CASE REPORT

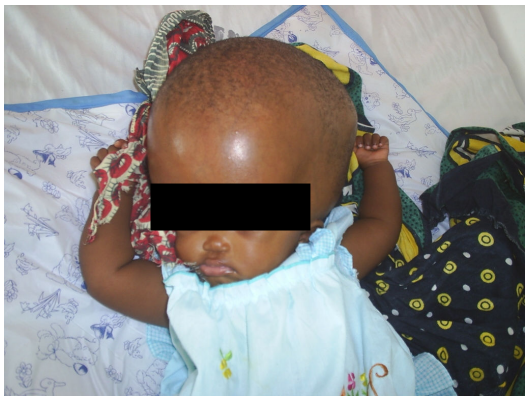
A 9 DAYS OLD, FEMALE BABY WITH CONGENITAL HYDROCEPHALUS

By Mgelea, Edward. (MD 4 – 2005/2006)

SUMMARY

A female infant, 3.5kg was born at Muhimbili National Hospital with a big head, Occipitofrontal Circumference (OFC) 49.5cm, and APGAR score 2 and 4 at first and fifth minutes respectively. She was admitted to ward 36 for the management of severe birth asphyxia. Cranial ultrasound revealed **Hydrocephalus** with bilateral ventriculomegally. Treatment is to perform **Ventriculoperitoneal shunting** at CCBRT hospital in Dar-es-Salaam.

Correspondence to: Edward Mgelea, +255-741-364-288; E-mail: edwardmgelea@yahoo.com.



INTRODUCTION

Hydrocephalus is a condition where there is an excessive accumulation of cerebrospinal fluid (CSF) under pressure and at times under no pressure resulting from impaired circulation and absorption of CSF or in some circumstances, from increased production by a choroid plexus papilloma[1].

Hydrocephalus may be of two (2) types; It may be **communicating** where there is no obstruction within the ventricular system of the brain (hollow space within the brain) or **non-communicating** (obstructive) where there is an obstruction of the ventricular system within the confinements of the brain[2]

Congenital anomalies may be etiologic in producing hydrocephalus or may be associated with hydrocephalus. Anomalies of ventricular flow system would produce hydrocephalus directly. The most common abnormality of ventricular flow is atresia of the aqueduct of Sylvius [3]. This blocks the fluid flow system between the third and fourth ventricles causing dilatation of the lateral ventricles and the third ventricle. The infant may be born with complete atresia of the aqueduct of Sylvius or may develop full occlusion of the aqueduct over the first few days or weeks of life. There is also a genetic sex - linked recessive form of hydrocephalus secondary to atresia of the aqueduct of Sylvius [4].

Another anomaly that will produce a non-communicating hydrocephalus is atresia of the foramen of Luschka and Magendie [5]. When it is associated with hypoplasia of the cerebellum and large cystic dilatation of the fourth ventricles it is referred to as Dandy-Walker anomaly.

There are other congenital anomalies of the brain and its bony coverings which are associated with hydrocephalus. The most common is Arnold-Chiari malformation where there is a displacement of the posterior fossa contents, the medulla or cerebellum into the cervical portion of the spinal cord on the basis of a multi-development of the cervical flexure during embryogenesis [6].

CASE PRESENTATION

A term female infant of 9 days old was born with a big head that was not noticed earlier in-utero. There was no history of a decrease or increase in size. She had neither convulsions nor fever. The blood vessels on the scalp were dilated. No abnormalities were noted except for the mucous vaginal discharge which is not unusual for a female baby.

Natal history, from rupture of membrane to the first dose of induction of labor by oxytoxin was about 7 hours, cervical dilatation was 2cm and 0.5cm long. The second dose five hours later ended with no changes in dilatation, where the decision to perform

emergency lower segment cesarean section was made. However there was a trial of pushing to deliver. On delivery the baby didn't cry immediately nor did she suck. She was just found to be a small baby with a big head.

After birth the APGAR score was 2 and 4 at first and fifth minutes respectively. That is a low score indication of severe birth asphyxia. The resuscitation was done by mask and then at neonatal ward kept under Oxygen therapy, the baby carried the next day with difficulty. In the ward the baby was fed expressed breast milk three hourly for three days. Until discharge, the baby was able to breastfeed.

On examination, anterior and posterior fontanelle had widen, Black curled hair, impaired up gaze (setting sun signs), dilation of scalp veins and hypertonic lower extremities and the face was broadened. No other abnormal features were seen.

Anthropometric measurement;
Occipitofrontal Circumference 49.5cm,
(normal 32-35cm conclusion, Hydrocephalus)
Length 49cm,
Weight 3.9kg.

On systemic examination of the central nervous system, the baby was alert with partial sucking reflex and a positive glabellar, rooting, grasp both hand and plantar, stepping, biceps, triceps, knee, ankle, tendon and Moro reflexes. The muscle tone was hypertonic. On examination of other systems no abnormality was detected.

The diagnosis was made after, cranial ultrasound scan (US) revealing huge hydrocephalus with bilateral ventriculomegally merging into one probably due to atresia of the aqueduct of Sylvius. Before the discharge (6 days) later, the Ultra-Sound scan was repeated showing the restriction of brain cortical growth due to fluid compression therefore the baby was surviving by midbrain.

During stay in the ward the baby was given IM vitamin K 1mg start, IM amplicillin, IM cloxacillin 195mg twice a day and IM gentamycin 20mg once a day all for seven days. Moreover, the baby kept warm with daily monitor of the respiratory rate, heart rate, temperature, Hypoxic Ischaemic Encephalopathy and OFC. The treatment of hydrocephalus is to shunt the CSF fluid to peritoneum (**ventrricoloperitoneal shunting**) where the mother was advised to attend clinic for possible surgical procedures at CCBRT.

DISCUSSION

Congenital Hydrocephalus is capable of producing brain atrophy[6], hence poor prognosis due to compression and may also be associated with severe mental retardation. Usually mental assessment together with other systems should be monitored cautiously because the birth asphyxia is associated with multi-organ damage. However this can be prevented in an up-to-date health centre by diagnosing hydrocephalus in uterine. The CT scans and US scan are the most important diagnostic tools.

We had to monitor complications which were associated with shunting such as plugging of the shunt, tube breakage or disconnection, infection, hematoma, thromboembolism, secondary craniosynostosis and isolated or trapped fourth ventricle. To monitor this we needed radiological evaluation of shunt malfunction such as plain films of the entire course of the shunt catheter, sequential CT or MRI scans for changes in the ventricular size or shunt position. Also abdominal Ultra-Sound evaluation may be useful to detect the presence of a CSF pseudo cyst or abscess secondary to an infected shunt. However in this child, there was a possibility of severe mental retardation and delayed milestones due to brain cortical growth failure, if the surgical procedures were delayed, therefore poor prognosis.

REFERENCES

1. Nelson , Text book of pediatrics, 17th Edition.

2. Danson, H, Dynamic aspects of cerebrospinal fluid. Deve med child Neuro, 14 (Suppl.G). 1 , 1972

3. Dandy, W. E. Diagnosis and treatment of strictures of the aqueduct of sylvius (causing hydrocephalus Arch surg (Chicago), 5, 1:1, 1945.

4. Bickers, D. S. and Adams, R. D : Hereditary stenosis of aqueduct of sylvius as a cause of congenital hydrocephalus. Brain 72.:245, 1949.

5. Dandy, W. E. Diagnosis and treatment of hydrocephalus due to occlusion of the foramina of Magendie and luschka. Surge Gynecol obstet, 32:112, 1921.

6. Peach, B. Arnold - Chiari Malformation : Morphogenesis, Arch Neuol, 12:527, 1955.