in larger series of Chiari type I and II malformations) are found in the literature (4, 10-12). The Chiari III malformation includes displacement of the medulla, cerebellum, occipital lobes, and meninges into a high cervical and/or low occipital cephalocele (13). The above features in combination with those seen in Chiari II malformations are believed to be characteristic of the type III malformations (14). A skull defect associated with herniation of the intracranial contents is termed a "cephalocele" (14). If the herniation contains both brain and meninges, it is referred to as an "meningoencephalocele"; herniation of only the meninges and CSF is called a "meningocele"; herniation of only brain tissues is called an "encephalocele" (15). Encephaloceles are believed to be slightly more common than meningoceles (1). In Chiari III malformations, an encephalocele (which may or may not contain meninges) must be present in combination with the brain anomalies present in the Chiari II malformations. Occipital encephaloceles can be isolated, or be associated with the Chiari malformations, Dandy-Walker complex, cerebellar dysplasias, diastematomyelia, and Kippel-Feil syndrome (14).

The embryologic origin of a low occipital (base-of-skull) cephaloceles probably involves the failure of induction of enchondral bone by incomplete closure of the neural tube, or failure of the ossification centers to fuse completely (16). A posterior cephalocele may involve the supraoccipital and parietal bones (membranous origin). This is likely to result from faulty induction or from pressure erosion by the adjacent mass of herniated tissues (16). It has been noted that supraoccipital encephaloceles occurring below the inion are the most common type seen in Chiari III malformations (1). The main features found in our nine patients with Chiari III malformations are discussed separately.

Cephaloceles, Skull, Dural Partitions, and Venous Anomalies

In all cases, the skull defects were located in the high cervical/occipital regions and extended to the parietal bones in four cases (Table 1 and Fig. 1). These encephaloceles contained cerebellum and occipital lobes in all cases. One encephalocele also contained the medulla and pons. In another case, only the cerebellum was herniated. The masses of herniated tissues were markedly distorted and we were unable to recognize any individual structures within them (Fig. 6). In four cases, surgical resection and pathology examination showed multiple areas of pressure necrosis in the herniated brain, gray matter heterotopias, gliosis, meningeal inflammation, and fibrosis. These findings were not individually demonstrated by either MR or CT, probably due to the marked disorganization of these tissues. Pathologically, the areas of gliosis were small and scattered, and we consider this the reason as to why gliosis was not identifiable by MR T2-weighted images. Because it is believed that the contents of the encephaloceles are composed of nonfunctioning tissues (17), exact recognition of these different components is not necessary and has no bearing on the clinical management of the patient. However, it is extremely important to recognize the position of the brain stem and the medulla so that they can be preserved. To avoid potential complications during surgery, it should also be kept in mind that anomalies of venous drainage (aberrant sinuses and deep veins) were seen in four (44%) patients in our series (Figs. 3 and 4A). Herniation of the lateral ventricles ac-

companied the brain parenchyma in three instances; in six of these, the fourth ventricle also herniated. In four cases, the herniated ventricles were dilated out of proportion to the intracranial ventricular system (Fig. 3). A possible explanation is that these ventricles contained choroid plexus and, therefore, produced CSF. Constriction of the encephalocele at the level of the skull defect could have accounted for lack of normal CSF drainage from the ventricle leading to dilatation, and secondarily resulting in gliosis and atrophy of the herniated tissues. Lacunar skull ("luckenschadel") was present in three cases. "Luckenschadel" is also commonly seen with isolated encephaloceles and with Chiari II malformations (3). Scalloping of the posterior petrous bones and clivus was present in five of our patients (Fig. 4B). In the type II malformation, this latter finding is believed to be secondary to pressure erosion from the cerebellum as it attempts to grow within the confines of a small posterior fossa (4). In the Chiari III malformation, the posterior skull defect should decompress the posterior fossa and, therefore, pressure erosion is an unlikely cause for bone scalloping. A more likely explanation is that this scalloping is a bone dysplasia similar to "luckenschadel." Aplasia of the posterior aspect of the falx cerebri was found at surgery in one patient.

Hindbrain, Midbrain, and Cervical Spine

Downward herniation of the cerebellar tonsils was present in three of our patients (30%). Traction of the hindbrain into the encephalocele probably accounted for this relatively low number of cases with this finding , since only those patients with relatively small encephaloceles showed tonsillar herniation. Medial and anterior overgrowth of the cerebellar hemispheres can be seen in up to 75% of patients with Chiari II malformation (Fig. 4A) (4). In our series, only two cases showed this feature (Table 1). The lack of cerebellar confinement in a small posterior fossa could have accounted for this relatively small number of cases. Also, the same hypothesis can help explain the absence of "towering" of the cerebellum through the tentorial incisura in our population. This finding is commonly noted in the type II malformation (5). In one patient, the cerebellum was not identified (Fig. 5). Extreme hypoplasia of the cerebellum is known to occur in patients with Chiari II (9). Beaking of the midbrain was clearly identified in only one of our patients. However, eight patients had a deformed quadrigeminal plate (probably due to fusion or distortion of the colliculi) and, therefore, were considered to have an abnormal midbrain (Fig. 5).

In three cases, the posterior elements of C1, C2, and C3 were absent (Figs. 1 and 5). In one case, the upper cervical spine was not evaluated. This anomaly is also commonly encountered in the type II malformation in which close to 70% of cases may show incomplete fusion of the posterior arch of C1 and other cervical vertebrae (16). In two of our patients, cord cavities were documented (Figs. 2 and 6). In one case, the syrinx extended into the pons (syringobulbia). Even though we do not have a good explanation for these findings, a possible hypothesis involves the failure of normal formation of the foramina of Luschka and Magendie (as part of the hindbrain dysgenesis) with subsequent alterations of the normal CSF dynamics at the level of foramen magnum leading to syrinx formation. These CSF flow abnormalities are also believed to be present in the type II Chiari malformations (18). Since the entire spine was not studied in our patients, it is conceivable that the incidence of syrinxes may have been even higher.

Ventricular System and Corpus Callosum

Marked intracranial dilatation of the ventricular system was seen in two instances (Fig. 1). Both of these patients had a deformed midbrain. The aqueduct of Sylvius was not clearly identified in these two patients (Fig. 5). Aqueductal stenosis can then be considered as a reason for the hydrocephalus. The septum pellucidum was present in all cases. Seven of our patients had dysgenesis of the corpus callosum (complete agenesis, four cases (50%); partial agenesis (always involving the splenium), two cases (25%); marked thinning (probably secondary to severe hydrocephalus), one case (12.5%)) (Table 3) (Figs. 1-3 and Fig. 5). Dysgenesis of the corpus callosum is commonly seen in the Chiari II malformations and can be seen in patients with isolated encephaloceles (19, 20). A normal corpus callosum was present in only two cases.

We do not have a good explanation for the development of Chiari III malformations. Many diverse hypotheses have been proposed to explain the varied abnormalities present in Chiari II malformations. The most recent (and probably one of the most popular) explanation suggests that the Chiari II anomalies are related to the

constant escape of CSF through the open neural tube defect during intrauterine life (21). This leads to failure of distension of the primitive ventricles with subsequent development of a small skull. As the brain attempts to grow in the reduced space, multiple cerebral abnormalities arise. We believe that this hypothesis can be used to explain the findings in the Chiari III patients. We also feel that it is appropriate to mention here that, even if our patients had no obvious source of CSF leakage at birth, CSF leakage in utero cannot be excluded. This may have lead to formation of the multiple abnormalities also present in the type II malformation.

In summary, Chiari III is a rare developmental condition with a very poor prognosis. Due to its distinct clinical and imaging features, it is easily distinguished from the more common Chiari I and II malformations and isolated encephaloceles. Due to its multiplanar capability, MR is more useful than CT to study preoperatively the content of the cephalocele. MR can readily establish the presence of solid tissues and of ventricular extension into the cephalocele. In our experience, MR was unable to distinguish many subtle abnormalities in the herniated brain; however, this issue is not critical because the tissues within the encephalocele are believed to be nonfunctioning. Nevertheless, the preoperative identification of the brain stem and medulla is mandatory to preserve respiratory function; this is easily accomplished with MR. To prevent complications when surgery is comtemplated, it should be kept in mind that anomalies of venous drainage are commonly present, but often not identified on MR studies. If surgery is performed in a patient with an encephalocele, resection of the herniated brain with preservation of the herniated venous structures accompanied by duroplasty and skin grafts is mandatory (GL Clifton, personal communication). Resection of the posterior two-thirds of the superior sagittal sinus is almost always fatal.

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