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The Neonatal Hand-Mouth Reflex its Persistence in a Case of Hydranencephaly

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The hand-mouth reflex is a response which can be regularly elicited in healthy new-born infants, disappearing at three or four months of age. It was first described by a Russian worker, Babkin, in 1953, but publication in English translation only took place in 1960. Lippman of Leipzig (1958) and Parmelee (1963), working in Paris, have made further detailed studies of this reflex in mature and premature infants.

Babkin (quoted by Parmelee) described the reflex as follows:

"It is elicited by pressure on the infant's hand and consists of opening of the mouth. In many new-borns, when pressure is exerted on the hand, there is, in addition to opening of the mouth, flexion of the forearm, flexion of the head and closing of the eyes. Sometimes the flexion of the head is so acute that it is raised six to seven centimetres from the table."

The reflex is best obtained by sudden strong pressure with the thumbs on the palms of both hands of the infant. Lippman (1958) found that closing of the eyes was irregularly present, but that in addition to flexion of the head and opening of the mouth, there was rotation of the head to the midline when turned to the side.

No reference has been found to the investigation of this reflex in abnormal infants or in older children with mental defect or brain damage. Its presence in a child of three years with hydranencephaly and virtual absence of both cerebral hemispheres may therefore be of some interest.

Case History

This girl, aged three years, is the only child of healthy parents. There was nothing significant in the family background and no history of attempted abortion, illness or drugs taken during pregnancy. Birth was normal and birth weight 6 lb. 2 oz. The progress after delivery was thought to have been in no way remarkable by the parents, with satisfactory feeding and weight-gain.

She was first seen by the writer at two months of age, when examination showed an active, normal-looking infant with good physical development and colour. She cried a great deal, but was readily stilled by a bottle, which she accepted voraciously. Skull circumference was

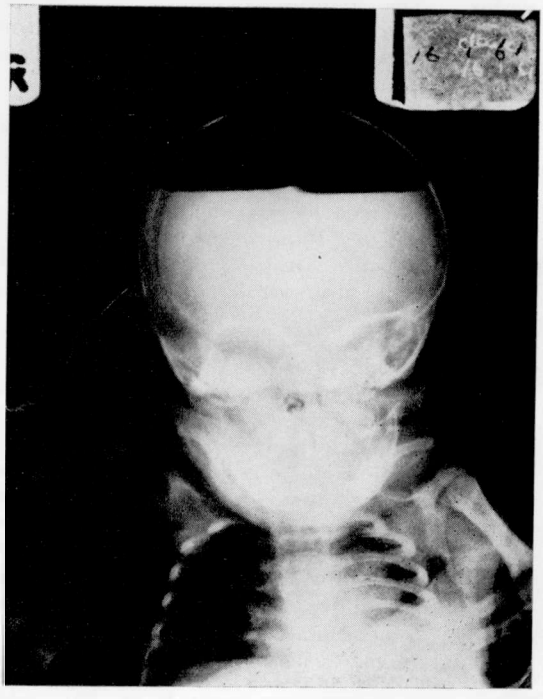

Fig. 1—Erect A.P.

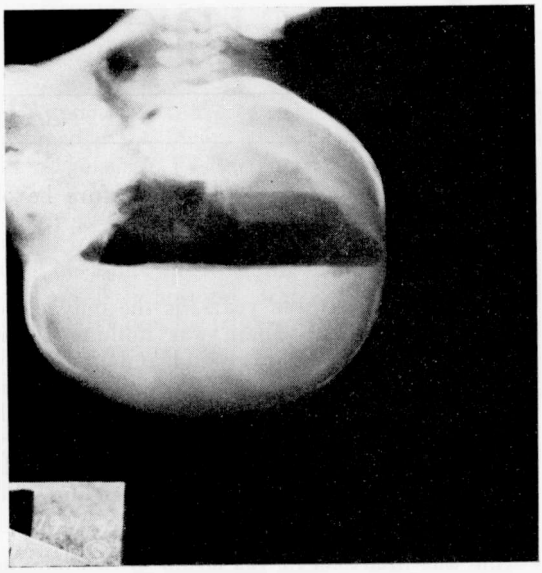

Fig. 2—Hanging head.

38.5 cm. This agreed with the mean expected circumference at two months for a birth weight of 6 lb. 2 oz. (O'Neill, 1961). The head was brachycephalic, with a rather steep, pointed vertex. The anterior fontanelle was widely patent, rather tense and cystic. There were no dilated scalp veins, however, nor was the shape of the skull suggestive of hydrocephalus.

The eyes moved concordinately, with normal roving movements, but there was no focusing or evidence of ability to see. On the other hand, she seemed to show momentary attention when spoken to. Pupils reacted to light. There was bilateral optic atrophy and the blood vessels were of small calibre, no choroidoretinitis being visible.

She showed a brisk Moro reflex to loud noises. Asymmetrical tonic neck reflexes of Magnus were not present. There was generalised increase in muscle tone, especially in the flexor groups of the upper limbs, hip adductors and extensors of the spine. All tendon reflexes were symmetrically brisk and plantar responses extensor. Crying caused bilateral tremulous movements of the upper limbs. The fingers, limbs and the head moved haphazardly and there was no sign of neuromuscular development past the neonatal stage.

No abnormality was noted in any other system.

Transillumination of the head in a dark room revealed brilliant translucency of the whole skull to the level of the ears on both sides. Through this translucency blood vessels and a central falx cerebri could clearly be seen. Light was visible also through the pupils and auditory meati (Figs. 1 and 2).

A full blood count and urine examination showed nothing abnormal. The Wasserman and Phenistix tests were negative.

A needle inserted through the anterior fontanelle encountered clear, colourless fluid immediately. This contained no cells and a protein of 22 mg./100 ml.: 120 ml. were removed and replaced by air. Radiographs (Figs. 3 and 4) showed that the calvarium contained two huge, symmetric fluid-filled cysts and that virtually

Fig. 3—Showing transillumination at three years of age.

no brain tissue was present apart from a small nubbin 3 cm. in diameter behind and above the sella turcica. The tentorium cerebelli could be seen clearly, but no air had entered below this.

The appearances were diagnostic of hydranencephaly.

Since these investigations she has been an inmate at the St. Francis Home, Bulawayo. Physical development is excellent (Fig. 5). She feeds well and has had none but the mildest of intercurrent infections. Neuromuscular development, however, has advanced little since she was first seen. When not sleeping or feeding, she lies placidly with open eyes and occasional limb movements. A moving figure in close proximity or speech close by appears to induce widening of the eyes and a slight turning of the head towards the stimulus. There is spontaneous smiling when she is contented. The Babkin hand-mouth reflex is active. Sudden pressure on both palms with the thumbs induces flexion of the head, opening of the mouth and a pursing movement of the lips (Fig. 6). Other reflex activity is summarised in the table.

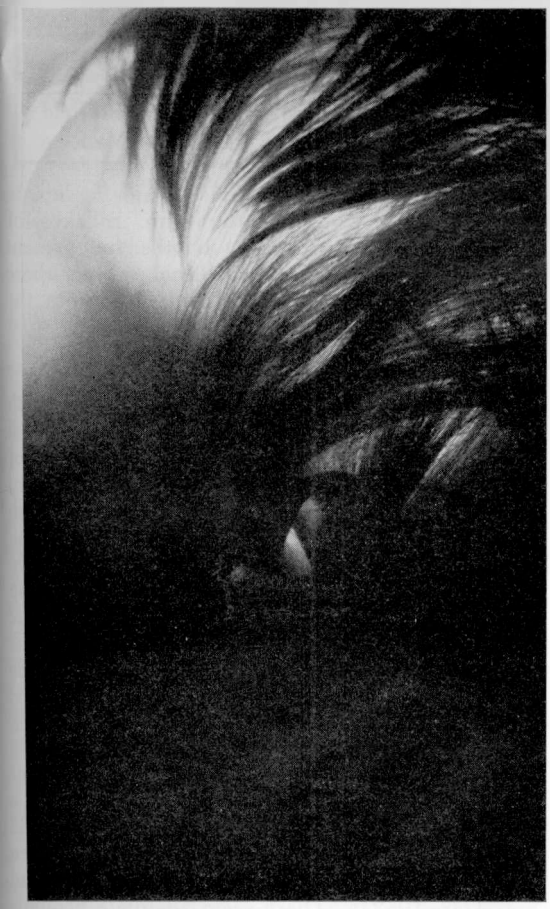

Fig. 4—Showing transillumination at three years of age.

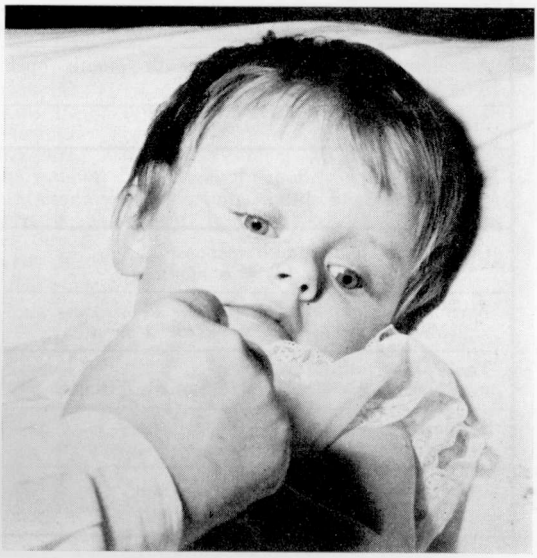

Fig. 5—Showing relatively normal appearances.

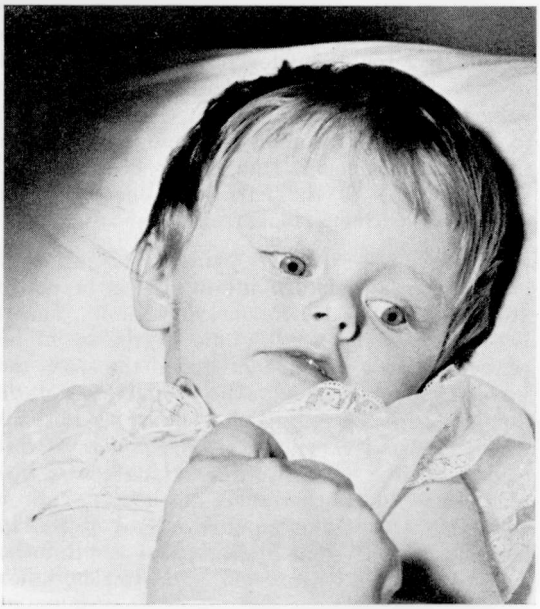

Fig. 6—The Babkin reflex. Note onset of head flexion and pursing of lips.

Table I

Neonatal Reflexes	Result (normal response in parenthesis)
Babkin hand-mouth reflex	Immediate, brisk.
"Rooting" reflex	Absent (mouth moves towards finger touching cheek).
Grasp reflex	Present + + bilaterally.
Strengthening of grasp during sucking (Bieber, 1940)	Present + + + bilaterally.
Associated hand movements with mouth opening (Alberts, 1955)	Absent.
Moro reflex	Absent.
Later Reflexes (Onset and disappearance in parenthesis)	
Conditioned hand-mouth response (Babkin, 1960) (3 → 6 months)	Absent (slow opening of mouth on flexion of forearm).
Asymmetrical tonic neck reflexes of Magnus (Magnus, 1926) (5 weeks → 4 months)	Present + + + bilaterally (extension of "face" arm flexion, abduction of "head" arm).
Tonic labyrinthine reflexes (5 weeks → 4 months) (Bobath & Bobath, 1955)	Present + + (maximal flexor tone when prone, maximal extensor tone when supine).
Supporting reaction (5 weeks → 4 months)	Present + + (rigid extension of legs when foot touches ground).
Landau reflex (6 months → 3 years) (Bobath & Bobath, 1956)	Absent (extension of head and spine when child held prone, supported beneath abdomen).

Discussion

This reflex is the most recently described and most constantly elicitable of a series of responses in the young infant which link the hand and the mouth in a senory motor reflex arc. Very little is known of the neurological mechanism involved in these responses, but they are thought to be mediated through the caudal part of the trigeminal nucleus in the upper cervical cord, which is one of the earliest to become functionally sensitive (Hooker, 1952).

The reflex is extremely primitive, being easily obtained in premature infants, even in one of 26 weeks gestation (Parmelee, 1963). That it is influenced at least to some degree by higher centre activity is evident from the fact that intensity of response varies with the state of the infant. The strongest response is obtained when the subject is awake and hungry, while testing during sleep or while crying elicits little reaction. In normal circumstances, and in common with other automatic and reflex behaviour, this response disappears at three or four months. As expressed by Parmelee, "there would appear to be a continuum from automatic responses to conditional and learned responses relative to hand and mouth interaction."

It is of interest that the tonic reflexes of Magnus were absent at two months, but present at three years of age. In normal infants these reflexes are usually clearly noted only from the fifth week onwards, when extensor tone first develops. They diminish after four months.

Clinical study of reflex activity in cases with developmental brain abnormalities may well prove fruitful in elucidating the mechanisms of normal development. In the case of hydranencephaly described there must be virtually complete absence of any functional connection between brain stem centres and the neopallium. Seventy-seven other mental defectives of varying age and aetiology were also tested, but none showed the Babkin hand-mouth reflex. On the other hand, I have recently seen a low grade defective African child who evinced this reflex. Encephalograms have not been obtained as yet.

Hydranencephaly is a relatively rare disorder which has been recognised for well over a century. In 1952 Moser could find only 32 recorded cases, and since then there have been at least seven further reports (Crome and Sylvester, 1958). There seem adequate grounds for differentiating it as a distinct entity from

hydrocephalus, anencephaly and other cystic encephalopathies.

The disorder is characterised by a relatively normal skull in which the cerebral hemispheres are replaced by thin-walled sacs containing cerebrospinal fluid. There is thus absence of all or the greater part of the solid cerebral tissue. The basal ganglia and the rostral parts of the mid-brain are also absent or degenerate in the most severe cases. Structures below this level are usually intact, showing merely agenesis of the long descending tracts (Crome and Sylvester, 1958).

The membrane lining the sacs consists of arachnoid and pia mater externally, and internally by a thin layer of glial tissue only. There is no ependymal lining to the cavity as in hydrocephalus. Hydranencephaly may be further differentiated from hydrocephalus by the absence of a characteristic "hydrocephalic" shape to the skull and, generally speaking, by the lack of excessive growth in head size. At least in some cases of hydranencephaly, however, there is a block in cerebrospinal fluid flow, with signs of raised intracranial pressure.

It seems generally agreed that hydranencephaly results from destruction and resorption of preformed solid cerebral tissue commencing before birth. Ischaemia is the mechanism favoured, perhaps from compression of carotid arteries by the umbilical cord.

Congenital syphilis, meningo-encephalitis, toxoplasmosis and severe maternal trauma have been suspected or incriminated in a few cases.

The whole subject and the relationship to hydrocephalus are well reviewed by Crome and Sylvester (1958).

The disorder is classically demonstrated clinically by transilluminating with a torch in a darkened room, when "the whole head lights up like an electric bulb." Such transillumination has recently been shown to occur in extensive hydrocephalus, porencephaly and subdural hygroma, though the translucency is usually more limited in extent (Dodge and Porter, 1961).

The fully developed condition is generally incompatible with long survival, almost all cases dying well within the first year of life. Length of survival, of course, is related primarily to the enthusiasm or otherwise of medical and nursing care. The oldest recorded case, that of Crome and Sylvester, died at 4½ years. The child here described is now 3½ years old and by present indications would seem likely to surpass this record with ease.

Summary

A three-year-old female child with hydranencephaly, diagnosed clinically, is reported.

She demonstrates a hand-mouth reflex heretofore described only in normal neonates.

The reflex was found also in one of 78 other mental defectives tested.

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Acknowledgments

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