The Egyptian Journal of Hospital Medicine (April 2018) Vol. 71 (3), Page 2651-2655

Congenital Hydrocephalus in Arar, Northern Saudi Arabia

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ABSTRACT

Background: Congenital hydrocephalus, an important cause of neurologic morbidity and mortality in children, is a medical condition characterized by an abnormal accumulation of cerebrospinal fluid in the brain. **Objectives:** To estimate the prevalence and the recent progresses in diagnosis of hydrocephalus as well as the changes in epidemiology and treatment outcomes of the disease. **Methods:** This is a descriptive study involved all neonates born in Maternity and child hospital in Arar city, Northern Saudi Arabia, KSA, during the period from 1 January to 31 December 2017. Among 6000 delivered infant in 2017; there was 23 cases of hydrocephalus. Data was collected by using predesigned questionnaire which include questions designed to fulfill the study objectives. **Results:** the prevalence of hydrocephalus among studied infants was 0.38%. Consanguinity between parents was reported among 60.9% of the cases. Other congenital anomalies reported, cardiac anomalies 39.1%, spina bifida 17.4% and cleft palate 8.7%. Cause of hydrocephalus was 65.2% hereditary and 34.8% infection (secondary cause). Swelling of the eyelid, increased vascular clarity on the skull, transparent skin in the head, the child's tendency to sleep, disorientation, irritability and nervousness, high crying, weakness of feeding and vomiting, delayed child skills, convulsions, increased size of the head, the sun's sign in the eye and the child is always looking down were the symptoms reported by the cases. 56.5% of the cases had shunt insertion and 43.4% had ordinary medical treatment. 30.4% of the shunt cases were complicated (17.4% had shunt blockage with infection and fever, 13% shunt blockage and 69.6% had other complications). Outcome of the case; 39.1% were stable, 13% worsen and 47.8% died (34.7% died by complications, 8.7% during operation and 4.3% did not receive treatment). **Conclusion:** The prevalence of hydrocephalus is comparable to that of other countries. More research is recommended because it is probable that the real prevalence of congenital hydrocephalus was underestimated and no account of how often abortions are performed among mothers of hydrocephalic fetuses. So more efforts from the ministry of health is needed.

Keywords: Hydrocephalus; prevalence; outcome; Arar; Northern Saudi Arabia.

INTRODUCTION

Hydrocephalus is broadly defined as a disturbance of formation, flow, or absorption of CSF that often is detected when it becomes symptomatic as a result of increased intracranial pressure [1]. Morbidity and mortality rates vary internationally but in general rising throughout the world, the highest incidence rates are found in Africa and Asia. In infants, hydrocephalus without an obvious extrinsic cause is usually referred to as congenital hydrocephalus, since it is often present at birth. When hydrocephalus occurs as a complication of another condition such as hemorrhage, infection or neoplasm, it is usually called acquired or secondary hydrocephalus [2]. Prevalence estimates for infantile hydrocephalus vary between one and 32 per 10,000 births, depending on the definition used and the population studied [3]. Management was described in (1923) by the use of endoscopic perforation of the floor of the third ventricle, endoscopic third ventriculostomy (ETV), but it was not a success because of technical problems and complications and for many decades ETV was not used [4]. Around (1960) the use of ventricular shunts became more common, as materials and techniques had improved, the use of modern shunts resulted in a reduction in mortality from 50% to about 10%during the last decade [5].

A previous study conducted in Sohag governorate, Egypt [6] reported that; incidence of hydrocephalus was 6.76/1000 living births. Consanguinity was present in 58% , family history was detected in 20.8%, drugs intake in 63.8%, fathers above 50 years at time of conception was detected in 30.4% of them (85.28%) were nonprofessionals, no antenatal care (51.7%), polyhydramnios (7.5%), oligohydramnios(7.1%), early vaginal bleeding (14.2%), preeclampsia (7.5%), twins was recorded (7.9%), breech presentation in (12.9%). Congenital hydrocephalus was treated by shunt

Received:12 / 1 /2018 DOI: **10.12816/0045824**
Accepted: 22/ 1 /2018

operation (85%), and prognosis was good in 70%.

The prevalence of CH was 0.81 per thousand. Diagnosis was performed prenatally in 41 cases. Forty-three (44.8%) of the cases had hydrocephalus without other malformations (isolated hydrocephalus), 18 (18.7%) infants had recognized chromosomal or non-chromosomal syndromes and 35 children (36.4%) had multiple malformations [7].

The aim of this study is to estimate the prevalence and the recent progresses in diagnosis of hydrocephalus as well as the changes in epidemiology and treatment outcomes of the disease.

PARTICIPANTS AND METHODS

Study design, setting, period and target population

This is a descriptive study involved all neonates born in Maternity and child hospital in Arar city, Northern Saudi Arabia, KSA, during the period from 1 January to 31 December 2017.

Data collection

Among 6000 delivered infant in 2017; there was 23 cases of hydrocephalus. Data was collected by using pre designed questionnaire which include questions designed to fulfill the study objectives.

- Socio-demographic characteristics of child including sex, mother and father educational level.
- Consanguinity between parents, other congenital anomalies like Cleft palate, Cardiac anomalies or Spina bifida.
- The questionnaire included also questions about cause of hydrocephalus (infection/ hereditary) and time of diagnosis (before or after delivery)
- Questions about reported symptoms, received management, outcome of the cases and cause of death in died cases.

Statistical analysis

Data were compiled and analyzed using statistical package for the social sciences (SPSS, version 16), results were analyzed with frequencies, and Chi-squared test as appropriate confidence level and p value were set at 95% and 5%, respectively.

Ethical considerations

Permission to conduct the study was obtained from the Research and Ethics Committee at the College of Medicine, Northern Border University, Arar, Saudi Arabia. The questionnaire had a brief introduction explaining the aims and significance of the study to parents. Parents of the neonates were informed about the study objectives. The participation was completely voluntary no name was recorded on the questionnaires.

RESULTS

Table 1: illustrate socio-demographic characteristics of hydrocephalus cases. 60.9% were males. 47.8% of mothers were highly educated (university or more), 17.4% secondary educated, 17.4% preparatory, 8.7% primary and 8.7% were illiterate. 39.1% of fathers were highly educated, 30.4% secondary educated, 13% preparatory, 8.7% primary and 8.7% were illiterate.

Figure 1: shows the prevalence of hydrocephalus among studied infants. 0.38% (23 infant) had hydrocephalus.

Table 2: illustrate consanguinity between parents, other congenital anomalies, reported causes of hydrocephalus and time of diagnosis of hydrocephalus. Consanguinity between parents was reported among 60.9% of cases. Congenital anomalies were reported by 65.2%, Spina bifida by 17.4%, cardiac anomalies 39.1% and cleft palate by 8.7%. Regarding cause of hydrocephalus; 65.2% hereditary causes and 34.8% infection (secondary cause).

Table 3: shows reported symptoms, outcome of the cases, cause of death, of HC cases.

Swelling of the eyelid, increased vascular clarity on the skull, transparent skin in the head, the child's tendency to sleep and his disorientation to the environment around the patient was reported by 30.4% of the cases, irritability and nervousness, high crying, weakness of feeding and vomiting, delayed child skills reported by 39.1% and convulsions, increased size of the head, the sun's sign in the eye: the child is always looking down were reported by 30.4% of the cases.

Regarding outcome of the case; 39.1% of the cases were stable, 13% worsen and 47.8% died (34.7% died by complications, 8.7% during operation and 4.3% did not receive treatment).

Table 4: illustrate treatment characteristics of HC cases. 56.5% of the cases had shunt insertion and 43.4% had ordinary medical treatment. 30.4% of the shunt cases were complicated (17.4% had shunt blockage with infection and fever, 13% shunt blockage and 69.6% had other complications).

Table (1): socio-demographic characteristics of hydrocephalus cases, Arar, KSA

Sex	Frequency (n=23)	Percent
Male	14	60.9
Female	9	39.1
Mother education		
Illiterate	2	8.7
Primary	2	8.7
Preparatory	4	17.4
Secondary	4	17.4
University or more	11	47.8
Father education		
Illiterate	2	8.7
Primary	2	8.7
Preparatory	3	13.0
Secondary	7	30.4
University or more	9	39.1

Figure (1): Prevalence of hydrocephalus among infants in Arar, Northern Saudi Arabia, 2017

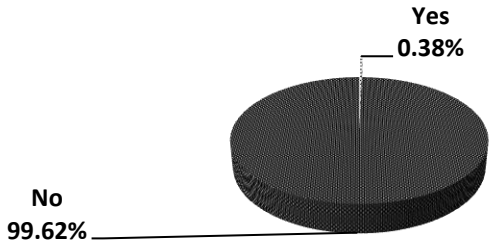

Table (2): Consanguinity between parents, other congenital anomalies, reported causes of hydrocephalus and time of diagnosis of hydrocephalus cases, Arar, KSA

Variable	No.	%
Consanguinity between parents		
Yes	14	60.9
No	9	39.1
Other congenital anomalies		
Yes	15	65.2
No	8	34.8
Spina bifida		
Yes	4	17.4
No	19	82.6
Cardiac anomalies		
Yes	9	39.1
No	14	60.9
Cleft palate		
Yes	2	8.7
No	21	91.3
Cause of hydrocephalus		
Infections	8	34.8
Hereditary causes	15	65.2
Time of diagnosis		
After birth	12	52.2
During pregnancy	11	47.8

Table (3): Reported symptoms, outcome of the cases, cause of death, of HC cases, Arar, KSA

Reported symptoms		
• Swelling of the eyelid, increased vascular clarity on the skull, transparent skin in the head, the child's tendency to sleep and his disorientation to the environment around him	7	30.4
• irritability and nervousness, high crying, weakness of feeding and vomiting, delayed child skills	9	39.1
• convulsions, increased size of the head, the sun's sign in the eye: the child is always looking down		30.4
Outcome of the case		
• Worsen	3	13.0
• Stable	9	39.1
• Died	11	47.8
Cause of death		
• During Operation	2	8.7
• Complications	8	34.7
• Not receiving treatment	1	4.3

Table (4): treatment characteristics of HC cases in Arar, KSA

		No.	**%**
Type of treatment	Medical treatment	10	43.4
	Shunt insertion	13	56.5
Complications of shunt	Yes	7	30.4
	No	6	26.1
Types of complications	Shunt blockage	3	13.0
	Shunt blockage with infection and fever	4	17.4

DISCUSSION

Hydrocephalus is a disorder in which an excessive amount of cerebrospinal fluid (CSF) accumulates within the cerebral ventricles and/or subarachnoid spaces, resulting in ventricular dilation and increased intracranial pressure (ICP) [8, 9].

This prospective study involves all neonates 6000 born in Arar city 2017, Northern Saudi Arabia, KSA. Our study reported that Hydrocephalus affected 23 infant from among 6000 registered total live births giving an overall prevalence of (0.38%), this percent was higher than a study conducted in southern Saudi Arabia (0.071%) [10]. Another study was conducted in Jordon a prevalence of congenital hydrocephalus was lower than our prevalence at 0.092% during 2004-2005 and 2008-2011 [11]. In Europe, a study was conducted in France and also showed prevalence which was lower than our study (0.081%) [12]. Also in Sweden a study was conducted and showed (0.82/1,000 live births in 1989–1998 versus 0.66/1,000 in 1999–2002 mainly resulting from a decreased incidence of Myelomeningocele [13, 14]). A similar trend was noticed in England and Wales [15] and in Japan [16]. On the other hand, other studies report the absence of significant changes in the incidence of congenital hydrocephalus with current figures ranging around 1/1,000 lives and stillborn (range, 0.2–3.5/1,000) according to the different countries [17, 18, 19, 20, 21].

Our study reported that two infants had cleft palate by a percentage of 8.7% which was higher than a study conducted in southern Saudi Arabia in which one infant was affected by a percentage of 1.6% [10]. A study was conducted in France, reported that 10 infants had cleft palate by a percentage of 28.6%, which was higher than our percentage [12]. Our study reported that 9 infant had cardiac anomalies by a percentage of 39.1% which was lower than a percentage in the French study (28.6%) [12].

Regarding treatment; our study reported that two types of treatment were used, medical treatment (43.4%) and shunt insertion (56.5%), in another study reported that 80.0% were shunt responders [23], also another study reported that 60 received control shunt systems [24]. Seven patients (30.4%) used complicated shunt. Our study showed that 17.4% were shunt blockage with infection and fever, in a study was conducted, reported that thirteen shunt infections were recorded [24], also another study reported that 25 (12%) experienced shunt infection [25], a study was conducted in Europe reported infection in 46 patients (8.2%) [26].

Our study reported that worsen cases were 13.0%, stable cases were 39.1% and died cases were 47.8%. In a study conducted in Sweden reported that cases extremely and very preterm were 37 cases, on the other hand, cases moderately preterm were 33. Seventeen of the 262 young children (6%) born in

1989-2002 died during their first years of life, ten with infantile hydrocephalus and seven with MMC which was lower than our died cases [22]. In another study reported that extremely preterm were 7 infants, very preterm were 6 infants and moderately preterm were 4 infants, six of the 54 infants, four by IH and two with MMC died with a percentage of 11% which was also lower than our study [13].

CONCLUSION AND RECOMMENDATIONS

The prevalence of congenital hydrocephalus is comparable to that of other countries. More research is recommended because it is probable that the real prevalence of congenital hydrocephalus was underestimated and no account of how often abortions are performed among mothers of hydrocephalic fetuses. So more efforts from the ministry of health is needed.

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