INFANTILE HYDROCEPHALUS IN SOUTHERN SAUDI ARABIA

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Objective: To study the prevalence and causes of infantile hydrocephalus. Methodology: Retrospective study of cases of infantile hydrocephalus comparing results with regional and international trends.

Results: Infantile hydrocephalus (IH) affected 62 infants from among 87,127 registered total live births giving an overall incidence of 0.71/1000. Eighteen cases (29%) with spinal dysraphism, 15 cases (24%) with aqueductal stenosis, 9 (14.5%) post meningsitis, 6 (9.7%) post haemorrhagic, 6 (9.7%) with structural CNS anomalies (holoprosencephaly 2, hemispheric cysts 2, brain dysgenesis 1, and vascular anomaly 1), 3 (4.9%) congenital idiopathic, 2 (3.2%) Dandy-Walker malformation, 2 (3.2%) toxoplasmosis and one case (1.6%) achondroplasia. Prenatal factors accounted for 46 cases (74.2%) of this series, while postnatal factors accounted for 16 cases (25.8%). Of the latter group, 9 (14.5%) were due to meningitis while 6 (9.7%) were post haemorrhagic.

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Of the postnatal group there were 8 preterm babies (4.7% of the total series and 53.3% of the postnatal group).

**Conclusion:** While the incidence of infantile hydrocephalus in this region remains statistically unchanged, new characteristics have emerged. It is interesting to notice the increased number of premature babies in the postnatal group. More associated CNS malformations have been noticed in the prenatal group. Still prenatal causes form the bulk of all cases.

**Key Words:** Infantile hydrocephalus, Southern Saudi Arabia.

**INTRODUCTION**

This paper explains the scarcity of information on the prevalence and incidence of infantile hydrocephalus in this part of the world and compares the different emerging trends (e.g. prevalence, causes, associated defects, etc) to international trends.

There has been a tremendous improvement in the social life, health care and other services in this region. This has resulted in improved maternal and childhood health but not without expected changing pattern of diseases. Hence the importance of studying the many factors that might significantly affect the well-being of children in this region. Infantile hydrocephalus (IH) remains an important problem as its causes both antenatal and postnatal are multi-fac-torial and always requires expensive management. The outcome is invariably unpredictable since a large proportion of the affected children end up with significant neurological and intellectual handicap. This places a heavy burden on the family and the community at large. In the west, the impact of many congenital diseases has been significantly reduced with the improvement in socioeconomic conditions and antenatal screening.

**MATERIAL AND METHODS**

Asir Central Hospital (ACH) is the only unit in this region that deals with management of hydrocephalus. All cases of IH are referred to this hospital for the necessary management. The case notes of all infants admitted with IH during the period 1/1/1990 to 31/12/1995 were analyzed. Chi square test was used for statistical analysis. IH is defined as significant ventricular dilatation with increased CSF pressure in the first year of life.

**RESULTS**

During the study period the total registered live birth in the region was 87,127. Sixty-two cases of IH were seen making a total incidence of 0.71/1000 births. The overall female to male ratio was 0.8:1.

**Prenatal causes (Table 1)**

There were 46 cases (74.2%) in this group. Spinal dysraphism constituted 18 cases (29%) (14 cases of myelomeningocele and 4 cases of encephalocoele). The female to male ratio was 1:0.6. The other associated malformation of this group were: dysmorphic facies in 3 cases of myelomeningocele and hydronephrosis in one case of encephalocoele (Meckel-Gruber’s syndrome (Table 2). Fifteen cases (24.2%) had aqueductal stenosis. The female to male ratio was 1:2. One case of this group had pulmonary stenosis with atrial septal defect while another case had tracheoesophageal fistula. Three cases (4.9%) were idiopathic. Two cases (3.2%) were due to Dandy-Walker malformation while another 2 cases (3.2%) were due to congenital toxoplasmosis. Six
other cases were associated with other CNS structural anomalies: 2 cases with holoprosencephaly, 2 cases with hemispheric cysts (one of them had agenesis of the corpus collosum), one case with brain dysgenesis and another with vascular anomalies.

**Postnatal factors**

There were 16 cases in this group representing 25.8% of the whole series. Nine cases (14.5%) were postmeningitic, 6 cases (9.7%) were posthaemorrhagic and one case (1.6%) was secondary to achondroplasia. Of all the postnatal factors there were 8 preterm babies (4.7% of the whole series and 53.3% of the postnatal group). Four of them were postmeningitic while the other four were posthaemorrhagic.

**Associated malformations**

The associated malformations detected by initial assessment were as follows: (1) dysmorphic facies - three myelomeningocele cases were associated with dysmorphic facies; (2) renal anomalies - one case of encephalocele had hydronephrosis (Meckel-Grubers syndrome) and another case of encephalocele had hypospadia; (3) Cardio-respiratory defects - the combination of tracheo-esophageal fistula, pulmonary stenosis, and atrial septal defect was noted in one case of acueductal stenosis. One case of Dandy-Walker malformations had the combination of patent ductus arteriosus (PDA) and coarctation of the aorta; (4) eye defects - one case of Dandy-Walker malformation had bilateral anophthalmia, one case of the idiopathic group had micro-ophthalmia; (5) other associated brain anomalies - the cases here had hydrocephalus (with no obvious cause) beside other brain anomalies, two cases had associated holoprosencephaly, one case was associated with brain dysgenesis and one case had agenesis of the corpus collosum.

**DISCUSSION**

The drop in the overall incidence of IH from 0.81/1000 in an earlier study to 0.71/1000 in our present study is not significant (P>0.5) meaning that it still remains an important medical problem. The predominance of prenatal factors over postnatal factors is still very noticeable. Nearly three quarters of all cases are due to prenatal factors. In this group, the contribution by spinal dysraphism has fallen from 39.3% to 29.0%. However, it is still the most prevalent cause of prenatal (congenital) hydrocephalus. This study indicated that the contribution of acueductal stenosis had increased from 16.4 to 24.2% and that primary CNS structural anomalies had increased. These have been associated with defects in other systems as well. It is hoped the incidence of neural tube abnormalities in this region will follow the international trend and decrease further. Negoro, et al in 1994 found the incidence of IH in Japan to be 0.58/1000 in live births with significant contribution by spinal dysraphism. It is interesting to notice the drop in incidence of IH (which implies a significant drop in the incidence of spinal dysraphism). In the West, antenatal screening coupled with selective termination have much reduced antenatal hydrocephalus. With the great improvement of socioeconomic conditions and the care of expectant mothers, it is expected that there would be a further reduction in the incidence of this problem. The administration of Periconceptional folic acid should reduce this incidence still farther.

We have observed that the high rate of consanguinity in this region accounts for the overall high rate of congenital malformation. Consequently, the high incidence of these primary CNS structural anomalies among the prenatal factors. It is impossible for all these defects to amplify the complexity of this disabling condition. The lack
Table 1: Infantine hydrocephalus: break down of cases

|                          | Number | %    | Females | Males | Female: Male ratio |
|--------------------------|--------|------|---------|-------|-------------------|
| **Prenatal**             |        |      |         |       |                   |
| Spinal dysraphism        | 18     | 29.0 | 11      | 7     | 1.6:1             |
| Acqueductal stenosis     | 15     | 24.2 | 5       | 10    | 0.5:1             |
| Congenital idiopathic    | 3      | 4.9  | 1       | 2     | 0.5:1             |
| Dandy-Walker malformation| 2      | 3.2  | 1       | 1     | 1:1               |
| Toxoplasmosis            | 2      | 3.2  | 1       | 1     | 1:1               |
| Associated with other    | 6      | 9.7  | 2       | 4     | 0.5:1             |
| structural CNS anomalies*|        |      |         |       |                   |
| **Postnatal**            | 16     | 25.8 | 7       | 9     |                   |
| Post menigitis           | 9      | 14.5 | 5       | 4     | 1.3:1             |
| Post haemorrhagic        | 6      | 9.7  | 2       | 4     | 0.5:1             |
| Achondroplasia           | 1      | 1.6  | 0       | 1     |                   |
| **Total**                | 62     | 100  | 28      | 34    | 0.8:1             |

*Holoprosencephaly 2, hemispheric cyst 2 (one with agenesis of copus colossum), brain dysgenesis 1, and vascular anomaly 1

Table 2: Associated non CNS malformations

| Malformation                        | No. of cases | Associated type of hydrocephalus |
|-------------------------------------|--------------|---------------------------------|
| Dysmorphic facies                   | 3            | Myelomeningocoele               |
| Hydrocephrosis                      | 1            | Encephalocele                    |
| Hypospadius                         | 1            | Idiopathic                       |
| Coarctation of aorta/patent ductus arteriosus | 1           | Dandy-Walker malformation        |
| Pulmonary stenosis/atrial septal defect | 1            | Acqueductal stenosis             |
| Tracheoesophageal fistula            | 1            | Acqueductal stenosis             |
| Cleft lip and palate                | 1            | Toxoplasmosis                    |
| Anophthalmia                        | 1            | Dandy-Walker malformation        |
| Microphthalmia                      | 1            | Idiopathic                       |
| Coloboma of eyes with hyperteleorism| 1            | Toxoplasmosis                    |
| Micognathia                         | 1            | Idiopathic                       |

of a proper registration system make it difficult to ascertain the family history of these defects.

In the postnatal group, we encountered more premature babies, (as neonatal units in the region are handling more premature babies with improving survival rates). However, the proportion of this group of infants does not reach that encountered in the West.7-10 In our study, intracranial hemorrhage affected 50% of preterm babies with IH. In Japan, 30% of the hydrocephalus in preterm babies was due to intracranial hemorrhage. This is expected to increase further with the improvement in the care of the preterm in this region.5 The most recent Swedish studies showed even higher contribution of intracranial hemorrhage in the preterm hydrocephalic population (89% of very preterm suffered confirmed intraventricular hemorrhage).11 Septic meningitis remains the most important causative factor in the postnatal group. Early detection with prompt medical treatment should greatly reduce its impact both in the neonatal period...
and thereafter. The administration of Hib vaccine should significantly reduce the incidence of meningitis in infancy.  

More males were affected in this series (female to male ratio 0.8:1). This is due to predominance of males in the aqueductal stenosis (which could be sex linked in some cases), posthaemorrhagic, and the structural CNS anomalies groups.

Among childhood neurodevelopmental disorders IH causes significant disability and needs continuous and expensive medical care 2,3,9 (shunting procedures, etc). Therefore, we should continue to observe its prevalence, causative factors and associated defects in order to implement effective preventive measures. Also a proper nationwide register for all congenital malformations should be instituted.

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