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Sonographic Findings in Infants with Macrocrania

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This study compares the sonographic and CT findings in a group of infants with macrocrania and correlates those findings with neurologic outcome to determine the diagnostic accuracy and prognostic value of sonography. Sonographic findings in 255 infants with macrocrania are described. Of the 195 term infants examined, 130 had normal sonograms, 11 (5.6%) had significant abnormalities, and 54 had increased intra- and/or extraaxial fluid spaces. Of the 60 former preterm infants, 33 had normal sonograms, four (6.7%) had significant abnormalities, and 23 had increased fluid spaces or small resolving germinal matrix hemorrhages. The patients with significant abnormalities usually had head circumferences greater than the 95th percentile and had neurologic abnormalities. There was good correlation between sonography and CT in 30 of the 36 patients evaluated by both. In six there was mild discrepancy in the volume of the extraaxial fluid. No significant abnormality was missed by sonography. CT did not contribute any additional information. Neurologic follow-up was available for 202 patients. Nineteen percent of the term infants and 24% of the former preterm infants were abnormal on neurologic follow-up. Most patients with normal sonograms were normal on follow-up. Twelve of the term and four of the preterm infants with normal sonograms were developmentally delayed on follow-up. Increased CSF in the ventricles and/or extraaxial spaces was a common abnormality, but it usually is associated with a normal neurologic outcome and represents "benign macrocrania."

We conclude that an infant with an enlarged or enlarging head should have a neurologic examination and head circumference measurement. If the patient has a head circumference greater than the 95th percentile, particularly if there are abnormal neurologic findings, further evaluation is indicated. Sonography is the initial procedure recommended since it accurately evaluates ventricular size, extraaxial fluid, and congenital malformations. If sonography is normal or shows mildly increased fluid spaces, then follow-up head circumference measurement and clinical evaluation will probably suffice. CT is indicated if there is a significant abnormality on sonography that requires further clarification.

The evaluation of excessive head growth during the first year of life is a problem familiar to all physicians who care for small children. It is important to establish the nature of any intracranial disease that may be present and, in particular, to detect any remedial condition or treatable complication such as hydrocephalus and subdural hematomas. The measurement of the maximum head circumference in children is standardized and a central part of clinical assessment. This measurement is compared with several published charts of normal head circumference according to age and sex [1]. In the past, evaluation of the status of an infant with a large head required invasive neuroradiologic techniques, including ventriculography and cerebral angiography. CT has made a major contribution to the solution of these problems [2-4], but it is still costly and exposes the infant to ionizing radiation. Sonography has had an increasingly important role in the evaluation of neurologic diseases in infants with an open anterior fontanelle. It has been particularly useful and accurate in diagnosing hydrocephalus, congenital malformations, and intracranial hemorrhage [5-7].

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The purpose of this study is to describe the sonographic findings in a group of infants with a large or enlarging head and to correlate them with CT findings. The presence of these sonographic findings and other clinical features was correlated with the neurologic outcome to determine the diagnostic accuracy and prognostic value of sonography in infants with macrocrania.

Materials and Methods

The sonograms were reviewed on 255 children examined between October 1978 and August 1984. The primary clinical indication for the examination was a large head. There were four groups: (1) those with a measured head circumference greater than the 95th percentile according to standards established by the National Center for Health Statistics [1]; (2) those with disproportionate head size compared with body length and weight; (3) those with head circumference that increased in percentile level on serial measurements; (4) and those with a large-appearing head by subjective observation. While the last category seems of less importance, these children present a common problem and are of significant concern to the practicing pediatrician. There were 195 term infants and 60 former premature infants in the study. Patients with known syndromes or obvious congenital malformations such as meningomyeloceles, history of previous trauma, or

cranial infection were excluded. The age range was newborn to 20 months.

Cranial sonography was performed with a sector real-time sonographic machine (Advanced Technology Laboratory) or with a contact B-mode gray scale sonographic machine (Picker 80L Digital) using 3–7.5 MHz transducers. Serial scans of the brain were obtained in coronal and sagittal planes, including magnified coronal views of the interhemispheric fissure and medial convexities to look for extraaxial fluid collections (Fig. 1). Patients older than 3 months occasionally required oral sedation with chloral hydrate in a dose of 50 mg/kg. No significant complications were associated with sedation.

All sonographic examinations were retrospectively reviewed without knowledge of the clinical course. Ventricular size, the presence of prominent extraaxial fluid, the presence of parenchymal abnormalities, and congenital malformations were noted. Those with slitlike ventricles or those containing minimal fluid were considered normal; anything greater than minimal was considered abnormal (Fig. 2). An absence or a minimal amount of extraaxial fluid was considered normal; anything greater than minimal was considered to be abnormal (Fig. 1).

CT examinations were available on 36 of the patients. These were reviewed without knowledge of the sonographic findings or clinical course. The ventricular size, presence of extraaxial fluid, the presence of parenchymal abnormalities, and congenital malformations were noted.

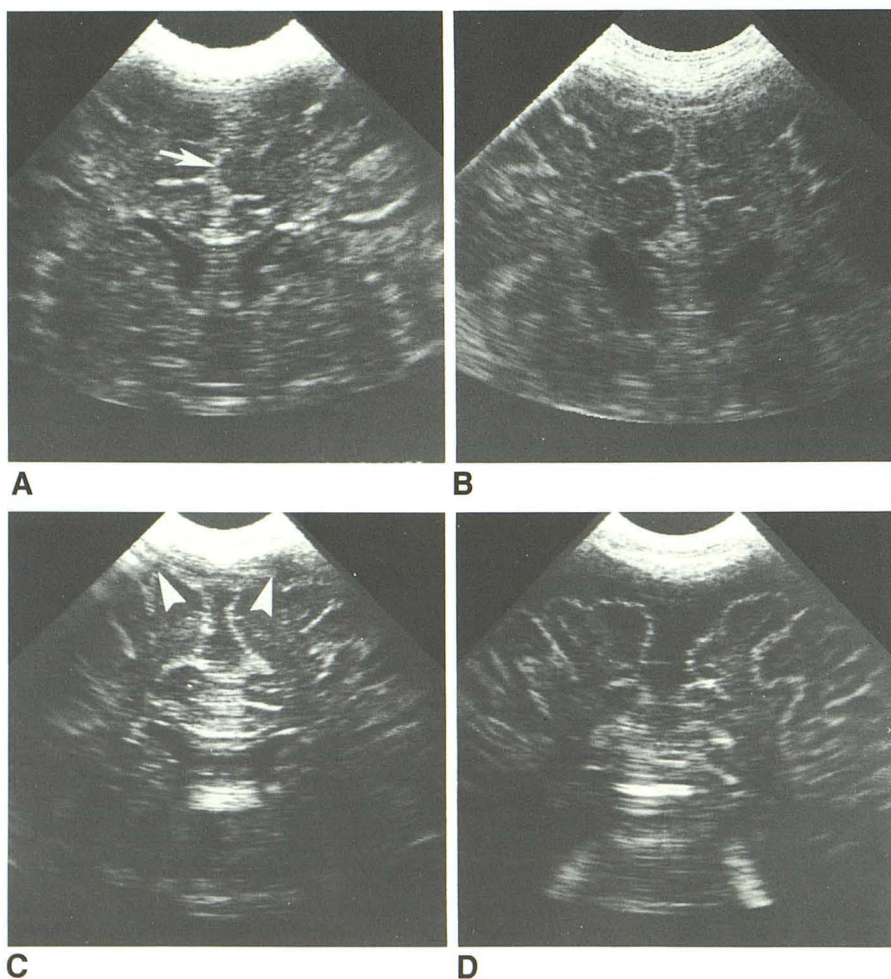


Fig. 1.—Extraaxial fluid collection (EAF), magnified coronal views of interhemispheric fissure and medial convexities with 7.5 MHz short focus transducer.

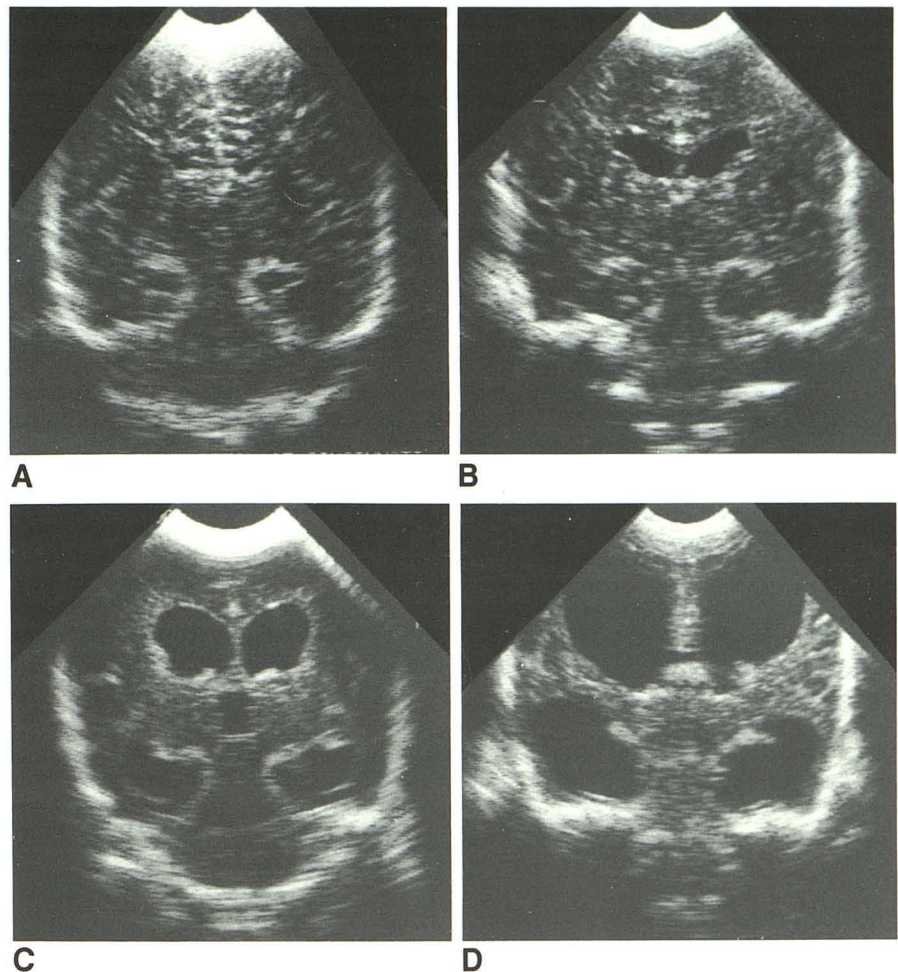
A, Normal: echogenic linear interhemispheric fissure (arrow) without evidence of EAF.

B, Normal: minimal widening of interhemispheric fissure.

C, Small EAF evidenced by slight widening of interhemispheric fissure and demonstration of brain gyral surface (arrowheads).

D, Moderate EAF over convexity with further separation between calvarium and brain.

Fig. 2.—Ventricular size, coronal scans.
A, Normal ventricles (minimal fluid).
B, Mild ventriculomegaly.
C, Moderate ventriculomegaly.
D, Marked ventriculomegaly.



Each infant's clinical findings and follow-up neurologic development was determined by reviewing patient charts and by contacting the physicians who were caring for the children. Information at the time of examination included head circumference and percentile, neurologic status and development, presence of seizures, a positive history for neurologic disease or for predisposing conditions such as prematurity, and body height and weight. Follow-up information regarding head size and neurologic and developmental status was obtained. Results of CT, surgery, or autopsy were recorded.

Results

The 255 infants examined were divided into two groups: those born at term and those who were born prematurely (less than 38 weeks gestation). There were 195 term infants and 60 former premature infants. The sonographic findings are reported in Tables 1 and 2.

Of the 195 term infants examined, 130 had normal sonograms and 65 were abnormal. Of the abnormal findings, 11 were significant, including five patients with aqueductal stenosis and moderate to marked hydrocephalus, two patients with severe hydrocephalus of other etiology, two patients with moderate to marked hydrocephalus with arachnoid

cysts, and two patients with agenesis of the corpus callosum (one of these also had marked hydrocephalus with an interhemispheric cyst). All 11 patients with severe abnormalities by sonography had head circumferences greater than the 95th percentile according to standards established by the National Center for Health Statistics [1]. The other 54 patients with abnormal sonograms had increased CSF in the ventricles (minimal to mild ventricular dilatation), and/or extraaxial fluid spaces (minimal to moderate) (Fig. 3).

Of the 60 former preterm infants examined, 33 had normal sonograms and 27 were abnormal. There were four with significant abnormalities, including two with marked hydrocephalus and aqueductal stenosis and two with marked communicating hydrocephalus. The remaining 23 patients with abnormal studies had increased CSF spaces (20) with minimal to mild ventricular dilatation and minimal to moderate extraaxial fluid or resolving small germinal matrix hemorrhages (three).

Sonographic and CT Correlation

Thirty-six patients had CT scans in addition to sonography. Twenty-seven were term infants and nine were preterm in-

fants. In 32 patients the CT was done within a month of the sonographic examination. In four patients the CT was done 1–7 months after the sonography. In 30 patients, the sonographic and CT findings were in agreement. There were six patients in whom the sonography and CT disagreed (Table 3) primarily in the amount of extraaxial fluid that was present (Figs. 4 and 5).

Key to Abbreviations in Tables 1–5

- A = Head circumference greater than 95th percentile
 B = Disproportionate head size compared with body length and weight
 C = Head circumference that increases in percentile level on serial measurements
 D = Large-appearing head by subjective observation
 1 = Abnormal neurologic findings or symptoms
 n/a = Neurologic status not available
 IAF = Intraaxial fluid (ventricles)
 EAF = Extraaxial fluid

Clinical Follow-up

Clinical follow-up was available for 202 of the 255 patients (149 of the 195 term infants [76%] and 53 of the 60 premature infants [88%]). The follow-up period ranged from 1 month to 5 years. Nineteen percent of the term infants and 25% of the preterm infants were neurologically or developmentally abnormal on follow-up (Tables 4 and 5).

Among the 149 term infants, 84 with normal sonograms were normal on clinical follow-up and 12 with normal sonograms were developmentally delayed on clinical follow-up. Thirty-seven infants had abnormal sonograms, mainly mildly increased CSF in the ventricles and extraaxial spaces, and were normal on clinical follow-up. Two of these also had agenesis of the corpus callosum without significant associated abnormalities. Sixteen had abnormal sonograms (11 significantly abnormal and five with excess fluid) and were abnormal on clinical follow-up.

Of the 53 preterm infants, 25 with normal sonograms were normal on clinical follow-up, and four with normal sonograms were developmentally delayed on follow-up. One of these had significant cerebral palsy. Fifteen patients had abnormal sonograms but were normal on follow-up. Of these, 14 had

TABLE 1: Sonographic Results of Term Infants

Finding ^a	Normal	Hydrocephalus, Congenital Malformations	Increased or Excess IAF ^a and/or EAF ^a Fluid	Total Abnormal	Total Normal Plus Abnormal
A	43	2	22	24	67
A ¹	6	9	4	13	19
B	11	0	4	4	15
B ¹	9	0	2	2	11
C	3	0	1	1	4
C ¹	0	0	0	0	0
D	9	0	6	6	15
D ^{n/a}	43	0	14	14	57
D ¹	6	0	1	1	7
Total	130	11	54	65	195

^a See Key to Abbreviations.

TABLE 2: Sonographic Results of Former Preterm Infants

Finding ^a	Normal	Hydrocephalus, Congenital Malformations	Increased or Excess IAF ^a and/or EAF ^a Fluid	Total Abnormal	Total Normal Plus Abnormal
A	4	3 ^b	5	8	12
A ¹	1	0	3	3	4
B	4	0	2	2	6
B ¹	2	0	2	2	4
C	6	0	0	0	6
C ¹	2	0	0	0	2
D	7	2 ^b	3	5	12
D ^{n/a}	4	1 ^b	4	5	9
D ¹	3	1	1	2	5
Total	33	7 ^b	20	27	60

^a See Key to Abbreviations.

^b Three of seven had resolving germinal matrix hemorrhage.

Fig. 3.—Benign macrocrania.
A, Coronal sonogram demonstrates mild ventricular enlargement and moderate extraaxial fluid over the convexities (arrowheads).
B, Axial CT scan shows similar findings.

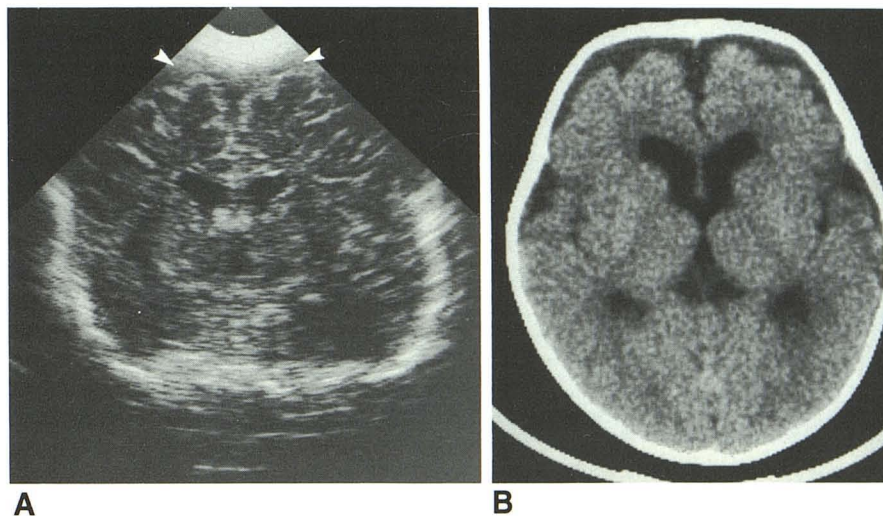


TABLE 3: Sonography/CT Discrepancy in Six Patients

Patient No.	Sonography	CT
1	Mild EAF ^a	Normal
2	Mild EAF ^a	Normal (7 months later)
3	Normal	Mild increased ventricular dilatation and EAF ^a
4	Normal	Mild EAF ^a (3 months later)
5	Moderate EAF ^a	Mild EAF ^a (1 month later)
6	Normal	Mild EAF ^a (2 months later)

^a See Key to Abbreviations, opposite.

increased fluid spaces and one had a small resolving germinal matrix hemorrhage. Nine patients had abnormal sonograms and were abnormal on clinical follow-up.

The group of infants with increased CSF spaces was looked at separately as to outcome. In the full-term group, 41 infants had increased fluid. Seven of these had neurologic abnormalities at the time of presentation. Thirty-six were normal on follow-up and five were abnormal. Of these, one had speech problems, one had subdural effusions that required tapping, two had mild developmental delay, and one had developmental delay and hyperreflexia complicated by meningitis. There was no correlation between the amount of extraaxial fluid and the neurologic outcome.

Of the premature infants, there were 17 with increased fluid. Thirteen of these were normal on follow-up and four were abnormal. Of these, one had spastic cerebral palsy, two were mildly developmentally delayed, and one was delayed with hypotonia.

Thirty-four term infants had "benign macrocrania"; that is, excess CSF in the ventricles (intraaxial) and/or extraaxial spaces but were neurologically and developmentally normal (the other seven of the 41 infants with increased fluid had abnormalities such as developmental delay at the initial pres-

entation). Thirty-two of these 34 were normal on clinical follow-up, and two had developmental delay.

Discussion

In these 255 infants referred because of enlarged or enlarging heads, the primary diagnostic objective was to identify those with intracranial disease and, in particular, to detect any remedial condition or treatable complication such as hydrocephalus or subdural hematomas. Eleven of the 195 term infants (5.6%) and four of the 60 former preterm infants (6.7%) had significant abnormalities that required further work-up and/or treatment. This is much less than the 75% reported in previous studies using CT to evaluate macrocrania [2, 4]. The increased cost and ionizing radiation involved in CT scanning probably influenced the selection of patients to be examined by CT in those studies.

Since a relatively small group of patients in our study had a significant abnormality, and since sonography did not miss significant abnormalities needing treatment, we conclude that the least invasive and least expensive method should be used for screening infants with a large head. In fact, all of the 11 term infants and two of the four preterm infants with significant abnormalities had head circumference greater than the 95th percentile by measurement. The tape measure is the cheapest means by which to identify those infants who are likely to have significant abnormalities.

In comparing the CT and sonographic studies in 36 patients who had both, there was good correlation between sonography and CT in the majority of cases. In a few cases, there was mild discrepancy in the amount of extraaxial fluid (mildly increased versus normal). There was no significant abnormality missed by sonography. Thus, sonography should be the first screening method used after the tape measure for evaluating the infant with a large head.

A small subdural hematoma, particularly if it is localized over the high convexity, might be missed on sonography. While we had no such patients in our group, any patient with

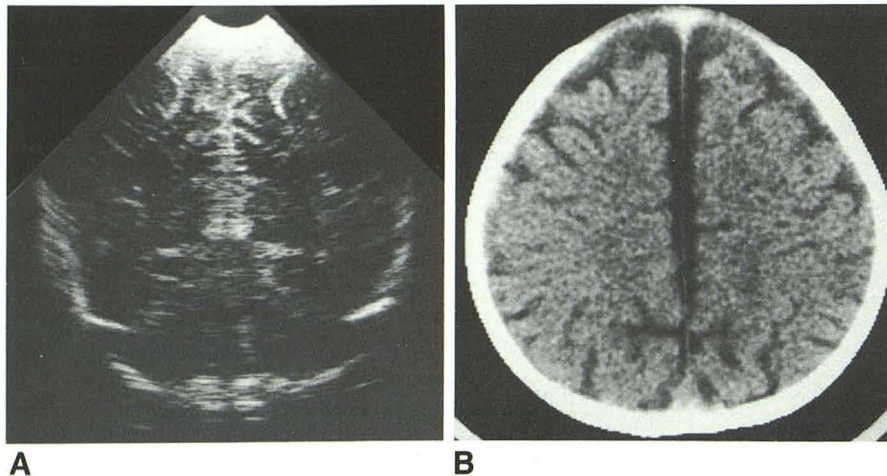


Fig. 4.—Discrepancy between sonography and CT.

A, Cranial sonogram is normal.

B, Axial CT shows mild extraaxial fluid.

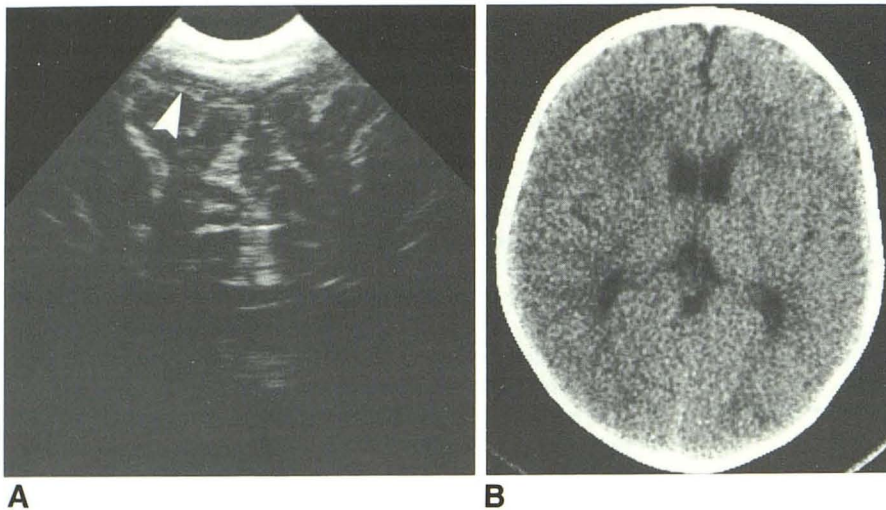


Fig. 5.—Discrepancy between sonography and CT.

A, Coronal sonogram shows mild extraaxial fluid (arrowhead).

B, Axial CT is normal.

a known history of trauma or suspected trauma should be examined by CT rather than sonography unless the fontanelle is exceptionally large and all portions of the intracranial contents can be adequately examined. This is particularly important in cases of suspected child abuse, in which even small hematomas or contusions would be important evidence in future legal proceedings. Also, patients with symptoms suggesting a brain tumor will likely be imaged more optimally with CT or MR.

A rapidly growing head in a former premature infant was not usually associated with a significant abnormality on sonography. Catch-up brain growth is known to occur after the initial postnatal problems have resolved and the patient has received good nutrition. Sonography may, however, miss parenchymal abnormalities, especially if peripheral.

Clinical follow-up information, which was available for 202 of the 255 patients, showed good correlation between the findings on sonography and the neurologic outcomes. Most patients with normal sonograms were normal on follow-up, although a few were developmentally delayed and one had significant cerebral palsy.

Increased CSF in the ventricles and/or extraaxial spaces

was a common abnormality but was usually associated with a normal neurologic outcome. Some of these patients continued to have a large head on follow-up although we had follow-up head measurements in only a few patients. Increased fluid spaces have been described previously as "benign macrocrania" or "chronic subdural fluid collections of childhood [8-14]. The exact pathophysiology of this process is not known, but it is thought to be a benign and mild form of communicating hydrocephalus for which an etiologic factor is not usually apparent. The natural history of this process has been described [14]. The patient generally presents at about 4 months of age with a large head and normal developmental milestones. On follow-up the head circumference usually parallels and/or returns to normal percentiles (39 of 40 patients in Kendall and Holland's group [9]). Additional invasive diagnostic procedures or treatment are not necessary.

In conclusion, an infant with an enlarged or enlarging head should have a neurologic examination, and the head circumference measurement should be compared with standards available from the National Center for Health Statistics. If the patient has a head circumference greater than the 95th percentile, and particularly if there are abnormal neurologic

TABLE 4: Neurologic Follow-up on Term Infants

Finding ^a	Total	Total Follow-ups	Normal Sonograms/ Normal Follow-up	Normal Sonograms/ Abnormal Follow-up	Abnormal Sonograms/ Normal Follow-up	Abnormal Sonograms/ Abnormal Follow-up	Total Abnormal Follow-ups
A	67	63	39	2	19	3	5
A ¹	19	18	3	3	2	10	13
B	15	15	10	1	4	0	1
B ¹	11	11	6	3	1	1	4
C	4	2	1	0	1	0	0
C ¹	0	0	—	—	—	—	0
D	15	13	8	0	4	1	1
D ^{n/a}	57	21	12	3	5	1	4
D ¹	7	6	5	0	1	0	0
Total	195	149 (76%)	84	12	37	16	28 (18.8%)

^a See Key to Abbreviations on page 310.

TABLE 5: Neurologic Follow-up on Premature Infants

Finding ^a	Total	Total Follow-ups	Normal Sonograms/ Normal Follow-up	Normal Sonograms/ Abnormal Follow-up	Abnormal Sonograms/ Normal Follow-up	Abnormal Sonograms/ Abnormal Follow-up	Total Abnormal Follow-ups
A	12	11	4	0	6	1	1
A ¹	4	4	1	0	1	2	2
B	6	6	3	1	2	0	1
B ¹	4	4	1	1	2	0	1
C	6	6	6	—	—	—	0
C ¹	2	2	2	—	—	—	0
D	12	10	3	2	2	3	5
D ^{n/a}	9	5	2	0	2	1	1
D ¹	5	5	3	0	0	2	2
Total	60	53 (88%)	25	4	15	9	13 (24.5%)

^a See Key to Abbreviations on page 310.

findings suggesting increased intracranial pressure, further evaluation is indicated. The infant should be studied initially by sonography, since it accurately evaluates ventricular size, extraaxial fluid, and congenital malformations (the exception would be cases of suspected trauma, child abuse, or brain tumor, in which CT or MR would be preferable). If sonography is normal or shows mildly increased intraaxial and/or extraaxial fluid, then follow-up head circumference measurements and clinical evaluation will suffice in most cases, since this usually represents benign macrocrania. CT is indicated when there is a significant abnormality on sonography that needs further clarification or if the patient is beyond the age at which the fontanelle is large enough to allow an adequate sonographic examination.

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