Demographic and Clinical Presentations of Pediatric Hydrocephalus in Medical City

Mahjoob N. Al Naddawi*,Numan Nafie Hameed * ,Amal A. Rasheed**

ABSTRACT:

BACKGROUND:

Hydrocephalus is distension of the ventricular system of the brain related to inadequate passage of cerebrospinal fluid from its points of production within the ventricular system to its points of absorption into the systemic circulation.

OBJECTIVE:

To study the demographic and clinical presentations of pediatric hydrocephalus in medical city complex , Baghdad.

PATIENTS AND METHODS:

This is a prospective study ,which was carried out on 100 children with a mean age+\- standard deviation (24.3 +/- 16.06)months and median 16 months, who were admitted to Children Welfare Teaching Hospital & neurosurgical department (Surgical Specialties Hospital) / Medical City complex, Baghdad in the period from April first 2009 to October first 2009. A specially designed questionnaires were used to aid the investigators in performing a family interview. General and neurological examinations, and investigations including neuroimaging studies were done.

RESULTS:

Of 100 patients,(62%)were males and (38%) were females, with male to female ratio of 1.6:1. Family history of congenital anomaly was positive in (26%) of patients, while hydrocephalus was positive in (14%). Seventy-Two percent of patients had congenital hydrocephalus. Eighty–Seven percent of patients were full term. Fifty-Seven percent of patients were products of NVD ,while (43%) were products of CS,(2%) of them were emergency CS& (41%) were elective. The macrocephaly was diagnosed or noted at birth in (32%)of patients. The study showed that U/S finding of hydrocephalus was positive in (49%).

CONCLUSION:

The majority of the patients had congenital hydrocephalus, but there was delay in the diagnosis of macrocephaly. Prenatal ultrasound diagnosis yield was low in this study. Family history of hydrocephalus and other neural tube defect was important to be elicited.

***KEYWORDS*:** hydrocephalus, pediatric, demographic, clinical presentation.

INTRODUCTION:

Hydrocephalus is an active distension of the ventricular system of the brain related to inadequate passage of CSF from its point of production within the ventricular system to its point of absorption into the systemic circulation. (1)

Normally, CSF flows from the lateral ventricles through the foramina of Monroe into the 3rd ventricle. It then traverses the narrow aqueduct of Sylvius to enter the 4th ventricle. The CSF exits the 4th ventricle through the paired lateral foramina of Luschka and the midline foramen of Magendie into the cisterns at the base of the brain. The CSF circulates from the basal cisterns posteriorly through the cistern system and over the convexities of the cerebral hemispheres. Hydrocephalus resulting from obstruction within the ventricular system is called obstructive or non communicating hydrocephalus, while hydrocephalus resulting from obliteration of the subarachnoid cisterns or malfunction of the arachnoid villi is called nonobstructive or communicating hydrocephalus**.** (2).

*Department of pediatrics, College of Medicine, Baghdad University.

** Children welfare Teaching Hospital, Medical City complex.

The incidence of neonatal hydrocephalus alone is unknown. When included in the diagnosis of spina bifida it occurs in 2-5 births in 1000.(3) The overall incidence of hydrocephalus is not known. Approximately 55% of all hydrocephalus are congenital. The etiology depends upon the age of the child. The clinical features are increase in the size of head, with wide anterior fontanelle, prominent scalp veins, sun-setting eyes, optic nerve atrophy, nystagmus and increased muscle tone in children up to 2 years. Children more than 2 years may present with these as hydrocephalus progresses; or if the fontanelles are closed, head size may be normal. These may present with optic atrophy or papilloedema, abnormal hypothalamic functions (short stature or gigantism, obesity, delayed puberty, primary amenorrhea or menstrual irregularity and diabetes insipidus) and spastic lower limbs. Performance IQ is worse than verbal IQ and learning problems are common.[4] The diagnostic procedures include measurement of head circumference, Plain X ray of head, Ventriculography, Pneumoencephalography, Ultrasonography, Computed Tomography and Magnetic Resonance Imaging, but Imaging plays a central role in establishing the diagnosis and guiding disposition and treatment of these patients.[5] Different types of treatment and operations with different outcomes were available.[6]

We aimed to study the demographical and clinical characteristics of pediatric hydrocephalus in medical city complex, Baghdad.

PATIENTS AND METHODS:

A descriptive study of 100 patients with hydrocephaly was carried out at Children Welfare Teaching Hospital & neurosurgical department (Surgical Specialties Hospital) / Medical City in Baghdad in the period from April first to October first 2009.The mean age+\- standard deviation was(24.3 +/- 16.06) months and median of 16 months. Age range was from one week to 14 years old. The patients were referred to the hospitals from health centers, GP(general physician) clinic, and hospitals.

The identification of children with hydrocephalus was based on the assumption that some children with these conditions had been referred to a pediatric clinic and registered with a diagnosis of hydrocephalus; others were diagnosed in the ward where the patient admitted. Both groups were examined by either of two specialists in the ward or the outpatient clinic. Children were included in the study according to the OFC measurement and the brain CT or brain ultrasound findings or brain CT alone. All included children had brain CT showing dilated ventricles.

Information included were name, age, sex, address, blood group (if not known by the family we did the investigation in our hospital labs), and other questions according to questionnaire designed to aid the investigator in performing a personal interview.

Examination of patient was done including general and neurological examination. Measuring OFC (using tape measure) , height or the length, and fontanel size (using tape measure), weight (using weight scale Secca).

Investigations in form of complete blood count (to exclude infection or anemia), TORCH (but was not always available in the teaching labs), brain CT scan (if recent film was available, a new one not requested), brain ultrasound (was done for those with opened fontanel), was done for all included patients.

The main diagnostic criterion for ventriculomegaly by antenatal ultrasound was the transverse diameter of the ventricular atrium at the level of the glomus of the choroid plexus measuring >10 mm, irrespective of gestational age. Hydrocephalus overt at birth was defined as congenital hydrocephalus with a head circumference larger than 2SD above the mean for gestational age Persson et al(2007)[7] or secondary to a pathology present at birth.

Maternal anemia was defined as blood hemoglobin level less than 11 mg/dl, we depended on documented data for blood tests done for mothers during pregnancy. Birth asphyxia was reported in the study according to APGAR score (if the score less than 8 for first 5 minutes of life).

For bleeding tendency , prolonged prothrombin time(PT)and partial thromboblastin time (PTT) was recorded{according to the control values of the labs}.

As the total number of patients with hydrocephalus were 100, so the statistics will be presented in percentages only.

RESULTS:

This study showed that (62%)of patients were males and (38%)were females with male to female ratio of 1.6:1.

The majority of the patients had congenital hydrocephalus (72%) , while (28%) had acquired one.

In this study, the prenatal data showed that maternal fever was present in (15%), rash (4%), previous history of toxoplasmosis (7%), maternal diabetes (7%), PET (10%), smoking (passive) (20%), anemia (19%), toxoplasmosis(7%).(table1)

The study showed that family history of congenital anomaly was positive in (26%) of patients, while hydrocephalus was positive in (14%).(table 2)

The study showed that U/S finding of hydrocephalus was positive in (49%), 26% of them were associated with other anomalies ,while (51 %) had normal prenatal U/S.

The study showed that (87%) of patients were full term ,while 7% & 6% were preterm & post term respectively .It also showed that (57%) were product of NVD ,while (43%) were product of C/S (2% of them were emergency & 41% were elective).

The study showed that neonatal jaundice was present in (47%), birth asphyxia (4%), bleeding tendency that was complicated by intraventricular hemorrhage reported in (4%) of cases , history of meningitis (16%), encephalitis (1%), and neonatal seizure (9%).

The macrocephaly was diagnosed or noted at birth in (32%)of patients , during first month of age in (11%), during first 6 months of life excluding first month in (24%), second half of their infancy period (> 6-12 months) in (3%) , and no patient had his first presentation with hydrocephalus beyond first year of life.(table 3)

Table 1 : Prenatal data

Family history (n =100)	%
Congenital anomaly	26
hydrocephalus	14
Spina bifida, meningocele	6
Mental retardation	5
Skin pigmentation	0

Table 2: Family history

Prenatal data (n=100)	%
Maternal fever	15
Rash	4
Previous history of toxoplasmosis	7
Maternal diabetes	7
PET	10
Smoking (passive)	20
Epilepsy	0
Anemia	19
Toxoplasmosis	7

Table 3: Hydrocephalus (macrocephaly) onset & duration

Age of onset(*n* =100)		%
Since birth		32
1st month		11
>1-6 month	Congenital	16
	acquired	8
	total	24
> 6- 12 month	Congenital	0
	acquired	3
	total	3
More than1 year		0

DISCUSSION:

This study showed that male to female ratio was 1.6:1 and this is consistent with Kadhom AH (1993)(8), but inconsistent with Murshid (2000)(9) who showed no sex preponderance.

Family history of hydrocephalus in the relatives was positive in (14%) patients and this was consistent with Murshid (2000) (15%)[9], Halliday (1986) (13%)[10] and Landingham (2008) (12.1%).[11]

Any of these figures is high enough to impress one that consideration of the X linked condition is important in counseling the parents of a male baby with hydrocephalus. Too often a positive family history is missed because doctors do not ask enough questions about early deaths in the mother's sibship, in her mother's sibship, or among the aunts' children.

The incidence of hydrocephalus was higher among consanguineous parents than unrelated parents. Our study showed that consanguinity was positive in (37%) of the cases , which is consistent with Murshid et al(2000).[9]

Majority of our patients were full term. History of prematurity was positive in (7%) patients and that was inconsistent with Murshid et al (2000) (62%)[9], Persson et al (2007) (44%)[7] and a study in Canada by Kulkarni et al(2007)(32%) (12). This may be justified by the fact that these studies included only patients less than one year of age while our study included older ages (up to 14 years), also it may be attributed mainly to the increased survival of extremely preterm neonates with a high risk of developing hydrocephalus after cerebral hemorrhage in the developed countries.

The mode of delivery was NVD in (57%) patients , a figure similar to that of Murshid et al (2000) (58%).[9]

In this study, (72%) patients diagnosed to have congenital hydrocephalus ,which is consistent with Persson et al (2007) (51%)[7], Adeloye et al(1997) (51%)[13], but inconsistent with Kulkarni et al (2007).[12]

The improved maternal nutrition and addition of folic acid before and early in pregnancy have probably contributed to the decrease in the prevalence of hydrocephalus and meningocele (Honein et al. 2006).(14) However in this study (17%) patients had history of regular maternal folic acid intake at least in first 3 months of gestation, while (36%) had not taken the folate at all. In comparison to Kadhom AH (1993) (8), only (4.8%) of mothers had regular intake of folic acid. This finding may be due to improved knowledge among people about the benefit of folate supplementation during pregnancy .

In this study (57%) patients were from urban areas, the rest were from rural areas. So there was no significant difference , which was similar to Kadhom AH (1993)[8], who found no significant difference in the residence of those patients. However there is vague definition of the rural and urban areas and difficulty in applying that in our country.

This study showed that mothers of (49%) patients had an antenatal ultrasonograghic evidence of hydrocephalus in the fetus, reported as ventriculomegaly, (26%) of them had associated anomalies documented in addition to the hydrocephalus. According to JOO study (2008)(15), (99%) of the patients were diagnosed to have hydrocephalus during antenatal period by ultrasonograghy. This difference could be explained by the lack of antenatal care of most of our mothers or by the inaccurate reports of sonographists.

History of meningitis in the neonatal period was reported in (16%) of patients. This was documented by cerebrospinal fluid analysis. A larger figure was given in Uganda by Warf BC et al (2001)(16),who showed that (68%) of infantile hydrocephalus was due to post neonatal meningitis.This could be explained by increased incidence of such infectious disorders at these areas or the insufficient infection screen in their country so they depend on a history of acute illness in previously well neonate.

We found that (70%) of the patients had macrocephaly,this is due to closure of fontanells that prevent enlargement of skull in children older than 3 years,who develop hydrocephalus due to any pathology as infection (e.g. meningitis), intracranial hemorrhage or brain tumor .Routine measurements of head circumference during the first year of life mainly detect infants with hydrocephalus or cysts; other expansive conditions yield other symptoms. Most children with increased head circumference as a symptom of intracranial expansion are identified during the first 10 months of life[17].

In this study, (17%)of the patients had ataxia either pre or post treatment(shunt) , related to cerebellar signs(i.e. posterior fossa tumor) or due to shunt obstruction. In both, there is an elevated intracranial pressure. Poor vision was detected in (15%) of the patients, either through history or clinical examination . The same figure was found by Persson

et al (2007) (7). It was difficult to perform ophthalmological examination in newborns. But all were advised to have eye examination later.

In this study, other symptoms of increased intracranial pressure as headache, irritability & convulsion were reported. Epilepsy of different types was reported in (34%) of the patients. The same figure was found by Persson et al (2007).(7)

CONCLUSION:

The majority of the patients had congenital hydrocephalus, but there was delay in the diagnosis of macrocephaly. Prenatal ultrasound diagnosis yield was low in this study. Family history of hydrocephalus and other neural tube defect was important to be elicited.

We Recommend to start antenatal screening program by accurate US examinations and measuring maternal serum alpha-fetoprotein (MSAFP) ,TORCH screen(toxoplasmosis, rubella, cytomegalovirus and herpes virus) for those women with recurrent abortions or mothers with previous baby having neural tube defect.

REFERERENCES:

1. Rekate HL.A contemporary definition and classification of hydrocephalus. Semin Pediatr Neurol. 2009;16:9-15.
2. Stephen L.K , Johnston M V: Congenital Anomalies of the Central Nervous system: Hydrocephalus. In: Kliegman RM, Behrman RE, Jenson HB, Stanton BF(editors), Nelson Text book of pediatrics, 18th ed. Philadelphia, Saunders; 2007: 2452-5.
3. Gomella TL, Cuuningham MD, Eyal FG. Hydrocephalus and Venticulomegaly, in: Lange Neonatology: Management, Procedures, On-Call Problems, Diseases, and Drugs,Chapter 90, 6th ed., 2009:483-87.
4. Rizvi R, Anjum Q. Hydrocephalus in children. J Pak Med Assoc. 2005;55:502-7.
5. Vertinsky AT, Barnes PD. Macrocephaly, increased intracranial pressure, and hydrocephalus in the infant and young child. Top Magn Reson Imaging. 2007;18:31-51.
6. Kadhim SF, Al-Shalchy AK. Study of hydrocephalus and complications of surgical treatment. J Faculty of Medicine/Baghdad 2009 ;51:248-50.
7. Persson EK, Anderson S, Wiklund LM, Uvebrant P. Hydrocephalus in children born in 1999-2002: Epidemiology and outcome and ophthalmological findings. Childs Nerv Syst. 2007; 23:1111-8.
8. Kadhom AH. Congenital hydrocephalus and neural tube defects. A thesis submitted to the scientific council of pediatrics in partial fulfillment for the degree of fellowship of the Iraqi commission for medical specializations in pediatrics Mosul 1993.
9. Murshid WR, Jarallah JS, Dad MI. Epidemiology of infantile hydrocephalus in Saudi Arabia: birth prevalence and associated factors. Pediatr Neurosurg 2000; 32:119-23.
10. Halliday J, Chowt CW, Wallaget D, Danks DM. X linked hydrocephalus: a survey of a 20 year period in Victoria, Australia. J Med Genet 1986; 23:23-31.
11. Landingham MV, Nguyen TV, Roberts A, Parent AD , Zhang J. Risk factors of congenital hydrocephalus: a 10 year retrospective study. J Neurol Neurosurg Psychiatr 2009; 80:213-7.
12. Kulkarni AV, Shams I. Quality of life in children with hydrocephalus: Results from the Hospital for Sick Children, Toronto. Neurosurg 2007;107: suppl:358–64.
13. Adeloye A, Khare R. Ultrasonographic study of children suspected of hydrocephalus at the Queen Elizabeth Central Hospital in Blantyre, Malawi. East Afr Med J 1997; 74:267-70.
14. Honein MA, Paulozzi LJ, Mathews TJ, Erickson JD, Wong LY. Impact of folic acid fortification of the US food supply on the occurrence of neural tube defects. Jama 2001; 285:2981-6.
15. Joó JG, Tóth Z, Beke A, Papp C, Toth-Pal E, Csaba A et al. Etiology, prenatal diagnostics and outcome of ventriculomegaly in 230 cases. Fetal Diagn Ther 2008; 24:254-63.
16. Warf BC, MacGowen L. Post infectious hydrocephalus: The most common cause of hydrocephalus in Uganda. Report of International federation for hydrocephalus and spina bifida (IF-HSB).Consultative status special category with Economic and Social Council of the United Nations Consultative status, Council of Europe, sep.,2001.

17. Zahl SM, Wester K. Routine measurement of head circumference as a tool for detecting intracranial expansion in infants: what is the gain? A nationwide survey. Pediatrics. 2008 ;121:e416-20.