The Course of Severe Untreated Infantile Hydrocephalus

Prognostic Significance of the Cerebral Mantle*

D. YASHON, M.D.,† J. A. JANE, M.D., AND O. SUGAR, M.D.

Department of Neurosurgery, University of Illinois, College of Medicine, Chicago, Illinois

The condition of infants with severe hydrocephalus who are not treated surgically may, in the long run, vary from death or complete disability to normal mental and physical performance. There is recent evidence that present day surgical therapy is more successful than no treatment at all. The decision as to surgical treatment thus has great practical significance. Many criteria including those of a social or philosophical nature must be evaluated. This paper is primarily concerned with one of these criteria, namely the problem of the relation between the thickness of the cerebral mantle (cerebral cortex and underlying white matter) and the final neurological status. Often the thickness of the cerebral substance has been a decisive factor in the determination of which patient should be subjected to surgical therapy. We will analyse 47 untreated patients who had severe hydrocephalus and were followed for an average period of 8 years; we will also evaluate other reports.

The determination of the thickness of the cerebral mantle in the living patient is made by ventricular tap, transillumination, or contrast study of the intracranial contents. Ventricular puncture gives an approximate measurement that is restricted to the area of needle insertion. The exact pallial thickness at which the head may be transilluminated is not known. On the basis of 5 cases examined at autopsy, von Bokay concluded that the average thickness of the cerebral mantle must be 1 cm. or less for transillumination to be possible. There can be no doubt, however, that when a skull transilluminates a very thin mantle exists.

The most accurate estimation of pallial thickness is obtained when air is utilized as a contrast medium. Air, introduced into the lumbar subarachnoid space, in addition to enabling one to measure the thickness of the pallium, may provide accurate diagnostic evidence concerning the nature of the hydrocephalus. In the event that ventricular filling does not occur, as in non-communicating hydrocephalus, air may be introduced by ventricular tap. Positive contrast material may also be used in this fashion for localization of the site of obstruction.

Clinical Material

The present series consists of long term follow-up studies of 47 infants who conform to 6 standards.

1. Presence of an Enlarged Ventricular System. The ventricular system was definitely enlarged as determined by air study. In most cases the thickness of the cerebral mantle was 2 cm. or less. Although transillumination was often performed and in many cases the depth at which the ventricle could be reached on puncture was measured, the estimation of pallial thickness for purposes of this study is restricted to those patients in whom air studies are available.

Between 20 and 40 cc. of air were introduced into the ventricular system, and a routine group of x-ray projections were then made. The frontal cerebral mantle was measured “brow up” in both antero-posterior and lateral projections. The occipital mantle was measured “brow down” in the lateral position. The parietal mantle was determined on lateral projection with the patient upright. The thickness of the cortex at the temporal tips was measured, when possible, on the “brow up” anteroposterior film. The cerebral mantle depth was also measured from the outer rim of bony calvarium to the closest contrast shadow due to air. The outer table was used because the relatively radiolucent bony edge of the inner table blends imperceptibly with the radio-

Received for publication December 14, 1964.

* Supported in part by Grant N 1504 A 64 from the Neurological and Sensory Disease Service Program, USPHS, and a Neurologic Birth Defects Grant from the National Foundation.

† Present address: Department of Neurosurgery, Cook County Hospital, Chicago, Illinois 60612.
lucent cerebral substance in these greatly thinned skulls. The separation between these two areas could not be made easily and consistently and the width of the bone was only a few millimeters in any event.

In a given patient the statement of average cerebral mantle thickness represents an average of these five projections or those which were available. The cerebral mantle thickness as measured on the x-ray film is approximately 10 per cent greater than the actual thickness due to the short distance of the roentgen tube from the infant’s head. This small difference is constant and it is not considered in the final statistics. The child’s head was in contact with the x-ray film in all cases.

2. Period of Follow-up. The follow-up period was at least 4 years in all cases and ranged up to 15 years; the average for the entire group of living patients (24) was 8 years. Estimation of the clinical course was made by one of the authors in most cases, particularly those in the “well” category. Other patients were evaluated by clinicians acquainted with the problem. Follow-up material was also obtained by questionnaire from parents and social service organizations.

3. Absence of Surgical Intervention. A major requirement for inclusion in this study was that the infant must have received no surgical therapy in the treatment of the hydrocephalic process. Moreover, no patient with other major disabilities which could influence survival was included. There were, however, 2 patients with associated small encephaloceles and 2 patients with myelomeningoceles; these seemed to have little influence with regard to survival. No obvious postmeningitic case is included in this group. The diagnosis of infantile meningitis may be difficult and only briefly noted as a febrile episode although communicating hydrocephalus may be the result.

4. Poor Prognosis as Judged by Clinical Appearance and Contrast Study. Perhaps the most significant, distinguishing feature of these 47 cases is the reason that they were denied surgical therapy. At present the policy at the Illinois Neuropsychiatric Institute is to treat with ventriculovenous shunt all infants having hydrocephalus (assuming that no other major abnormality is present). Cases in the present series were seen for the most part in the era before this type of shunt was available and when only those cases which were considered particularly favorable were subjected to surgical therapy. Thus, the present group includes mostly patients that were judged to have a poor prognosis, either because of thin mantles, large heads, or generally unimposing appearance. They were infants believed to be in such poor condition as to preclude the probability of social adaptability. It is of passing interest that the mortality in this group was lower than that among those treated with the surgical techniques available at that time.

5. Time of Original Evaluation. All children were evaluated originally as infants (the oldest was 36 months at the time of air study and the average age of the entire group at the time of air study was 6 months).

6. Progressive Nature of Disorder. For the most part these patients had actively progressive hydrocephalus at the time of the air study. One child (T.B.) was thought to have arrested hydrocephalus when first evaluated and indeed his head did not enlarge in size. However, an air study 5 years later revealed the cerebral mantle to be thinner than before. This case illustrates the fact that exact criteria must be maintained when the diagnosis of arrested hydrocephalus is made. Of the remaining 8 cases in which progression was not certain at the time of air study (average mantle 3.25 cm.) 2 have died, 3 are doing poorly and 3 are normal or near normal. These patients demonstrate that even with a thick mantle prognosis should be guarded.

Methods

The living patients were categorized into three broad groups on the basis of social acceptability. The estimation “poor” refers to those patients who have little or no voluntary motor power and show severe mental retardation. Measurement of mental capacity was not obtainable in this group. It is of interest that many of these children have far outlived clinical expectations in chronic disease hospitals. The “fair” category refers to those patients who are intermediate in social acceptability. These children have voluntary motor function and are usually able to ambulate but are not educable. In the main they adjust well in a home environment. The patients in the “well” classification have achieved normal to near-normal social acceptability. Motor power is normal in most cases. These patients are usually educable and in some instances normally so. Intelligence and psychological testing was performed in the “fair” and “well” groups. Surprising discrepancies in these children exist when social acceptability (behavior and appearance) is compared with mental capacity (IQ). Thus, the IQ itself may not be an entirely accurate measure of the final neurological and mental status.

Clinicians in the past have noted, but never documented, that verbal performance in hydrocephalic children is higher than one might expect when compared to general intelligence. In this group of patients we also noted this to be true. We were suprised in many instances when a child spoke well and seemed to comprehend but whose IQ fell short of what was expected.

Postmortem studies were performed at this