IN THIS study we are presenting an anatomicoclinical picture which results from increased intracranial pressure in patients with extensive dysplasia of the brain.

Wilson’s use of the generic term encephalo-dysplasia, a “comprehensive term for the multitudinous aberrant faults of development to which the brain is subject,” is inclusive of all brain malformations. Certain malformations present anatomicoclinical and surgical reasons for consideration as a separate group, and we have therefore suggested the term hydroencephalo-dysplasia as applying to those patients who show signs of increased intracranial pressure at any period in their development.

For diagnostic purposes hydrocephalus in infants was differentiated according to the nature of the lesion, and the conclusion was reached that porencephaly should be considered as a related anomaly. Porencephaly has not been well defined. Heschel considered porencephaly a cerebral defect that allowed communication between the subarachnoid space and the ventricles. However contemporary authors are inclined to consider this condition as any malformation characterized by extensive loss of brain tissue whether in communication or not with the subarachnoid space or the ventricles. Intracranial pressure has been reported in patients who at varying times in life manifest symptoms of a porencephalic cyst. Cruveilhier noted that patients with a large part of the brain missing could be either microcephalic or hydrocephalic. The latter condition he called “hydro-anencephaly” and felt certain that the “atrophy” was not the result of the pressure of fluids. Similar instances in which the cerebral hemispheres were missing have been reported by others.

External hydrocephalus, that is, the accumulation of cerebrospinal fluid under increased pressure on the external surface of the brain, is unusual as a primary condition. Hygromas and subdural hematomas have been described under this heading in ancient times. According to Dandy, external hydrocephalus may occur as follows: In cases of communicating internal hydrocephalus, the cerebrospinal fluid may so distend the cisterns that they bulge and compress the brain, and even the arachnoid membrane may break and allow the fluid to escape to the subdural space where it cannot be ab-
sorbed, thus causing gradually increasing compression of the brain. This seldom happens spontaneously although it is a not infrequent complication of operations for hydrocephalus when the arachnoid membrane is opened or the cortex is divided.

In a series of 135 consecutive cases of hydrocephalus we have not found a single instance of primary external hydrocephalus as described by Dandy. Furthermore, wherever there was an accumulation of fluid over the surface of the brain there was extensive encephalodysplasia. This condition we have named hydroencephalodysplasia. Cushing (1928) stated that true external hydrocephalus may be associated with congenital anomalies. Probably his illustration in Osler’s Modern Medicine (p. 257) is of this condition.

In a previous study of 50 cases of hydrocephalus, the authors found 11 such malformations, then designated “hydrodysplasia.” These cases are included in the present study. The criteria are the same as in the previous study but the designation, hydroencephalodysplasia, (hydro, water; enkefalo, head content; dysplasia, maldevelopment) is considered more applicable to the condition. Twelve cases have been added, making a total of 23 cases of hydroencephalodysplasia in this study of 135 hydrocephalics.

**CLINICAL SYMPTOMS**

The ages of the patients ranged from 23 days to 2 years; 17 of the patients were males, 6 females; 21 were white, 2 mulatto (white and negro). Although the proportion of negroes in Cuba is as high as 20 per cent in certain areas there were no pure negroes in any of the groups.

**GENERAL SYMPTOMS**

The two most outstanding and constant findings were deformity of the head and psychomotor arrest. These symptoms were present to some degree in all patients. The increase in size of the head was similar to that seen in hydrocephalus and measurements varied from 41 cm. in circumference at 1 month to 56 cm. at 4 months. The fontanels bulged slightly and the shape of the head was not unlike that seen in congenital hydrocephalus (Fig. 1). There was one example of turricephaly which was probably the result of a certain amount of premature synostosis of the parietotemporal and lambdoid sutures. One patient was definitely microcephalic with a head circumference of 41 cm. at 2 years of age. Probably, increased cerebrospinal fluid pressure was present after completion of the premature closure of the cranial sutures.

In a previous review, we mentioned the phenomenon of transillumination of the skull or fontanels by a strong beam of light. More recently Hamby has described and beautifully illustrated this phenomenon.