Journal of the Institute of Medicine 1999; **21**: 1 –270 Case Report

Hydrancephaly: report of two cases

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ABSTRACT

Hydrancephaly is a rare brain malformation in which there is near total absence of cerebral hemispheres. The child may appear normal at birth but usually has a large head. The diagnosis is established by CT or MRI and there is no known treatment. This is a report of two cases of hydrancephaly diagnosed at our hospital.

Keywords: hydrancephaly; vascular insult; hydrocephalus.

INTRODUCTION

Hydrancephaly is a rare congenital malformation of the brain with devastating complications. We report two cases of hydrancephaly diagnosed at our hospital in the last four years.

Case 1

A seventy-day old male child, delivered by cesarean section for cephalopelvic disproportion with an apparently normal APGAR score after birth, was brought to the Tribhuvan University Teaching Hospital neurosurgery clinic for evaluation. The child had been sucking well, moving all limbs and had no urinary dribbling or respiratory problems. On examination, the occipitofrontal circumference (OFC) of the head was 48 cm; there were very large fontanels and the sutures were diastased. The patient moved all extremities with noxious stimuli. There was a setting sun sign and the head brilliantly transilluminated. A provisional diagnosis of Hydrancephaly was made and a CT scan was done. The parents were counselled and no specific treatment was offered.

Case 2

A nineteen-day old male child, delivered by cesarean section for foetal distress with an apparently normal APGAR score after birth, was brought to our neurosurgical clinic for evaluation. On examination the child was afrebile, had normal pupils, OFC 44 cm, large fontanels and diastasis of sutures. The patient moved all extremities to pain. There was no setting sun sign but the head was brilliantly translucent. Our clinical diagnosis of Hydrancephaly was confirmed by the CT scan.

DISCUSSION

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Hydrancephaly is one of the rare but devastating congenital malformations of the brain consisting of nearly complete absence of both cerebral hemispheres. Its incidence is unknown. The exact aetiology of the abnormality is still unknown but is believed to be a significant vascular insult in the territories of both internal carotid arteries after the fourth month of gestation, so that the portion of the brain supplied by the anterior and middle cerebral arteries is replaced by cerebrospinal fluid. The patient has a normal cerebellum and brainstem supplied by the vertebrobasilar system. The cranium is intact and there is a well formed and somewhat thickened sac consisting of an outer leptomeningeal layer and rudimentary representation of cortex. There is no evidence of normal ventricular configuration or ependymal lining but the ventricular surface is smooth and covered by a narrow layer of condensed glial fibrils (the membrana glial limitans). The condition is uniformly fatal; only rarely will the child survive beyond infancy. Before the CT era the condition used to be diagnosed by bilateral carotid angiograms in which there was absence of filling of both internal carotid arteries. From the management point of view, this condition should be differentiated from massive hydrocephalus, where there is thin cortical mantle, whereas in hydrancephaly there is no such cortical layer. There is no treatment for this condition. In cases where the head is very large, shunting the CSF reduces the size but does not alter the survival.

ACKNOWLEDGEMENT