The Pattern of Central Nervous Disease in Children in King Khalid University Hospital in Riyadh, Saudi Arabia

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Summary

Two hundred and sixty children with CNS disease in the age groups 3 months to 15 years were studied. There were 133 males (51 per cent) and 127 females (49 per cent). The aetiological factors studied showed prenatal origin in 28 per cent, perinatal in 13 per cent, post-natal in 32 per cent and mixed causes in 4 per cent. The aetiology could not be defined in the remainder. The pattern of disease was studied and the following were the presenting diagnostic problems:

Convulsive disorders 125 patients (48 per cent). There were 68 (26 per cent) patients with febrile convulsions and 53 (20 per cent) patients with epilepsy and 4 (2 per cent) with various metabolic causes. The cerebral palsies were seen in 53 (20 per cent) patients and these included the usual known types, but there was a higher incidence in females with a female:male ratio of 19:33. All the other diagnostic groups showed a higher incidence in males.

Head injuries were seen in 29 (11 per cent) patients. The types of injuries are not discussed.

Congenital malformations excluding hydrocephalus were present in eight patients (3 per cent).

Hydrocephalus was seen in 10 patients (4 per cent).

The 'other' group, 36 patients in total (14 per cent) constituted heredodegenerative disorders, microcephaly, tumours, rheumatic chorea, spinal cord injuries, and some miscellaneous problems.

Introduction

The literature available on CNS disease of children in Saudi Arabia is limited and, so far, not much work has been done. A lot has been said about birth hazards, home deliveries, and inherited disorders. The most common types of disorders still have to be outlined if any success is to be achieved through prevention.

This paper describes the nature of these disorders. The aetiology, diagnostic categories, and sex incidence are studied and it is thought that although it is a small sample, the King Khalid University Hospital being the teaching hospital of King Saud University in Riyadh, drains a wide area of the city as well as the peripheral areas. It is also a referral centre for other hospitals for specialist consultation clinics and therefore, the sample can be quite representative on a small scale.

The main diagnostic categories of febrile convulsions, epilepsy, and cerebral palsy are studied in detail. On reviewing the literature, accepted and recent definitions are used.[1,2]

Materials and Methods

The medical records of 260 children aged 3 months to 15 years referred for consultation and/or admitted to the King Khalid University Hospital, King Saud University, Riyadh with neurological or neuromuscular disorders were reviewed and analysed. These children were seen over a 15-month period from August 1982 to November 1983.

Data collected for age, sex, diagnosis, and aetiological factors, including, when available, the birth histories of these children.

The laboratory diagnostic facilities and the radiological procedures available at King Khalid University Hospital are of international standards and almost all investigations are possible. Our search did not require additional investigations other than the

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TABLE 2
Distribution of cases with hydrocephalus

Type	No.	%
Congenital hydrocephalus	3	30
Post-menigitic hydrocephalus	3	30
Hydrocephalus with cerebral palsy	4	40
Tumour	1	10
Total	10	100

(0.76 per cent), tuberous sclerosis one patient (0.38 per cent).

Brain tumours were diagnosed in four children (2 per cent) and consisted of one case of pituitary adenoma, one of haematoma, one optic nerve glioma, and one craniopharyngioma (Table 1). Intra-uterine infections had affected two children (1 per cent), (congenital syphilis and congenital toxoplasmosis).

Congenital malformations were diagnosed in eight patients (3 per cent). Six of these were spina bifida with (myelomenigocoele) (2 per cent). Two were cases of Sturge Weber Syndrome.

The miscellaneous group comprised 11 patients (4 per cent) as follows: rheumatic chorea in four (2 per cent), spinal cord injury in two patients (1 per cent), intra-uterine infections in two (1 per cent), breath-holding attacks in one, cyclical vomiting in 2 (1 per cent), Werdnig–Hoffman in two (1 per cent), headache in one. The types of epilepsy were grand mal epilepsy as the most common type, (26 patients) infantile spasm in eight, myoclonic epilepsy in two, minor motor seizures in two, nocturnal epilepsy in two, temporal lobe epilepsy in three, and unclassified epilepsy in five (Table 3).

The sex incidence in the convulsive disorders group was 57 per cent males to 43 per cent females (Table 4). Among the febrile convulsions, there were 51 per cent males to 49 per cent females. In the epilepsy group, there were 66 per cent males to 34 per cent females. In the cerebral palsy group, spastic quadriplegia, diplegia and hemiplegia were the most common types in

TABLE 3
Distribution of various types of epilepsy

Type	No.	%
Grand mal	26	49.5
Infantile spasms	8	15.09
Focal epilepsy	5	9.43
Temporal lobe epilepsy	3	5.66
Myoclonic epilepsy	2	3.77
Minor motor seizures	2	3.77
Nocturnal epilepsy	2	3.77
Unclassified	5	9.43
Total	53	100

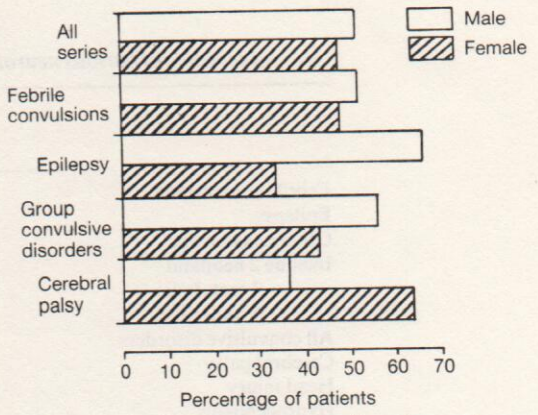

FIG. 2. Sex incidence–convulsive disorders and cerebral palsy.

that order and constituted more than half the patients. Diplegia was present in 10 (19 per cent) (Table 5). Almost one-third of the children with cerebral palsy had an abnormal birth history. Other types of cerebral palsy included less frequently paraplegia in five (10 per cent), athetosis in two (4 per cent), hypotonia in six (12 per cent). Spastic, but type not specified in four (8 per cent), type not specified at all in six (12 per cent). Of the total 52 cerebral palsy children, there were 19 males and 33 females, a sex incidence of 37 and 63 per cent, respectively. The sex incidence in the group of head injuries was 17 males to 12 females, 59 and 41 per cent, respectively (Table 4).

Microcephaly was seen in six patients (2 per cent), three of them had associated blindness and two were part of the cerebral palsy syndrome. The associated factors in those presenting with fever and convulsion were; prenatal in 25 (37 per cent), perinatal in 12 (18 per cent), postnatal in 10 (15 per cent), while mixed factors were seen in five patients (7 per cent). Unknown factors were seen in 16 (24 per cent) (Table 6, Fig. 3).

In the epilepsy group, aetiological factors were prenatal in 19 (36 per cent), perinatal factors in 10 (19 per cent) and postnatal factors in five (9 per cent). There were 19 cases (36 per cent) where the aetiology was unknown.

In the cerebral palsy group, the prenatal factors were 3 (6 per cent), birth hazards 11 (21 per cent), postnatal factors in 18 (35 per cent) and in five patients (10 per cent). There was more than one aetiological factor as the cause; miscellaneous in eight (15 per cent) and in 18 the cause was unknown (35 per cent).

Table 7 summarizes results from various studies on the aetiology of cerebral palsy.

Discussion

The population of Saudi Arabia in the last census of

TABLE 1
Incidence of various neurological disorders diagnostic groups

			Males		Females	
	No	%	No	%	No	%
Febrile convulsions	68	26.15	35	51.47	33	48.5
Epilepsy	53	20.38	35	66.03	18	33.9
Other convulsions	4	1.53	1		3	
Include 2 neonatal						
1 metabolic						
1 undecided						
All convulsive disorders	125	48.07	71		54	
Cerebral palsy	52	20.00	19	36.53	33	63.4
Head injury	29	11.15	17	58.62	12	41.3
Hydrocephalus	10	3.85	7	70.00	3	30.0
Microcephaly	6	2.3	2		4	
Heredo-degenerative disorders	11	4.23				
Include SSPE	7	2.69	4		3	
*Neurofibromatosis	2	0.76			2	
*Tuberous sclerosis	1	0.38	1			
*Freidreich's ataxia	1	0.38			1	
Congenital malformations	8	3.07				
*Myelomeningocoele	6	2.30	2		4	
*Sturge Weber Syndrome	2	0.76			2	
Tumours	4	1.53	2		2	
Rheumatic chorea	4	1.53	2		2	
Spinal cord injury	2	0.76	1		1	
Intra-uterine infections	2	0.76				
*Congenital syphilis	1	0.38			1	
*Congenital toxoplasmosis	1	0.38	1			
Miscellaneous group	7	2.69				
*Breath-holding attack	1	0.38			1	
*Cyclical vomiting	1	0.38			1	
*Sub-dural effusion	1	0.38	1			
*Developmental aphasia	1	0.38	1			
Werding Hoffmann	2	0.76	1		1	
*Headache	1	0.38	1			
Total	260		133	51.5	127	48.84

ones the patients had while in the hospital ward or the pediatric consultation clinics.

The various data are analysed and conclusions are made.

Results and Conclusions

The incidence of different neurological diseases is presented in Table 1 (Fig. 1). The first group is the 'convulsive disorders' consisting of 125 patients (48 per cent). There were 68 patients (26 per cent) with febrile convulsions, 53 patients (20 per cent) with epilepsy, and four children with metabolic and neonatal convulsions (2 per cent).

The second diagnostic category was the 'cerebral palsy' group consisting of 52 patients (20 per cent).

The group of 'head injuries' comprised 29 patients (11 per cent). Hydrocephalus was present in 10 patients (4 per cent), while microcephaly was seen in six patients (2 per cent).

In the hydrocephalus group, the majority (six) were

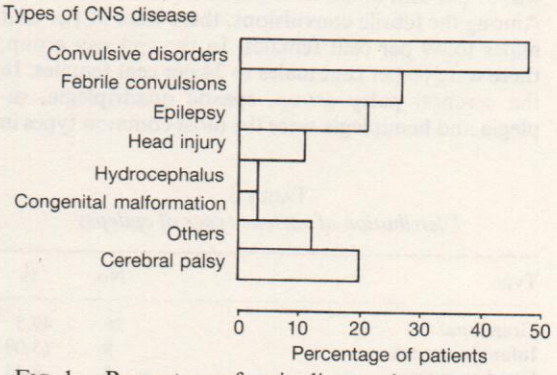

FIG. 1. Percentage of main diagnostic categories.

congenital in origin (Table 2). Heredo-degenerative disorders were seen in 11 patients (4 per cent). These included subacute sclerosing panencephalitis in seven patients (3 per cent), neurofibromatous two patients

TABLE 6
Number and percentage of aetiological factors, certain diagnostic groups and all the series

	% all series	No.	% febrile conv. Group 1	No.	% EP Group 2	No.	% GP Group	No.
Prenatal	27.96	72	36.76	25	35.84	19	5.76	3
Perinatal	12.68	33	17.64	12	18.86	10	21.15	11
Postnatal	32.30	84	14.70	10	9.43	5	13.64	7
Mixed factors	3.88	10	7.35	5	—	—	9.61	5
Unknown	20.38	53	23.52	16	35.84	19	34.61	18
Unsure	3.07	8	—	—	—	—	15.38	8
Total	100	260	68	100	53	100	100	52

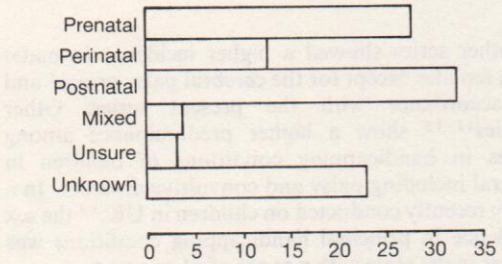

FIG. 3. Percentage of aetiological factors in CNS disease.

normal signs that in isolation were predictive, but clusters of abnormal signs correlated directly with outcome. These findings were in accordance with those of Nelson and Ellenberg (1981),[22] and in accordance with our own results where 13 per cent only of the children had perinatal causes.

The aetiological factors in 1503 patients with cerebral palsy lists prematurity as the most prominent factor[24] (28 per cent) whereas 39 per cent of the causes were prenatal in origin, 46 per cent natal and 15 per cent postnatal.

There has been a striking decrease in the incidence of erythroblastosis as a factor which dropped to nil in the 1970s. There have been lesser decreases in encephalitis, dystonia, and idiopathic factors.[25]

The first diagnostic group is that of 'convulsive disorders' (48 per cent), with febrile convulsions being 26 per cent and epilepsy 20 per cent. The second largest group is that of 'cerebral palsy', at 20 per cent. The third group is 'head injuries' at 11 per cent. Hydrocephalus accounted for 4 per cent and all the other groups together for 17 per cent. The types of handicapping conditions are similar to other studies in Saudi Arabia and those reported from other parts of the world. Figures in the series of 1983[5] included 42 per cent febrile convulsions, 21 per cent epilepsy, and 16 per cent cerebral palsy.

The number of patients among the epilepsy group is too small to consider the various types, and so does not offer a wide representation of the problem. The incidence of grand mal epilepsy in our series is, however, similar to the Bristol study[26] where half of the children with epilepsy had grand mal fits. Also the incidence of grand mal epilepsy is 50 per cent in both uncomplicated epilepsy and with structural brain disease in the study of Graham *et al.* (1970),[8] where focal epilepsy is represented as 5 and 13 per cent as infantile spasm.

The various types of cerebral palsy from various countries can be seen in Table 8.

In our series, there were no patients with the 'mixed' cerebral palsy, tremor or ataxia. The hypotonic group 12 per cent was quite marked whereas the spastic and athetoid types were not much different from other studies.

The types of head injury were not discussed as it was considered to be beyond the scope of this paper.

The group with Heredo-degenerative disorders (4 per cent) is interesting in that it included subacute sclerosing pan encephalitis (3 per cent), quite a high figure for this small series and neurocutaneous syndrome (1 per cent). If we were to include the two children with Sturge Weber Syndrome, the neurocutaneous syndrome will comprise 2 per cent, quite a representative figure for this small series. In comparison, out of 2.1 million patients seen over 25 years at the University College, Ibadan, Nigeria, only 25 suffered from heredo-degenerative disorders of the nervous system. These included hereditary ataxia, essential tremor, and ataxia telengiectasia.[27] It is also noted that neuromuscular diseases constitute only 1 per cent of the series and taking into consideration the degree of consanguinity in Saudi Arabia one could look for higher figures.

There is no significant difference in the major diagnostic categories from those reported in the literature.

The percentage of children with intracranial tumours at 2 per cent is high.

Recommendations

It is hoped that other colleagues specializing in this

TABLE 4
Sex incidence

	Males		Females		
	%	No.	%	No.	Total
All the series	51.15	133	48.84	127	260
Febrile convulsions	51.47	35	48.52	33	68
Epilepsy	66.03	35	33.96	18	53
All convulsive disorders	56.8	71	43.2	54	125
Cerebral palsy	36.53	19	63.46	33	52
Head injury	58.62	17	41.37	12	29
Hydrocephalus	70.00	7	30.00	3	10
Microcephaly		2		4	6
Spina bifida		2		4	6

TABLE 5
Distribution of different types of cerebral palsy

Type	No.	%
Spastic hemiplegia	10	19.23
Spastic paraplegia	5	9.61
Spastic diplegia	10	19.23
Spastic quadriplegia	11	21.15
Hypotonia	6	11.53
Athetosis	2	3.84
CP type not specified	6	11.53
Spastic, but not specified	4	7.96
Total	52	100

1974[3] was estimated at 7.9 million where about 52 per cent are children aged between 0 and 15 years.[4]

The age groups of the children studied included 3 months to 15 years, well within the paediatric scope.

No statistics in the literature on the frequency of handicaps of specific neurological problems are available in Saudi Arabia but over a one year period in King Khalid University Hospital, out of 891 admissions in 1982/83, there were 174 children admitted with a neurological, neuromuscular, or muscular problem and/or mental retardation due to various causes. This is equal to 20 per cent of the paediatric hospital population for that year.[5]

The young male is more susceptible than the young female to a wide variety of biological hazards. He is more vulnerable to all manner of infections in childhood[6,7] and more commonly has specific delays in development.[8] Herlitz and Redin (1975)[9] reported an incidence of 55 per cent in males in these cases of cerebral palsy. Graham *et al.* (1970)[8] also recorded a higher incidence of other brain disorders including epilepsy amongst males.[8] Our present study included 51.15 per cent males and 48.84 per cent females. This is in accordance with the study by Taha and Mahdi (1984).[10] The ratio between males and females among severely affected cerebral palsy children is 3:2. Another series showed a higher incidence in males than females except for the cerebral palsy group[5] and in accordance with the present series. Other studies[11–15] show a higher predominance among males in handicapping conditions of children in general including palsy and convulsive disorder. In a study recently conducted on children in UK,[16] the sex incidence in principal handicapping conditions was higher in the male with a range of 51 per cent.

In our present series, the incidence of hydrocephalus was 70 per cent in males, and microcephaly 40 per cent. This is in accordance with Bradshaw and Lawson (1985),[16] but the number of patients is too small to draw conclusions.

Prematurity and neonatal insults are predominant in the aetiology of cerebral palsy.[17] In infants where resuscitation took place within 15 minutes or more, 18 per cent of motor sequelae are reported.[18]

Another study[10] carried out on children in the same university, but different hospital on severely affected children with cerebral palsy showed the aetiological factors being 24 per cent prenatal, 48 per cent perinatal, 28 per cent postnatal, and 7 per cent mixed causes with 13 per cent unknown.

The Holm Study (1982)[19] showed prenatal factors to be 50 per cent, perinatal factors 38 per cent, postnatal 10 per cent and mixed 7 per cent. A relationship between perinatal risk, i.e. obstetric complications and cerebral palsy were documented more than 120 years ago by J. Little in 1961.[20] Much more recently, similar interesting findings are discussed by Nelson and Ellenberg in 1984.[21, 22] Among infants of normal birth weight (2500 g) the main finding was a relatively low risk for cerebral palsy regardless of obstetrics complications as long as the 5-minute Apgar score was more than 7. Similarly, the risk for cerebral palsy was not particularly high with a very low 5 minutes Apgar score of less than 3 unless one or more complications were present and the 5 minutes Apgar score was low.

Dubowitz *et al.* (1984)[23] found no individual ab-

TABLE 7
The summarized results from various studies

Author	Year	Prenatal	Perinatal	Postnatal	Mixed	Unknown
Taha and Mahdi, S. Arabia[10]	1984	23.5	48.0	28.4	7.0	13.0
Hilm V. E., USA	1982	50.0	33.0	10.0	7.0	—
Al Naquib, N., Iraq[15]	1981	24.5	27.0	15.5	2.5	30.5
O'Reilly and Walentynowicz, USA[24]	1981	38.5	46.4	15.2	—	—
Denhoff and Robinault, USA	1960	(10–40)	33.0	16.0	—	—
Present Series, S. Arabia		27.96	12.68	32.30	3.88	20.38

TABLE 8
Cerebral palsy by type, figures from various studies

Author	Percentage						
	Spastic	Athetoid	Mixed	Tremor	Ataxia	Hypotonia	Others type
Present Series 1985, S. Arabia	69.22	7.96	—	—	—	11.53	—
Taha and Mahdi 1984, S. Arabia	61.8	2.0	13.7	—	2.0	19.6	—
Al Naquib 1983, S. Arabia	78.57	3.57	—	—	7.14	10.71	—
Al Naquib 1981, Iraq	80.00	6.0	12.0	2.0	4.0	8.0	—
O'Reilly and Walentynowicz, 1981	—	3.0	—	—	—	3.4	—
Al Naquib 1975, UK	55.00	14.0	9.0	3.2	7.7	—	—
Illingworth 1974	72.04	15.6	1.57	2.99	9.18	—	—
O'Reilly 1971	59.3	13.9	12.1	0.3	4.6	—	—
Isle of Wight Study 1970	94.3	2.9 dyskinesia	—	—	2.9	—	—
Dundee Survey 1961, Ref. 41	80.00	8.4 dyskinesia	—	—	1.7	—	10
Edinburgh Survey 1955	83.7	8.2 dyskinesia	—	—	7.2	—	10
Hopkins, Bice and Colton 1954	45.9	23.7	—	—	—	—	—
Dunsdon 1952	81.8	10.4	—	—	—	—	—
Pohl 1950	66.00	19.0	—	—	—	—	—

field will become interested in similar work in order to throw more light on the epidemiology of neuro and neuromuscular problems of children in Saudi Arabia.

In addition to improving the care during the perinatal period, efforts towards prevention of prenatal incidents and postnatal factors would be worthwhile. The unknown factors are still represented in a high figure suggesting further scope for research.

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