Evaluation of Congenital Hydrocephalus Association With Aqueduct Stenosis in Mosul Pediatric Patients

Zhraa Abd-Al Kader Taboo

ABSTRACT:

BACKGROUND:

Congenital hydrocephaly is one of the most common central nervous system anomalies. This generally refers to a condition that exists prenatally and excludes other neural tube defects ; this condition usually develops by the twentieth week of gestation , and defect can occur either alone, in association with spina bifida, or as part of a greater syndrome, such as Dandy-Walker Syndrome. There are various types or classifications of congenital hydrocephaly. Aqueductal stenosis is a type of hydrocephaly that results from narrowing of the aqueduct of Sylvius, an opening connecting the third and fourth ventricles in the brain.

OBJECTIVE:

There are few studies evaluating the incidence of aqueduct stenosis out of hydrocephalus patient in pediatric population, in Mosul city ,therefore, this topic was considered in present study .

MATERIAL AND METHOD:

The study was conducted in Ebn Sena Teaching Hospital, Department of Neurosurgery and Radiology in Iraq Mosul city, as a retrospective study for 250 cases collected between October 2007 and 31 December 2008, and prospective for 250 patients are studied between June 2009 and January 2011. Spiral computed tomography of the brain was performed for all patients. five hundred patients were analyzed by spiral computed tomography.

RESULTS:

During the analyses the peak age incidence of congenital hydrocephalus association with aqueduct stenosis was below 2 years old (39%). The male to female ratio was (1.3:1). The incidence of aqueduct stenosis was (63.9%) out of congenital hydrocephalus and (16.6%)out of acquired cases. In this series the non-communicating type of hydrocephalus was forming about (65.6%) and the communicating type forming about (16.2%), the lest type is compensatory (10.4%).

CONCLUSION:

The most common cause of hydrocephalus was congenital in origin (56%), and it was mostly due to congenital aqueduct stenosis (63.9%).

KEYWORDS: hydrocephalus - congenital - aqueduct stenosis .

INTRODUCTION:

The cerebral aqueduct of Sylvius is a structure within the brainstem that connects the third ventricle to the fourth. It is located between the pons and the cerebellum [(1)]. A blockage in this duct is the most common cause of hydrocephalus in infancy and childhood [(2)]. The causes of this obstruction are divided as congenital causes (atresia of aqueduct congenital infection brainstem malformation) [(3, 4)] and acquired causes (infective, traumatic) [(5,6)],neoplastic causes (benign, malignant) and idiopathic. [(7).]

Congenital Hydrocephalus results from a complex interaction of genetic and environmental factors and is present at birth. It is important to remember that the term genetic does not imply that it is hereditary. Often the exact cause of congenital Hydrocephalus cannot be determined. Though it might not be recognised and diagnosed immediately, congenital Hydrocephalus is often diagnosed before birth through routine ultrasound. Hydrocephalus diagnosed in adulthood may have existed since birth and can still be considered congenital and may be referred to as compensated Hydrocephalus.[(8)]

Hydrocephaly is the first complication of aqueduct stenosis which is defined as an enlargement of head caused by an abnormal

Department of Anatomy-Embryology -Nineveh Medical College.

accumulation of cerebrospinal fluid in the cranium due to an imbalance between the production and the absorption of cerebrospinal fluid. This forces the ventricles to enlarge, which in turn exerts pressure on the surrounding brain tissue, causing the brain tissue to shrink and the head to enlarge [9].

The word hydrocephalus is a Greek word *Hydro* means water and *cephalus* means head [4]. Hydrocephalus is usually associated with a rise intracranial pressure [10].

According to the National Institute of Health (NIH) website, there is an estimated number as 700,000 children and adults living with hydro-cephalus. Pediatric hydrocephalus affects one in every 500 live births, making it one of the most common developmental disabilities. It is the leading cause of brain surgery for children in United States. There are over 180 different causes of the condition, Pediatric hydrocephalus may also be a heritable condition and the most common of them is aqueductal stenosis, and mainly affects males [3].

Large head size, episodic vomiting, sleepiness, reduced activity, poor feeding, failure to thrive, irritability, sun setting and seizure disorders are considered as symptoms in infants [6,9]. while symptom in children are headache, blurred vision, diplopia, stunted growth, gait disturbance, urine incontinence, lethargy, and eventually deterioration of the level of consciousness leading to coma and death in severe cases [6,9].

There are two types of hydrocephalus(non-Communicating Hydrocephalus, Communicating Hydrocephalus) depending on whether it was a reabsorption problem or a blockage somewhere within the ventricular

One of the most common causes of non-communicating hydro-cephalus is "aqueduct stenosis". [3]. While Communicating Hydrocephalus (non-obstructive) can be caused by meningitis, sub-arachnoid hemorrhage, bilateral subdural haematoma, anoxia, leukemia, lymphoma and disease of connective tissue. [11,12, 13].

Congenital aqueductal stenosis is the most common of the developmental causes of congenital hydrocephalus and it develops about the 6th week of gestation. Other possible associated causes are Arnold Chiari malformation, Dandy walker cyst, agenesis of foramen of Monro and others. Furthermore, brain tumors, abscesses and haematoma , hemorrhage Infection, could be a causes of acquired aqueduct stenosis and hydrocephalus. [3,4,5].

AIM OF THE STUDY:

there are few studies evaluating the incidence of aqueduct stenosis out of congenital and acquired hydrocephalus patient in pediatric population, in Mosul city ,therefore, this topic was considered.

PATIENTS AND METHODS:

The present study is based on CT (computerized tomographic) scans to detect the intracranial abnormality as hydrocephalus by means of measures the size of the fluid spaces, or ventricles, within the skull. Five hundred patients under 10 years old were studied after admission in the department of Neurosurgery in Ebn Sena Teaching Hospital in Mosul city. The study was combined prospective and retrospective . The prospective study for 250 patients are studied between June 2009 and January 2011 and retrospective for 250 patients are collected between October 2007 and 31 December 2008. The questions as patients name, age, sex, occupation , clinical presentation, the sign and symptom was taken and reported for each patient on a special forma. The patients were divided according age into five groups, sex into two groups and also according to the radiological finding into many subdivisions. All the patients are examined in the department of Radiology, in the spiral computed tomography unit in the same center. The spiral computed tomography used in examination of patients is siemens somatom plus 4 made in Germany . The CT scanner takes a series of x-ray images which can be used to view "slices" of the body (or any other object), or to create a 3D image. This type of image can be used to view the ventricles, to diagnose many of the causes leading to hydrocephalus, to check the shunt catheter placement, etc. Often the CT scanning will involve the use of contrast dye to create better images of the scanned area.

Statistical method:

Statistical analysis of data were carried out and data were expressed as number and percentage. Chi-Square or Fissure Exact test of proportions for comparison between various groups was obtained for assessing the level of significance. The statistical results were considered significant at P-value equal or less than 0.05.

RESULT:

The analytic results were as follow:

The table (1)shows that the cases were grouped according to their ages into five groups. The pattern of age distribution within these groups were 0-2, 3-4, 5-6, 7-8, 9-10years,respectively. Out of 500 cases, hydrocephalus due to various causes had the largest population 92.2 versus7.8 cases were normal. The peak incidence of age in this study was below 2 years old (39%), there was a gradual non-significant decrease with age till the lowest age group affected between 7-8year. The Chi square is 9.44 and the P-value is 0.002 <0.05 (significant). The incidence of congenital hydrocephalus is greater in males than in females, in a ratio of (1.3:1), as shown in Table (2)

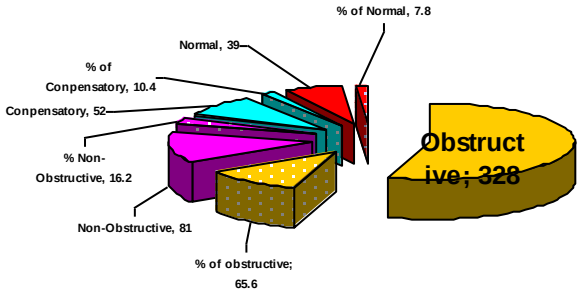
The incidence of obstructive (non-communicating) hydrocephalus was 328 patients (65.6%) where as the non-obstructive (communicating) hydrocephalus was 81 patients (16.2%), and the compensatory type was 52 patients (10.4%) out of 500 patients, added to that 39 patients (7.8%) were normal. In all these types the peak age incidence was below 2 years, the Chi-square is 12.201 and the P-value is 0.001<0.05 (significant), pie chart (1).

The distribution of congenital hydrocephalus versus acquired was 280 patients (56%) and 181 patients (36.2%) respectively and the rest was the 39 normal patients (7.8%) all are out of 500 patients, the Chi-square is 10.52, the P-value is0.003<0.05 (significant). Pie Chart (2).

The incidence of the congenital aqueduct stenosis was (63.9%) out of 280 patients, however the incidence of acquired aqueduct stenosis in 181 patients with acquired hydrocephalus was (16.6%). The statistic study in this series show that the prevalence of congenital aqueduct stenosis was 0.73, odds ratio was 0.781, Chi-square was 16.32, accuracy was 0.71, and the predictive value was 0.003<0.05 (significant). Table (3).

Regarding the grade of aqueduct obstruction(complete, partial) and level of obstruction , it was found that the complete obstructed in (50.2%) patients while partially stenosis in(49.8%)patient. The level of obstruction was distributed in upper 3rd (17.3%) patients, middle 3rd (25.7%) patients, lower 3rd 5 (31.3%) patients and whole length (25.7%)patients ,Table (4).

The most common causes of congenital hydrocephalus are aqueduct stenosis (63.9%),the Arnold Chari malformation(14.3%)and Dandy Walker cyst(10%)in comparison with other congenital causes as shown in Table (5)



Pie chart 1: The incidence of type of hydrocephalus in 500 patients

Table 1: Test comparison between hydrocephalic cases and normal cases for various age groups and its% out of 500 patients.

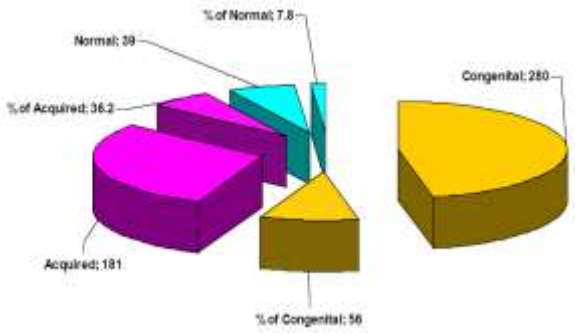
Age group in years	Abnormal cases (Hydrocephalic)		Normal cases		Total No. of patients	% out of 500 patients
	No. of patients	% out of 500 patients	No. of patients	% out of 500 patients		
0-2	195	39	5	1	200	40
3-4	130	26	10	2	140	28
5-6	61	12.2	9	1.8	70	14
7-8	30	6	8	1.6	38	7.6
9-10	45	9	7	1.4	52	10.4
Total	461	92.2	39	7.8	500	100

Chi-square = 9.44 , P-value = 0.002 < 0.05 significant

Table 2: Percentage of sex distribution of the study sample

Age group	No. of patients	% out of 500
Male	284	56.8
Female	216	43.2
Total	500	100

Male to Female Ratio: 1.3 : 1



Pie Chart 2: Analysis of hydrocephalus according to the cause (congenital or acquired)out of 500 patients.

Table 3: Test comparison between congenital and acquired hydrocephalus with respect to positive aqueduct stenosis for various age groups.

Age groups in years	Aqueduct stenosis out of all congenital causes (280)	%	Aqueduct stenosis out of all acquired causes (181)	%
	No.		No.	
0-2	95	33.9%	16	8.8%
3-4	40	14.3%	4	2.2%
5-6	18	6.4%	4	2.2%
7-8	15	5.3%	3	1.7%
9-10	11	4%	3	1.7%
Total	179	63.9%	30	16.6%

The prevalence: 0.73 ,Odds ratio: 0.781, Chi-square: 16.32, Accuracy: 0.71, Predictive value 0.003 < 0.05 (significant)

Table 4: Grade and site of aqueduct stenosis according to Ct-Scan.

No. of patients	Grade of obstruction			Level of obstruction				
	Complete	Partial	Total	Upper 3rd	Middle 3rd	Lower 3rd	Whole length	Total
No.	90	89	179	31	46	56	46	179
%	50.2	49.8	100	17.3	25.7	31.3	25.7	100

Table 5: Congenital causes of hydrocephalus in 280 patients.

Congenital Causes	No. of patients	%
Aqueduct stenosis	179	63.9
Septum pellucidum cyst + corpus & cerebellum agenesis	3	1.07
Arnold Chiari (pure)	40	14.3
Dandy walker cyst	28	10
Menegocele & Meningomyelocele	2	0.71
Obstruction of foramina of Monro	4	1.43
Agenesis of frontal lobe	1	0.36
Interventricular cyst	1	0.36
Holoprosencephaly	2	0.71
aneurysm of vein of Galen	2	0.71
Arachnoid cyst	9	3.21
Trigeminal cistern cyst	1	0.36
Giant cisterna magna	1	0.36
Congenital autosomal leukodystrophy	7	2.5
Total	280	100

DISCUSSION :

Many surveys have been carried out in the recent years to determine the incidence of hydrocephalus and other central nervous system abnormalities. In this series the peak age incidence of hydrocephalus was below 2 years old (39%) which was mostly of congenital type and the aqueduct stenosis formed the main cause. However such findings were parallel to (Alberto J. Espay 2009)[5]which said that the incidence of human hydrocephalus presents a bimodal age curve. One peak occurs in infancy and is related to the various forms of congenital malformations, another peak occurs in adulthood. This can be due to the premature infants have an increased risk of intraventricular hemorrhage in which severe bleeding within the ventricles of the brain can lead to hydrocephalus. Other problems that can occur during pregnancy may increase an infant's risk of developing hydrocephalus, including intrauterine infection or a disorder involving incomplete closure of an infant's spinal column and myelomeningocele. Some authors have identified no sex preponderance in hydrocephalus [14,15], but other have found a male preponderance[16,17,18]. ; there was a mild male domination(56.8% vs. 43.2.%) in our study.

Regarded to the incidence and type of hydrocephalus our result was in the same direction in comparison with the other authors who were divided hydrocephalus into communicating an non-communicating. [3,12], The author Losowska-Kaniewska D,Ole's A in Poland in (2007), mentioned that the commonest type of hydrocephalus is obstructive[19]

Concerning the incidence of congenital and acquired hydrocephalus: Losowska and Ole's in Poland (2007) stated that about (55%) of hydrocephalus are congenital in origin [20]. Other authors like Chumas et al., in United Kingdom and Ireland stated that the prevalence of congenital hydrocephalus were between 0.48and 0.81 per 1000 birth (60.5%), [21]. That is to say the percentage of congenital hydrocephalus in Iraq Mosul city is less than the incidence of hydrocephalus in west countries and its nearer to the incidence of this disease in the east countries.

Hydrocephalus due to congenital stenosis of aqueduct of sylvius is listed as rare disease by the office of rare diseases of the national institutes of health. This mean that hydrocephalus due to congenital stenosis of aqueduct of sylvius affects less than200,00people in the US population[22]. Another study done in Iran (2007)indicated that the incidence of congenital aqueduct stenosis(58%)while acquired aqueduct stenosis caused by space occupying lesions and calcified arteriovenous malformation were(37.5%)[23]. In 2010 John and Collin in USA Minnesota city found the incidence of congenital aqueduct stenosis was (43%).[24]Although this result was slightly lower than our finding but agreement with us that the commonest cause of obstructive congenital hydrocephalus are aqueduct stenosis. In fact there is no explanation about increasing incidence of congenital anomalies in infancy and childhood ,although this could be due to stress and uranium used weapons in the war against Iraq.

Though estimates of the grade and sit of aqueduct stenosis, patients with severe hydrocephalus generally have a stenosis in the proximal aqueduct, either at the level of the superior colliculi or at the entrance to the aqueduct immediately inferior to the posterior commissure. In patient with mild hydrocephalus, the level of the obstruction is often more distal. In distal aqueductal stenosis, the proximal aqueduct is dilated and the tectum is displaced posteriorly and stretched. Primary neoplasm that fill the aqueduct rather than compress it are uncommon [20].

Robert, (2011)found in their research that (90.9%)cases of aqueduct stenosis had complete occlusion and (9.1)had high grade stenosis [25]. In our series it's mostly complete obstruction (50.2%) and the level of obstruction was the most likely at the lower third. This difference is due to the high quality of imaging in different axes and plan performed there in the countries of the previous mentioned authors (axial, sagital and coronal) which is not usually done for each patient in Mosul imaging centers.

Hydrocephalus is the most common congenital malformation of the brain and aqueduct stenosis is the most common cause of it. It may be a solitary malformation or may be associated with other entities such as Arnold-Chiari malformation and meningomyelocele. Aqueduct stenosis may be inherited as an x-linked recessive[26,27].Furthermore these results were actually observed in the patient surveyed with

this study. It was noticed that the most common causes of congenital hydrocephalus was pure aqueduct stenos is(50%),Arnold Chiari malformation(14.3%)and dandy-walker cyst(10%).

CONCLUSION:

1-The most common cause of hydrocephalus was congenital in origin (56%) and it was mostly due to congenital aqueduct stenosis (63.9%).

2-The majority of hydrocephalus was obstructive (non-communicating) (65.6%) while non-obstructive type (communicating) was about (16.2%).

3-Hydrocephalus occurred frequently during the neonatal to late infancy that is below 2 years.

4- In general hydrocephalus is slightly more in male than female 1.3:1

5- The incidence of aqueduct stenosis was (63.9%) out of congenital hydrocephalus and (16.6%)out of acquired cases.

6-The second most common cause of congenital hydrocephaly was Arnold Chiari malformation which was (14.3%).

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