Congenital Communicating Hydrocephalus

LARS GRANHOLM, M.D., AND CLAES RÄDBERG, M.D.

Department of Neurosurgery* and Roentgen Department II,† Sahlgrenska Sjukhuset, Gothenburg, Sweden

Hydrocephalus, by current definition, means dilatation of the ventricular system, coupled with raised intracranial pressure, present or past. Dilatation of the ventricular system without increased pressure is ascribed to atrophy. The fundamental problems of hydrocephalus were elucidated further by the investigations of Dandy and Blackfan. They proved in experiments on the canine that the dilatation of the cerebrospinal-fluid pathways will extend only as far as the site of an experimental occlusion. They divided clinical hydrocephalus into communicating and non-communicating types, depending on whether or not dye injected intraventricularly appeared in the lumbar subarachnoid fluid. In non-communicating hydrocephalus there is an obstruction of the intracerebral cerebrospinal-fluid pathways. In some cases of communicating hydrocephalus a tumor, a midline defect, or a postmeningitic arachnoiditis will explain the disorder. But in most cases of communicating hydrocephalus it has not been possible to demonstrate any occlusion of the pathways with the prevalent technique of examination. This fact has caused much speculation. Communicating hydrocephalus frequently has been characterized as congenital. Anatomical abnormalities in the choroid plexus, hypersecretion of the cerebrospinal fluid, and stenoses or atresias of the intracranial venous sinuses all have been described as plausible explanations.

Surgeons, e.g. Dandy and Blackfan, Fraser and Dott, and pathologists, e.g. Russell and Laurence have described series of patients with communicating hydrocephalus and arachnoiditis, i.e. changes in the subarachnoid space following meningitis or intracranial haemorrhage. Intracranial haemorrhage is not unusual during a traumatic delivery. Radiologically a dilatation of the basal cisterns has been demonstrated in isolated cases in lumbar pneumoencephalograms and in a series of ventriculograms. No general conclusions have been drawn from all this evidence.

In the majority of patients with so-called "congenital communicating hydrocephalus," a dye test revealing communication is regarded as sufficient for diagnosis; and in most centers a ventriculogram with direct puncture of a lateral ventricle is considered the method of choice when an air study is indicated. On a ventriculogram the basal cisterns frequently are not at all or only partially air-filled, with resultant poor filling of the rest of the subarachnoid space. On the other hand, lumbar injection of gas allows one to direct gas deliberately into the cisterns with subsequent filling of the rest of the intracranial part of the subarachnoid space.

Selection of Material, Definitions and Interpretations

1. This investigation has been limited to patients in whom the neurological disorder was suspected before the age of 18 months. The age limit is quite arbitrary but will give a reasonably homogeneous group in which the clinical picture appears uniform.

2. Dilatation of the ventricular system has been estimated according to Evans. His ratio is the greatest width of the anterior horns divided by the greatest internal cranial diameter on the anteroposterior projection. A value of 0.30 or more indicates definite dilatation.

3. Cases have been classified as communicating when there was passage into the ventricular system from a lumbar injection of gas, or passage from the ventricles to the

Received for publication October 19, 1962.
* Director: Professor Gösta Norlén, M.D.
† Director: Docent Ingmar Wickbom, M.D.
cisterns during ventriculography. No other test of communication has been used.

4. The term "arachnoiditis" has been used for practical reasons throughout the paper, even if we have no pathological confirmation of the diagnosis. Cases were assigned to the group "arachnoiditis" when no gas or only a localized small amount was present over the cerebral convexities in the air study. Absence of air over the convexities has not been interpreted as indicating generalized "arachnoiditis." It is taken only as proof of the absence of communication between the basal cisterns and the convexities.

5. The amount of gas listed as "normal" in Table 2 means gas in several sulci over both convexities. The amount described as "small" means a localized, single air-bubble in the Sylvian fissure in a hydrocephalic patient and in one sulcus or a few sulci over the convexities in the controls.

**Technique**

Fractional lumbar encephalography has been done according to the principles of Robertson and Lindgren. General anesthesia was given to all patients. At the beginning of the examination the patient is seated leaning slightly forward with the head somewhat flexed and with the forehead resting against the vertically adjusted objectable of a skull-stand. The head is held up with a bandage of adhesive plaster or with a Glisson-sling, attached by bands to a metal arm, mounted on the upper border of the objectable. With the bands, flexion of the head is controlled.

The lumbar puncture is performed with a thin needle with a two-way stopcock. Only a few drops are allowed to drip off in order to make certain that the needle is placed correctly. The gas is injected in portions of 7–10 cc. At first the gas is directed into the ventricular system by using a suitable flexion of the head. When the anatomy of the 4th ventricle, the aqueduct and the posterior part of the 3rd ventricle has been demonstrated on the films and a sufficient amount of gas has passed on to the lateral ventricles, a certain quantity of cerebrospinal fluid is evacuated through the needle, although this must not exceed half the amount of gas injected. It usually is not necessary to inject more than 15 cc. of gas into an undilated ventricular system, but in cases of communicating hydrocephalus considerably greater amounts of gas are injected—up to 150 cc.—to make possible estimation of the lateral ventricles. Even when so great an amount of gas is injected, only a relatively small amount of fluid is evacuated. If the gas does not pass into the 3rd and lateral ventricles or if a stenosis of the aqueduct is present the encephalography is combined with ventriculography.

After the ventricular system has been filled sufficiently, the extracerebral part of the cerebrospinal pathways is investigated. Gas then is injected with more extension of the head than when the ventricular system is filled, and the gas usually will pass to the medullary, pontine, pontocerebellar, interpeduncular, ambient, and quadrigeminal cisterns. In addition, the chiasmatic cistern is often but not always filled. From the cisterns the gas passes up to the convexity, usually parietally in the sitting position. By injecting with extreme flexion of the head, the gas sometimes may pass to the upper surface of the cerebellum and vermis and to the quadrigeminal cistern. Gas is injected until the cisterns in question are sufficiently gas-filled or until obliteration of one or more of the cisterns is demonstrated convincingly.

The examination then is continued with the patient recumbent. In the supine position the gas usually passes from the interpeduncular to the chiasmatic cistern. From the chiasmatic cistern the air passes to the Sylvian fissure and to neighboring parts of the convexity and via cisterna laminae terminalis to the convexity over the frontal pole.

No serious complications attributable to this technique have occurred.

**Our Series**

All lumbar pneumoencephalograms on patients below 3 years of age done in Gothenburg between 1953 and 1962 were reviewed. Every patient with increased intracranial pressure, past or present, and with ventricular dilatation (according to Evans) was assigned to the group of hydrocephalus. Remaining studies form the control material.

A few older patients, qualified because of their age at the onset of the disorder, were included in the group of hydrocephalus.

In the years 1953 to 1962, 69 patients in this group have been admitted to hospitals in Gothenburg with the clinical diagnosis of hydrocephalus, later verified by lumbar pneumoencephalogram. No less than 51 of these 69 patients proved to have communicating hydrocephalus. The material is listed in Table 1. The series of 51 cases of communicating hydrocephalus was reduced to 37 when 14 patients with tumors, midline de-