Nigerian Journal of Paediatrics 1996; 23 (1): 21 - 4

Childhood Neuro-ophthalmic Diseases

FB Akinsola*

Summary

Akinsola FB. Childhood Neuro-ophthalmic Diseases. *Nigerian Journal of Paediatrics 1996; 23 (1): 21.* A 12-month prospective study of neuro-ophthalmic diseases in 56 children, aged between 10 days and 15 years has shown an approximately equal sex distribution. The three commonest diseases seen were 18 cases of optic atrophy, 17 cases of nystagmus and 12 cases of cortical blindness. Less common cases included myasthenia gravis, migraine and hypoplastic discs. There were 44.6 percent cases of blindness. Preventive measures as well as early diagnosis and treatment of predisposing conditions are recommended in order to reduce the incidence of these diseases, while rehabilitation of the visually handicapped should be vigorously pursued.

Introduction

STUDIES on general paediatric ophthalmic diseases among Nigerian children have been undertaken by Majekodunmi [1] who in a hospital-based study in Lagos, found refractive error to be the commonest. Similarly, Abiose, [2] working in the northern part of the country, studied 500 children over a six-month period and found the largest number of the subjects to have conjunctivitis. Other studies on specific paediatric eye diseases such as glaucoma, conjunctivitis and uveitis both in Nigeria and elsewhere, have also been reported.[3-6] However, to the best of our knowledge, there has been little or no studies on neuro-ophthalmic diseases among our childhood population. The aim of the present communication was therefore, to analyse the paediatric neuro-ophthalmic diseases seen in Lagos University Teaching Hospital (LUTH) and also to highlight their role in the aetiology of blindness and its preventive measures.

College of Medicine, University of Lagos

Department of Surgery

* Consultant Ophthalmologist

Patients and Methods

One thousand consecutive children, aged 16 years and below, seen at the general eye clinics at LUTH from January to December 1989, formed the subjects of the present study. This number included 110 hospitalized children referred to the eye clinic and others who presented directly to the clinic, or were referred from other clinics. Using detailed clinical protocol, the following information was recorded

about each patient: age, sex, family history, antenatal and parental history, post-natal period, developmental milestones where appropriate and clinical features. Specific examinations carried out included visual acuity, external eye examination, slit lamp biomicroscopy and direct and indirect dilated fundoscopy with an ophthalmoscope. When indicated, examination under anaesthesia with or without refraction was also undertaken. Haemogram, bacteriological studies, skull radiographs and other appropriate investigations were carried out when available, in order to confirm the diagnosis.

Results

Of the 1,000 patients seen in the eye clinic over the one-year period, 56 children (29 males and 27 females) had neuro-ophthalmic diseases. The ages of the 56 patients ranged between 10 days and 15 years, mean age being four and a half years. Twentyeight of the patients were hospitalized. Of the 56 patients, 39 (69.6 percent) were aged five years and below, while 11 (19.6 percent) were aged between six and 10 years. As can be seen in the Table, optic atrophy was the commonest neuro-ophthalmic disease, being present in 18 (32.1 percent) of the 56 cases; this was followed by 17 (30.4 percent) cases of nystagmus and 12(21.4 percent) of cortical blindness. The causes of optical atrophy in the 18 cases included pyogenic meningitis in four cases, birth asphyxia in three, hydrocephalus in two, pyrexia of unknown origin in one and eight of unknown causes. Nystagmus was associated with congenital cataract in 75 percent of the cases and with albinism in 25 percent. The albinism was of the tyrosinase negative, oculocutaneous type. All the cases of cortical blindness had normal pupillary reaction to light with no ocular pathology. Cortical blindness was associated with pyogenic meningitis which manifested with pyrexia and repeated seizures in four cases, asphyxia and repeated seizures in four cases, asphyxia in four, traumatic head injury in two while the cause was unidentified in two others.

TABLE

Neuro-ophthalmic Diseases in 56 Patients

Disease	*No of Cases*	*Percent of Total*
Optic atrophy	18	32.1
Nystagmus	17	30.4
Cortical blindness	12	21.4
Myasthenia gravis	5	8.9
Migraine	2	3.6
Hypoplastic discs	2	3.6
Total	56	100.0

The five cases of myasthenia gravis were primarily ocular without systemic clinical signs. Other associated features included progressive ptosis that tended to be worse in the evening and, in two cases, palsy of the medial and superior recti. There were two (3.6 percent) cases each, of migraine and hypoplastic discs. The former occurred in males aged 12 years and 15 years and had no significant effect on the vision. There was no associated ptosis or ophthalmoplegia and the haemoglobin genotype was AA in each case. The two cases of hypoplastic discs had marked diminution of vision.

Blindness in one or both eyes occurred in

25 (44.6 percent) of the 56 cases with neuro-ophthalmic disorders. The causes of the blindness included optic atrophy in 16 (64.0 percent), cortical blindness in eight (32.0 percent) and hypoplastic discs in one (4.0 percent).

Discussion

To our knowledge, this is the first study on neuro-ophthalmic diseases in our children. Out of the 56 cases, optic atrophy accounted for 32.1 percent, but although it was the commonest neuro-ophthalmic disease in the series, it was second to cataract as a cause of blindness among the 1,000 patients that presented to the eye clinic during the study period. This is in contrast to the earlier findings of Olurin[7] and Elebesunu[8] in which measles keratopathy was the commonest cause of blindness, while optic atrophy was reported by these workers as the third cause. It seems likely that the expanded programme on immunization introduced about a decade ago, has contributed to the reduction in the incidence of blindness from measles. Optic atrophy due to meningitis and birth asphyxia is preventable through health education of the mothers who should be advised to seek early medical care for their sick children or when they themselves are pregnant. Improved socioeconomic conditions will also help to reduce blindness caused by meningitis.

Nystagmus which constituted 30.4 percent of the neuro-ophthalmic cases, was commonly associated with congenital cataract and albinism. Cataract causes sensory deprivation, while there is hypoplasia or aplasia of the macula in albinism resulting in sensory nystagmus. Early diagnosis and surgery for cataract in these cases may also reduce the incidence of blindness among these children, but unfortunately, out of ignorance, most parents will not consent to their children undergoing this operation. An enlightenment programme on the visual benefits of surgery as well as the visual rehabilitative measures available after surgery, should help in reducing or abolishing the incidence of nystagmus and sensory deprivation associated with congenital cataract. Children with albinism are visually handicapped and refractive errors in such children should be corrected early, in order to minimise their disability.

Cortical blindness which occurred in 12 children was associated with high fever and meningitis in four cases. The fact that both meningitis and high fever associated with malaria, especially cerebral malaria, could result in cortical blindness probably informed the treatment of these children with both antibiotics and antimalarials. It is interesting to note that none of these cases was due to sickle-cell disease which is also known to cause cortical blindness. Early diagnosis and treatment of febrile conditions such as malaria and improved health care delivery are some of the measures that should reduce the incidence of cortical blindness.

In keeping with reports from Benin,[9] all our cases of myasthenia gravis had the primary ocular type; furthermore, the oldest in our series was 11 years old, while four of the six cases from Benin were under the age of 20 years. Our two cases of migraine had no associated ptosis or ophthalmoplegia as reported by Osuntokun and Osuntokun[10] The visually handicapped cases which accounted for 21.4 percent will need to be referred to special schools for the blind and the school for the handicapped, for

primary and vocational education. Reduction of the incidence of blindness and visual handicap from neuro-ophthalmic disorders will require multidisciplinary efforts. The obstetrician, paediatrician, health educator and ophthalmologist will need to work together from the antenatal period through adolescent life to achieve this goal.

Acknowledgements

I wish to thank Professor A A Majekodunmi for useful suggestions and Mr R A Morakinyo for secretarial assistance.

Refetences

1 Majekodunmi S. Analysis of ophthalmic diseases in Nigerian children. *J Pharm Med Sci* 1979;5: 127-9.

2 Abiose A. Paediatric ophthalmic problems in Nigeria. *J Trop Paediatr* 1985; **31:** 30-5.

3 Rice NSC. Management of infantile glaucoma. *Br J Ophthalmol* 1974; 56: 294-9.

4 Majekodunmi AA. Uveitis in Nigerian children. *Nig Med J* 1978; 8: 425-7

5 Majekodunmi S. Vernal conjunctivitis in Nigerian children. *J Paediatr Ophthalmol Strabismus* 1978; 15: 176-8.

6 Conrad L, Giles MD. Uveitis in childhood. *Ann Ophthalmol* 1989; 21: 13-28.

7 Olurin OO. Etiology of blindness in Nigerian children. *Am J Ophthalmol* 1970; **70:** 533-40.

8 Elebesunu M. Aetiology of childhood blind ness in Benin city, Nigeria. *Pub Hlth J* 1987; **101:** 59-62.

9 Ayanru JO. Ocular myasthenia gravis in Nigeria *Nig Med J* 1978; 8: 137- 40.

10 Osuntokun BO, Osuntokun O. Complicated migraine and haemoglobin AS in Nigerians. *Br Med J* 1972; 2: 621-2.