Analysis of medical records of patients with "other neurological diseases" admitted to Lady Ridgeway Hospital

S S Disanayaka[1], H G N De Zoysa[1], S Senaratne[2], K R D de Silva[3]

Sri Lanka Journal of Child Health, 2008; **37**: 118-121

(Key words: Medical records; diseases of nerve and muscle; genetic diagnosis)

Abstract

Objective To assess the frequency of diseases categorized under "other neurological diseases" in the Indoor Morbidity and Mortality Registry (IMMR) of the Medical Record Unit of infants and children admitted to the Lady Ridgeway Hospital for Children (LRH) and coded and indexed according to the 10th revision of the International Classification of Diseases (ICD 10) of the World Health Organization.

Design Retrospective data analysis

Method Data of in-ward patients, obtained from the medical record section of LRH for a period of two years (2004 and 2005), were analysed

Results Two hundred and ninety nine (0.21%) [157 (52.5%) males and 142 (47.5%) females] had been categorized under "other neurological diseases". The four major contributors were hydrocephalus 59 (20%), polyneuropathy & disorders of peripheral nervous system 44 (15%), Guillain Barre' syndrome 39 (13%) and disorders of myoneural junction & muscle 29 (10%). A family history was present in a patient with Duchenne muscular dystrophy (DMD) and a patient with limb-girdle muscular dystrophy.

Introduction

Medical record keeping is of crucial importance in managing health care; it not only helps to improve the quality of medical care but also provides easy access to necessary information required in research and statistical surveys when planning health care systems. However, no database for the prevalence and the incidence of separate diseases of the nervous and muscular system of children is available in Sri Lanka. Of 49,092 discharges due to diseases of the nervous system during the year 2003 from hospitals throughout Sri Lanka, 26% were paediatric discharges (<1 year 3%, 1-4 years 7% and 5-16 years 16%)[1]. Lady Ridgeway Hospital for Children (LRH) is a tertiary referral national paediatric hospital in Sri Lanka with an annual admission of 73,554 and 69,399 of children and infants in 2004 and 2005 respectively[2].

Objective

The aim of this study is to assess the frequency of diseases categorized under "other neurological diseases" in the Indoor Morbidity and Mortality Registry (IMMR) of the Medical Record Unit of infants and children admitted to LRH and coded and indexed according to the 10th revision of the International Classification of Diseases (ICD 10) of the World Health Organization (WHO)[3]. "Other neurological diseases" comprise the following:

ICD number	*Disorder*
G10–G13	Systemic atrophies primarily affecting the central nervous system
G20–G26	Extrapyramidal and movement disorders
G30–G32	Other degenerative diseases of the nervous system
G60–G64	Polyneuropathies and other disorders of the peripheral nervous system
G70–G73	Diseases of myoneural junction and muscle
G80–G83	Cerebral palsy and other paralytic syndromes
G90–G99	Other disorders of the nervous system

[1]*Research Assistant, Department of Anatomy, Faculty of Medical Sciences, University of Sri Jayewardenepura, Nugegoda*, [2]*Deputy Director, Lady Ridgeway Hospital for Children, Colombo,* [3]*Senior Lecturer, Department of Anatomy, Faculty of Medical Sciences, University of Sri Jayewardenepura, Nugegoda..*

(Received on 25 March 2008. Accepted on 20 April 2008))

Method

Medical Record Unit receives the Bed Head Tickets (BHTs) after discharge of in-ward patients; thereafter diagnosis is indexed according to ICD 10. IMMR is prepared quarterly by the Medical Record Unit. Case sheets from the BHTs and IMMR of patients categorized under "other neurological diseases" were obtained from the medical record section of the LRH for a period of two years from 1st January 2004 to 31st December 2005. Retrospective data analysis was done. Multiple admissions of the same patients were excluded from records. Myopathies had been diagnosed by haematoxylin and eosin (H & E) staining only; specific diagnosis of each disorder by immunohistochemistry had not been done due to limited resources. Ethical clearance for the study was obtained from the Ethical Committee of LRH.

Results

A total of 299 (157 males and 142 females) patients covering all the provinces in Sri Lanka was obtained from a total of 29,990 BHTs categorized under "other neurological diseases" by the IMMR. The age distribution of the cohort was: 81 (27%) cases <1 year of age, 121 (40.5%) cases in the 1-4 year and 97 (32.4%) in the 5-15 year age groups. Demographic characteristics of these patients are shown in Table 1. A detailed report of neurological disorders at LRH in 2004 & 2005 are shown in table 2.

Table 1
Demographic characteristics of the cohort

	2004	2005	Total Number
Total admissions	73,554	69,399	142,953
Male	42,116 (57%)	39,139 (56%)	81,255 (57%)
Female	31,438 (43%)	30,260 (44%)	61,698 (43%)
"Other neurological disorders"	127 (0.17%)	172 (0.25%)	299 (0.21%)
Male	66 (52%)	91 (53%)	157 (52.5%)
Female	61 (48%)	81 (47%)	142 (47.5%)
Age groups			
<1 year	34 (26.8%)	47 (27.3%)	81 (27.1%)
1-4 years	54 (42.5%)	67 (39.0%)	121 (40.5%)
5-16 years	39 (30.7%)	58 (33.7%)	97 (32.4%)
"Other neurological disorders"			
G10–G13 Systemic atrophies primarily affecting the central nervous system	04 (3.1%)	08 (4.6%)	12 (4.0%)
G20–G26 Extrapyramidal and movement disorders	00 (0.0%)	05 (2.9%)	05 (1.7%)
G30–G32 Other degenerative diseases of the nervous system	10 (7.9%)	08 (4.6%)	18 (6.0%)
G60–G64 Polyneuropathies and other disorders of the peripheral nervous system	19 (15.0%)	25 (14.5%)	44 (14.7%)
G70–G73 Diseases of myoneural junction and muscle	11(8.7%)	18 (10.5%)	29 (9.7%)
G80–G83 Cerebral palsy and other paralytic syndromes	02(1.6%)	01 (0.6%)	03 (1.0%)
G90–G99 Other disorders of the nervous system	48(37.8%)	62 (36.0%)	110 (36.8%)
Others	33(26.0%)	45 (26.2%)	78 (26.1%)

The category *G90-G99 (Other diseases of the nervous system)* included the following: hydrocephalus 59 (54%), encephalopathy 26 (24%), cerebral oedema 09 (8%), intracranial hypertension 05 (4.5%), Reye's syndrome 06 (5.5%) and arachnoids cyst 03 (3%).

The category G70-G73 (Diseases of myoneural junction and muscle) included 8 males (5 Sinhalese, 2 Tamils, 1 Moor) with Duchenne muscular dystrophy (DMD), 2 Sinhalese males with Backer muscular dystrophy (BMD), 1 Sinhalese female with facio-scapulo-humeral dystrophy, 7 patients (M=3, F=4; Sinhalese 3, Tamils 4) with myopathies, 7 patients (M=4, F=3; Sinhalese 5, Tamils 2) with congenital myopathy and one male Moor patient with nemaline rod myopathy. Family history was present in a male Tamil patient with DMD and a male Moor patient with limb-girdle muscular dystrophy. 78 patients were categorized as "others" as the IMMR has grouped all these patients under *"other neurological diseases"* but we could not find a specific diagnosis in the BHT falling under the said ICD 10 classification

Table 2
Detailed report of neurological disorders at LRH in years 2004 & 2005

ICD 10 Classification	Disorder	2004	2005	Total
Systemic atrophies primarily affecting the central nervous system (G10 – G 13)	Hereditary spastic paraplegia	1	4	5
	Anterior horn cell disease	0	1	1
	Spinomuscular atrophy	3	3	6
	Sub Total	**4**	**8**	**12**
Extrapyramidal and movement disorders (G20 – G26)	Abnormal movements	0	1	1
	Benign familial chorea	0	1	1
	Myoclonus	0	1	1
	Myoclonus jerk	0	1	1
	Benign infantile myoclonus	0	1	1
	Sub Total	**0**	**5**	**5**
Other degenerative diseases of the nervous system (G30 – G32)	Cerebral atrophy	5	7	12
	Degenerative brain disease	1	0	1
	Neurodegenerative disease (grey matter)	1	1	2
	X linked spinocerebellar degeneration	3	0	3
	Sub Total	**10**	**8**	**18**
Polyneuropathies and other disorders of the peripheral nervous system (G 60 - G64)	Guillain Barre syndrome	19	20	39
	Sensory motor polyneuropathy	0	3	3
	Acute flaccid paralysis	0	1	1
	Demyalinating polyneuropathy	0	1	1
	Sub Total	**19**	**25**	**44**
Diseases of myoneural junction and muscle. (G70 – G73)	Myasthenia gravis	1	2	3
	Primary disorders of muscle			
	Muscular dystrophy	1	2	3
	Duchenne muscular dystrophy	2	6	8
	Limb girdle muscular dystrophy	1	1	2
	Facioscapulohumeral dystrophy	0	1	1
	Becker muscular dystrophy	1	1	2
	Congenital myopathy	5	2	7
	Other specified myopathies	0	0	0
	Unspecified myopathy	0	3	3
	Sub Total	**11**	**18**	**29**
Cerebral palsy and other paralytic syndromes (G80 - G83)	Cerebral palsy	2	1	3
	Sub Total	**2**	**1**	**3**
Other disorders of the nervous system (G90 - G99)	Hydrocephalus	25	34	59
	Other disorders of brain			
	Encephalopathy	13	13	26
	Reye syndrome	3	3	6
	Arachnoid cyst	2	1	3
	Cerebral oedema	3	6	9
	Intracranial hypertension	0	5	5
	Other disorders of central nervous system	2	0	2
	Sub Total	**48**	**62**	**110**
Others		**33**	**45**	**78**
	Final Total	**127**	**172**	**299**

Discussion

A retrospective data analysis of cases obtained from the medical record section of the hospital reveals that diseases categorized under *"other neurological diseases"* of the IMMR comprise less than 1% of total admissions of infants and children to LRH during the years 2004 and 2005 (Table 1). The four major

contributors were hydrocephalus 59 (20%), polyneuropathy & disorders of peripheral nervous system 44 (15%), Guillain Barre' syndrome 39 (13%) and disorders of myoneural junction & muscle 29 (10%). Whilst the majority of these patients were suffering from life threatening, life long, incurable and incapaciting diseases, hydrocephalus is a treatable condition where early diagnosis is essential for surgery to be of value[4].

Of the 29 patients recorded under "diseases of myoneural junction and muscle" (G70-G73) 8 were DMD and 2 were BMD. DMD and BMD are the most common form of hereditary muscular dystrophies, with an incidence of about 1/3,300 male births[5].

Sri Lanka is a multiethnic, mutireligious, multilinguistic country with cultural diversity comprising Sinhalese 73.9%, Sri Lankan Tamils 12.7%, Indian Tamils 5.5%, Moors 7%, Burghers 0.3%, Malays 0.3% and others 0.2%[6]. Family history was present in one (12.5%) Tamil patient with DMD and in one (50%) Moor patient with limb-girdle muscular dystrophy. A positive family history is a strong sign of a genetic disease though not all familial disorders are genetic and not all genetic disorders are familial[7]. DMD and BMD are X-linked recessive allelic disorders[8,9]. Limb-girdle muscular is usually sporadic[10] but could be inherited as an autosomal dominant or recessive trait[11]. Congenital muscular dystrophies are inherited as autosomal dominant or recessive traits[12].

The method used for the establishment of diagnosis of these patients was based on written diagnosis of the IMMR; under presentation of data is possible as patients diagnosed as having disorders of the nervous system could be possibly categorized under the index of a different chapter in ICD 10 depending on the incidental diagnosis of the presenting complaint. Confirmation of diagnosis either by immunohistochemical methods or genetic diagnosis of these cases had not been stated in the BHTs possibly due to unavailability of such service in the State sector. Establishing a genetic diagnosis confirms the inheritance pattern of the disorder and allows the risk of other family members to be determined[13].

Maintaining medical records adequately and accurately and establishing confirmation of diagnosis by immunohistochemical methods and genetic diagnosis by the state sector for patients with "other neurological diseases" will be an important step toward effective prevention: by detection of carriers, genetic counselling and antenatal diagnosis of affected fetuses.

Acknowledgments

We thank Prof. E. P. Hoffman, Director of Research Centre for Genetic Medicine Children's National Medical Centre Washington DC for his advice, Dr. C.S. Perera, Consultant Pathologist, Director, LRH and staff of the Medical Records Unit for their assistance.

References

1. Annual Health Bulletin, Ministry of Health Sri Lanka 2003.

2. Annual Bulletin, Lady Ridgeway Hospital 2005.

3. WHO. International Classification of Diseases 10th Revision (ICD 10). Available from: www.who.int/classifications/icd/en/

4. Weinman D. Hydrocephalus. *Ceylon Journal of Child Health* 1965; **3**:13-19.

5. Emery A E H. Duchenne and other X-linked muscular dystrophies. In: Rimoin D L, Connor J M, Pyeritz R E, Korf B R, editors. Emery and Rimoin's Principles and Practice of Medical Genetics, 4th ed. London: Harcourt Publishers Limited; 2002: 3266-84.

6. Census 2001 Sri Lanka. Available from www.rootsweb.com/~lkawgw/index4.html

7. Bird T D. Approaches to the patient with neurogenetic disease. *Neurologic Clinics of North America* 2002; **20**: 619-26.

8. Kingston H M, Thomas N S, Pearson P L, Sarfarazi M, Harper P S. Genetic linkage between Becker muscular dystrophy and a polymorphic DNA sequence on the short arm of the X chromosome.*J Med Genet* 1983; **20**:255-8.

9. Kingston H M, Sarfarazi M, Thomas N S, Harper P S. Localisation of the Becker muscular dystrophy the short arm of the X chromosome by linkage to cloned DNA sequences. *Human Genet* 1984; **67**:6-17.

10. Yamanouchi Y, Arikawa E, Arahata K, et al: Limb-girdle muscular dystrophy: clinical and pathologic re-evaluation. *J Neuro Sci* 1995; **129**:15.

11. Richard I, Broux O, Allamand V, et al: Mutations in the proteolitic enzyme calpain 3 cause limb-girdle muscular dystrophy type 2A. *Cell* 1995:**81**: 27.

12. Gilroy J. Basic Neurology. 3rd ed. USA: Macgraw Hill companies:; 2000..

13. Krajewski M K and Shy M E. Genetic testing in neuromuscular disease, *Neurologic Clinics* 2004; **22**: 481-508.