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TRANSILLUMINATION OF THE INFANT HEAD DEMONSTRATED BY A CASE OF HYDRANENCEPHALY

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Transillumination of the infant head is a sign which deserves greater prominence. It is simple to elicit and requires only a darkened room and a source of illumination, with the head held in the beam of light. A pocket torch has been used with success, but the stronger the light used the more striking is a positive result.

Normally, and where the cerebral cortex is more than 1 cm. thick,[1] the test is negative and the head does not transilluminate. When hydranencephaly is present or when the cerebral cortex is very thin, a light placed behind the head in a darkened room will light up the entire head with an orange-red light, giving an appearance rather like a Chinese lantern. The vessels of the scalp and the basal structures of the brain stand out in dark contrast to the reddish glow of light through the head.

Soon after this sign was first brought to my attention[2] I found it most useful. Two Bantu mothers whose very young babies had large heads were seen on the same afternoon. As the nearest neurosurgical unit was 20 miles away, the journey costly and beds at a premium, it was decided to transilluminate the patients' heads as a simple screening procedure to determine whether operative interference was likely to be helpful.

The transillumination test was negative in the one infant with hydrocephalus, and this mother undertook the journey to the hospital, where it was decided that the baby was an early and suitable case for operation.

The second baby showed a strikingly positive test (Fig. 2) and was spared the journey to hospital. She is presented below as a case of hydranencephaly, believed to be the first reported in the South African Bantu. The prognosis is hopeless.

CASE REPORT

In October 1957 a female child of 4 months was brought to the clinic because her mother had noticed at the age of 2 months that the head was big. She stated that pregnancy and delivery were normal and the baby, who was the 5th child, cried at birth, and during the next few weeks appeared quite as normal in every way as the mother's preceding 4 infants. However, at 2 months of age it was noticed that the baby's head was unusually large. She had always sucked well at the breast and slept well at night, but during the day she was restless and cried a great deal. She had thrived and gained weight steadily (infant welfare clinic records were available to confirm this), but had not yet smiled, nor did she follow a light with her eyes or hold up her head.

The patient was a plump infant with a large head (Fig. 1), measuring 19¾ inches in circumference. There was no craniotabes; the anterior fontanelle was large, but the cranial sutures were not unduly separated; no tympanitic percussion note was obtained. Transillumination of head showed diffusion of light in all directions (Fig. 2). The pupils were equal and responded to light, but the child did not appear to appreciate light and the eyes did not follow a light or other moving objects. There was bilateral optic atrophy. The Moro reflex was still present in response to loud noises, or pulling the blanket from beneath the child. Reflexes were all present; plantars were extensor. Apart from the size of the baby, she resembled a vigorous newborn infant. An electro-encephalograph (Fig. 3) showed little or no evidence of electrical activity on the left side, and on the right high-voltage delta activity was recorded. This tracing supported the diagnosis of hydranencephaly.

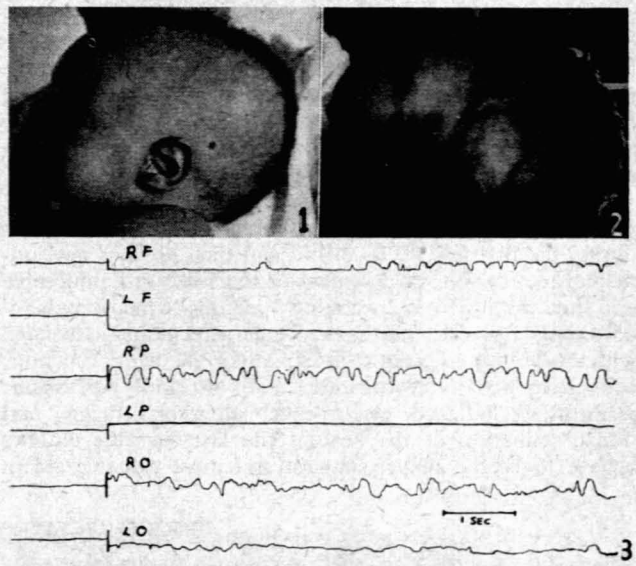

Fig. 1. Appearance of the patient's head at 4 months of age.
Fig. 2. The appearance of the patient's head when transilluminated. The source of light is behind the head. *Fig.* 3. Electro-encephalograph showing flat tracing on left side.

Five months later the child had shown no neurological improve-

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ment, irritability had increased, and the head had enlarged to a circumference of 22½ inches.

DISCUSSION

Hydranencephaly is a remarkable cerebral defect, probably due to the conversion of the cerebral hemispheres into single thin-walled, sac-like chambers due to a destruction of most, or all, of the cerebral cortex and white matter.[3] The basal ganglia, thalamus, and mid-brain are often to some extent preserved, and symptoms may be so inconspicuous in the early weeks of life as to give the impression of a relatively normal infant. After several months, failure of mental development, paralysis and epileptic manifestations become prominent symptoms.[4] In babies who live beyond the 3rd month an important feature is an abnormal increase in the size of the head, although this may occur earlier. The view is held that hydranencephaly constitutes a pathological process which is similar in kind to multiple cystic encephalomalacia[3] but more severe in degree. Yakovlev and Wadsworth, however, consider the defect to be a true cerebral agenesia.[5]

The condition was first described by Cruveilhier in 1835.[6] In 1950 Hamby *et al.*[7] reported the diagnosis of 7 cases during life seen in a period of 2 years, and made a plea for the frequent use of the transillumination test. One of their cases in whom this test was not used early had been passed for adoption at the age of 1 week by a competent paediatrician, but at 2 weeks of age the baby became irritable and the head was noticeably enlarged by 6 weeks. The foster-mother noticed on one occasion that the head was transilluminated by the light from a near-by floor lamp. These authors have tried the transillumination test on several children with communicating hydrocephalus but have found the residual brain tissue sufficiently opaque to prevent transillumination, except in one case where the cerebral layer was only 0·3 cm. thick.

In reviewing their 7 cases of hydranencephaly, Hamby *et al.* found the following clinical pattern: Normal behaviour during the first few weeks of life, but then obvious developmental retardation; enlargement of the head, first noticeable at a time varying from 2 weeks to 3 months; hyper-irritability becoming apparent with the head enlargement and associated with the Moro and grasp reflexes; abnormal ocular function; convulsive activity; tympanitic note on skull percussion; a diffusely flattened electro-encephalograph tracing and transillumination of the head. The last sign the authors believe to be the single, simplest and most valuable aid in diagnosis. Its value is lost if the fluid within the meninges is made opaque by blood, as after operation or needling, or by intracranial infection.

The prognosis for patients with hydranencephaly is poor, most cases dying before the age of 1 year, although Edinger and Fischer described one child who lived for 3½ years.[8]

As a screening procedure, preliminary to further expensive investigations or as part of the routine examination of small babies for adoption, the transillumination sign deserves greater prominence. It may be especially helpful in areas where radiological and other facilities are not available, as it proved to be in the case reported above.

SUMMARY

The infant head will transilluminate when hydranencephaly is present or if the cerebral cortex is very thin. Transillumination of the infant head deserves greater prominence as a screening procedure preliminary to further expensive investigations, or as part of the routine examination of small babies for adoption.

The sign, which is easy to elicit, is described, discussed, and illustrated photographically. A patient is described and hydranencephaly is briefly discussed. This is apparently the first case of this condition described in the South African Bantu.

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