Epidemic optic neuropathy in primary school children in Dar es Salaam, Tanzania

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Abstract

***Background*—An epidemic of bilateral optic neuropathy is affecting large numbers of people aged between 10 and 40 in Dar es Salaam, the capital city of Tanzania. The disease is characterised by acute onset of bilateral visual impairment, bilateral impairment of colour vision, and a characteristic temporal pallor of the optic discs. The disease often occurs in association with peripheral neuropathy and sensorineural hearing loss. This report presents the first data on disease prevalence in adolescents, based on a rapid assessment of schoolchildren.**

***Methods*—Three schools in Dar es Salaam were visited and all children aged between 10 and 16 were screened for the disease.**

***Results*—The prevalence of bilateral optic neuropathy among the schoolchildren is estimated to be 1.0% (95% CI 0.5–1.4%). This suggests that 5000 children (95% CI 2600–7300) aged 10–16 in Dar es Salaam may have the disease.**

***Conclusion*—The results of this rapid assessment indicate this epidemic is a major public health problem. The prevalence of the disease in the community is likely to be far higher than found in this survey because children with the disease may have withdrawn from school. As the disease predominantly affects young adults, resulting in impaired vision and hearing, the economic and social consequences are enormous.**

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An epidemic of bilateral optic neuropathy is affecting large numbers of people aged between 10 and 40 in Dar es Salaam, the capital city of Tanzania.[1] The disease is characterised by an acute onset of bilateral visual impairment, bilateral impairment of colour vision, and a characteristic temporal pallor of the optic discs. The disease often occurs in association with peripheral neuropathy and sensorineural hearing loss. A detailed clinical description of the optic and peripheral neuropathy, including fundus photographs appears elsewhere.[2] The Tanzanian optic neuropathy cases closely resemble a recent optic neuropathy epidemic in Cuba in which at least 50 000 cases were reported[3–6] and other epidemics previously described in the Caribbean and Africa.[7 8]

The Tanzanian outbreak appears to have been ongoing for 10 years. A small number of optic neuropathy cases were noticed by local ophthalmologists in 1988 and reported in a letter to the *Lancet*.[9] New cases have continued to occur. Around 1500 incident cases presented to community eye clinics in Dar es Salaam during 1996, indicating the outbreak has reached epidemic proportions. This report presents the first data on disease prevalence in adolescents, based on a rapid assessment of schoolchildren.

Methods

Three primary schools were surveyed, two in Ilala District and one in Kinondoni District of the city. The three schools were selected because their catchment areas covered large parts of the city. It was not feasible in this rapid assessment to examine a random sample of students from a larger number of schools because each school principal wanted all their students examined.

At each school all children in classes III to VI (ages 10–16) had their visual acuity measured by an ophthalmologist using a Snellen chart and pinhole correction. All those with visual acuity 6/9 or worse for both eyes were flagged to be seen by a senior ophthalmologist who visited the schools over the following 4 weeks.

At the follow up visit flagged students had their colour vision sensitivity assessed using Tokyo Medical College (TMC) pseudo-isochromatic plates (eight plates) to identify those with bilateral reduced colour vision. TMC plates are of similar sensitivity to the Ishihara pseudo-isochromatic plates.[10] Fundus examination was carried out after pupil dilatation, using a direct ophthalmoscope with white and with red-free light. A clinical history was taken to determine whether symptoms of hearing loss or stomatitis were present. Neurological examinations to detect peripheral neuropathy were not carried out in this field study.

A student was classified as a definite case of bilateral optic neuropathy compatible with epidemic neuropathy if all three of the following features were present:

(1) visual acuity with pinhole correction of 6/9 or worse in both eyes, but a difference no greater than three lines of acuity between the eyes;

(2) bilateral impairment of colour vision (failure of at least 1 test plate in each eye) of an acquired pattern; and

(3) bilateral optic atrophy of characteristic type showing principally a loss of the caeco-central projection ("papillomacular bundle") of retinal nerve fibres.

Results

A total of 1715 students were examined across the three schools of whom 109 (6.4%) were flagged as having visual acuity of 6/9 or worse

Table 1 Characteristics of cases of optic neuropathy

Age	*Sex*	*Vision (R:L)*	*Colour deficit*	*Optic atrophy*	*Hearing loss*	*Stomatitis*
13	F	6/36:6/60	+	+	+	−
10	F	6/24:6/36	+	+	−	−
15	M	6/9:6/9	+	+	+	+
12	F	6/9:6/9	+	+	−	+
14	M	6/18:6/18	+	+	+	−
12	M	6/18:6/36	+	+	−	−
16	F	6/9:6/9	+	+	−	−
12	F	6/36:6/36	+	+	−	−
14	F	6/36:6/36	+	+	−	−

Table 2 Best visual acuity for cases of optic neuropathy identified in school survey and for cases presenting to community eye clinics

Best visual acuity	*School survey (n=9) (%)*	*Community eye clinics (n=234) (%)*
6/9	33.3	5.6
6/12	0.0	6.4
6/18	22.3	12.8
6/24	11.1	17.9
6/36	33.3	36.8
6/60 or worse	0.0	20.5

for both eyes. Fifty nine (54.1%) of 109 flagged students were examined by the senior ophthalmologist. The remaining 50 flagged students were absent from school on each of the subsequent days the senior ophthalmologist visited.

Nine definite cases of bilateral optic neuropathy were found among the 59 flagged cases examined. It is not known how many cases were present among the 50 flagged students who did not reattend. We believe disease occurrence among the 50 flagged cases not examined by the ophthalmologist is likely to be similar to that among the 59 flagged cases who were examined, as is more fully discussed below. Based on this assumption, the overall prevalence of disease among the schoolchildren is 1.0% (95% CI 0.5–1.4%).

There was no difference in disease prevalence between boys and girls, among the three schools, or among the various residential areas from which the children originated. Table 1 shows clinical details of the nine cases of optic neuropathy. All had unequivocal bilateral optic atrophy on funduscopy, and one also had symptoms suggestive of peripheral neuropathy.

Two cases reported hearing impairment. It was not possible to undertake audiometric assessment. However, audiometry on clinic based cases has indicated that two thirds of cases with sensorineural hearing impairment (20+ dB broad frequency loss) do not self report a hearing problem. A higher proportion of these children may have hearing loss than is suggested by self reporting.

The estimated population of Dar es Salaam was 3.0 million in 1996, of which 17% were aged 10–16. Applying the prevalence finding of 1.0% to this population suggests that around 5000 children aged 10–16 (95% CI 2600–7300) in the city may have optic neuropathy.

Discussion

Our estimate of 1.0% disease prevalence among the schoolchildren is likely to be an underestimation of disease prevalence in the community. The severity of vision loss among the nine school cases is less than among similar aged cases presenting to community eye clinics (Table 2): 55.5% of the school cases had best visual acuity of 6/18 or better, compared with only 24.8% of those attending community eye clinics. None of the school cases had best visual acuity below 6/36, whereas 20.5% of the optic neuropathy cases presenting to the community eye clinic had best visual acuity of 6/60 or worse. This suggests that more severely affected cases had withdrawn from school and only the remaining milder cases were seen in this survey.

The optic neuropathy epidemic in Dar es Salaam predominantly affects the poor. It is generally the poor who fail to complete their schooling or attend intermittently because of economic hardship. Thus, those at highest risk of having the disease are least likely to attend school, or at best attend intermittently. A complete listing was obtained from each class teacher of children who registered at the start of the year. A total of 2455 students enrolled, but only 1715 (70%) were attending when we surveyed the schools. Similarly, 50 of the 109 flagged students were absent from school on all of the days when the senior ophthalmologist visited to examine the flagged cases. While some were sick, most were attending school intermittently. These attendance patterns lead us to believe the prevalence of disease among adolescents in the city is higher than the 1.0% reported here.

The disease definition used in this rapid assessment only included those cases with visual acuity of 6/9 or worse for both eyes plus failure of at least one colour vision test plate for both eyes. This is a more stringent definition than that used in Cuba.[4] Thus, cases of bilateral optic neuropathy with visual acuity of 6/6 or better in both eyes and cases with normal colour vision would not have been detected in the present study.

The Tanzania cases previously described from the Muhimbili Medical Centre[1 2] underwent extensive clinical examinations to exclude alternative underlying causes such as demyelinating disease, post-infectious optic neuritis, or genetically determined optic neuropathies. The clinical examination undertaken at the schools could not exclude these alternative underlying causes. However, such other causes of bilateral optic neuropathy have rarely been seen in Dar es Salaam. The disease seen in the schoolchildren and in cases presenting to the community eye clinics is characterised by its symmetrical involvement of the optic nerves and the loss of the papillomacular bundle of the nerve fibre layer. It is most likely that the cases of optic neuropathy identified in the schools survey have epidemic optic neuropathy.

Since undertaking this survey, we have established a screening programme for the disease. All people aged between 10 and 40 who attend one of the city's community eye clinics are screened to identify those with bilateral reduced vision and bilateral loss of colour sensitivity. During an initial 3 month period (August–October 1996) over 5% of all new patients presenting to the eye clinics were flagged using these criteria and confirmed by an ophthalmologist as having epidemic optic

neuropathy. This supports the school survey results and indicates the epidemic has become a major public health problem in Dar es Salaam. A population based survey to provide estimates of disease prevalence in the community has recently been undertaken.

The cause of the Tanzanian epidemic is currently unknown. A number of epidemiological studies are under way and full results are expected to be available shortly. Cyanide intoxication and mitochondrial DNA mutations associated with Leber's hereditary optic neuropathy have been ruled out.[11] Weight loss and stomatitis have been common accompanying features in Tanzanian cases, the latter occurring in five of the nine cases identified in the present study. Preliminary micronutrient studies have shown early acute cases are deficient in B group vitamins, indicating a nutritional role in the disease. Interestingly, blood samples from healthy controls have also shown similar deficiencies. We suspect the Tanzania epidemic has a multi-factorial aetiology. Micronutrient deficiencies are probably a necessary factor, but not sufficient in themselves to produce the disease. We believe some other genetic, dietary, or environmental factor is playing a role.

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